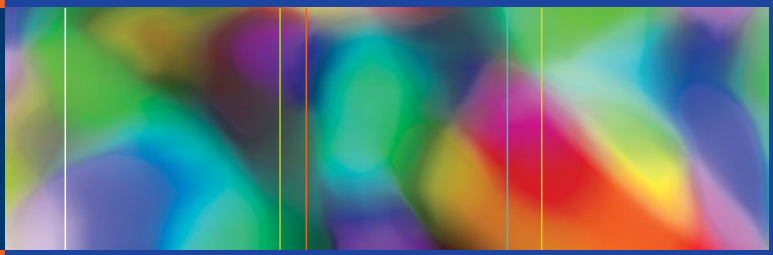
Vincent E. Friedewald



Clinical Guide to Cardiovascular Disease

Springer

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Vincent E. Friedewald

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Vincent E. Friedewald
Division of Cardiology
UT Health Science Center at Houston Division of Cardiology
Houston, Texas, USA

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To our patients – our best teachers, when we listen

Preface

The *Clinical Guide to Cardiovascular Disease* culminates over 70 years of disease data collection, begun by my father, Vincent E. Friedewald, Sr. M.D., when he was awarded patent rights for the first medical computer – a mechanical index card-sorting machine – for differential diagnosis and other elements of medical decision support (Figs. 1 and 2). Today these data reside within the largest medical relational database in the world,[1] comprising a unified lexicon of thousands of confirmed clinical manifestations of human disease. This massive collection of information is the foundation for the *Clinical Guide to Cardiovascular Disease* and the preceding *Clinical Guide to Bioweapons and Chemical Agents* (Friedewald VE, Springer-Verlag, 2006).

Unlike traditional books, the *Clinical Guide* is specifically designed for rapid access to disease information, segregated into keyword data elements organized under 20 separate headings relevant to clinical care. In addition, external links are provided for supplemental and updated information.

The bulk of content in the *Clinical Guide* is focused on information essential to correct disease diagnosis, for good reason. According to the Institute of Medicine (IOM),[2] "*diagnosis—and, in particular, the occurrence of diagnostic errors—has been largely unappreciated in efforts to improve the quality and safety of health care. The result of this inattention*

[1] COR Medical Technologies, Inc. https://www.cormedicaltechnologies.com/landing.aspx.

[2] National Academies of Sciences, Engineering, and Medicine. *Improving diagnosis in health care*. Washington: The National Academies Press; 2015.

is significant: The committee concluded that most people will experience at least one diagnostic error in their lifetime, sometimes with devastating consequences." The IOM report further points out that:

- Five percent of adults in the USA seeking outpatient care experience a diagnostic error.
- Diagnostic errors contribute to 10 % of deaths.
- 6–17 % of adverse hospital events are due to diagnostic errors.
- Diagnostic errors are the leading cause of malpractice claims in the USA.

The *Clinical Guide* directly addresses the challenges of diagnostic accuracy with eight sections of information relevant to diagnosis in every disease chapter:

- Signs and Symptoms
- Predisposing/Comorbid Conditions
- Differential Diagnosis
- ECG
- Genomics
- Imaging
- Laboratory
- Other Tests

While the main emphasis of the *Clinical Guide* content is on diagnosis, treatment is presented in a more generic form. The reasons for this less-granular information about treatment are threefold:

1. Treatment recommendations are extremely dynamic, constantly changing as new outcome studies for current treatments are completed and as new modalities emerge, thereby greatly reducing the shelf life of treatment information.
2. Treatment is more and more being personalized according to individual patient preferences, circumstances, comorbidities, and other factors, all of which cannot be accommodated in one book.

3. Treatment recommendations are exquisitely defined and openly accessed in major Guidelines – especially those written by the American College of Cardiology/American Heart Association and by the European Society of Cardiology; they are linked to each disease in the *Clinical Guide* when they exist and are relatively current.

In addition to diagnostic and treatment information, other information that is often important to patient management is included in separate sections, such as demographics, pathophysiology, and clinical course. The style of the *Clinical Guide* is designed for easy use on mobile devices, as well for rapid access in its print form. This design includes extensive use of abbreviations, keywords, short phrases, and external links to both professional and patient information.

All of the content in the *Clinical Guide* was made possible by thousands of researchers worldwide via their contributions to the many excellent cardiovascular and general medical journals we are fortunate to have at our disposal. To them, I offer my deepest thanks, and an apology: because this book is so content-rich, it would take a second book just to accommodate standard referencing, and even then many of these primary authors would likely be slighted. Thus, I have chosen to list only a relatively few, select articles in the section Professional Information, along with their links, that I encourage readers to access for additional information.

I acknowledge and thank the many authors of major Guidelines, especially Guidelines written by the American College of Cardiology and American Heart Association, and by the European Society of Cardiology. Such Guidelines are remarkable documents – in my opinion, far too underutilized by practitioners – and a rich source of information for this book. In places, I have gone so far as to extract exact language from Guidelines, with the source specified.

I thank some of the many persons who assisted in compiling the *Clinical Guide* information, especially Doctor Patrick Finnigan, Mr. Ryan Carbone, my daughter Natalie Nieto, and the cardiology Fellows at The Cleveland Clinic, selected for

me by my friend and colleague, Doctor James Young. Those Fellows are Doctor Mohammed B. Elshazly, Doctor Samuel Horr, Doctor Manju Pai, Doctor Grant Reed, Doctor Brett Sperry, and Doctor Amanda Vest.

As further testament to the digital age, I thank Mr. John Scott – who does not even pretend to understand a word in this book, nor do I have even the most remote notion of what he does – for building the software program that so greatly facilitated writing this book.

Finally, I offer a great big Texas-size mountain of gratitude to my publisher at Springer-Verlag, Mr. Grant Weston, for his patience, which is a vanishing virtue.

Houston, TX, USA

Vincent E. Friedewald
MD, FACC, FACP

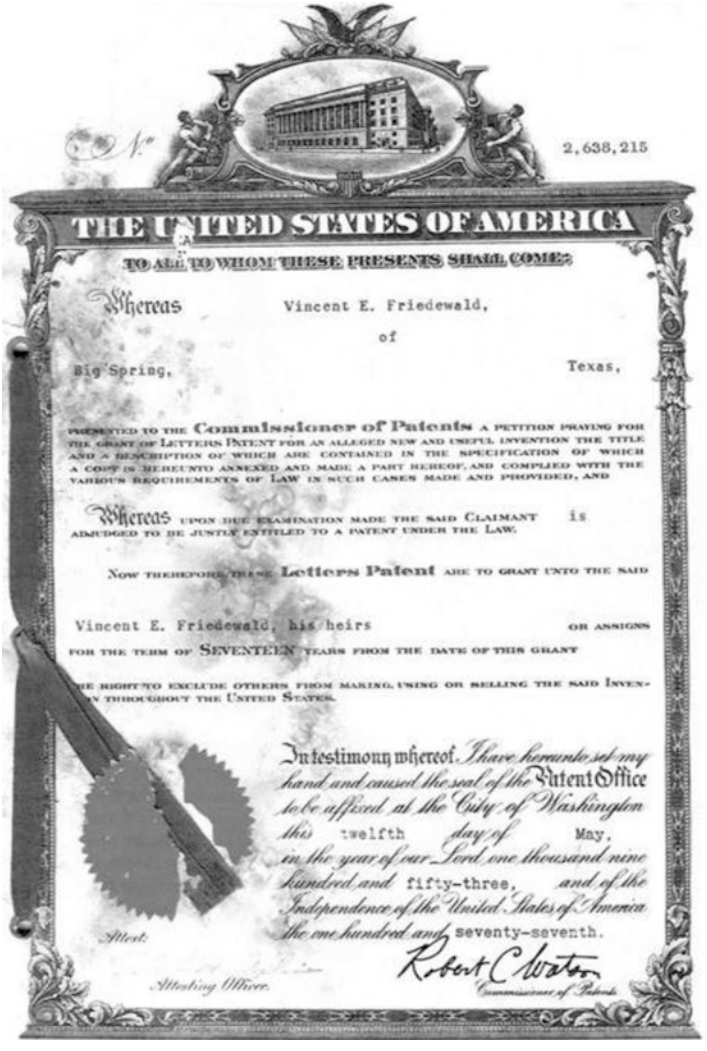

FIG. 1 United States patent award in 1953 to Vincent E. Friedewald, Sr, M.D., for the first medical computer

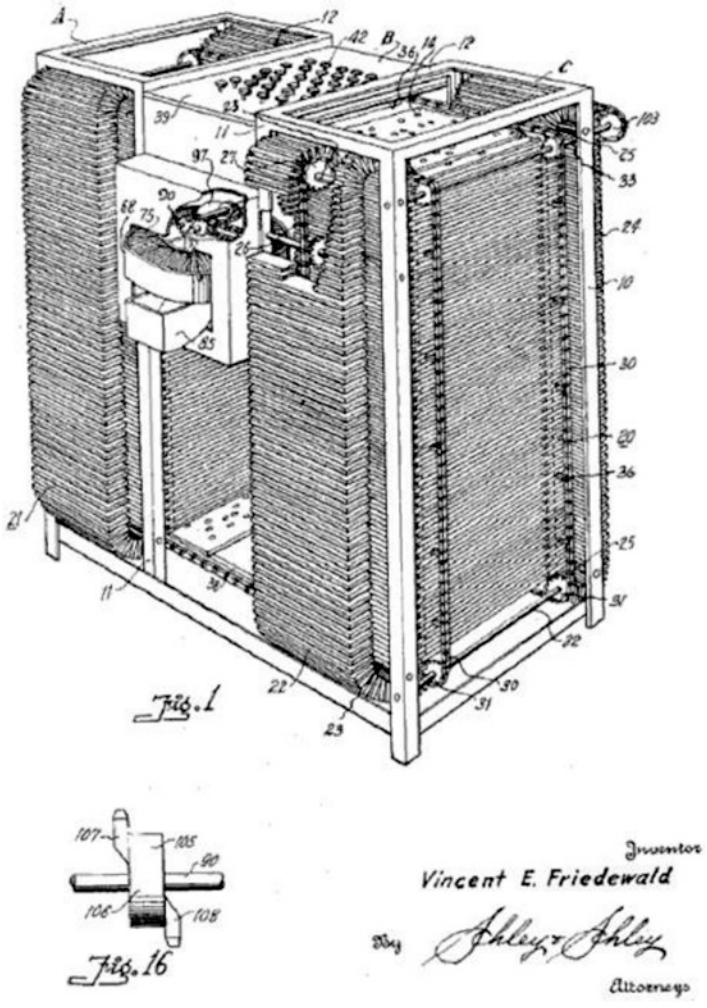

Fig. 2 Exterior of Dr. Friedewald, Sr's, invention of the first medical computer. Note the keys at the top center of the machine, where clinical information such as signs and symptoms were entered for differential diagnosis

Abbreviations

A2	Aortic valve second heart sound
AAA	Abdominal aortic aneurysm
AATS	American Association for Thoracic Surgery
ABD	Abdominal
ACC	American College of Cardiology
ACCF	American College of Cardiology Foundation
ACCP	American College of Chest Physicians
ACEI(S)	Angiotensin converting enzyme inhibitor(s)
ACS	Acute cardiac syndrome
AED	Automated external defibrillator
AF	Atrial fibrillation
AHA	American Heart Association
AMI	Acute myocardial infarction
ANT	Anterior
AOS	Aneurysms-osteoarthritis syndrome
APOB	Apolipoprotein B
AR	Aortic regurgitation
ARB(S)	Angiotensin receptor blocker(s)
ARVD	Arrhythmogenic right ventricular dysplasia
AS	Aortic stenosis
ASA	Aspirin, acetylsalicylic acid
ASD	Atrial septal defect
AT	Atrial tachycardia
ATP	Adenosine triphosphate
ATVR	Atrioventricular
AV	Aortic valve
A-V	Arterio-venous
AVN	Atrioventricular node

AVNRT	Atrioventricular node reentry tachycardia
AVR	Aortic valve replacement
AVRT	Atrioventricular reentry tachycardia
AVSD	Atrioventricular septal defect
BBB	Bundle branch block
BP	Blood pressure (arterial)
BPM	Beats per minute
BS	Breath sounds
BUN	Blood urea nitrogen
BVH	Biventricular hypertrophy
CABG	Coronary artery bypass graft surgery
CAD	Coronary artery disease
CAS	Carotid artery stenosis
CAF	Coronary arteriovenous fistula
CAV	Cardiac allograft vasculopathy
CCA	Circumflex coronary artery
CCB(S)	Calcium channel blocker(s)
CKD	Chronic kidney disease
CKMB	Ck-Mb fraction
CMRI	Cardiac magnetic resonance imaging
COA	Coarctation of aorta
CONT	Continuous
COPD	Chronic obstructive pulmonary disease
CPAP	Continuous positive airway pressure
CPR	Cardiopulmonary resuscitation
CPVT	Catecholaminergic polymorphic ventricular tachycardia
CRT	Cardiac resynchronization therapy
CT	Computed tomography
CVA	Cerebrovascular accident
CVD	Cardiovascular disease
CXR(S)	Chest X-ray(s)
DCM	Dilated cardiomyopathy
DECR	Decreased
DEPR	Depression(s)
DESC	Descending
DIAS	Diastolic, diastole
DIL	Dilation, dilated

DM	Diabetes mellitus
DSA	Digital subtraction angiography
DVT	Deep vein (venous) thrombosis
DYSRHY	Dysrhythmia
ECG	Electrocardiogram
ECHO	Echocardiogram (includes Doppler, transesophageal)
ECMO	Extracorporeal membrane oxygenation
EF	Ejection fraction
EG	For Example
ELEV	Elevation(s)
EMB	Endomyocardial biopsy
EMF	Endomyocardial fibrosis
EMG	Electromyogram
EP	Electrophysiology test
ERS	Early repolarization syndrome
ESC	European Society of Cardiology
ESP	Especially
EXT	External
FFR	Fractional flow reserve
FMC	First medical contact
FMD	Fibromuscular dysplasia
GCM	Giant cell myocarditis
GDMT	Guideline directed medical therapy
HB	Heart block
HCM	Hypertrophic cardiomyopathy
HDL-C	High-density lipoprotein cholesterol
HEFH	Heterozygous familial hypercholesterolemia
HF	Heart failure
HFpEF	Heart failure preserved ejection fraction
HFrEF	Heart failure reduced ejection fraction
HIV	Human immunodeficiency virus
HOCM	Hypertrophic obstructive cardiomyopathy
HOEF	Homozygous familial hypercholesterolemia
HR	Heart rate
HTN	Hypertension
IART	Intraatrial reentrant tachycardia
ICD(S)	Implantable cardiac defibrillator(s)

ICD-10	International Classification of Diseases, Tenth Revision
ICS	Intercostal space
IOC	Iron overload cardiomyopathy
IE	Infective endocarditis
INCR	Increased
INF	Inferior
INSP	Inspiration
INT	Internal
IST	Inappropriate sinus tachycardia
IV	Intravenous
IVC	Inferior vena cava
IVS	Interventricular septum
JVP	Jugular venous pulse/pulsation
L	Left
LA	Left atrium
LAA	Left atrial appendage
LAD	Left anterior descending coronary artery
LAT	Lateral
LBB(B)	Left bundle branch (block)
LCA	Left coronary artery
LDL-C	Low-density lipoprotein cholesterol
LE	Lower extremity
LEAD	Lower extremity artery disease
LGE	Late gadolinium enhancement
LLQ	Lower left quadrant
LMWH	Low molecular weight heparin
LQTS	Long QT syndrome
L-R	Left to right
LSB	Left sternal border
LUQ	Left upper quadrant
LV	Left ventricle
LVAD	Left ventricular assist device
LVEDP	Left ventricular end-diastolic pressure
LVEDV	Left ventricular end-diastolic volume
LVH	Left ventricular hypertrophy
LVOT	Left ventricular outflow tract
LVSV	Left ventricular stroke volume
M1	Mitral valve first heart sound

MACE	Major adverse cardiovascular/cerebrovascular events
MALE	Major adverse limb events
MAP	Mean arterial pressure
MPI	Myocardial perfusion imaging
MR	Mitral regurgitation
MRA	Magnetic resonance angiography
MRI	Magnetic resonance imaging
MS	Mitral stenosis
MUR	Murmur
MV	Mitral valve
MVP	Mitral valve prolapse
MYOCARD	Myocardial, myocardium
NA	Not applicable
NEG	Negative
NO	Nitric oxide
NS	Nonspecific/no specific
NSTEMI	Non ST segment elevation myocardial infarction
NSVT	Nonsustained ventricular tachycardia
O2	Oxygen
OSA	Obstructive sleep apnea
P2	Pulmonic valve second heart sound
PA	Pulmonary artery
PAC(S)	Premature atrial contraction(s)
PAD	Peripheral arterial disease
PAH	Pulmonary arterial hypertension
PAROX	Paroxysmal
PAT	Paroxysmal atrial tachycardia
PCI	Percutaneous coronary intervention
PCR	Polymerase chain reaction
PDA	Patent ductus arteriosus
PET	Positron emission tomography
PFT	Pulmonary function test
PH	Pulmonary hypertension
PHEO	Pheochromocytoma
PPCM	Peripartum cardiomyopathy
PPV	Positive pressure ventilation
PRESS	Pressure

PROX	Proximal
PS	Pulmonary stenosis
PSVT	Paroxysmal supraventricular tachycardia
PTCA	Percutaneous transluminal coronary angioplasty
PV	Pulmonary valve
PVC(S)	Premature ventricular contraction(s)
PVR	Pulmonary vascular resistance
QOL	Quality of life
QTC	Corrected QT interval
RA	Right atrium
RAA	Right atrial appendage
RAAS	Renin aldosterone angiotensin system
RAS	Renal artery stenosis
RBB(B)	Right bundle branch (block)
RCA	Right coronary artery
RCM	Restrictive cardiomyopathy
RF	Radiofrequency
RHF	Right heart failure
R-L	Right to left
RLQ	Right lower quadrant
RSB	Right sternal border
RUQ	Right upper quadrant
RV	Right ventricle
RVEDP	Right ventricular end-diastolic pressure
RVEDV	Right ventricular end-diastolic volume
RVH	Right ventricular hypertrophy
RVOT	Right ventricular outflow tract
S/S	Signs and symptoms
S1	First heart sound
S2	Second heart sound
S3	Third heart sound (gallop)
S4	Fourth heart sound (gallop)
SAH	Systemic arterial hypertension
SCD	Sudden cardiac death
SD	Sudden death
SIHD	Stable ischemic heart disease
SLE	Systemic lupus erythematosus

SQTS	Short QT syndrome
STEMI	ST segment elevation myocardial infarction
SubAS	Subvalvular aortic stenosis (discrete)
SVA	Sinus of Valsalva aneurysm
SVAS	Supravalvular aortic stenosis
SVC	Superior vena cava
SVT	Supraventricular tachycardia
SX(S)	Sign(s)
SYMP(S)	Symptom(s)/symptomatic
SYS	Systolic, systole
T1	Tricuspid valve first heart sound
TAVR	Transcatheter aortic valve replacement
TEE	Transesophageal echocardiogram
TG(S)	Triglyceride(s)
TGA	Transposition of great arteries
TIA	Transient ischemic attack
TIC	Tachycardia-induced cardiomyopathy
TIMI	Thrombolysis in myocardial infarction
TNF	Tumor necrosis factor
TNG	Tri-nitroglycerin
TOF	Tetralogy of Fallot
TR	Tricuspid regurgitation
TS	Tricuspid stenosis
TSH	Thyroid stimulating hormone
TTE	Transthoracic echocardiogram
TV	Tricuspid valve
TVP	Tricuspid valve prolapse
UA	Unstable angina, urinalysis
UE	Upper extremity
UTI	Urinary tract infection
VF	Ventricular fibrillation
VAD	Ventricular assist device
VMA	Vanillylmandelic acid
VSD(S)	Ventricular septal defect(s)
VT	Ventricular tachycardia
WHO	World Health Organization
WPW	Wolff-Parkinson-White syndrome
WS	Williams syndrome

Contents

Chapter 1
Abdominal Aortic Aneurysm

Management Keys

- Early screening/diagnosis/repair as high mortality rate occurs with rupture
- Consider possible CAD in patients undergoing surgery for AAA [22]
- Urgent repair with symptom onset
- Post-rupture BP/HR control
- Long-term surveillance after endovascular repair for aneurysm sac reperfusion/late rupture
- One-time screening with abdominal ultrasound in males age 65–75 years with history of tobacco use may decrease mortality
- Stop tobacco use [2]

ICD-10 Code

I71.4 Without rupture
I71.3 With rupture

Alternate Names/Abbreviation

AAA

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_1,

Description/Etiology

Segmental saccular full-thickness dilatation of abdominal aorta

>50 % of N aorta diameter
Threshold: 3.0 cm

Location: between diaphragm and aortic bifurcation; most often involves segment between renal arteries and aortic bifurcation

Suprarenal: involves origin of 1/more visceral arteries
Pararenal: involves origin of renal arteries
Infrarenal: involves aorta below renal arteries (85 %); often involv iliac arteriess

Less common types:

Familial [1]
Inflammatory [24]
Mycotic [20]

Comorbid Conditions [3]

ANEURYSMS OF OTHER MAJOR ARTERIES [10]
CORONARY ARTERY DISEASE [22]
IgG4-RELATED DIS
PERIPHERAL ARTERY DISEASE
TOBACCO USE [2]

Demography

M > F
Advanced age [4]
Ethnicity: more common in whites than African-Americans, Asians, Hispanics
Incidence decreasing, perhaps due to declining tobacco use

Pathophysiology [21]

Degradation of abdominal wall (all layers) including:

- Elastin/collagen destruction in media/adventitia by proteases
- Loss of smooth muscle cells/thinning of media
- Neovascularization
- Wall infiltration by lymphocytes/macrophages

Progressive aortic enlargement in accordance with Laplace Law by 0.2–0.5 cm/year

Renin-aldosterone-angiotensin system may play important developmental role [25]

Signs/Symptoms [9] [13]

ABDOMEN – BRUIT
ABDOMEN – FULLNESS
ABDOMEN – MASS, PULSATING [5]
ABDOMEN – MASS, TENDER
ABDOMEN – PAIN
APPETITE – DECR (ANOREXIA) [8]
BACK – PAIN [6]
BOWEL MOVEMENTS – CONSTIPATION [8]
BREATHING – DIFF (DYSPNEA) [8]
CHEST – PAIN [8]
FLANK – BRUIT
FLANK – PAIN [7]
NAUSEA [8]
VOMITING (EMESIS) [8]

Differentiation

Other causes of abdominal pain
Other causes of back pain
Paraaortic/other abdominal masses (pseudoaneurysm)

Complications

AV fistula with high output cardiac failure [11]
Disseminated Intravascular Coagulation
Expansion/obstruction of IVC
Rupture [12] [15] [16]

Laboratory

BLOOD, D-DIMERS – INCR [23]

ECG

Often abnormal due to comorbidities [especially CAD]

Imaging [18]

AORTA, ABD, SIZE – INCR [15]
AORTA, ABD, WALL – CALCIUM [14]
ULTRASOUND SCREENING [19]

Other Tests

Aortography
Coronary angiography if concomitant CAD suspected [22]
Digital subtraction angiography

Treatment: Nonpharmacologic

DC tobacco use [2]

Treatment: Pharmacologic

Hypertension control
Lipid control (especially with statins)

Treatment: Surgical/Invasive [17]

Coronary artery revascularization in select patients [22]
Endovascular repair
Surgical repair

Prevention

Diet high in fruits/vegetables
DC tobacco use [2]
Hypertension control
Regular exercise
Statins/cholesterol control
Ultrasound screening for select patients age 65–75 years [19]

Course

Highly variable
Rupture mortality: 85–90 %

Notes

[1] Possibly autosomal dominance with incomplete penetrance
[2] Tobacco use particularly strong risk factor; also associated with increased rate of expansion/rupture
[3] Except DM, which has negative relationship
[4] 5 % among men age >65 years by screening ultrasound
[5] Lateral systolic expansion on palpation
[6] May radiate to posterior LEs
[7] May radiate to anterior left thigh or scrotum with left genitofemoral nerve impingement
[8] Less common unless rupture has occurred
[9] Sudden onset of symptoms, especially severe pain, may indicate rupture
[10] Especially iliac artery (pulsatile groin mass) and popliteal artery
[11] Following rupture into IVC, iliac vein, renal vein

[12] Usually retroperitoneal but also into peritoneal cavity, adjacent vessels, GI tract; sudden abdominal/back pain, circulatory collapse
[13] Most patients asymptomatic until rupture; incidentally detected on exam or ultrasound
[14] Mainly lateral view; detectable only if walls calcified
[15] Note: CT measures 3–9 mm > ultrasound; risk of rupture (m): <4.0 cm – 0.5 %; 4.0–4.9 cm–1.5 %; 5.0–5.9 cm – 6.5 %
[16] Females rupture 3× more often than males and at smaller diameter; rupture also more likely in tobacco users and pts with hypertension
[17] Elective: males ≥5.5 CM; females ≥4.5–5.0 CM
[18] Echo, MRI useful for initial diagnosis and serial monitoring of size
[19] USPSTF grade B for males age 65–75 years who have ever smoked; grade C for those who have never smoked; insufficient evidence for females who have ever smoked; not recommended for females who have never smoked
[20] Mycotic aneurysm prevalence: 0.7–3 % of all aortic aneurysms; agents most often S aureus, salmonella, pseudomonas; high risk of expansion/rupture; also involve thoracic aorta, mesenteric branches, iliacs; often misdiagnosed early as presenting features may be only fever, malaise, leukocytosis
[21] Role of atherosclerosis less important than once believed although risk factors except DM similar
[22] Symptomatic/asymptomatic CAD occurs in 31–71 % of patients with AAA
[23] Plasma D-dimers may have role in diagnosis and prognosis
[24] Related to chronic periaortitis, including thoracic/abdomonal periaortitis and retroperitioneal fibrosis (Ormonds dis), IGG4-related disease
[25] Strong animal evidence that RAS over-activation promotes both thoracic and abdominal aneurysm development; many proposed mechanisms, including effects of angiotensin II on a diverse array of cell types and mediators

Guidelines

2011 ACCF/AHA focused update of the guideline for the management of patients with peripheral artery disease (Updating the 2005 Guideline)
Circulation. 2011;124:2020–45. http://circ.ahajournals.org/content/124/18/2020.
2014 ESC guidelines on the diagnosis and treatment of aortic diseases
Eur Heart J. 2014;35;2873–2926. http://eurheartj.oxfordjournals.org/content/ehj/35/41/2873.full.pdf.

Patient Information

Images

http://www.nlm.nih.gov/medlineplus/ency/imagepages/1149.htm.
http://www.nlm.nih.gov/medlineplus/ency/imagepages/18072.htm.

Medlineplus

ENGLISH

http://www.nlm.nih.gov/medlineplus/ency/article/000162.htm.

ESPANOL

http://www.nlm.nih.gov/medlineplus/spanish/ency/article/000162.htm.

Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/abdominal-aortic-aneurysm/basics/definition/con-20023784.

Cleveland Clinic

http://my.clevelandclinic.org/services/heart/disorders/aortic-aneurysm/hic-Abdominal-Aortic-Aneurysm.

Merck

http://www.merckmanuals.com/home/SearchResults?query=Abdominal+Aortic+Aneurysms++(AAA).

CDC-Aortic Aneurysm Fact Sheet

http://www.cdc.gov/dhdsp/data-statistics/fact-sheets/docs/fs-aortic-aneurysm.pdf.

Professional Information

Review

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Aortic Pseudoaneurysm (Case Report)

Circulation. 2013;128:674–5. http://circ.ahajournals.org/content/128/6/674.full.

Atherosclerosis

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D-Dimer

Eur Heart J. 2011;32:354–64. http://eurheartj.oxfordjournals.org/content/32/3/354.

Diabetes Mellitus

Eur J Vasc Endovasc Surg. 2014;47:243–61. http://www.sciencedirect.com/science/article/pii/S1078588413007405.

Disseminated Intravascular Coagulation

Eur J Int Med. 2005;16;551–60. http://www.sciencedirect.com/science/article/pii/S095362050500292X.

Disseminated Intravascular Coagulation

J Vasc Surg. 1986;4;184–6. http://www.sciencedirect.com/science/article/pii/0741521486904210.

Endovascular Repair (Review)

Eur Heart J. 2016;37:145–51. http://eurheartj.oxfordjournals.org/content/37/2/145.

Endovascular Repair; Patients Ineligible for Open Repair

N Engl J Med. 2010;362:1872–80. http://www.nejm.org/doi/full/10.1056/NEJMoa0911056.

Epidemiology: Global

Circulation. 2014;129:747–53. http://www.ncbi.nlm.nih.gov/pubmed/24249717.

Familial Type

J Vasc Surg. 2003;37:340–5. http://www.ncbi.nlm.nih.gov/pubmed/12563204.

Fruit/Vegetable Consumption

Circulation. 2013;128:795–802. http://circ.ahajournals.org/content/128/8/795.full.

Genetic Basis

Eur J Vasc Endovasc Surg. 2007;33:381–90. http://www.sciencedirect.com/science/article/pii/S1078588406006137.

IgG4-Related Disease

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Inflammatory with Retroperitoneal Fibrosis

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Inflammatory Type

JAMA. 2007;297:395–400. http://jama.jamanetwork.com/article.aspx?articleid=205226.

Inflammatory Type

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Octogenarians: Endovascular Repair

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Pathogenesis: RAAS

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Risk Factors

J Vasc Surg. 2010;52:539–48. http://www.jvascsurg.org/article/S0741-5214(10)01302-9/abstract.

Risk Factors

Circulation. 2009;119:2202–8. http://circ.ahajournals.org/content/119/16/2202.

Rupture/Dissection Risk Based on Size

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Rupture: Survival

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Screening: Recommendations

USPSTF

http://www.uspreventiveservicestaskforce.org/Page/Document/RecommendationStatementFinal/abdominal-aortic-aneurysm-screening.

Surveillance: Small AAA

J Vasc Surg. 2007;46:190–5. http://www.ncbi.nlm.nih.gov/pubmed/17540533?dopt=Abstract.

Tobacco

Arterioscler Thromb Vasc Biol. 2013;33:1473–7. http://atvb.ahajournals.org/content/33/7/1473.long.

Updates and More

https://clinicalguidecvd.com/aaa

Chapter 2
Acute Myocardial Infarction (Coronary Syndrome) (Heart Attack)

Management Keys

PREHOSPITAL

Monitor continuously and treat ventricular arrhythmias/cardiac arrest

Perform ECG by first medical contact within 10 min

Recognize atypical symptoms

Transport immediately by EMS to PCI-capable hospital

Treat pain, dyspnea, anxiety

HOSPITAL

Treat Type 2 AMI by correcting underlying cause

Implant ICD when appropriate for primary/secondary prevention of cardiac arrest/sudden death

Perform coronary reperfusion therapy in 90 min or less from time of first medical contact on all eligible patients with symptom onset within prior 1 h

Treat both STEMI and non-STEMI patients with aggressive Guideline-Directed Therapy

Treat, when appropriate, with drugs that improve long-term outcomes, including:

ACEIs
ARBs
ASA
Beta-Blockers

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_2,

Calcium Channel Blockers
Dual Antiplatelet therapy
Statins

Treat with fibrinolytics in non-PCI capable hospitals when PCI is delayed and in certain situations as part of pre-hospital care [55]

PRE-DISCHARGE
Provide lifestyle/risk factor modification to patients and caregivers

POST-DISCHARGE
Pay careful attention to patient medication adherence after discharge
Provide reinfarction/primary prevention education for patients and caregivers such as AHA/NIH "Act in Time to Heart Attack Signs"
Consider ICD implantation in patients with EF <35 % after optimal medical therapy for at least 40 days post-AMI, regardless of age [42]

ICD-10 Code

I21.3

Alternate Names/Abbreviation

Acute Coronary Syndrome
AMI
Heart Attack (LAY)
MI
Non-ST-Elevation Myocardial Infarction (non-STEMI) (NSTEMI)
Q-Wave Myocardial Infarction
ST-Elevation Myocardial Infarction (STEMI)
Transmural Myocardial Infarction

Description/Etiology

Clinical syndrome comprising symptoms of myocardial ischemia, ECG changes and subsequent release of myocardial biomarkers [19]
Myocardial cell death due to prolonged ischemia
Clinical forms:

STEMI

25–40 % of all AMIs
ECG findings: new elevation at J point in at least 2 contiguous leads of 2 mm or greater in males or 1.5 mm or greater in females in leads v2–v3/1.5 mm or greater in other contiguous chest leads or limb leads; most evolve to Q wave infarction [30]
Elevated cardiac biomarkers

NSTEMI

Elevated cardiac biomarkers without ECG findings of ST elevation [31] [32]

Most common cause: coronary atherosclerosis, including

Plaque dissection
Plaque erosion
Plaque fissuring
Plaque rupture
Associated with intraluminal thrombus leading to coronary flow obstruction/distal platelet embolus and resultant myocardial necrosis

Nonatherosclerotic causes (atherosclerosis may be contributing factor):

Allergic Acute Coronary Syndrome (Kounis Syndrome)
Amphetamines [2]
Anemia [2]
Type A Aortic Dissection involving aortic root
Arteritis (eg, SLE, Giant Cell, Rheumatoid Arthritis, Bechet Disease, Takayusu Disease)

CABG
Carbon Monoxide Poisoning
Cocaine
Coronary congenital anomaly
Coronary dissection

Spontaneous [24]
Trauma

Coronary embolus, including

Infective/noninfective Endocarditis
Air embolism
Mural thrombus, paradoxical

Coronary external compression/entrapment
Coronary spasm
Coronary stent thrombosis
Coronary trauma (blunt chest, PCI)
Disseminated Intravascular Coagulation
Hypercoagulable states
Hypertension (with/without LVH)
Hyperthyroidism [2]
Hyperviscosity syndromes
Hypotension
Kawasaki Disease
Maccallum plaque
Moyamoya Disease
Respiratory failure
Pheochromocytoma [2]
Polycythemia Vera
Raynaud's Disease
Tachycardia/bradycardia arrhythmias

Clinical classification of AMI (3rd Universal Classification of Myocardial Infarction):

Type 1: Spontaneous
Type 2: Secondary to ischemic imbalance [36]

Decreased O2 supply

Anemia
Bradyarrhythmia
Coronary embolus
Resp failure
Shock

Increased O2 demand

Hypertensive pulmonary edema
Severe Systemic Arterial Hypertension with LVH

Supraventricular Tachyaarhythmia
Ventricular tachyarrhythmia

Type 3: Resulting in death when biomarker values unavailable
Type 4: Related to PCI
Type 4b: Related to stent thrombosis
Type 5: Related to CABG

Silent myocardial infarction: asymptomatic patients who develop:

New pathologic Q waves during routine ECG followup
Imaging findings of myocardial infarction that cannot be attributed to coronary revascularization

Chronic inflammatory disease significantly increases AMI risk, including:

Psoriatic Arthritis
Rheumatoid Arthritis
Severe Psoriasis

Comorbid Conditions [51]

ANEMIA
ARTHRITIS

ATHEROSCLEROSIS IN OTHER CV AREAS
ATRIAL FIBRILLATION
CATARACT
CEREBROVASCULAR DISEASE
CHRONIC INFLAMMATORY DISEASE
CHRONIC KIDNEY DISEASE [4]
CHRONIC OBSTRUCTIVE PULMONARY DISEASE (EMPHYSEMA) [41]
CONGENITAL ABSENCE OF PERICARDIUM [IMPINGEMENT ON CORONARY ARTS]
DEPRESSION
DIABETES MELLITUS [1] [29] [35]
DYSLIPIDEMIA
ENDOMETRIOSIS
FABRY DISEASE
FAMILY HX: PREMATURE ATHEROSCLEROSIS
GOUT
HEART FAILURE
HUMAN IMMUNODEFICIENCY VIRUS INFECTION
HYPERCHOLESTEROLEMIA – FAMILIAL
HYPERCHOLESTEROLEMIA – PHENOTYPE
HYPERTENSION – SYSTEMIC ARTERIAL
INFLAMMATION
INFLUENZA
METABOLIC SYNDROME
OBESITY
PERIODONTITIS
PNEUMONIA – COMMUNITY-ACQUIRED
PSYCHOSOCIAL FACTORS [5]
RHEUMATOID ARTHRITIS
SEDENTARY LIFESTYLE
SITOSTEROLEMIA
SPONTANEOUS CORONARY ARTERY DISSECTION
SYSTEMIC SCLEROSIS
TOBACCO USE

Demography

Leading cause of death in North America and Europe; increasing incidence of STEMI/decreasing incidence NSTEMI in recent years
Increased incidence with aging
Males predominate [3]
Males present at younger age [38]
Incidence of AMI and related death dramatically decreased over past 30 years, believed in part due to improved CVD prevention and coronary reperfusion therapy

Pathophysiology

Myocardial necrosis due to prolonged ischemia, associated with adjacent viable but noncontractile/hypocontractile myocardium (myocardial stunning and hibernation)
Innate immunity, especially involving T cells, may play role in ischemia-reperfusion injury, myocardial remodeling, healing

Signs/Symptoms [6]

ABDOMEN – PAIN
BLOOD PRESSURE, ARTERIAL – INCREASED/ELEVATED
BREATHING – DIFF (DYSPNEA)
BREATHING – RAPID (TACHYPNEA)
CHEST – FRICTION RUB
CHEST – PAIN [20]
CHEST – PALPITATIONS
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)
COUGH

EARS, BILAT, EARLOBE CREASE, DIAGONAL
EXTREM, UPPER – PAIN
FACE, JAW – PAIN
FATIGUE
FEVER
HEART, LV, APEX – IMP, PRESYS
HEART, LV, APEX – MURMUR, SYS
HEART, LV, APEX, IMP – FORCEFUL/SUSTAINED
HEART, RATE – RAPID (TACHYCARDIA)
HEART, RATE – SLOW (BRADYCARDIA)
HEART, RHYTHM – IRREG
HEART, RSB, LOWER – MURMUR, SYS
HEART, S2, SPLIT – REVERSED (PARADOXICAL)
HEART, S3 LV
HEART, S4 LV
HEART, SOUNDS, INTENSITY – DECR
HICCUPS
HYPOTENSION (BLOOD PRESSURE – DECREASED/LOW)
JOINT, SHOULDER – PAIN
JOINT, WRIST – PAIN
MENTATION – CONFUSION
MENTATION – FEELING OF DOOM
MENTATION – WEAKNESS (MALAISE)
MOOD – ANXIOUS
MOOD – DEPRESSED
MOOD – RESTLESS/IRRITABLE/COMBATIVE
NAUSEA
NECK, ANT – PAIN
NECK, JVP – ELEV
SKIN, COLOR – BLUE (CYANOSIS)
SKIN, COLOR – PALE (PALLOR)
SKIN, TEMP – DECR
SWEATING–INCR(DIAPHORESIS/HYPERHIDROSIS)
THROAT – PAIN/TIGHTNESS
VOMITING (EMESIS)

Differentiation [39]

Acute Cardiac Sarcoidosis
Acute Pulmonary Embolism
Allergic Acute Coronary Syndrome Kounis Syndrome
Arrhythmogenic Right Ventricular Dysplasia/Cardiomyopathy
Aortic Dissection
Barium Poisoning
Benzene Poisoning
Brugada Syndrome
Cardiac Contusion
Cardiomyopathy – Takotsubo
Celiac Artery Spontaneous Dissection
Coronary Artery Spasm
Cyanide Poisoning
Dengue Fever(Myocarditis)
Dropped Left Shoulder Syndrome
Early Repolarization Syndromes
Hyperkalemia
Left Bundle Branch Block
LVH
Lyme Disease
Myocarditis
Other causes of chest pain
Pericarditis – Acute
Sodium Monofluoroacetate Poisoning
Spinal Epidural Abscess
Thallium Poisoning

Complications

AF
Atrial Infarction
Cardiac Tamponade
Cardiogenic Shock [47]
Cognitive Impairment

DVT
Dressler's Syndrome
Dysrhythmia – Atrial Premature Beats
Dysrhythmia – AV Dissociation
Dysrhythmia – Complete Heart Block
Dysrhythmia – First Degree Block
Dysrhythmia – Monomorphic Ventricular Tachycardia
Dysrhythmia – Second Degree Block, Type I
Dysrhythmia – Second Degree Block, Type Ii
Dysrhythmia – Sick Sinus Syndrome
Dysrhythmia – Sinus Bradycardia
Dysrhythmia – Sinus Tachycardia
Dysrhythmia – Ventricular Fibrillation
Dysrhythmia – Ventricular Parasystole
Dysrhythmia – Ventricular Premature Beats
Dysrhythmia – Ventricular Tachycardia
Hand-Shoulder Syndrome
HF [45] [48]
LBBB
Major Depression
Mitral Regurgitation
Myocardial free wall rupture
Papillary muscle rupture
Pericarditis
Pneumonia
Pulmonary Embolism
Renal Failure
RBBB
Right Ventricular Infarction [17]
SCD
Systemic Embolism (Cerebral, Renal, Extremity, Ocular, Intestinal, Splenic)
Unplanned rehospitalization [46]
Ventricular Aneurysm
Ventricular Pseudoaneurysm
VSD
Ventricular Septum Rupture

Laboratory

BLOOD, C-REACTIVE PROTEIN (CRP) – INCR
BLOOD, CHOLESTEROL, LDL (LDL-C) – INCR
BLOOD, CKMB – INCR [19]
BLOOD, COPEPTIN – INCR
BLOOD, ENZYMES, CARD – INCR [19]
BLOOD, ESR – INCR
BLOOD, GLUCOSE – INCR (HYPERGLYCEMIA) [37]
BLOOD, LDH – INCR
BLOOD, MYOGLOBIN – INCR
BLOOD, NT-PROBNP – INCR
BLOOD, ST2 – INCR
BLOOD, TRIGLYCERIDES – INCR
BLOOD, TROPONIN – INCR [19] [39]
BLOOD, WBC – INCR (LEUKOCYTOSIS)

ECG [11] [33] [34] [35]

AV COND – 1ST DEGREE BLOCK
AV COND – 2ND DEGREE BLOCK, MOBITZ II
AV COND – 3RD DEGREE BLOCK
DYSRHYTHMIAS – ATRIAL (PACS/OTHERS)
DYSRHYTHMIAS – VENTRICULAR (PVCS/OTHERS)
J POINT – DEPRESSED [49]
Q WAVE – ABN
QRS – LBBB/LBBB PATTERN [30]
QRS – LONG, NS
QRS – R PROGRESSION, POOR
QRS, AMP – DECR
RATE – DECREASED (SINUS BRADYCARDIA)
ST SEGMENT – DEPR [7) (STEMI) [21])] [22] [49]
ST SEGMENT – ELEV [8] [22]
T WAVE – INVER, ABN [7]
T WAVE – TALL/PEAKED [18] [49]

Imaging [10]

ART, CORONARY – CALCIUM
LV, MYOCARD – LGE [SUGGESTS SCAR]
LV, MYOCARD, PERFUSION – DECR
LV, WALL MOTION, SEG – DECR/AKINETIC [9]
LV, WALL MOTION, SEG – INCR/HYPERDYNAMIC [NONINFARCT AREAS]
PERICARD – FLUID [44]

Genomics [40]

There are at least 50 genetic risk varients associated with CAD, including:
9p21
CDKN2B [CHR 9p21/SNP rs4977574]
rs646776]
1p13
ANRIL
CDKN2A [CHR 9p21/SNP rs4977574]
rs4977574
SORT1 [CHR 1p13/SNP rs646776]

Other Tests

Continuous ECG monitoring
Coronary arteriography
Noninvasive testing for ischemia prior to discharge

Treatment: Nonpharmacologic [54]

EMS direct transport to PCI-capable hospital [12] [13]
Oxygen
Cardiac rehabilitation after discharge
Lifestyle and diet modification
Tobacco cessation

Treatment: Pharmacologic

Anticoagulants:

- Bivalirudin
- Fondaparinux (contraindicated as sole therapy due to risk of catheter thrombosis)
- Unfractionated heparin

Antiplatelets:

- ASA 162–325 mg before primary PCI and maintained post-PCI (81 mg)
- p2y12 inhibitor prior to or at time of PCI [14] [27]

Beta-blockers
CCBs
Fibrinolytics: only in STEMI (contraindicated in NSTEMI or unstable angina) [16] [25] [26]
Glucose-insulin-potassium infusion (controversial)
GPIIb/IIa inhibitor (IV) at time of PCI [15]
Morphine [23]
Nitrates
RAAS inhibitors

- ACEIs
- ARBs

Statins

Treatment: Surgical/Invasive

CABG [54]
Hypothermia protocol for VT/VF/cardiac arrest
ICD when EF <35 % after optimal medical therapy [42]
Mechanical circulatory support, eg, intra-aortic balloon pump or veno-arterial extracorporeal membrane oxygenation or tandem heart or LVAD
Reperfusion – fibrinolytic therapy
Reperfusion – primary PCI with intracoronary stenting: [55] [56]

NSTEMI [28]
STEMI [12]

Stem cell/regenerative therapy (investigational) [52]
Temporary pacing [53]

Prevention (Secondary)

ASA
Beta-blockers
RAAS inhibitors [50]

ACEIs
ARBs

Statins (high dose)
Predischarge initiation/education about atherosclerotic risk factor and lifestyle modification including: [46]

BP control
Diet
Glucose control
Physical activity
Tobacco cessation

Course

In-hospital mortality: 5–6 %
1 year mortality: 7–18 %
Independent predictors of death in STEMI include:

Age
Anterior infarct location
DM
Hypotension [47]
Killip class
Markedly elevated biomarkers

Prior AMI
Renal dysfunction
Tachycardia
Tobacco use
Unplanned rehospitalization [46]

Notes

[1] In USA 23 % of patients with AMI have DM; 75 % of deaths in patients with DM are related to CAD
[2] Due to increased myocardial O_2 demand
[3] 30 % of patients with STEMI are female, who have 2x risk of dying during hospitalization, less likely to receive reperfusion therapy, and about 50 % greater risk of subsequent death
[4] Contrary to evidence of benefit, patients with CKD/on dialysis less likely to receive guideline-recommended therapy
[5] Eg, work/home stress, financial stress, recent adverse life events, depression
[6] 1/3 of patients do not have chest pain and have atypical symptoms, especially women, elderly, critically ill, postop patients, diabetics
[7] $\geq$0.05 mv new horizontal or down-sloping in 2 contiguous leads and/or $\geq$0.1 mv T wave inversion in 2 contiguous leads with prominent R wave or R/S ratio >1
[8] New ST elevation at J point in 2 contiguous leads with cut-points: $\geq$0.1 mv in all leads other than V1-V2; in V1-V2 $\geq$0.2 mv in men $\geq$40 year, $\geq$0.25 mv in men <40 year, $\geq$0.15 mv in women
[9] Not diagnostic of infarction as other myocardial abnormalities, eg, prior infarction, myocardial fibrosis, can also cause segmental wall motion abnormalities
[10] In setting of possible AMI, optimal use is when ECG and biomarkers are nondiagnostic; echo, radionuclide ventriculography, myocardial perfusion scintigraphy

using single photon emission CT (SPECT) and MRI are commonly used for myocardial perfusion, wall motion and thickness, LV function, myocyte viability, and serially for changes in LV function and complications; also (including CT) used for helping differentiate AMI from other disease states, such as Acute Pulmonary Embolism and Dissecting Aortic Aneurysm; normal findings of wall motion and perfusion have high negative predictive value for AMI, but normal biomarkers, when appropriately measured, take precedence over imaging results

[11] Recommended at time of first contact by EMS when AMI is possible; correlates with rapid reperfusion times and improved outcomes

[12] Ideal first medical contact-to-device time of 90 min or less

[13] EMS transport recommended rather than friends/relatives because of frequent cardiac arrest during transport

[14] Clopidogrel 600 mg loading dose followed by 75 mg daily; or prasugrel 60 mg loading dose followed by 10 mg daily (greater benefit for diabetics; avoid in patients with prior TIA/stroke or >75 years old or weight < 60 kg); or ticagrelor 180 mg loading dose followed by 90 mg twice daily (superior to clopidogrel; avoid in patients with prior TIA/stroke or age >75 years or high dose ASA)

[15] Abciximab, high-bolus-dose tirofiban, double-bolus eptifibatide

[16] Prehospital fibrinolytic therapy may lower AMI mortality; more widely used in Europe and UK but lack of training in rural areas limits use in USA

[17] Occurs in 1/3 of inferior wall STEMIs; signs include elevated JVP, low BP, clear lungs, 1 mm ST elevation in leads V1 and V4r

[18] Hyperacute T waves may be (rarely) recorded in early phase AMI prior to ST elevation

[19] ESC/ACCF/AHA/WHO universal definition of myocardial infarction: detection of rise and/or fall of cardiac

biomarker values (preferably troponin) with at least one value above 99th percentile of upper reference limit and with at least one of following:

(1) Symptoms of ischemia
(2) New or presumably new significant ST-T changes or new LBBB
(3) Development of pathological Q waves on ECG
(4) Imaging evidence of new loss of viable myocardium, or new regional wall motion abnormality
(5) Identification of intracoronary thrombus by angiography or autopsy

[20] Lasts ≥20 min and does not respond to NTG
[21] ≥0.05 mv in V1-V3 suggests STEMI of inferior-posterior basilar LV (distribution of left circumflex coronary artery)
[22] Broad ST depression (2 or more surface leads) with ST elevation in AVR/V1 suggests left main coronary artery or multivessel obstruction
[23] For relief of pain, dyspnea, anxiety, which also increase sympathetic activation, causing vasoconstriction and increased cardiac workload
[24] Most common cause in pregnancy-related AMI
[25] Fibrinolytic therapy has better outcomes when patient presents very soon after onset of chest pain (within 12 h but most benefit within 3 h); patients should undergo rescue PCI if fibrinolytic therapy fails; if fibrinolytic therapy is successful, patient should have pharmacoinvasive strategy involving early cardiac catheterization but not within first 2–4 h after fibrinolysis
[26] Successful fibrinolysis defined as resolution of ST segment elevation by > 70 % and/or resolution of chest pain and/or accelerated idioventricular rhythm
[27] Avoid using P2Y12 inhibitors if triple vessel CAD requiring CABG or if emergent CABG is expected
[28] In NSTEMI, intermediate and high risk patients (using TIMI or grace scores) benefit from early invasive strategy within 12–24 h after hospitalization; very low risk

patients may be managed conservatively with medical therapy and non-invasive testing

[29] Often undiagnosed (up to 17 % of STEMI/NSTEMI patients)

[30] New onset LBBB sometimes considered a STEMI equivalent: often not possible to determine whether "new"; alone insufficient for diagnosis of AMI

[31] Transient ST elevation, ST depression, marked T wave inversion may occur but not required for diagnosis of NSTEMI

[32] In absence of elevated biomarkers in appropriate clinical context, patient may be diagnosed as having unstable angina

[33] Causes of false positive ECG findings, including Q waves:

Acute Pulmonary Embolism/Cor Pulmonale
Cardiomyopathy, including Amyloidosis, Takotsubo, DCM
Cholecystitis
Early repolarization
J point elevation syndromes, eg, Brugada
Left anterior hemiblock
LBBB
LVH
Lead transposition
Malposition of precordial ECG electrodes
Metabolic abnormalities, eg hyperkalemia
Myocarditis
Pericarditis/Myocarditis
Persistent juvenile pattern
Phenothiazines
Preexcitation, eg, Wolff-Parkinson-White syndrome
RBBB
RVH
Subarachnoid Hemorrhage
Tricyclic antidepressants

[34] Causes of false negative ECG findings:

LBBB
Prior AMI with Q waves/persistent ST elevation
RV pacing

[35] Silent myocardial infarction detected on ECG in 1/6 newly diagnosed DM patients in UK

[36] Characteristics of Type 2 AMI in unselected hospitalized patients include:

25 % of all AMIs
Most frequent mechanisms:

Anemia
Respiratory failure
Tachyarrhythmias

50 % in departments other than cardiology
50 % have no CAD

[37] Hyperglycemia at time of admission: common, including non-diabetics; strongly correlates with increased risk for adverse events in both diabetic and non-diabetic patients (underlying pathophysiology uncertain)

[38] However, women age <50 years are underrecognized and undertreated group for AMI and themselves less likely to seek care/recognize prodromal/acute AMI symptoms

[39] Other causes of increased cardiac troponin include:

Acute Pulmonary Embolism
Acute stroke
Adriamycin
Cardiac contusion
Cardiac surgery
Cardiac trauma
Connective tissue disease
Coronary artery embolism
Coronary artery trauma
Coronary artery spasm
Coronary vasculitis
End stage renal failure

Extreme endurance exercise
Fluorouracil
HF
Herceptin
Hypothyroidsm
Multiple organ failure
Myocarditis
Radiofrequency catheter ablation
Sepsis/septic shock
Snake envenomation
Supraventricular tachycardia
Sympathomimetics
Systemic Lupus Erythematosus

[40] Studies identifying tag snps in loci associated with these genes of interest in patients with AMI/CAD

[41] Patients with COPD/AMI more likely to have delayed diagnosis and suboptimal treatment, including angiography, reperfusion, and use of secondary prevention strategies

[42] Only 10 % of patients age >65 years who are candidates receive post-AMI ICD implants

[43] Long term follow-up (GRACE study):

1/4 pts readmitted for CV reasons
1/14 die by 2 years
1/16 develop HF
1/22 re-infarct

[44] Pericardial fluid post-AMI usually mild; moderate-large pericardial effusion found with CMR reported in >20 % and may be marker for poorer short-term prognosis

[45] Heart Failure: frequent complication of first AMI, both during acute phase and shortly after hospital discharge

[46] Unplanned rehospitalization: common after AMI; almost 1/3 observational; predischarge/postdischarge assessments of overall (not just CV) strategies to optimize patient functional status may help

[47] Cardiogenic shock post-AMI hospital survivors: increased risk of death/hospitalization in 1st year post-discharge; risk is time-dependent and clustered in early post-discharge period, after which prognosis is similar to patients without cardiogenic shock

[48] In 1 study among patients with severe systolic dysfunction following AMI with an EF <35 %, 57 % had EF recovery to >35 %

[49] Combination of J point/upsloping ST depression and symmetrical peaked T waves in precordial leads signifies high grade proximal LAD occlusion (deWinter ECG changes)

[50] Post-AMI treatment with ACEIs/ARBs associated with improved long-term survival, regardless of underlying renal function, with low rates of adverse renal events

[51] Chronic inflammatory disease significantly increases risk, including:

Psoriatic arthritis
Rheumatoid arthritis
Severe psoriasis

[52] No evidence for short term efficacy based on limited data

[53] Pacing for AMI: extracted/modified from 2013 ACCF/AHA Guideline for the Management of ST-Elevation Myocardial Infarction (J Am Coll Cardiol 2013;61:e78–e140)

Temporary pacing is indicated for symptomatic bradyarrhythmias unresponsive to medical treatment. (*Level of Evidence: C)*

Sinus bradycardia:

- Common early after STEMI, particularly with inferior location
- Mediated through increased vagal tone, usually self-limited, generally requires no treatment
- May be necessary to withhold beta blockers until bradycardia resolves

Symptomatic or hemodynamically important sinus bradycardia should be treated with atropine or temporary pacing if not responsive

AV block and intraventricular conduction delays:

Associated with extent of infarction
Incidence of abnormal conduction has decreased substantially in reperfusion
AV block of varying degree and persistent BBB develop in approximately 7 % and 5 % of patients with STEMI, respectively
High-grade (i.e., second- or third-degree) AV block and persistent BBB are independently associated with worse short- and long-term prognosis in both inferior/posterior and anterior/lateral MI but are more ominous in anterior/lateral MI because of a relatively greater extent of myocardial injury

First-degree AV block:

Does not require treatment
High-grade AV block with inferior/posterior STEMI usually transient and associated with a narrow complex/junctional escape rhythm that can be managed conservatively
Application of transcutaneous pacing pads for potential use is reasonable
Prophylactic placement of temporary pacing system recommended for high-grade AV block and/or new BB (especially LBBB) or bifascicular block in patients with anterior/lateral MI

[54] CABG for STEMI: extracted/modified from 2013 ACCF/AHA Guideline for the Management of ST-Elevation Myocardial Infarction (J Am Coll Cardiol 2013;61:e78-e140)
Recommendations

CLASS I

1. Urgent CABG is indicated in patients with STEMI and coronary anatomy not amenable to PCI who have ongoing or recurrent ischemia, cardiogenic shock, severe HF, or other high–risk features. (Level of Evidence: B)
2. CABG is recommended in patients with STEMI at time of operative repair of mechanical defects. (Level of Evidence: B)

CLASS IIa

1. The use of mechanical circulatory support is reasonable in patients with STEMI who are hemodynamically unstable and require urgent CABG. (Level of Evidence: C)

CLASS IIb

1. Emergency CABG within 6 hours of symptom onset may be considered in patients with STEMI who do not have cardiogenic shock and are not candidates for PCI or fibrinolytic therapy. (Level of Evidence: C)

[55] Reperfusion at a PCI-capable hospital: extracted/modified from 2013 ACCF/AHA Guideline for the Management of ST-Elevation Myocardial Infarction

CLASS I

1. Primary PCI should be performed in patients with STEMI and ischemic symptoms of less than 12 hours' duration. (Level of Evidence: A)
2. Primary PCI should be performed in patients with STEMI and ischemic symptoms of less than 12 hours' duration who have contraindications to fibrinolytic therapy, irrespective of the time delay from FMC. (Level of Evidence: B)
3. Primary PCI should be performed in patients with STEMI and cardiogenic shock or acute severe HF, irrespective of time delay from MI onset. (Level of Evidence: B)

CLASS IIa

1. Primary PCI is reasonable in patients with STEMI if there is clinical and/or ECG evidence of ongoing ischemia between 12 and 24 hours after symptom onset. (Level of Evidence: B)

CLASS III: HARM

1. PCI should not be performed in a noninfarct artery at the time of primary PCI in patients with STEMI who are hemodynamically stable. (Level of Evidence: B)

[56] Reperfusion at a non-PCI-capable hospital: extracted/modified from 2013 ACCF/AHA Guideline for the Management of ST-Elevation Myocardial Infarction

Fibrinolytic Therapy When There Is an Anticipated Delay to Performing Primary PCI Within 120 Minutes of FMC: Recommendations

CLASS I

1. In the absence of contraindications, fibrinolytic therapy should be given to patients with STEMI and onset of ischemic symptoms within the previous 12 hours when it is anticipated that primary PCI cannot be performed within 120 minutes of FMC. (Level of Evidence: A)

CLASS IIa

1. In the absence of contraindications and when PCI is not available, fibrinolytic therapy is reasonable for patients with STEMI if there is clinical and/or ECG evidence of ongoing ischemia within 12 to 24 hours of symptom onset and a large area of myocardium at risk or hemodynamic instability. (Level of Evidence: C)

CLASS III: HARM

1. Fibrinolytic therapy should not be administered to patients with ST depression except when a true pos-

terior (inferobasal) MI is suspected or when associated with ST elevation in lead aVR. (Level of Evidence: B)

[57] Listed treatments apply to Type I AMI; Type 2 treatment is to correct the underlying cause

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Updates and More

https://clinicalguidecvd.com/ami

Chapter 3
Acute Pulmonary Embolism (Venous Thromboembolism)

Management Keys

- Utilize services of Multidisciplinary Pulmonary Embolism Response Team when available [34]
- Adhere to current guidelines in evaluation to avoid unnecessary/costly tests
- Initially treat patients in shock/hypotensive (high risk) with IV anticoagulation (unfractionated heparin) and thrombolytic therapy
- Have high index of suspicion for this diagnosis in patients presenting with cardiac arrest, hypotension/shock, cardiac/respiratory complaints as Acute Pulmonary Embolism is missed in up to 70 % of hospitalized cases
- Perform surgical embolectomy in high risk patients with contraindication to thrombolysis or when thrombolytic treatment fails
- Rule out this diagnosis in patients with suspected Acute Myocardial Infarction who have nonobstructive lesions on coronary angiography
- Use established guidelines/pulmonary embolism rule out criteria [Appendix A] to avoid unnecessary tests
- Use blood D-dimer to rule out this diagnosis in patients with high clinical probability [12]

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 63
DOI 10.1007/978-1-4471-7293-2_3,

Use CT angiography for diagnosis as first line test in patients with high clinical probability and second line in patients with elevated D-dimer blood level
Continue anticoagulation for at least 3 months after first occurrence
Continue anticoagulation indefinitely in patients with unprovoked Acute Pulmonary Embolism recurrence

ICD-10 Code

I26.0

Alternate Names/Abbreviation

PE
Venous Thromboembolism (VTE)

Description/Etiology

Embolic obstruction of pulmonary artery, usually due to dislodgement of thrombus in distal vein
Acute form of VTE with clinical manifestations ranging from none to Cardiac Arrest/sudden death
<10 % of fatal cases diagnosed antemortem
Associated with non-O blood group [36]

Comorbid Conditions [5] [20] [24] [25]

ACUTE MYOCARDIAL INFARCTION (WITHIN LAST 3 MONTHS)
ARTHROSCOPIC KNEE SURG
ATRIAL FIBRILLATION
AUTOIMMUNE DISEASES (ESP SLE)
BLOOD TRANSFUSION
CANCER (ACTIVE/REMISSION) [15]
CHEMOTHERAPY [18]

CHRONIC OBSTRUCTIVE PULMONARY DISEASE (EMPHYSEMA)
DEEP VEIN THROMBOSIS [4]
DISEASE-MODIFYING BIOLOGIC DMARDS
ERYTHROPOIETIN-STIMULATING AGENTS
FAMILY HX: DEEP VENOUS THROMBOSIS
FAMILY HX: THIS CONDITION
FRACTURE – LOWER LIMB
HEART FAILURE
HIP REPLACEMENT SURG
HORMONE USE (M/F: OCS, ESTROGENS, OTHER)
HX: PULMONARY EMBOLUS
IMMOBILITY (ESP LIMB)
IN VITRO FERTILIZATION
INDWELLING VENOUS CATHETER
INFECTION [21]
INFLAMMATORY BOWEL DISEASE
KNEE REPLACEMENT SURG
MAJOR TRAUMA
OBESITY
ORAL CONTRACEPTIVE/ESTROGEN REPLACEMENT THERAPY
PAGET-SCHROETTER SYNDROME [37]
PARALYSIS
POSTPARTUM [31]
PREGNANCY [33]
PROTHROMBOTIC STATE
RECENT HOSPITALIZATION (ESP SURGERY/TRAUMA)
RESPIRATORY FAILURE
RHEUMATOID ARTHRITIS
SICKLE CELL DISEASE/TRAIT
SPINAL CORD INJURY
STROKE (ESP PARALYTIC)
SUPERFICIAL VEIN THROMBOSIS
SURGERY
THROMBOPHILIA
TOBACCO USE
TRAUMA

Demography

All ages [3]
Genders: about equal
All ethnicities
Venous thromboembolic disease is 3rd most common form of CVD in developed countries
Hospitalization rates increasing, especially among blacks

Pathophysiology

Clinically relevant hemodynamic changes occur with 30–50 % embolic occlusion of pulmonary arterial bed

Signs/Symptoms [24] [25]

ABDOMEN – PAIN
BLOOD PRESSURE, ARTERIAL – INCREASED/ELEVATED [10]
BREATH SOUNDS – CRACKLES (RALES)
BREATH SOUNDS – WHEEZES
BREATHING – DIFF (DYSPNEA) [7]
BREATHING – RAPID (TACHYPNEA)
CHEST – FRICTION RUB
CHEST – PAIN PLEURITIC/NON-PLEURITIC [6]
CHEST, LOCAL – PAIN, PLEURITIC
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)
COUGH
DIZZY/LIGHTHEADED/PRESYNCOPE
EXTREM, LOWER, UNILAT – PAIN [8]
EXTREM, LOWER, UNILAT – TENDERNESS [8]
EXTREM, LOWER, UNILAT, TEMP – INCR [8]
EXTREM, UNILAT – EDEMA [8]
FEVER [10 % OF CASES]
HEART, P2, INTENSITY – INCR
HEART, RATE – RAPID (TACHYCARDIA)

HYPOTENSION (BLOOD PRESSURE – DECREASED/LOW) [9]
MOOD – ANXIOUS
SEIZURES [30]
SKIN, COLOR – BLUE (CYANOSIS)
SPUTUM – BLOOD (HEMOPTYSIS)
SWEATING – INCR (DIAPHORESIS/ HYPERHIDROSIS)

Differentiation

Acute Myocardial Infarction
Aortic Dissection
Other causes of acute cyanosis
Other causes of acute dyspnea
Other causes of chest pain

Complications [29]

Acute right heart failure [32]
Cardiac Arrest
Dysrhythmias (most often atrial fibrillation)
HF
Hypoxemia aggravated by patent foramen ovale
Pulmonary Hypertension [11]
Pulmonary infarction
RV infarction
Shock
Sudden death

Laboratory [26] [Appendix A and B]

BLOOD, ARTERIAL PO2 – DECREASED (HYPOXIA)
BLOOD, D-DIMERS – INCR [12]
BLOOD, TROPONIN – INCR

ECG [22] [26]

AV COND – 1ST DEGREE BLOCK
DYSRHYTHMIAS – ATRIAL (PACS/OTHERS)
P WAVE – TALL/PEAKED
Q WAVE – ABN [1]
QRS – RVH pattern
QRS – RBBB/RBBB PATTERN
QRS, AXIS – L
QRS, AXIS – R
RATE – INCREASED (SINUS TACHYCARDIA)
ST SEGMENT – ELEV [19]
ST-T WAVE – ABN, NS [1]
T WAVE – INVER, ABN [2]

Imaging [13] [14] [17] [26] [Appendix A and B]

ART, PUL – OCCL [16]
CARDIOMEGALY
DIAPHRAGM, HEMI – ELEV
LUNGS – ATELECTASIS
LUNGS, PERIPH – OPACITY, WEDGE-SHAPED [27]
PA, MAIN, SIZE – INCR [23]
PA, R DESCEND – ENLARGED [28]
PLEURA – FLUID
PUL – EDEMA
PUL, VASCULARITY – DECR [23]

Other Tests [26]

V/Q scan

Treatment: Nonpharmacologic [26]

(Acute phase)
ECMO [35]

Mechanical ventilation (with caution)
O2
Volume expansion (modest, with caution)

Treatment: Pharmacologic [26] [34]

(Acute phase)
Anticoagulants
Parenteral
- LMWH
- Fondaparinux

Oral: Vitamin K antagonists
Oral factor Xa inhibitors
- Apixaban
- Dabigatran
- Edoxaban
- Rivaroxaban

Thrombolytics
Vasodilators
Vasopressors

(Post-acute/long term)
Antithrombotic options:

- Antiplatelet agents
  - ASA
  - Clopidogrel

Direct thrombin inhibitors
- Bivalrudin
- Dabigatran

Oral factor Xa inhibitors
- Apixaban
- Dabigatran
- Edoxaban
- Rivaroxaban

Warfarin

Treatment: Surgical/Invasive [26] [34]

(Acute phase)
Embolectomy – percutaneous
Embolectomy – surgical
IVC filter

Prevention [26]

Anticoagulation
ASA
IVC filter
Statins

Course [26]

(Among overall VTE episodes)
All-cause mortality:

30 days: 9–11 %
3 months: 9–18 %

Early recurrence (may be lower with use of newer anticoagulants):

2 weeks: 2 %
3 months: 6.4 %
6 months: 8 %

Late recurrence (after 6 months/usually after discontinuing anticoagulants):

1 years: 13 %
5 years: 23 %
10 years: 30 %

Chronic PAH: highly variable incidence (about 1.5 %) with most cases appearing within 2 years after acute event

Notes

[1] Usually III, AVF, sometimes anterior leads; may resemble Acute Inferior MI
[2] Right precordial leads; most common finding
[3] Mean age >60 years; risk doubles in each decade after age 40
[4] Although often silent, Deep Vein Thrombosis can be detected in 70 % of patients with Acute Pulmonary Embolism using sensitive techniques
[5] No identifiable predisposing factor in 20–30 %
[6] May be retrosternal and resemble Acute Myocardial Infarction
[7] May be sudden onset or rapidly progressive over days-weeks; in patients with HF, may be manifest as worsening dyspnea, and may be missed
[8] Signs of Deep Vein Thrombosis, most often detected in calf by physical exam
[9] Signifies large event
[10] Due to prior hypertension or anxiety
[11] Especially with recurrent pulmonary embolism; termed Chronic Thromboembolic Pulmonary Hypertension; due to nonresolving fibrothrombotic obstruction of large pulmonary arteries, combined with small-vessel arteriopathy in some patients; both proximal and small-vessel obstruction increases PVR, leading to progressive Pulmonary Hypertension, RHF, death
[12] 97 % sensitive but nonspecific, especially with advanced age
[13] Also: lower limb compression ultrasonography
[14] V/Q scan for excluding pulmonary embolism
[15] Especially metastatic forms and cancer of:

Blood (especially multiple myeloma, with 46x relative risk)
Brain (20x relative risk)
GI tract
Lung
Pancreas (16x relative risk)

[16] CT angiogram: imaging method of choice for Acute Pulmonary Embolism diagnosis, with 96 % specificity, 83 % sensitivity (PIOPED II trial)

[17] Doppler/echo: absence of RV dysfunction/overload excludes pulmonary embolism in patients with shock or hypotension

[18] Includes but not limited to bevacizumab, sorafenib, sunitinib, erlotinib, thalidomide, lenalidomide, tamoxifen, cisplatin, vorinostat

[19] ST segment elevation rare and suggests massive pulmonary embolism

[20] Additional (weak) risk factors:

Advanced age
Bed rest >3 days
DM
Immobility due to prolonged sitting (prolonged auto/air travel)
Laparoscopic surgery
Obesity
Systemic Arterial Hypertension
Varicose veins

[21] Especially pneumonia, urinary tract infection, HIV

[22] ECG is normal in up to 45 % of cases

[23] Westermark sign: combined prominent central pulmonary artery with decreased pulmonary vascularity

[24] Wells clinical prediction rules for PE (simplified)

(Item)	(Points)
Previous PE or DVT	1
Heart rate >110 bpm	1
Surg/immobilization in last month	1
Hemoptysis	1
Active cancer	1
Clin Sx of DVT	1
Alternative Dx less likely than PE	1

Clin probability:
PE unlikely 0-1
PE likely >1

[25] Geneva clinical prediction rules for PE (simplified)

(Item)	(Points)
Previous PE or DVT	1
Heart rate >110 bpm	1
Surg/immobilization in last month	1
Hemoptysis	1
Active cancer	1
Clin Sx of DVT	1
Alternative Dx less likely than PE	1

Clin probability:
PE unlikely 0-1
PE likely >1

[26] Management strategies/course for patients with confounding conditions, eg, pregnancy, cancer, may differ and specific protocols should be consulted for patients with these conditions

[27] CXR: Hamptons hump

[28] CXR: Pallas sign

[29] BOVA score predictors of complications in intermediate - risk patients with acute symptommatic PE

HR ≤110 bpm
Increased cardiac troponin
RV dysfunction
Systolic BP 90–100 mm Hg

[30] Believed due to transient global cerebral hypoperfusion/ hypoxia

[31] Low birth weight associated with 3x increased risk for thromboembolism

[32] Acute RHF with associated decreased cardiac output: leading cause of mortality

[33] Pulmonary embolism is leading cause of pregnancy-related mortality in developed countries, especially postpartum and after C-section

[34] Multidisciplinary Pulmonary Embolism Response Team comprises variety of specialties, including Vascular Medicine, Interventional Cardiology, Cardiothoracic Surgery, Pulmonology, Hematology, Echocardiography, and Radiology, for real-time patient evaluation/ enhanced clinical decision making/treatment

[35] ECMO may be indicated for respiratory/hemodynamic support in patients with massive pulmonary embolism and:

Have failed other therapies or
Other therapies contraindicated or
Too unstable to tolerate other interventions

[36] Non-O blood groups associated with >30 % of venous thromboembolic events, but clinical utility of this observation unresolved

[37] Axillary-subclavian vein thrombosis due to strenuous / repetitive UE activity

Appendix A

Pulmonary Embolism Ruleout Criteria

Age >49 years
HR >99
Pulse oximetry <95 % (room air)
History of DVT/PE requiring anticoagulation
History of surgery/trauma past 4 weeks requiring general anesthesia/hospitalization

Unilateral leg swelling
Hemoptysis in past week
Hormone use (oral contraceptives, estrogen, others in males/females)

Appendix B

American College Of Physicians Best Practices

(from ACP Guidelines, above)

Best Practice Advice 1: Clinicians should use validated clinical prediction rules to estimate pretest probability in patients in whom acute PE is being considered.

Best Practice Advice 2: Clinicians should not obtain D-dimer measurements or imaging studies in patients with a

low pretest probability of PE and who meet all Pulmonary Embolism Rule-Out Criteria.

Best Practice Advice 3: Clinicians should obtain a high-sensitivity D-dimer measurement as the initial diagnostic test in patients who have an intermediate pretest probability of PE or in patients with low pretest probability of PE who do not meet all Pulmonary Embolism Rule-Out Criteria. Clinicians should not use imaging studies as the initial test in patients who have a low or intermediate pretest probability of PE.

Best Practice Advice 4: Clinicians should use age-adjusted D-dimer thresholds (age × 10 ng/mL rather than a generic 500 ng/mL) in patients older than 50 years to determine whether imaging is warranted.

Best Practice Advice 5: Clinicians should not obtain any imaging studies in patients with a D-dimer level below the age-adjusted cutoff.

Best Practice Advice 6: Clinicians should obtain imaging with CTPA in patients with high pretest probability of PE. Clinicians should reserve V/Q scans for patients who have a contraindication to CTPA or if CTPA is not available. Clinicians should not obtain a D-dimer measurement in patients with high pretest probability of PE.

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ENGLISH

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Updates and More

https://clinicalguidecvd.com/ape

Chapter 4
Allergic Acute Coronary Syndrome (Kounis Syndrome)

Management Keys

- Consider this diagnosis in any patient with suspected/proven acute coronary syndrome and concurrent allergic manifestations, eg, pruritus, hives, wheezing
- Consider this diagnosis in any patient with prior intracoronary stent implantation and acute coronary syndrome and allergic manifestations
- Caution when using epinephrine, morphine, beta-blockers in patients with this syndrome

ICD-10 Code

NS

Alternate Names/Abbreviation

- Allergic angina
- Allergic Myocardial Infarction

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_4,

Description/Etiology

Acute coronary syndrome accompanying mast cell activation caused by allergic, hypersensitivity, or anaphylactoid reaction

May consist of transient ischemia without elevated enzymes or progress to Acute Myocardial Infarction

Variants:

- Type I: due to coronary artery vasospasm, with normal/near normal coronary arteries [1]
- Type II: culprit/quiescent preexisting atheromatous coronary disease
- Type III: coronary stent thrombosis in which eosinophils/mast cells demonstrated in aspirated thrombus

Possible drug causes include:

- Antibiotics
- Anticoagulants
- Analgesics
- Antineoplastics
- Contrast media
- Corticosteroids
- IV anesthetics
- NSAIDs
- Skin disinfectants
- Thrombolytics

Possible environmental causes include:

- Ant sting
- Bee sting
- Grass cutting
- Jellyfish sting
- Latex contact
- Limpet ingestion
- Millet allergy
- Scromboid fish poisoning
- Shellfish ingestion

Viper venom
Wasp sting

Comorbid Conditions

ANGIOEDEMA
ASTHMA
EXERCISE INDUCED ANAPHYLAXIS
FOOD ALLERGY
IDIOPATHIC ANAPHYLAXIS
INTRACORONARY STENT
SERUM SICKNESS
SYSTEMIC MASTOCYTOSIS
URTICARIA

Demography [4]

NS

Pathophysiology

Release of inflammatory mediators/cytokines via mast cell activation, causing coronary vasospasm/atherosclerotic plaque erosion or rupture

Mediators:

Chymase
Cytokines
Histamine [2]
Platelet-activating factor
Prostaglandins
Tryptase

Decreased cardiac output during allergic episode due to:

Decreased venous return
Systemic vasodilatation
Volume loss from increased vascular permeability [3]

Signs/Symptoms [5]

ABDOMEN – PAIN
BLOOD PRESSURE, ARTERIAL – INCREASED/ELEVATED
BREATH SOUNDS – WHEEZES
BREATHING – DIFF (DYSPNEA)
BREATHING – RAPID (TACHYPNEA)
CHEST – FRICTION RUB
CHEST – PAIN
CHEST – PALPITATIONS
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)
COUGH
DIZZY/LIGHTHEADED/PRESYNCOPE
EXTREM, UPPER – PAIN
FACE, JAW – PAIN
FATIGUE
FEVER
HEART, LV APEX – MURMUR, SYS
HEART, LV, APEX – IMP, PRESYS
HEART, RATE – RAPID (TACHYCARDIA)
HEART, RATE – SLOW (BRADYCARDIA)
HEART, RHYTHM – IRREG
HEART, RSB, LOWER – MURMUR, SYS
HEART, S2, SPLIT – REVERSED (PARADOXICAL)
HEART, S3 LV
HEART, S4 LV
HEART, SOUNDS, INTENSITY – DECR
HICCUPS
HYPOTENSION (BLOOD PRESSURE – DECREASED/LOW)
JOINT, SHOULDER – PAIN
JOINT, WRIST – PAIN
MENTATION – CONFUSION
MENTATION – FEELING OF DOOM
MENTATION – WEAKNESS (MALAISE)
MOOD – ANXIOUS
MOOD – DEPRESSED

MOOD – RESTLESS/IRRITABLE/COMBATIVE
NAUSEA
NECK, ANT – PAIN
NECK, JVP – ELEV
SKIN – HIVES
SKIN – ITCHING (PRURITUS)
SKIN, COLOR – BLUE (CYANOSIS)
SKIN, COLOR – PALE (PALLOR)
SKIN, TEMP – DECR
SWEATING–INCR(DIAPHORESIS/HYPERHIDROSIS)
VOMITING (EMESIS)

Differentiation

Atherosclerotic CAD
Cardiomyopathy – Takotsubo
Coronary Allograph Vasculopathy
Hypersensitivity myocarditis [8]

Complications [10]

Atrial Fibrillation
Acute Kidney Injury
Acute Pulmonary Embolism
Atrial Infarction
Atrioventricular Heart Block
Cardiac Arrest
Cardiogenic Shock
Cardiac Tamponade
Cognitive impairment
Coronary Artery Aneurysm (Post-Stent Thrombosis)
Deep Vein Thrombosis
Dressler Syndrome
Dysrhythmias – Atrial
Dysrhythmias – Ventricular
Hand-shoulder Syndrome

Heart Failure
Major Depression
Mitral Regurgitation – Acute
Myocardial free wall rupture
Papillary muscle rupture
Pericarditis – Acute
Pneumonia
Right ventricular Infarction
Systemic Embolism
  Cerebral
  Extremity
  Intestinal
  Ocular
  Renal
  Splenic
Ventricular Aneurysm
Ventricular Pseudoaneurysm
Ventricular Septal Defect
Ventricular Septal Rupture

Laboratory

BLOOD, C-REACTIVE PROTEIN (CRP) – INCR
BLOOD, CKMB – INCR
BLOOD, COPEPTIN – INCR
BLOOD, ENZYMES, CARD – INCR
BLOOD, EOSINOPHILES – INCR (EOSINOPHILIA)
BLOOD, ESR – INCR
BLOOD, GLUCOSE – INCR (HYPERGLYCEMIA)
BLOOD, MYOGLOBIN – INCR
BLOOD, NT-PROBNP – INCR
BLOOD, TRIGLYCERIDES – INCR
BLOOD, TROPONIN – INCR
BLOOD, WBC – INCR (LEUKOCYTOSIS)

ECG

AV COND – 1ST DEGREE BLOCK
AV COND – 2ND DEGREE BLOCK
AV COND – 3RD DEGREE BLOCK
DYSRHYTHMIAS – ATRIAL (PACS/OTHERS)
DYSRHYTHMIAS – VENTRICULAR (PVCS/OTHERS)
Q WAVE – ABN
QRS – LBBB/LBBB PATTERN
QRS – LONG, NS
QRS – R PROGRESSION, POOR
QRS, AMP – DECR
RATE – DECREASED (SINUS BRADYCARDIA)
RATE – INCREASED (SINUS TACHYCARDIA)
ST SEGMENT – DEPR
ST SEGMENT – ELEV
T WAVE – INVER, ABN
T WAVE – TALL/PEAKED

Imaging

LV, MYOCARD – LGE
LV, MYOCARD, PERFUSION – DECR
LV, WALL MOTION, SEG – DECR/AKINETIC
LV, WALL MOTION, SEG – INCR/HYPERDYNAMIC

Other Tests

Myocardial biopsy [8]

Treatment: Non-pharmacologic

NS

Treatment: Pharmacologic

Current guidelines do not address this syndrome specifically, but may apply on a case-by-case basis
Antihistamines
Corticosteroids
Caution using beta-blockers [7]
Caution using epinephrine [9]
Caution using morphine [6]

Treatment: Surgical/Invasive

Current Guidelines do not address this syndrome specifically, but may apply on a case-by-case basis
Caution in implanting drug eluting stents, especially in atopic patients

Prevention

Allergen avoidance/hyposensitization

Course

Variable per extent of myocardial damage

Notes

[1] May be variant of endothelial dysfunction/microvascular angina
[2] Cardiac mast cell-derived histamine:

Activates platelets
Constricts coronary arteries
Induces proinflammatory cytokine production from endothelial cells
Modulates activity of inflammatory cells

Potentiates aggregatory response of other agonists (e.g., adrenaline, 5- hydroxytryptamine, thrombin)
Sensitizes nerve endings close to adventitial mast cells in atherosclerotic coronary arteries
Upregulates P-selectin on endothelial cell surface

[3] Severe allergic reactions may cause up to 35 % decrease in circulating blood volume within 10 min due to transfer of fluid from intravascular to extravascular space
[4] True prevalence of this syndrome may be increasing, but adequate reporting mechanisms do not exist
[5] Signs/symptoms due to mix of allergic reaction/acute coronary syndrome
[6] Morphine stimulates histamine release from mast cells
[7] Beta-blockers may promote release of anaphylaxis mediators and have adverse effects on end organs
[8] Myocardial biopsy typically normal in this syndrome; when abnormal shows atypical lymphocytes, eosinophils, giant cells in hypersensitivity myocarditis
[9] Due to cardiac effects
[10] List comprises known complications of Acute Myocardial Infarction in general, but not specifically with Kounis syndrome, except atrioventricular heart block

Guidelines

NS

Professional Information

Review

Clin Ther. 2013;35:563–71. http://www.clinicaltherapeutics.com/article/S0149-2918(13)00078-7/abstract.

Review

Sou Med J. 2010;103:1148–55. http://sma.org/southern-medical-journal/article/kounis-syndrome/.

Bee Sting (Case Report)

J Am Coll Cardiol. 2015;65(10-S). doi:10.1016/S0735-1097(15)60579-4. http://content.onlinejacc.org/article.aspx?articleid=2198217.

Coronary Aneurysm Post-Intracoronary Stent

J Am Coll Cardiol Intv. 2014;7:e173–6. doi:10.1016/j.jcin.2014.05.031. http://interventions.onlinejacc.org/article.aspx?articleid=1936117.

Drug Eluting Stent/Hypersensitivity

J Am Coll Cardiol. 2006;47:175–81. http://www.sciencedirect.com/science/article/pii/S0735109705027191.

Drug Eluting Stent/Thrombosis

J Am Coll Cardiol Intv. 2009;2:583–93. http://interventions.onlinejacc.org/article.aspx?articleid=1110900#topLocation.

Fish Consumption/Histamine-Induced Coronary Spasm (Editorial)

Int J Cardiol. 2015;193:39–41. http://www.sciencedirect.com/science/article/pii/S0167527315010578.

Heart Block

J Pharmacol Pharmacother. 2013;4:161–2. http://www.ncbi.nlm.nih.gov/pmc/articles/PMC3669582/.

Scromboid Fish Poisoning

Australas Med J. 2015;8:96–9. http://amj.net.au/index.php?journal=AMJ&page=article&op=viewFile&path%5B%5D=2310&path%5B%5D=1305.

Updates and More

https://clinicalguidecvd.com/aacs

Chapter 5
Aneurysms-Osteoarthritis Syndrome

Management Keys

- Patient management by multidisciplinary team including clinical geneticists, cardiologists, orthopedic surgeons, radiologists, neurologists, vascular/cardiothoracic surgeons
- Consider this diagnosis in all patients with thoracic aortic aneurysms/dissections without molecular diagnosis or known cause and test these patients for SMAD3 mutations
- Consider early elective repair of dilated ascending aorta in patients with this diagnosis

ICD-10 Code

NS

Alternate Names/Abbreviation

AOS

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_5,

Description/Etiology

Autosomal-dominant connective tissue disorder with multi-arterial aneurysms, often first manifest as early-onset osteoarthritis

Often associated with:

Craniofacial abnormalities
Cutaneous abnormalities
LVH [3]

Etiology: heterozygous mutations of SMAD3 gene

Comorbid Conditions

AORTIC VALVE STENOSIS [BICUSPID AORTIC VALVE]
ATRIAL FIBRILLATION [2]
ATRIAL SEPTAL DEFECT – SECUNDUM
MITRAL REGURGITATION – CHRONIC [1]
MITRAL REGURGITATION – ACUTE [1]
MITRAL VALVE PROLAPSE [1]
PATENT DUCTUS ARTERIOSUS

Demography

Clinical onset age about 40 years (range 25–50 years)

Pathophysiology

Aneurysms and tortuosity of arteries, especially:Aortic root (most common)

Abdominal aorta
Celiac
Cerebral

Descending thoracic aorta
Hepatic
Iliac (common, external and internal iliac)
Pulmonary trunk
Splenic
Superior mesenteric

Cardiac structural abnormalities common, especially involvement of MV

Signs/Symptoms

ABDOMEN – BRUIT [4]
ABDOMEN – MASS, PULSATING [4]
ABDOMEN, UMBILICUS – HERNIA
ARTERY, CAROTID – BRUIT [4]
ARTERY, FEMORAL – BRUIT [4]
ARTERY, ILIAC – BRUIT [4]
BACK, CURV – LAT (SCOLIOSIS)
EXTREM, ANKLES – PAIN [5]
EXTREM, ELBOW – PAIN [5]
EXTREM, FEET – FLAT (PES PLANUS)
EXTREM, FINGERS – LONG (ARACHNODACTYLY)
EXTREM, HIP – PAIN [5]
EXTREM, KNEE – PAIN [5]
EXTREM, WRIST – PAIN [5]
EYES – XANTHELASMAS [10]
EYES. SEPARATION – WIDE (HYPERTELORISM)
FACE – LONG
FACE, CHEEKS – HYPOPLASTIC
FACE, SUPRAORBITAL RIDGES – FLAT
FATIGUE [6]
FLANK – BRUIT [4]
GROIN – HERNIA, INGUINAL
HEADACHE [7]
HEART, LV, APEX – CLICK(S), SYS [8]
HEART, LV, APEX – MURMUR, SYS [8]
JOINTS – LAX

JOINTS – PAIN (ARTHRALGIA)
MOUTH, PALATE – HIGH/ARCHED
MOUTH, UVULA – BIFID [9]
MOUTH, UVULA – BROAD [9]
PELVIS – PROLAPSE [11]
SKIN – ATROPHY
SKIN – BRUISING, EASY
SKIN – FRIABLE
SKIN – SCARS
SKIN – STRIAE
SKIN – THIN [12]
SKIN – VARICES
STERNUM, CURV – ANT (PECTUS CARINATUM)
STERNUM, CURV – POST (PECTUS EXCAVATUM)

Differentiation

Ehlers-Danlos Syndrome
Loeys-Dietz Syndrome
Marfan Syndrome

Complications

Aortic Dissection [13]
Aortic rupture
Intervertebral Disc Degeneration
Meniscus lesions
Osteochondritis Dissecans
Spondylosis/Spondylolisthesis
Sudden death

Laboratory

NS

ECG

DYSRHYTHMIAS – ATRIAL (PACS/OTHERS) [13]
QRS – LVH PATTERN

Imaging

AORTA, ABD, SIZE – INCR
AORTA, ARCH, SIZE – INCR
AORTA, ASCEND, SIZE – INCR
AORTA, DESCEND, SIZE – INCR
ART – TORTUOUS
ART, BASILAR – ANEURYSM
ART, CAROTID, EXT – ANEURYSM
ART, ILIAC, COMMON – ANEURYSM
ART, MESENTERIC – ANEURYSM
ART, SPLENIC – ANEURYSM
BACK, VERTEBRAE, DISCS – DEGEN
JOINTS – OSTEOCHONDRITIS
LV, MYOCARD, WALL THICKNESS – INCR (HYPERTROPHY)
MV, FLOW – REGURG
MV, LEAFLETS – PROLAPSE

Genomics

SMAD3

Other Tests

NS

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic

Strict hypertension control

Treatment: Surgical/Invasive

Early intervention for aortic dilatation

Prevention

Hypertension control

Notes

[1] MV abnormalities reported in about 50 % of affected persons
[2] AF reported in about 20 % of affected persons
[3] LVH common in absence of Systemic Arterial Hypertension or Aortic Stenosis
[4] Multiple aneurysm sites may occur
[5] Due to osteoarthritis, which may occur at early age
[6] Reported in 40 %; chronic/intermittent
[7] Migraine/severe headache reported in 50 %
[8] MV abnormalities ranging from mild MVP to severe Mitral Regurgitation common and may appear in second decade
[9] Uvula abnormalities occur in >50 %
[10] Occur in absence of dyslipidemia
[11] May include uterus, bladder
[12] Described as “velvety”
[13] Both Stanford types A and B reported

Guidelines

2014 ESC Guidelines on the diagnosis and treatment of aortic diseases
Eur Heart J. 2014;35:2873–926. http://eurheartj.oxfordjournals.org/content/35/41/2873.

Patient Information

NIH
http://www.nhlbi.nih.gov/health/health-topics/topics/arm/treatment.html.

Medlineplus

ENGLISH. http://www.nlm.nih.gov/medlineplus/aneurysms.html.
ESPANOL. http://www.nlm.nih.gov/medlineplus/spanish/aneurysms.html.

Genetics Home REF

http://ghr.nlm.nih.gov/condition/loeys-dietz-syndrome.

Professional Information

Review

J Am Coll Cardiol. 2012;60:397–403. http://content.onlinejacc.org/article.aspx?articleid=1206858.

Genetic Testing

J Am Coll Cardiol. 2012;60:404–7. http://content.onlinejacc.org/article.aspx?articleid=1206857#bib19.

Phenotypic Spectrum

J Med Genet. 2012;49:47–57. http://jmg.bmj.com/content/49/1/47.full.

SMAD3 Mutations

Nat Genet. 2011;43:121–6. http://www.nature.com/ng/journal/v43/n2/full/ng.744.html.

Updates and More

https://clinicalguidecvd.com/aos

Chapter 6
Aortic Dissection

Management Keys

- Prompt recognition of acute aortic dissection, which may present with atypical symptoms and findings [16]
- Knowledge of risk factors associated with development of thoracic aortic dissection
- Recognize clinical presentation clues to avoid missed diagnoses, including:
  - High-risk medical/family history
  - Sudden onset severe chest, back, abdominal pain
  - Syncope/acute neurological complaints
- Treat as surgical emergency

ICD-10 Code

I71.00

Alternate Names/Abbreviation

AOD

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 105
DOI 10.1007/978-1-4471-7293-2_6,

Description/Etiology

Disruption of media layer of aorta with bleeding within and along aortic wall
May occur with or without associated aneurysm
The term "dissecting aortic aneurysm" should be used only when dissection occurs in an aneurysmal aorta
Debakey classification (according to origin):

- Type 1: origin in ascending aorta with extension into aortic arch
- Type 2: origin in ascending aorta without extension into aortic arch
- Type 3: origin in descending aorta with distal and (rarely) proximal extension

Stanford classification (according to involvement of ascending aorta):

- Type A: involves ascending aorta
- Type B: does not involve ascending aorta

Component of closely-related acute aortic syndrome:

- Aortic dissection
- Intramural hematoma
- Penetrating atheromatous ulcer

Syndromic forms associated with genetic mutations [17] [18][19]

Comorbid Conditions

ANNULOAORTIC ECTASIA
AORTIC ROOT DILATATION
ATHEROSCLEROSIS IN OTHER CV AREAS [3]
AUTOIMMUNE/CONNECTIVE TISSUE DISEASE [1]
BICUSPID AORTIC VALVE
CABG SURGERY
CARDIAC INVASIVE PROCEDURES
CARDIAC SURGERY [ESP CABG]

CEREBRAL HEMORRHAGE [17]
CLEFT PALATE [17] [18] [19]
CLUBFOOT [17] [18] [19]
COARCTATION OF AORTA
CRANIOSYNOSTOSIS [17]
DUAL ECTASIA [17][18] [19]
EHLERS-DANLOS SYNDROME
EXOTROPIA [17] [18] [19]
EXTREME PHYSICAL ACTIVITY [2]
GIANT CELL ARTERITIS
HERNIA [17] [18] [19]
HYPERTELORISM [17] [18] [19]
HYPERTENSION – SYSTEMIC ARTERIAL [3]
LOEYS-DIETZ SYNDROME
MARFAN SYNDROME [10–25 % OF PTS WITH TYPE A; 5–15 % WITH TYPE B]
OSTEOARTHRITIS [17] [19]
PHEOCHROMOCYTOMA
PNEUMOTHORAX
PROSTHETIC AORTIC VALVE
RETROGNATHIA SURGERY [17] [18] [19]
SCOLIOSIS [17][18] [19]
SHPRINTZEN – GOLDBERG SYNDROME
SPINE, CERVICAL – UNSTABLE [17] [19]
SPONTANEOUS PNEUMOTHORAX [17] [18]
SUBSTANCE ABUSE: [1]

- AMPHETAMINES
- COCAINE

SYSTEMIC LUPUS ERYTHEMATOSUS
TAKAYASU ARTERITIS
TERTIARY SYPHILIS VASCULITIS
UVULA, BIFID [17] [18] [19]

Demography

Males 2:1
Average age: 65 years

Pathophysiology

Usual acute sequence:

- Aortic intima tear
- Formation of pressurized hematoma within media
- Rapid blood propagation along length of aorta (in false lumen)
- Compromise of branch vessels/aortic valve function/aortic regurgitation
- Rupture (because of weak outer wall of false lumen) common into:
  - Pericardial space
  - Pleural space
  - Mediastinum

Signs/Symptoms [7]

- ABDOMEN – PAIN [4]
- ARTERIAL PRESSURE, UE, SYS – R > L
- ARTERIAL PULSE – DEFICIT [30 % OF TYPE A/15–25 % OF TYPE B
- ARTERIAL PULSE PRESSURE – INCR
- ARTERIAL PULSE, BRACHIAL – DECR/ABSENT [6]
- ARTERIAL PULSE, CAROTID – DECR/ABSENT [6]
- ARTERIAL PULSE, FEMORAL – DECR/ABSENT [6]
- BACK – PAIN [4]
- BLOOD PRESSURE, ARTERIAL – INCREASED/ELEVATED
- BREATH SOUNDS – WHEEZES
- BREATHING – DIFF (DYSPNEA)
- CHEST – PAIN [4]
- CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)
- DIZZY/LIGHTHEADED/PRESYNCOPE
- EARS, UNILAT – PAIN

EXTREM, LOWER, BILAT – PARALYSIS (PARAPLEGIA)
FACE, JAW – PAIN
FEVER
FOCAL NEUROL DEFICITS [8]
HEART, LSB, MID – MURMUR, DIAS [5]
HEART, LV, APEX – MURMUR, DIAS [5]
HEART, RSB, MID – MURMUR, DIAS
HEART, RSB, UPPER – MURMUR, DIAS [5]
HYPOTENSION (BLOOD PRESSURE – DECREASED/LOW)
JOINT, STERNOCLAVICULAR – PAIN
JOINT, STERNOCLAVICULAR – PULSATION
MOOD – ANXIOUS
MOOD – RESTLESS/IRRITABLE/COMBATIVE
NAUSEA
NECK, ANT – PAIN
SPUTUM – BLOOD (HEMOPTYSIS)
SWALLOWING – DIFFICULT (DYSPHAGIA)
SWEATING – INCR (DIAPHORESIS/HYPERHIDROSIS)
SYNDROME – HORNER, IPSILATERAL [11]
SYNDROME – SUP VENA CAVAL
TEETH – PAIN
THROAT – PAIN/TIGHTNESS
VOICE – HOARSE (LARYNGITIS) [10]
VOMITING – BLOOD (HEMATEMESIS)

Differentiation

Acute Pulmonary Embolism
AMI
Cholecystitis
Costochondritis
Esophageal spasm
Fibromyalgia
Mediastinal tumor
Mesenteric ischemia

Other causes of Acute Aortic Regurgitation
Other causes of back pain
Other causes of chest pain
Pancreatitis
Peptic ulcer dis
Pleuritis
Pneumothorax
Spinal Epidural Abscess
Thymic tumor

Complications

Acute Aortic Regurgitation
Acute Myocardial Infarction
Cardiac Tamponade
HF
Hemorrhage into lung tissue/pleural space/pericardial space
Shock
Spinal cord injury [9]
Stroke
Sudden death
Tracheal compression

Laboratory

BLOOD, D-DIMERS – INCR
BLOOD, TROPONIN – INCR [ACUTE]

ECG [21]

Q WAVE – ABN [12]
QRS – LVH PATTERN
ST SEGMENT – ELEV

Imaging [13][14]

AORTA, ARCH, SIZE – INCR

AORTA, ASCEND, CONTOUR – ABN
AORTA, ASCEND, SIZE – INCR
AORTA, INTIMA – FLAP
AV, FLOW – REGURG
MEDIASTINUM – WIDE
PERICARD – FLUID
PLEURA – FLUID
TRACHEA/NASOGASTRIC, POSITION – DEVIATION, R

Genomics

COL3A1
FBN1
MYH11
TGFB3
TGFBR1
TGFBR2
ACTA2 [20]

Other Tests

Aortography

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic [15]

Analgesia
Antihypertensives

Treatment: Surgical/Invasive [15]

Endovascular repair

Surgical repair
See Guidelines for specific surgical recommendations for:

- Pregnancy
- Takayusu Arteritis

Prevention

General medical and risk factor management including:

- Blood pressure
- Dyslipidemia
- Tobacco use

Family screening

Course

Overall mortality

- Type A: 24.4 %
- Type B: 10.7 %

Notes

[1] Especially age <40 years
[2] E.g., weight lifting
[3] Most common factor after age 40 years; overall 77 % have history of hypertension, which is slightly more common in Type A
[4] Acute, severe
[5] Aortic regurgitation
[6] Pulse deficit: 20 mmHg difference compared to opposite side; presence depends on whether dissection involves artery

[7] Clinical features of cardiac tamponade may also be present with rupture into pericardial space, such as paradoxical pulse, elevated JVP
[8] Involvement of cerebrospinal arteries
[9] E.g., sudden LE paralysis
[10] Impingement on recurrent laryngeal nerve
[11] Impingement on sympathetic chain
[12] Especially inferior leads due to proximal dissection involving RCA
[13] CXR: normal in 29 % of type A/36 % of Type B
[14] TEE, CT, MRI comparable for diagnosis
[15] Refer to guidelines
[16] Dissection suspected at initial presentation in <50 % of patients with this final diagnosis; clinical features associated with delayed diagnosis include:

Female gender
Absence of abrupt or significant chest/back pain
Presence of fever

[17] Associated with TGFBR1, TGFBR2, SMAD3 mutations
[18] Associated with TGFB2 mutations
[19] Associated with TGFB3 mutations
[20] Associated with with 75 % lifetime risk for aortic event
[21] ECG: Normal in 36 % of Type A/38 % of Type B

Guidelines

2010 ACCF/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/STS/SVM Guidelines for the diagnosis and management of patients with thoracic aortic disease
Circulation. 2010:121;1544–79. http://circ.ahajournals.org/content/121/13/e266.full.pdf.
2014 ESC Guidelines on the diagnosis and treatment of aortic diseases
Eur Heart J. doi:10.1093/eurheartj/ehu281. https://www.nvvc.nl/media/richtlijn/187/2014_ESC_Guidelines_on_Diagnosis_and_Treatment_of_Aortic_Disease.pdf.

Patient Information

AHA: Activity Post-dissection
Circulation. 2014;130:e140–2. http://circ.ahajournals.org/content/130/16/e140.full.

Images

http://www.nlm.nih.gov/medlineplus/ency/imagepages/1149.htm.
http://www.nlm.nih.gov/medlineplus/ency/imagepages/18072.htm.
http://www.nlm.nih.gov/medlineplus/ency/imagepages/18073.htm.

Medline Plus

ENGLISH

http://www.nlm.nih.gov/medlineplus/ency/article/000181.htm.

ESPANOL

http://www.nlm.nih.gov/medlineplus/spanish/ency/article/000181.htm.

Genetics Home Reference

http://ghr.nlm.nih.gov/condition/familial-thoracic-aortic-aneurysm-and-dissection.

Cleveland Clinic

http://my.clevelandclinic.org/heart/disorders/aortic-dissection-video-transcript.aspx.

Merck

http://www.merckmanuals.com/home/heart-and-blood-vessel-disorders/aneurysms-and-aortic-dissection/aortic-dissection.

Professional Information

Review

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Review

Lancet. 2008;372:55–64. http://www.sciencedirect.com/science/article/pii/S0140673608609940.

Review: Endovascular Repair

Heart.2015;101:586–91.http://heart.bmj.com/content/101/8/586?etoc.

Review: Management

Heart. 2014;100:1491–7. http://heart.bmj.com/content/100/19/1491.abstract.

Review: Management in Marfan/Other Cong Dis

Heart. 2014;100:1571–6. http://heart.bmj.com/content/100/20/1571.abstract.

Acute Aortic Syndrome

Circulation. 2013;128:1122–7. http://circ.ahajournals.org/content/128/10/1122.full.

Acute Aortic Syndrome

Circulation. 2005;112:3802–13. http://circ.ahajournals.org/content/112/24/3802.full.

Acute Aortic Syndrome

Radiographics. 2003;23:S93–S110. http://pubs.rsna.org/doi/full/10.1148/rg.23si035507.

Acute Myocardial Infarction Presentation

Circulation. 2015;131:e503–4. http://circ.ahajournals.org/content/131/21/e503.extract.

African-Americans

Am J Med. 2013;126:909–15. http://www.sciencedirect.com/science/article/pii/S0002934313004919.

Aortic DIL/Bicuspid AV

N Engl J Med. 2014;370:1920–9. http://www.nejm.org/doi/full/10.1056/NEJMra1207059.

Bicuspid AV and Type A Dissection: PT Characteristics

Heart. 2013;99:1668–74. http://heart.bmj.com/content/99/22/1668.abstract.

Chest Pain: Differential Diagnosis

Emerg Med Clin North Am. 2009;27:685–712. http://www.ncbi.nlm.nih.gov/pubmed/19932401.

Cocaine

Am J Emerg Med. 1997;15:507–9. http://www.sciencedirect.com/science/article/pii/S0735675797901960.

Cocaine

J Vasc Surg. 2007;46:427–33. http://www.sciencedirect.com/science/article/pii/S074152140700969X.

Cocaine

Am J Med. 2014;127:878–85. http://www.sciencedirect.com/science/article/pii/S000293431400391X.

Diagnosis/Treatment

Heart. 2007;93:266–70. http://heart.bmj.com/content/93/2/266.full.

Delayed Diagnosis/Treatment

Circulation. 2011;124:1911–8. http://circ.ahajournals.org/content/124/18/1911.full.pdf+html.

Delayed Diagnosis: Risk Factors

Am J Cardiol. 2008;102:1399–406. http://www.ajconline.org/article/S0002-9149(08)01216-2/fulltext.

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Endovascular Repair: Ascending Dissection

J Am Coll Cardiol. 2013;61:1917–24. http://content.onlinejacc.org/article.aspx?articleID=1387692.

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Circulation. 2014;130:2287–94. http://circ.ahajournals.org/content/130/25/2287.full.

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Heart. 2013;99:1030–3. http://heart.bmj.com/content/99/14/1030.abstract.

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Circ Cardiovasc Genet. 2015;8:457–64. http://circgenetics.ahajournals.org/content/8/3/457.abstract?.

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Geometrical Change

J Am Coll Cardiol. 2014;63:1311–19. http://content.onlinejacc.org/article.aspx?articleID=1828677.

Imaging: Acute Aortic Syndrome

Radiol Assistant. 2006. http://www.radiologyassistant.nl/en/p441baa8530e86/thoracic-aorta-the-acute-aortic-syndrome.html.

International Registry (IRAD)

JAMA. 2000;283:897–903. http://jama.jamanetwork.com/article.aspx?articleid=192401.

Loeys-Dietz Syndrome

Heart.2016;102:626–32.http://heart.bmj.com/content/102/8/626?etoc.

Marfan Syndrome: Surg

Circulation. 2013;127:1569–75. http://circ.ahajournals.org/content/127/15/1569.full?sid=da46d891-b599-42a8-989d-3f61d1686fd2.

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Circulation. 2014;129:1381–86. http://circ.ahajournals.org/content/129/13/1381.full.

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Outcomes

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Chest. 1987;91:71–4. http://www.sciencedirect.com/science/article/pii/S0012369215428661.

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JAMA Cardiol. 2016. doi:10.1001/jamacardio.2015.0338. http://cardiology.jamanetwork.com/article.aspx?articleID=2511292.

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J Am Coll Cardiol. 2015;65:1324–36. http://content.onlinejacc.org/article.aspx?articleID=2211962.

Trends: Presentation, DX, Outcomes

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Troponin

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Updates and More

https://clinicalguidecvd.com/aod

Chapter 7
Aortic Regurgitation: Acute

Management Keys

Diagnose/treat as surgical emergency when caused by IE or aortic dissection, especially if there is hypotension, pulmonary edema, or evidence of low flow
Intra-aortic balloon counterpulsation is contraindicated
Use beta-blockers with caution if at all [17]

ICD-10 Code

I35.1

Alternate Names/Abbreviation

Acute AR

Description/Etiology

Sudden mechanical disruption of AV diastolic integrity due to variety of causes including:

Aortic Prosthetic Valve Dysfunction
Aortic Dissection (Type A)

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 123
DOI 10.1007/978-1-4471-7293-2_7,

Blunt chest trauma
Iatrogenic injury
Infective Endocarditis
Noninfective Endocarditis
Ruptured fenestration

Comorbid Conditions

AORTIC DISSECTION [TYPE A] [1]
CHEST TRAUMA
ENDOCARDITIS
PROSTHETIC AORTIC VALVE

Demography

Variable per etiology

Pathophysiology

Increased LV filling and end-diastolic pressure due to delivery of large blood volume (increased preload) to relatively non-compliant left ventricle

Signs/Symptoms [2]

ARTERIAL PULSE PRESSURE – DECR
BREATHING – DIFF (DYSPNEA)
BREATHING – RAPID (TACHYPNEA)
CHEST – PAIN [3]
CONSCIOUSNESS – ALTERED
DIZZY/LIGHTHEADED/PRESYNCOPE
EXTREM, HANDS/FEET, COLOR – BLUE (ACROCYANOSIS) [16]
FEVER [5]

HEART, A2, INTENSITY – DECR/ABSENT [7]
HEART, LSB, MID – MURMUR, DIAS [4]
HEART, LV, APEX – MURMUR, DIAS [4]
HEART, P2, INTENSITY – INCR
HEART, RATE – RAPID (TACHYCARDIA)
HEART, RSB, UPPER – MURMUR, DIAS [4]
HEART, S1, INTENSITY – DECR/ABSENT [6]
HEART, S3 LV
HYPOTENSION (BLOOD PRESSURE – DECREASED/LOW)
MENTATION – WEAKNESS (MALAISE)
SKIN, TEMP – DECR [16]

Differentiation

Other causes of acute hypotension/HF

Complications

AMI
Circulatory collapse
Dysrhythmias
Electromechanical Dissociation
HF
Pulmonary Edema

Laboratory

NS

ECG

RATE – INCREASED (SINUS TACHYCARDIA)
ST SEGMENT – DEPR [8]
ST-T WAVE – ABN, NS

Imaging [12]

AORTA, ABD, FLOW – REVERSED
AORTA, DESCEND, FLOW – REV
AR DIASTOLIC – TIME < 300 MS [11]
AV/ANNULUS MORPHOLOGY [14]
ECHO/DOPPLER [10]
LV SIZE/FUNCTION N
MEDIASTINUM – WIDE [9]
MV DECELERATION TIME < 150 MS [11]
MV, CLOSURE – PREMATURE
MV, FLOW – REVERSED
PUL – EDEMA
VENA CONTRACTA >6 MM

Other Tests

Cardiac Catheterization [13]

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic [17]

Antibiotics for IE [15]
Inotropes
Nitroprusside

Treatment: Surgical/Invasive

AV/aorta replacement/repair

Course

Variable per etiology

Notes

[1] Caused by Marfan Disease, bicuspid AV, atherosclerosis
[2] In addition to signs/symptoms due to underlying cause, e.g., Aortic Dissection, Infective Endocarditis; auscultatory findings are reduced or absent; marked differences from chronic AR due largely to normal LV size with consequent increased LVEDP, decreased cardiac output, and decreased systolic BP
[3] Signifies possible Aortic Dissection, especially if severe; may also be due to acute coronary insufficiency
[4] Aortic regurgitation murmur: soft/early as opposed to holosystolic decrescendo murmur of Chronic Aortic Regurgitation
[5] Signifies possible Infective Endocarditis
[6] Due to premature MV closure
[7] Due to failure of AV leaflet coaptation
[8] Myocardial ischemia
[9] Aortic Dissection
[10] May underestimate regurgitation severity
[11] Indicators of rapid equilibration of aortic pressure and LVEDP
[12] TEE findings diagnostic but CT/MRI preferred when attained faster than echo, as acute AR is surgical emergency
[13] Rarely required
[14] In Infective Endocarditis, intraoperative TEE exam of annulus important to determine if intact and for presence of abscess
[15] For hemodynamically stable and mild AR secondary to Infective Endocarditis, only antibiotics may be required
[16] Peripheral vasoconstriction

[17] Beta-blockers used only with caution if at all, because they block compensatory tachycardia and can precipitate marked fall in BP

Guidelines

2014 AHA/ACC guideline for the management of patients with valvular heart disease
J Am Coll Cardiol. 2014;63:e57–e185. http://content.onlinejacc.org/article.aspx?articleid=1838843&resultClick=3.

Guidelines on the management of valvular heart disease (version 2012)
Eur Heart J. 2012;33:2451–96. http://eurheartj.oxfordjournals.org/content/33/19/2451.full.pdf.

Patient Information

Images

http://www.nlm.nih.gov/medlineplus/ency/imagepages/18074.htm.

Medline Plus

ENGLISH

http://www.nlm.nih.gov/medlineplus/ency/article/000179.htm.

ESPANOL

http://www.nlm.nih.gov/medlineplus/spanish/ency/article/000179.htm.

Cleveland Clinic

http://my.clevelandclinic.org/heart/disorders/valve/valve-types.aspx.

Merck

http://www.merckmanuals.com/home/SearchResults?query=Aortic+Regurgitation&icd9=424.1%3b395.1.

Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/aortic-valve-regurgitation/basics/symptoms/con-20022523.

AHA

http://www.heart.org/HEARTORG/Conditions/More/HeartValveProblemsandDisease/Problem-Aortic-Valve-Stenosis-UCM-450437-Article.jsp?gclid=Cj0KEQjwg9-vBRCK7L7wmO2u0JcBEiQA-tzoaKzLLapbFnIE6goIxNLW7A8HZ-84qpW2U-av2QMxZJMaAvzx8P8HAQ.

Professional Information

Review

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Review

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J Am Coll Cardiol. 2013;62:1488. http://content.onlinejacc.org/article.aspx?articleID=1731144.

LA/LV/Arterial Pressure

Cardiosource Image. http://www.cardiosource.org/Science-And-Quality/Clinical-Images/L/LV-LA-and-Arterial-Pressure-in-Acute-Aortic-Regurgitation.aspx?w-nav=Search&WT.oss=acute%20aortic%20regurgitation&WT.oss-r=157&.

LV Ejection Dynamics

Cardiosource Image. http://www.cardiosource.org/Science-And-Quality/Clinical-Images/T/The-LV-Ejection-Dynamics-in-Acute-Aortic-Regurgitation.aspx?w-nav=Search&WT.oss=acute%20aortic%20regurgitation&WT.oss-r=157&.

Prosthetic Valve Dysfunction

Eur Heart J Cardiovasc Imaging. 2008;9:2 291–3. http://ehjcimaging.oxfordjournals.org/content/9/2/291.full.

Trauma

Heart. 2005;91:568–70. http://heart.bmj.com/content/91/5/568.full.

Updates and More

https://clinicalguidecvd.com/ara

Chapter 8
Aortic Regurgitation: Chronic

Management Keys

Perform TTE for management decisions at all stages of Chronic AR [9]
Perform TTE to detect/evaluate possible aortic regurgitation in patients with other conditions commonly associated with AR [10]
Closely monitor changing symptoms in patients with previously documented AR as these may be indicators for aortic valve replacement
Treat arterial hypertension according to current guideline recommendations
Perform AV replacement in symptomatic patients with severe AR regardless of LV function

ICD-10 Code

I35.1

Alternate Names/Abbreviation

AR
Aortic Insufficiency

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 131
DOI 10.1007/978-1-4471-7293-2_8,

Description/Etiology

Failure of AV leaflet coaptation due to:

Annulus abnormality
Dilatation/altered geometry of aortic root
Leaflet deformities (acquired, congenital)
Trauma (e.g., fibrous band)

Comorbid Conditions

ANTIPHOSPHOLIPID SYNDROME
AORTIC DISSECTION
AORTIC STENOSIS – SUBVALVULAR
AORTIC STENOSIS – VALVULAR
AORTIC VALVE REPLACEMENT
AORTIC VALVOTOMY
APPETITE-SUPPRESSANT DRUGS [2]
AUTOIMMUNE/CONNECTIVE TISSUE DISEASE [1]
CHEST TRAUMA
EHLERS-DANLOS SYNDROME
HYPERTENSION – SYSTEMIC ARTERIAL
IDIOPATHIC AORTIC ROOT DILATATION [3]
INFECTIVE ENDOCARDITIS
MARFAN SYNDROME
OSTEOGENESIS IMPERFECTA
RHEUMATIC FEVER [SEE Appendix A]
SYPHILITIC AORTITIS
SYSTEMIC LUPUS ERYTHEMATOSUS

Demography

M > F

Pathophysiology

Characterized by increased preload and afterload
LV progressively dilates to maintain cardiac output, with increased end-diastolic and end-systolic volumes; elevated LV afterload results from increased LV end-diastolic volume, causing increased wall stress, and from increased SV that is ejected into high-impedance aorta, causing systolic hypertension
Volume overload due to AR causes compensatory eccentric hypertrophy, and pressure overload causes concentric hypertrophy; increased systolic wall stress and afterload stimulate further concentric hypertrophy; combination preload reserve and continued compensatory hypertrophy allow LV to maintain normal wall stress, with normal ejection
Symptoms develop when LA pressure increases late in disease process
Failure of compensatory hypertrophy causes afterload mismatch and ensuing systolic dysfunction
Systolic dysfunction is at first reversible if valvular dysfunction is corrected, eliminating afterload mismatch; when untreated, permanent contractile dysfunction can occur

Signs/Symptoms

ARTERIAL PRESSURE, DIAS – DECR
ARTERIAL PULSE – DOUBLE (BISFERIENS)
ARTERIAL PULSE PRESSURE – INCR
ARTERIAL PULSE, FALL – RAPID
ARTERIAL PULSE, RISE – RAPID
ARTERY, CAROTID – THRILL
ARTERY, FEMORAL, SOUND, SYS/DIAS – BOOMING (TRAUBE SIGN)
BLOOD PRESSURE, ARTERIAL – INCREASED/ELEVATED, SYS
BREATHING – DIFF (DYSPNEA)
BREATHING – DIFF, NOCTURNAL (DYSPNEA, NOCT)

CHEST – PAIN
CHEST – PALPITATIONS
EXTREM, DIGITS, NAILS – PULSATIONS (QUINCKE SIGN)
HEAD – BOBBING (DE MUSSET SIGN)
HEADACHE
HEART, A2, INTENSITY – DECR/ABSENT
HEART, LSB, MID – MURMUR, DIAS [10][11]
HEART, LSB, MID – MURMUR, SYS [12]
HEART, LSB, MID – THRILL, DIAS
HEART, LV APEX – MURMUR, SYS [12]
HEART, LV, APEX – IMP, DIFFUSE
HEART, LV, APEX – IMP, INF DISPLACEMENT
HEART, LV, APEX – IMP, LAT DISPLACEMENT
HEART, LV, APEX – MURMUR, DIAS [10][11]
HEART, RSB, UPPER – MURMUR, DIAS [10][11]
HEART, RSB, UPPER – MURMUR, SYS [12]
HEART, RSB, UPPER – THRILL, SYS
HEART, S1, INTENSITY – DECR/ABSENT
HEART, S2, SPLIT – REVERSED (PARADOXICAL)
HEART, S3 LV
MOUTH, UVULA – PULSATIONS (MUELLER SIGN)
NECK, SUPRASTERNAL NOTCH – THRILL, SYS
SWEATING, NOCT – INCR (DIAPHORESIS, NOCT)

Differentiation

Other causes of diastolic murmur at LV apex
Other causes of diastolic murmur at mid LSB
Other causes of diastolic murmur at upper RSB
Other causes of wide pulse pressure

Complications

Aortic Regurgitation – Acute
Heart Failure

Infective Endocarditis
Progression of predisposing/comorbid condition
Sudden death

Laboratory

NS

ECG

QRS – LBBB/LBBB PATTERN
QRS – LVH PATTERN
QRS, AMP – INCR
QRS, AXIS – L
ST SEGMENT – DEPR

Imaging [9]

AORTA, ASCEND, SIZE – INCR
AV, FLOW – REGURG
AV, LEAFLETS – CALCIUM [6]
AV, LEAFLETS – THICK [6]
AV, LEAFLETS, MORPH – ABN [6]
AV, LEAFLETS, MOTION – ABN [6]
CARDIAC CATH WITH AORTIC ROOT ANGIO [7]
CARDIOMEGALY
CORONARY ANGIO [4]
LV, CHAMBER, SIZE – INCR
LV, EF – INCR [8]
LV, WALL MOTION – INCR/HYPERDYNAMIC [8]

Other Tests

Exercise test with radionuclide angiography [7]

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic

Vasodilators
Hydralazine
Nifedipine

Treatment: Surgical/Invasive [13]

Aortic root reconstruction
Aortic valve replacement/repair

Course

Non-surgically treated symptomatic patients have mortality >10 %/year

Notes

[1] Especially Ankylosing Spondylitis
[2] Dexfenfluramine, Phentermine, Fenfluramine
[3] Often associated with bicuspid AV
[4] Evaluate for CAD in patients requiring surgical intervention
[5] To reduce systolic BP; diastolic BP generally low in significant AR and care must be taken to avoid further reduction, which could compromise coronary artery blood flow; not recommended for normotensive asymptomatic patients with normal LV function
[6] When AR is due to AV disease

[7] When noninvasive tests are unsatisfactory or discordant with clinical features
[8] When compensated
[9] TTE indicated in all stages of AR for:

Cause of regurgitation
LV size/systolic function
Severity of regurgitation
Timing of valve intervention

[10] AR commonly occurs, but may be clinically silent by auscultation, in association with:

Bicuspid AV
Dilated aortic sinuses
Dilated ascending aorta

[11] Longer murmur correlates with lesser severity of AR
[12] Flow-related systolic murmur may be predominant finding as diastolic murmur may be difficult to discern
[13] Timing/type of surgical intervention not well defined and controversial, in part due to complexities/diverse causes of chronic AR

Appendix A

World Heart Federation Echo Criteria for Diagnosis of Rheumatic Fever

INDIVIDUALS AGE <20 YEARS:
Definite RHD (either A, B, C, or D):

(A) Pathological MR and at least two morphological features of RHD of the MV
(B) MS mean gradient = 4 mmHg*
(C) Pathological AR and at least two morphological features of RHD of the AV‡
(D) Borderline disease of both the AV and MV§

Borderline RHD (either A, B, or C):

(A) At least two morphological features of RHD of the MV without pathological MR or MS
(B) Pathological MR
(C) Pathological AR

Normal echocardiographic findings (all of A, B, C, and D):

(A) MR that does not meet all four Doppler echocardiographic criteria (physiological MR)
(B) AR that does not meet all four Doppler echocardiographic criteria (physiological AR)
(C) An isolated morphological feature of RHD of the MV (for example, valvular thickening) without any associated pathological stenosis or regurgitation
(D) Morphological feature of RHD of the AV (for example, valvular thickening) without any associated pathological stenosis or regurgitation

INDIVIDUALS AGE 20+ YEARS:
Definite RHD (either A, B, C, or D):

(A) Pathological MR and at least two morphological features of RHD of the MV
(B) MS mean gradient = 4 mmHg*
(C) Pathological AR and at least two morphological features of RHD of the AV, only in individuals aged <35 years‡
(D) Pathological AR and at least two morphological features of RHD of the MV

*Congenital MV anomalies must be excluded. Furthermore, inflow obstruction due to nonrheumatic mitral annular calcification must be excluded in adults. ‡Bicuspid AV, dilated aortic root, and hypertension must be excluded. §Combined AR and MR in high prevalence regions and in the absence of congenital heart disease is regarded as rheumatic. Abbreviations: AR, aortic regurgitation; AV, aortic valve;

MR, mitral regurgitation; MS, mitral stenosis; MV, mitral valve; RHD, rheumatic heart disease; WHF, World Heart Federation.

Guidelines

2014 AHA/ACC Guideline for the management of patients with valvular heart disease
J Am Coll Cardiol. 2014;63:e57–185. http://content.onlinejacc.org/article.aspx?articleid=1838843&resultClick=3.
Guidelines on the management of valvular heart disease (version 2012)
Eur Heart J. 2012;33:2451–96. http://eurheartj.oxfordjournals.org/content/33/19/2451.full.pdf.

Patient Information

Images

http://www.nlm.nih.gov/medlineplus/ency/imagepages/1056.htm.
http://www.nlm.nih.gov/medlineplus/ency/imagepages/9380.htm.
http://www.nlm.nih.gov/medlineplus/ency/imagepages/18074.htm.

Medlineplus

ENGLISH

http://www.nlm.nih.gov/medlineplus/ency/article/003266.htm.
http://www.nlm.nih.gov/medlineplus/ency/article/000179.htm.

ESPANOL

http://www.nlm.nih.gov/medlineplus/spanish/ency/article/003266.htm.
http://www.nlm.nih.gov/medlineplus/spanish/ency/article/000179.htm.

Cleveland Clinic

http://my.clevelandclinic.org/heart/disorders/valve/valve-types.aspx.

Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/aortic-valve-regurgitation/basics/definition/con-20022523.

Merck

http://www.merckmanuals.com/home/SearchResults?query=Aortic+Regurgitation&icd9=424.1%3b395.1.

Professional Information

Review

Lancet. 2016;387:1312–23. http://www.sciencedirect.com/science/article/pii/S0140673616005869.

Review

N Engl J Med. 2004;351:1539–46. http://www.nejm.org/doi/full/10.1056/NEJMcp030912.

Review: Valve Repair

Circulation. 2013;128:854–63. http://circ.ahajournals.org/content/128/8/854.full.

Combined AS/AR

J Am Coll Cardiol. 2013;61:1489–95. http://content.onlinejacc.org/article.aspx?articleID=1673463.

Echo: Severity

J Am Soc Echocardiogr. 2003;16:777–802. http://www.onlinejase.com/article/S0894-7317(03)00335-3/abstract.

Etiolology: Patients Requiring AVR

Circulation. 2006;114:422–9. http://circ.ahajournals.org/content/114/5/422.full.

NT-PROBNP

Int J Cardiol. 2008;127:321–7. http://www.sciencedirect.com/science/article/pii/S0167527307018013.

Outcomes

Circulation. 1999;99:1851–7. http://circ.ahajournals.org/content/99/14/1851.full.

Post-AVR (Low EF): Outcomes

Circulation. 2002;106:2687–93. http://circ.ahajournals.org/content/106/21/2687.full.

Post-transcatheter AVR: Outcomes

J Am Coll Cardiol. 2013;61:1585–95. http://content.onlinejacc.org/article.aspx?articleID=1676139.

Transcatheter AVR

J Am Coll Cardiol. 2013;61:1577–84. http://content.onlinejacc.org/article.aspx?articleid=1654982&resultClick=3.

Trauma: Fibrous Band

Eur Heart J Cardiovasc Imaging. 2015;16:465. http://ehjcimaging.oxfordjournals.org/content/16/5/465.full?etoc.

Vasodilator Therapy

N Engl J Med. 2005;353:1342–9. http://www.nejm.org/doi/full/10.1056/NEJMoa050666.

WHF Echo Criteria for RHD

Nat Rev Cardiol. 2012;9:297–309. http://www.nature.com/nrcardio/journal/v9/n5/box/nrcardio.2012.7-BX1.html.

Updates and More

https://clinicalguidecvd.com/arc

Chapter 9
Aortic Stenosis: Discrete Subvalvular

ICD-10 Code

Q24.4

Alternate Names/Abbreviation

SubAS
Congenital Fixed Subaortic Stenosis
Discrete Subaortic Stenosis

Description/Etiology

Discrete fibrous ring or fibromuscular narrowing:

- Distinct from hypertrophic cardiomyopathy with dynamic LVOT obstruction
- Ring may extend onto anterior mitral leaflet
- Usually solitary but may be superimposed on other congenital heart defects

Usually congenital
Acquired under certain circumstances (eg, VSD patching) [5]
Familial in some cases, such as Shones Syndrome [1]

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, DOI 10.1007/978-1-4471-7293-2_9,

Usually fixed obstruction but secondary dynamic component may occur secondary to myocardial hypertrophy and dynamic LV ejection.
In some cases accessory mitral tissue or anomalous chords may cause SubAS

Comorbid Conditions

AORTIC REGURGITATION – CHRONIC [7]
ATRIOVENTRICULAR SEPTAL DEFECT [4]
BICUSPID AORTIC VALVE
CARDIOMYOPATHY – HYPERTROPHIC
COARCTATION OF AORTA
CONOTRUNCAL ABNORMALITIES
DOUBLE OUTLET RIGHT VENTRICLE
LEFT SUPERIOR VENA CAVA
NOONAN SYNDROME
PATENT DUCTUS ARTERIOSUS
SHONES SYNDROME [1]
VENTRICULAR SEPTAL DEFECT [4]

Demography

M 2:1

Pathophysiology

Anatomical:

Discrete fibrous ring or Fibromuscular Hyperplasia
Fibrous membrane
Tunnel-like fibromuscular band
Abnormal MV insertion
Progressive AV damage

Physiological: hemodynamics due to AS/AR and progressive LVH

Signs/Symptoms

ARTERIAL PULSE PRESSURE – DECR
ARTERIAL PULSE, DOWNSLOPE – GRADUAL
ARTERIAL PULSE, PEAK – SUSTAINED
ARTERIAL PULSE, RISE – SLOW
ARTERY, CAROTID – THRILL
BREATHING – DIFF (DYSPNEA)
CONSCIOUSNESS – LOSS, SUDDEN, EFFORT (EFFORT SYNCOPE)
DIZZY/LIGHTHEADED, EFFORT
EYES, SEPARATION – WIDE (HYPERTELORISM)
FATIGUE
HEART, A2, INTENSITY – DECR/ABSENT
HEART, APEX, LV – MURMUR, SYS
HEART, LSB, MID – MURMUR, DIAS
HEART, LSB, MID – MURMUR, SYS [2]
HEART, LV, APEX, IMP – FORCEFUL/SUSTAINED
HEART, LV, APEX, IMP – FORCEFUL/SUSTAINED
HEART, LV, APEX, IMP – PRESYS
HEART, RSB, UPPER – MURMUR, SYS
HEART, S2, SPLIT – REVERSED (PARADOXICAL)
MENTATION, LEARNING, DEVELOPMENT – DECR
NECK, JVP, A WAVE – INCR/LARGE (CANNON WAVE)
NECK, SUPRASTERNAL NOTCH – THRILL, SYS
SWEATING – INCR (DIAPHORESIS/HYPERHIDROSIS)

Differentiation

Aortic Stenosis – Valvular
Aortic Stenosis – Supravalvular
Cardiomyopathy – Hypertrophic (with aortic outflow obs)

Complications

Heart Failure
Infective Endocarditis
Sudden death

Laboratory

NS

ECG [3]

DYSRHYTHMIAS – ATRIAL (PACS/OTHERS)
DYSRHYTHMIAS – VENTRICULAR (PVCS/OTHERS)
QRS – LVH PATTERN

Imaging [3] [8]

AORTA, ASCEND, SIZE – INCR
ECHO/DOPPLER FOR SUBAORTIC GRADIENT; ASSD AV ABN; LVH/FUNCTION
LV, CHAMBER, SIZE – INCR
LV, MYOCARD, WALL THICKNESS – INCR (HYPERTROPHY)

Other Tests

Cardiac catheterization: for other possible lesions; angiography unreliable for detecting subaortic membrane

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic

Infective endocarditis prophylaxis

Treatment: Surgical/Invasive

Surgical resection [9]

Course

Often progressive
Unrepaired: progressive AV damage, ventricular dysfunction, Infective Endocarditis, SCD

Notes

[1] Shones Syndrome:

Supravalvular stenosing ring
Parachute MV
Subaortic stenosis
Coarctation of Aorta

[2] May or may not radiate to carotids; ejection click absent
[3] May be normal in absence of significant AS or AR
[4] May occur after patch closure of VSD or AVSD
[5] Does not appear in embryologic development and infrequent in neonates
[6] E.g., VSD, AV Septal Defect, Double Outlet RV
[7] Mild-moderate in 60 % of cases
[8] Transthoracic 2D Echo-Doppler: initial diagnostic method of choice for:

LV outflow anatomy
Subaortic gradient
Associated AV abnormality
AR quantification
Ascending aorta diameter
MV involvement
Assess LV hypertrophy and function
TEE may add valuable preoperative/intraoperative anatomic detail

[9] Guidelines:ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease (J Am Coll Cardiol 2008;52(23):e143–e263)

Subaortic Stenosis

Recommendations for Surgical Intervention

Class I

1. Surgical intervention is recommended for patients with SubAS and a peak instantaneous gradient of 50 mmHg or a mean gradient of 30 mmHg on echocardiography-Doppler. *(Level of Evidence: C)*
2. Surgical intervention is recommended for SubAS with less than a 50-mmHg peak or less than a 30-mmHg mean gradient and progressive AR and an LV dimension at end-systolic diameter of 50 mm or more or LV ejection fraction less than 55 %. *(Level of Evidence: C)*

Class IIb

1. Surgical resection may be considered in patients with a mean gradient of 30 mmHg, but careful follow-up is required to detect progression of stenosis or AR. *(Level of Evidence: C)*
2. Surgical resection may be considered for patients with less than a 50-mmHg peak gradient or less than a 30-mmHg mean gradient in the following situations:
   (a) When LV hypertrophy is present. *(Level of Evidence: C)*
   (b) When pregnancy is being planned. *(Level of Evidence: C)*
   (c) When the patient plans to engage in strenuous/competitive sports. *(Level of Evidence: C)*

Class III

1. Surgical intervention is not recommended to prevent AR for patients with SubAS if the patient has trivial LVOT obstruction or trivial to mild AR. *(Level of Evidence: C)*

Guidelines

ACC/AHA 2008 Guidelines for the management of adults with congenital heart disease
J Am Coll Cardiol. 2008;52:e143–e263. http://content.onlinejacc.org/article.aspx?articleid=1188032#tab1.

ESC Guidelines for the management of grown-up congenital heart disease (new version 2010)
Eur Heart J. 2010;31:2915–57. http://www.escardio.org/guidelines-surveys/esc-guidelines/Pages/grown-up-congenital-heart-disease.aspx.

Patient Information

Images

http://www.nlm.nih.gov/medlineplus/ency/imagepages/18075.htm.

Medlineplus

ENGLISH

http://www.nlm.nih.gov/medlineplus/ency/article/000178.htm.

ESPANOL

http://www.nlm.nih.gov/medlineplus/spanish/ency/article/000178.htm.

Genetics Home Reference

http://ghr.nlm.nih.gov/condition/familial-hypertrophic-cardiomyopathy.

Cleveland Clinic

http://my.clevelandclinic.org/heart/disorders/valve/valve-types.aspx.
http://my.clevelandclinic.org/childrens-hospital/health-info/diseases-conditions/heart/hic-pediatric-congenital-heart-defects.aspx.

Professional Information

Balloon Dilatation: Long-Term Outcomes

Circulation. 2011;124:1461–8. http://circ.ahajournals.org/content/124/13/1461.full?sid=6b5ccfb5-acc6-4765-99b0-93adeefa19a8.

Natural History: Adults

Eur Heart J. 2013;34:1548–56. http://eurheartj.oxfordjournals.org/content/34/21/1548.full?sid=d529391a-c121-45ab-b54a-d9cab0bdbd3d.

Prevalence/Progression

J Am Coll Cardiol. 2001;38:835–42. http://content.onlinejacc.org/article.aspx?articleid=1127431.

Surgery: Long-Term Outcomes

Circulation. 2013;127:1184–91. http://circ.ahajournals.org/content/127/11/1184.full.

Surgery: Long-Term Outcomes

J Card Surg. 2005;20:16–21. http://www.ncbi.nlm.nih.gov/pubmed/15673405?dopt=Abstract.

Updates and More

https://clinicalguidecvd.com/assub

Chapter 10
Aortic Stenosis: Supravalvular

Management Keys

- Annual follow up of both operated and unoperated patients with SupraAS at a regional ACHD center
- Screen all available/undetected family members, who may be at risk for hypertension, CAD, or stroke
- Counsel patients with SupraAS and significant obstruction, coronary involvement, or aortic disease about pregnancy

ICD-10 Code

Q25.3

Alternate Names/Abbreviation

SupraAS

Description/Etiology

- Fixed obstruction just above sinus of Valsalva, extending a variable distance along aorta [3]

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_10,

Origin of coronary arteries usually proximal to obstruction, causing high intracoronary systolic pressure/limited diastolic flow; sometimes associated with partial or complete coronary artery ostial obstruction, ectasia, or aneurysm
Often associated with Williams Syndrome and other forms of congenital cardiovascular disease

Comorbid Conditions

BICUSPID AORTIC VALVE
HYPERCALCEMIA [WILLIAMS SYNDROME]
HYPOPLASTIC DESCENDING AORTA
NOONAN SYNDROME
PULMONARY STENOSIS – SUPRAVALVULAR
RENAL ARTERY STENOSIS
WILLIAMS SYNDROME [4]

Demography

Variable according to associated condition

Pathophysiology

Morphological abnormalities include:

Diffuse or focal intimal and medial fibrosis
Hyperplasia and dysplasia
Adventitial fibroelastosis
Intramedial dissection (more common in adults)

In adults, coronary insufficiency and early onset of CAD due to anatomic obstruction/myocardial hypertrophy that restricts intramyocardial coronary flow

Signs/Symptoms [7]

ARTERIAL PRESSURE, UE, SYS – R > L [5]

ARTERIAL PULSE PRESSURE – DECR
ARTERIAL PULSE, CAROTID – R > L
ARTERIAL PULSE, DOWNSLOPE – GRADUAL
ARTERIAL PULSE, PEAK – SUSTAINED
ARTERIAL PULSE, RISE – SLOW
ARTERIAL PULSE, UE – ASYMMETRIC [5]
ARTERY, CAROTID – THRILL [1]
BREATHING – DIFF (DYSPNEA)
CONSCIOUSNESS – LOSS, SUDDEN, EFFORT (EFFORT SYNCOPE)
DIZZY/LIGHTHEADED, EFFORT
FATIGUE
HEART, LV, APEX, IMP – FORCEFUL/SUSTAINED
HEART, RSB, UPPER – MURMUR, SYS [6]
HEART, S2, SPLIT – REVERSED (PARADOXICAL)
HEART, S4 LV
NECK, JVP, A WAVE – INCR/LARGE (CANNON WAVE)
NECK, SUPRASTERNAL NOTCH – THRILL, SYS
SWEATING – INCR (DIAPHORESIS/HYPERHIDROSIS)

Differentiation

Other causes of LV outflow obstruction

Complications

CAD [8]
HF
SCD
Stroke
Systemic Arterial Hypertension

Laboratory

NS

ECG

DYSRHYTHMIAS – ATRIAL (PACS/OTHERS)
DYSRHYTHMIAS – VENTRICULAR (PVCS/OTHERS)
QRS – LVH PATTERN [2]
ST SEGMENT – DEPR [8]
T WAVE – INVER, ABN [8]

Imaging [9]

AORTA, ASCEND, SYS – GRADIENT
AORTA, ASCENDING – SMALL [9]
LV, MYOCARD, WALL THICKNESS – INCR (HYPERTROPHY)

Genomics

CLIP2 [WILLIAMS SYNDROME]
ELN [WILLIAMS SYNDROME]
GTF2I [WILLIAMS SYNDROME]
GTF2IRD1 [WILLIAMS SYNDROME]
LIMK1 [WILLIAMS SYNDROME]

Other Tests

Stress test for CAD
Cardiac cath

Treatment: Nonpharmacologic

Counseling:

- Pregnancy
- Psychosocial

Treatment: Pharmacologic

NS

Treatment: Surgical/Invasive

Guideline-directed therapy [10]
CABG
Complex aorta patching
Reconstruction of coronary ostia

Course

Variable according to severity/associated lesions

Notes

[1] Right > left
[2] ECG changes do not correlate with degree of obstruction
[3] Composed of intimal and media fibrosis, hyperplasia, adventitial fibroelastosis, intramedial dissection
[4] Including supravalvular AS, PA stenosis, teeth/jaw malformations, mental retardation, hypercalcemia, joint abnormalities, small stature, renal artery stenosis, systemic arterial hypertension, nephrocalcinosis
[5] Due to Coanda effect: preferential blood flow into right brachiocephalic artery
[6] Crescendo-decrescendo; ejection click absent; radiates to right carotid artery
[7] Exam should also include auscultation for murmurs of peripheral PA stenosis (back) and RA stenosis (flank, abdominal bruit)
[8] Myocardial ischemia secondary to coronary ostial stenosis/LVH

[9] Echo (TTE and TEE) for assessing:

Aortic sinus diameter and anatomy
Sinotubular ridge
Proximal ascending aorta
Origins of coronary arteries
Systolic gradient across obstruction
Degree of LV hypertrophy
MRI/CT used precisely define anatomy of aorta and branches and pulmonary arteries

[10] ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease (J Am Coll Cardiol. 2008;52(23):e143–e263)

Supravavular Aortic Stenosis

Class I

1. Operative intervention should be performed for patients with supravalvular LVOT obstruction (discrete or diffuse) with symptoms (i.e, angina, dyspnea, or syncope) and/or mean gradient greater than 50 mmHg or peak instantaneous gradient by Doppler echocardiography greater than 70 mmHg. *(Level of Evidence: B)*
2. Surgical repair is recommended for adults with lesser degrees of supravalvular LVOT obstruction and the following indications:
   (a) Symptoms (i.e, angina, dyspnea, or syncope). *(Level of Evidence: B)*
   (b) LV hypertrophy. *(Level of Evidence: C)*
   (c) Desire for greater degrees of exercise or a planned pregnancy. *(Level of Evidence: C)*
   (d) LV systolic dysfunction. *(Level of Evidence: C)*
3. Interventions for coronary artery obstruction in patients with SupraAS should be performed in ACHD centers with demonstrated expertise in the interventional management of such patients. *(Level of Evidence: C)*

Guidelines

ACC/AHA 2008 Guidelines for the management of adults with congenital heart disease
J Am Coll Cardiol. 2008;52:e143–e263. http://content.onlinejacc.org/article.aspx?articleid=1188032#tab1.

ESC Guidelines for the management of grown-up congenital heart disease (new version 2010)
Eur Heart J. 2010;31:2915–57. http://www.escardio.org/guidelines-surveys/esc-guidelines/Pages/grown-up-congenital-heart-disease.aspx.

Patient Information

Schematic: ACC

http://www.acc.org/education-and-meetings/image-and-slide-gallery/media-detail?id=cc346a36856c49a18e4f7af058ccae73.

Images

http://www.nlm.nih.gov/medlineplus/ency/imagepages/18075.htm.

Medlineplus

ENGLISH

http://www.nlm.nih.gov/medlineplus/ency/article/000178.htm.

ESPANOL

http://www.nlm.nih.gov/medlineplus/spanish/ency/article/000178.htm.

Genetics Home Reference

http://ghr.nlm.nih.gov/condition/supravalvular-aortic-stenosis.

Cleveland Clinic

http://www.clevelandclinicmeded.com/medicalpubs/diseasemanagement/cardiology/aortic-valve-disease/.

Professional Information

Image: Case Report

Heart. 2010;96:1808. http://heart.bmj.com/content/96/22/1808.extract.

Image: Hourglass Type

J Am Coll Cardiol. 2010;56:e13. http://ac.els-cdn.com/S0735109710021030/1-s2.0-S0735109710021030-main.pdf?-tid=7ecfa826-e734-11e3-ae1d-00000aab0f26&acdnat=1401370020-018e4944a1900702c851b5983e21db28.

Pathophysiology

Circ Cardiovasc Genet. 2012;5:692–6. http://circgenetics.ahajournals.org/content/5/6/692.full.pdf.

Williams Syndrome

Circulation. 2013;127:2125–34. http://circ.ahajournals.org/content/127/21/2125.full?sid=dbdc5d06-8014-4258-bd23-b81f88aab788.

Updates and More

https://clinicalguidecvd.com/assup

Chapter 11
Aortic Stenosis: Valvular

Management Keys

- Regular reevaluation, including echo, at 1–3 year intervals dictated by disease severity once diagnosis of AV sclerosis/stenosis is made
- Interpret AV invasive/noninvasive hemodynamics with meticulous attention to detail because can be severe with lower AV gradients and velocities [17]
- Expert, accurate echo measures of aortic velocity, mean transaortic gradient, valve area for establishing disease severity, progression monitoring, management decisions [27]
- Consider additional diagnoses when symptoms occur in patients with documented mild-moderate AS, which is hemodynamically well-tolerated
- Immediate AV replacement consideration with onset of any symptoms attributable to AS
- Perform coronary angiography in all patients prior to AV replacement
- Closely monitor certain subsets of patients with comorbidities for rapid disease progression including those with renal dysfunction and history of mediastinal radiation therapy
- Diagnose/treat conventional CVD risk factors according to current guidelines, especially Systemic Arterial Hypertension, which places additional load on LV [9]

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 159
DOI 10.1007/978-1-4471-7293-2_11,

Patient education about AS, especially early symptoms that may be missed or passed off without focused questions, including tolerance to physical exertion
Avoidance of strenuous/competitive exercise in patients with severe AS

ICD-10 Code

I35.0

Alternate Names/Abbreviation

AS
Aortic Valve Sclerosis
Calcific Aortic Valve Disease (CAVD)

Description/Etiology

Causes:
- Calcification of trileaflet AV
- Calcification of congenital bicuspid AV [12]
- Rheumatic heart disease (uncommon in developed countries)

Stages (with valve anatomy):

A: At risk of AS

Bicuspid AV/other congenital valve abnormality
Aortic Valve Sclerosis

B: Progressive AS

Nonrheumatic: mild-mod leaflet calcification of valve leaflets with some reduction in systolic motion
Rheumatic: leaflet changes with commissural fusion

C1: Asymptomatic severe [27]

Severe leaflet calcification or
Congenital stenosis with severely reduced leaflet opening

C2: Asymptomatic severe with LV dysfunction

Severe leaflet calcification or
Congenital stenosis with severely reduced leaflet opening

D1: Symptomatic severe high-gradient AS

Severe leaflet calcification or
Congenital stenosis with severely reduced leaflet opening

D2: Symptomatic severe with low-flow/low-gradient as with reduced LVEF [17]

Severe leaflet calcification with severely reduced leaflet motion

D3: Symptomatic severe low-gradient AS with normal LVEF or paradoxical low- flow severe AS [17]

Severe leaflet calcification with severely reduced leaflet motion

Aortic Valve Sclerosis [15]: focal areas of leaflet thickening or mild calcification detected by echo/CT in absence of LVOT obstruction; associated factors:

Diabetes Mellitus [32]
Increased LDL-C [32]
Male gender
Metabolic Syndrome
Older age
Short height
Systemic Arterial Hypertension [32]
Tobacco use [32]

Mild/moderate alcohol use may be associated with lower risk

Initial diagnosis usually incidental by auscultation/echo

Comorbid Conditions

ALKAPTONURIA
AORTIC REGURGITATION – CHRONIC
AORTIC STENOSIS – SUBVALVULAR
AORTIC STENOSIS – SUPRAVALVULAR
AORTOPATHY [28] [BICUSPID AV]
ATHEROSCLEROSIS IN OTHER CV AREAS
ATRIAL FIBRILLATION [40]
COARCTATION OF AORTA
DYSLIPIDEMIA
HYPERCHOLESTEROLEMIA [14]
HYPERCHOLESTEROLEMIA – FAMILIAL [14]
HYPERTENSION – SYSTEMIC ARTERIAL
MITRAL REGURGITATION [39]
PATENT DUCTUS ARTERIOSUS
PORCELAIN AORTA [36]
RHEUMATIC FEVER
SHONE SYNDROME [13]
SINUS OF VALSALVA ANEURYSM
TOBACCO USE
VENTRICULAR SEPTAL DEFECT
VON WILLEBRAND SYNDROME [6] [ACQUIRED]

Demography

Increased prevalence with age; moderate-severe form present in 5 % of persons age >75 years in USA
M > F under age 75 years; gender equal >75 years
Rheumatic: increased incidence in developing countries

Pathophysiology

Non-rheumatic [1] [26]

Initiation phase
Valvular lipid deposition
Injury
Inflammation

Propagation phase
Pro-calcific factors
Pro-osteogenic factors

Rheumatic: commissural fusion, scaring, calcification [2]

Compensatory changes for LV overload to maintain card output:
Concentric LVH [3]
Increased myocardial contractility

Compensatory changes over several decades eventually overwhelmed by:
Decreased diastolic compliance
Exhausted myocardial contractile reserve
Irreversible myocardial fibrosis
Subendocardial ischemia
Vasodilation (baroreceptor-activated)

AV sclerosis: progressive leaflet thickening leading to LVOT obstruction in 10 % [16]

Hemodynamically significant MR often present and is factor in outcomes in AV valve replacement [39]

Signs/Symptoms [4]

ARTERIAL PULSE – SHUDDER
ARTERIAL PULSE PRESSURE – DECR
ARTERIAL PULSE, AMP – DECR/ABS [20]
ARTERIAL PULSE, DOWNSLOPE – GRADUAL
ARTERIAL PULSE, RISE – SLOW [20]
ARTERY, CAROTID – THRILL
BLEEDING, PROPENSITY – INCR [6]
BREATHING – DIFF (DYSPNEA)
CHEST – PAIN [42]
CHEST – PALPITATIONS

CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE) [31]
CONSCIOUSNESS – LOSS, SUDDEN, EFFORT (EFFORT SYNCOPE)
DIZZY/LIGHTHEADED, EFFORT
EYES, VISION – DECR/LOSS
FATIGUE
HEART, A2, INTENSITY – DECR/ABSENT
HEART, LSB, MID – MURMUR, DIAS
HEART, LSB, MID – MURMUR, SYS [18] [19] [33]
HEART, LV APEX – MURMUR, SYS [18] [19] [33] [38]
HEART, LV, APEX – CLICK(S), SYS [CONG BICUSPID AS]
HEART, LV, APEX – IMP, PRESYS
HEART, LV, APEX, IMP – FORCEFUL/SUSTAINED
HEART, RSB, UPPER – CLICK, SYS
HEART, RSB, UPPER – MURMUR, SYS [18] [19] [33]
HEART, S1, INTENSITY – DECR/ABSENT
HEART, S2 – SINGLE [SEVERE; ONLY P2 AUDIBLE]
HEART, S2, SPLIT – REVERSED (PARADOXICAL) [21]
NECK, SUPRASTERNAL NOTCH – THRILL, SYS
NOSE – BLOOD (EPISTAXIS) [6]
SKIN – PETECHIAE/ECCHYMOSES/PURPURA [6]

Differentiation

Aortic Sclerosis
Papillary Fibroelastoma
Subvalvular AA
Supravalvular AS

Complications

Aortic Dissection [12] [28]
Bleeding [6]
Heart Failure
Infective Endocarditis
Sudden Death [5]

Laboratory

BLOOD, CHOLESTEROL, LDL (LDL-C) – INCR [14]
BLOOD, HGB/HCT – DECR (ANEMIA) [6]
BLOOD, NT-PROBNP – INCR [EARLY HEMODYNAMIC COMPROMISE]
BLOOD, PLATELET FUNCTION – ABN [WITH NORMAL PLATELET COUNT]
BLOOD, ST2 – INCR
BLOOD, TROPONIN – INCR

ECG [10]

DYSRHYTHMIAS – ATRIAL (PACS/OTHERS)
DYSRHYTHMIAS – VENTRICULAR (PVCS/OTHERS)
QRS – LVH PATTERN [34]
QRS, AMP – INCR
QRS, AXIS – L
ST SEGMENT – DEPR [29]

Imaging [17] [24] [27] [43]

AV, FLOW, VELOCITY – INCR [27]
AV, GRADIENT, TRANSAORTIC – INCR [27]
AV, LEAFLETS – CALCIUM [26]
AV, LEAFLETS – THICK
AV, LEAFLETS, MORPH – ABN
AV, VALVE AREA – DECR [27]
LV, EF – DECR [MAY BE N]
LV, MYOCARD, WALL THICKNESS – INCR (HYPERTROPHY) [34]
MV, FLOW – REGURG [23]

Genomics

LPA [37]

Other Tests

Dobutamine stress [11]; indications:

Calcified AV with reduced sys opening
LVEF <50 %
Calculated valve area 1 cm or less
Aortic velocity <4 m/s or mean press gradient <40 mmHg

Exercise test [8]

Cardiac catheterization: indicated for estimation of AS severity when:

Noninvasive data nondiagnostic
Discrepancy between echo and clinical evaluation

Coronary angiography indications before valve intervention [25]:

Angina pectoris
CAD risk factors, including:

Males <40 YEARS
Females post-menopause

Decreased LV systolic function

History of CAD

Objective evidence of myocardial ischemia

Treatment: Nonpharmacologic

NS [35]

Treatment: Pharmacologic

Systemic Arterial Hypertension: careful control in patients with stages A-C

Vasodilator treatment: (with invasive hemodynamic monitoring) in acute treatment of patients with severe decompensated AS (stage D)/NYHA Class IV symptoms

Treatment: Surgical/Invasive

AV replacement [44]

- Surgical AVR [45]
- TAVR [45]

Balloon valvuloplasty [43]

Prevention

Endocarditis prophylaxis
Rheumatic Fever prophylaxis until age 40 years in patients with rheumatic form of AS
Atherosclerotic CVD risk factor modification [9] [32]

Course

Progressive disease with inevitable increase in hemodynamic severity
Average survival 2–3 years after symptom onset without AV replacement, with mortality up to 50 % at 1 year

Notes

[1] Similar to atherogenesis
[2] Usually with MS
[3] May be excessive with aging, especially in females
[4] Symptom onset varies with etiology, as early as age 50 years with congenital bicuspid form and as late as age 80 years with calcific form; most patients asymptomatic at time of initial diagnosis; symptom onset is critical point in natural history and may require definitive (i.e., surgical) intervention; asymptomatic patients should be closely monitored;

symptoms in presence of documented mild-moderate AS should prompt consideration of additional causes

[5] Usually occurs in presence of prior symptoms but uncommonly occurs in asymptomatic patients

[6] Heyde Syndrome: increased bleeding due to acquired Type 2A von Willebrand Syndrome; also GI, gingival, increased menopausal bleeding; 20 % of patients with advanced AS; related to AS severity; improved by AV replacement

[7] Coronary/CT angiography may be indicated for echo/clinical discrepancy and/or when symptoms may be due to CAD

[8] Contraindicated in symptomatic patients

[9] No data support efficacy in affecting AS progression; however, should be used for hypercholesterolemia according to standard GDMT

[10] May be normal even in severe AS

[11] High risk test, only performed in experienced centers; to determine transvalvar gradient in select patients

[12] Bicuspid AV: 1 % of population; underlying cause of 50 % of AVRs in USA; also associated with AR, aortic root dilatation in absence of AS

[13] Parachute MV, Coarctation of Aorta, supravalvular mitral membrane, Subaortic Stenosis

[14] Genetic predisposition to increased LDL-C has been associated with valvular AS and AV calcification

[15] Aortic Sclerosis: associated with 50 % increased risk of CV death/AMI over 5 years; may be marker for subclinical CAD

[16] Progression may be accelerated by severe hypercholesterolemia, renal failure, radiation therapy, other unidentified factors

[17] Patients with normal/decreased LVEF pose special diagnostic and treatment challenge due to LV systolic dysfunction or small hypertrophied LV with low SV, resulting in low transaortic flow volume

[18] Radiates to carotids

[19] Murmur intensity grade 3–4 with thrill correlates with severe AS, but lesser intensity does not exclude severe AS

[20] Prominent carotid impulse may occur with severe AS when concurrent carotid atherosclerosis/increased vascular stiffness present

[21] Normal split S2 excludes severe AS
[22] TTE indications in setting of possible AS:

History of bicuspid AV
Single S2
Symptoms consistent with AS
Unexplained systolic murmur

[23] MR common with AS; presence of moderate/severe MR at time of AV replacement surgery associated with poor prognosis
[24] Echo/Doppler also useful for assessing LV diastolic function, estimating PA systolic pressure, diagnosing concurrent valve lesions/other cardiac and aortic abnormalities
[25] Presence of severe CAD may help in determining surgical versus transcatheter AV replacement
[26] Degree of calcification: strong predictor of progression/outcome
[27] Severe AS defined by echo:

Mean gradient >40 mmHg
Peak aortic jet velocity >4.0 m/s
Aortic area <0.6 cm^2/m^2

[28] Aortopathy associated with bicuspid AV including dilated aortic sinuses/proximal ascending aorta and increased risk of Aortic Dissection
[29] ST segment depression present in 80 % of cases with severe AS
[30] Considered appropriate because of shorter life expectancy/longer durability in older adults
[31] Syncope due to AS: sign of serious disease requiring immediate intervention
[32] Clinical risk factors of AS and atherosclerotic CVD are similar
[33] Long murmur duration/late intensity accentuation correlate with severity
[34] LVH may be more maladaptive than beneficial
[35] Consultation / education about physical exertion
[36] Circumferential calcification of ascending aorta due to atherosclerosis; may complicate AV replacement surgery
[37] Valve calcification

[38] AS systolic murmur at LV apex may have blowing quality and be mistaken for concomitant MR (double valve disease) (Gallavardin Effect)
[39] MR associated with increased early/late mortality following TAVR
[40] AF: major predictor of mortality in both surgical/non-surgically treated patients with AS
[41] AV calcium detected in almost 100 % of pts with HOFC (prevalence in HEFC unknown); surgical intervention for AV stenosis often needed
[42] Angina associated with abnormal coronary microvascular function in AS
[43] Balloon aortic valvuloplasty: AS recurrence 80 % after 6–12 months; requires further treatment; does not improve survival
[44] ACC/AHA Guidelines: Aortic Valve Replacement ***

Class I

1. AVR is recommended in symptomatic patients with severe AS (stage D1) with *(Level of Evidence: B)*:
   (a) Decreased systolic opening of a calcified or congenitally stenotic aortic valve; and
   (b) An aortic velocity 4.0 m per second or greater or mean pressure gradient 40 mmHg or higher; and
   (c) Symptoms of HF, syncope, exertional dyspnea, angina, or presyncope by history or on exercise testing.
2. AVR is recommended for asymptomatic patients with severe AS (stage C2) and an LVEF less than 50 % with decreased systolic opening of a calcified aortic valve with an aortic velocity 4.0 m per second or greater or mean pressure gradient 40 mmHg or higher *(Level of Evidence: B)*
3. AVR is indicated for patients with severe AS (stage C or D) when undergoing cardiac surgery for other indications when there is decreased systolic opening of a calcified aortic valve and an aortic velocity 4.0 m per second or greater or mean pressure gradient 40 mmHg or higher *(Level of Evidence: B)*

Class IIa

1. AVR is reasonable for asymptomatic patients with very severe AS (stage C1) with *(Level of Evidence: B)*:

   (a) Decreased systolic opening of a calcified valve;
   (b) An aortic velocity 5.0 m per second or greater or mean pressure gradient 60 mmHg or higher; and
   (c) A low surgical risk.

2. AVR is reasonable in apparently asymptomatic patients with severe AS (stage C1) with *(Level of Evidence: B)*:

   (a) A calcified aortic valve
   (b) An aortic velocity of 4.0 m per second to 4.9 m per second or mean pressure gradient of 40–59 mmHg; and
   (c) An exercise test demonstrating decreased exercise tolerance or a fall in systolic blood pressure (BP).

3. AVR is reasonable in symptomatic patients with low-flow/low-gradient severe AS with reduced LVEF (stage D2) with a(66–68)*(Level of Evidence: B):*

   (a) Calcified aortic valve with reduced systolic opening;
   (b) Resting valve area 1.0 cm^2or less;
   (c) Aortic velocity less than 4.0 m per second or mean pressure gradient less than 40 mmHg;
   (d) LVEF less than 50 %; and
   (e) A low-dose dobutamine stress study that shows an aortic velocity 4.0 m per second or greater or mean pressure gradient 40 mmHg or higher with a valve area 1.0 cm^2or less at any dobutamine dose.

4. AVR is reasonable in symptomatic patients with low-flow/low-gradient severe AS (stage D3) with an LVEF 50 % or greater, a calcified aortic valve with significantly reduced leaflet motion, and a valve area 1.0 cm^2or less only if clinical, hemodynamic, and anatomic data support valve obstruction as the most likely cause of symptoms and data recorded when the patient is normotensive (systolic BP <140 mmHg) indicate *(Level of Evidence: C)*:

(a) An aortic velocity less than 4.0 m per second or mean pressure gradient less than 40 mmHg; and
(b) A stroke volume index less than 35 mL/m^2; and
(c) An indexed valve area 0.6 cm^2/m^2or less.

5. AVR is reasonable for patients with moderate AS (stage B) with an aortic velocity between 3.0 m per second and 3.9 m per second or mean pressure gradient between 20 and 39 mmHg who are undergoing cardiac surgery for other indications. *(Level of Evidence: C)*

Class IIb

1. AVR may be considered for asymptomatic patients with severe AS (stage C1) with an aortic velocity 4.0 m per second or greater or mean pressure gradient 40 mmHg or higher if the patient is at low surgical risk and serial testing shows an increase in aortic velocity 0.3 m/s or greater per year. *(Level of Evidence: C)*

[45] ACC/AHA 2014 Guidelines: Choice of Intervention ***

Class I

1. Surgical AVR is recommended in patients who meet an indication for AVR with low or intermediate surgical risk. *(Level of Evidence: A)*
2. For patients in whom TAVR or high-risk surgical AVR is being considered, a Heart Valve Team consisting of an integrated, multidisciplinary group of healthcare professionals with expertise in VHD, cardiac imaging, interventional cardiology, cardiac anesthesia, and cardiac surgery should collaborate to provide optimal patient care. *(Level of Evidence: C)*
3. TAVR is recommended in patients who meet an indication for AVR who have a prohibitive risk for surgical AVR and a predicted post-TAVR survival greater than 12 months. *(Level of Evidence: B)*

Class IIa

1. TAVR is a reasonable alternative to surgical AVR in patients who meet an indication for AVR and who have high surgical risk for surgical AVR. *(Level of Evidence: B)*

*** Extracted from 2014 AHA/ACC Guideline for the Management of Patients With Valvular Heart Disease (J Am Coll Cardiol 2014;63:e57–e185)

Guidelines

2014 AHA/ACC Guideline for the management of patients with valvular heart disease

J Am Coll Cardiol. 2014;63:e57–e185. http://content.onlinejacc.org/article.aspx?articleid=1838843.

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Medlineplus

ENGLISH

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ESPANOL

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Review: CAD

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Review: LVH

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Transcatheter Replacement: Timing in Symptomatic Patients with Low-Flow Aortic Stenosis

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Eur Heart J. 2014;35:2639–54. http://eurheartj.oxfordjournals.org/content/35/38/2639.

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Am Heart J. 2014;168:150–59. http://www.ncbi.nlm.nih.gov/pubmed/25066553.

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Updates and More

https://clinicalguidecvd.com/asval

Chapter 12
Aortocoronary Saphenous Vein Graft Aneurysm

ICD-10 Code

NS

Alternate Names/Abbreviation

SVGA

Description/Etiology

Abnormal dilatation of SVG post-CABG, most often to RCA, followed in frequency by conduits to LAD, obtuse marginal, and left circumflex coronary arteries [1]

SVGAs have varied clinical presentations; most often:

Chest pain/angina (46.4 %)
Shortness of breath (12.9 %)
AMI (7.7 %)

Almost 1/3 of cases discovered incidentally, often first as abnormalities on routine CXR or at autopsy

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_12,

Etiology – early formation (days-months post-CABG): indefinite, but may be due to SVG infection, undetected varicosities causing intrinsic wall weakness, technical factors related to surgery

Etiology – late formation (average 13 years post-CABG): indefinite, but generally believed that atherosclcerosis, graft endothelial dysfunction, wall stress/ischemia, secondary medial smooth muscle changes may all contribute

Comorbid Conditions

CABG

Demography

M > F
Average age 65–70 years

Pathophysiology

Early development: several possible factors may contribute, including:

Infection of implanted graft
Intrinsic weakness of venous wall
Technical factors relating to conduit harvesting, preparation, and grafting, such as:

Conduit injury
Anastomotic suture disruption
Failure to reverse SVG at time of grafting

Late development: several factors may contribute, including:

SVG atherosclerotic degeneration, causing vessel wall weakening/graft dilatation

Vessel wall ischemia due to disruption of vasa vasorum during harvesting/grafting process
Abrupt change in wall stress when graft undergoes high pressure and pulsatile flow of arterial system
Graft endothelial dysfunction
Changed medial smooth muscle cell orientation in vicinity of valve sites

Signs/Symptoms [2] [3]

BREATHING – DIFF (DYSPNEA)
CHEST – PAIN, EFFORT (ANGINA PECTORIS)
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)
SPUTUM – BLOOD (HEMOPTYSIS)

Differentiation

Other causes of extra-cardiac masses
Progression of native CAD

Complications [5]

Acute Myocardial Infarction
Dysrhythmia
Heart Failure
Hemothorax
Impingement on adjacent structures [4]
Rupture
Severe Sepsis/Septic Shock
Shock

Laboratory

NS

ECG

NS

Imaging

AORTOCORONARY SAPH VEIN GRAPH, SIZE – INCR [7] [6]
CHEST – MASS, EXTRACARD [CHEST X-RAY]
RA, CHAMBER – IMPINGEMENT, EXT

Other Tests

NS

Treatment: Nonpharmacologic [8]

NS

Treatment: Pharmacologic [8]

NS

Treatment: Surgical/Invasive [8]

Surgical resection
Surgical ligation
Revascularization
Coil embolization

Course

Variable

Notes

[1] Aneurysmal dilatation generally defined as 1.5x reference size; mean reported size >60 mm for both SVG true aneurysms (entire vessel wall) and pseudoaneurysms (1–2 layers)
[2] Often asymptomatic and detected incidentally or with complication
[3] Does not include findings due to complications
[4] Especially RA compression, RV compression, fistula formation to RA and RV; also, PA compression, SVC/IVC obstruction; LA compression
[5] SVGAS of any size continue to dilate and may lead to complications
[6] May be negative on coronary angiography, due to intraluminal thrombus
[7] Usually best diagnosed on CT or MRI
[8] Most reports are of surgical and percutaneous intervention but due to rarity of SVGAS, no guidelines established; treatment may depend on presence of symptoms, with some centers using conservative approach in asymptomatic patients

Guidelines

NS

Professional Information

Review

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Case/Images

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Giant Aneurysm: Image

J Am Coll Cardiol. 2009;54:1899. http://content.onlinejacc.org/article.aspx?articleid=1140176.

Image

N Engl J Med. 2001;344:1139. http://www.nejm.org/doi/full/10.1056/NEJM200104123441505.

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Updates and More

https://clinicalguidecvd.com/asvga

Chapter 13
Arrhythmogenic Right Ventricular Dysplasia (ARVD/Naxos Disease)

Management Keys [27]

Sudden death prevention strategy after diagnosis, especially:

- Antiarrhythmics
- Beta-blockers
- ICD (including early concealed phase)
- Physical exercise restriction

- Diagnose/treat only in centers highly experienced with ARVD
- Diagnose using task force major/minor criteria
- Consider ARVD in any young person (especially adolescents) with palpitations, syncope, aborted SCD
- Screen family members of patients with ARVD [23]
- Differentiate from Athlete Heart in young athletically active persons
- Counsel patients/asymptomatic family members regarding exercise/competitive athletics

ICD-10 Code

I42.8

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 191
DOI 10.1007/978-1-4471-7293-2_13,

Alternate Names/Abbreviation

ARVD
ARRHYTHMOGENIC RIGHT VENTRICULAR CARDIOMYOPATHY
NAXOS DISEASE (RECESSIVE FORM)

Description/Etiology [27]

Genetic autosomal dominant desmosomal cardiomyopathy [2][26]
Right ventricular (sometimes extends to LV) fibro-fatty infiltration; clinically manifest by ventricular arrhythmias and progressive ventricular dysfunction
Typical presentation: age 20–40 years with stable monomorphic VT originating in RV
Myocardial cell separation may be increased by exercise, sometimes causing unexpected SCD in young athletes, which may be 1st clinical manifestation
Highly variable disease expression, even in same family
Sudden death risk factors:

Competitive sports
LV Involvement
Malignant family history
Previous cardiac arrest
QRS Dispersion 40 msc
Severe RV dysfunction
Syncope
VT
Young age

Comorbid Conditions

FAMILY HX: ARVD [2][26]
FAMILY HX: SUDDEN DEATH [20]
INFECTION [POSSIBLE]
INFLAMMATION [POSSIBLE]
UHL ANOMALY [POSSIBLE MILD FORM]

Demography

Mean age of 1st diagnosis about age 30 years; rare <12 years/>60 years
M > F
1/1,000–5,000 in Caucasians (may be low estimate due to underreporting)

Pathophysiology

Abnormal desmosome function [16]
Progressive replacement of RV myocardium by adipose and fibrous tissue:

- Beginning in epicardium/mid-myocardium, then becoming transmural
- Progressing to wall thinning/aneurysms, most often RV inferior, apical, infundibular walls

Phases:

- Early concealed (subclinical): asymptomatic but at risk for SCD, especially with exercise; morphological abnormalities present
- Electrical: changes localized to RV inflow/outflow tracts, apex ("triangle of dysplasia"); symptomatic reentrant ventricular dysrhythmias, abnormal electrical conduction with epsilon waves, RBBB, late potentials
- RV failure: diffuse RV abnormalities and extension to LV (50 % of cases; typically posterior lateral wall)
- Biventricular failure

Signs/Symptoms

BREATHING – DIFF (DYSPNEA)
CHEST – PAIN
CHEST – PALPITATIONS
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)

DIZZY/LIGHTHEADED/PRESYNCOPE
EXTREM, LOWER, BILAT – EDEMA
HAIR – THICK, WOOL-LIKE [14]
HEART, S4 LV
NAUSEA
SKIN, PALMS/SOLES – THICK (HYPERKERATOSIS) [14]
SWEATING – INCR (DIAPHORESIS/HYPERHIDROSIS)
VOMITING (EMESIS)

Differentiation

Athlete Heart
Atrial Septal Defect
Brugada Syndrome
Cardiac Sarcoidosis
Cardiomyopathy – Dilated (Idiopathic)
Chagas Disease
Ebstein Anomaly
Idiopathic RVOT VT [18]
Left-right shunts (Especially Partial Anomalous Venous Return, Atrial Septal Defect)
Mahaim-Pre-excited AV Reentry Tachycardia
Myocarditis
Other causes of dysrhythmias
Other causes of RH disease[5]
Other causes of syncope
Pulmonary Arterial Hypertension
Uhl Disease/Anomaly [24]

Complications

LV Failure [4]
Myocarditis
RV failure [1]
SCD [20]
VT [3]

Laboratory

NS

ECG [12] [19]

DYSRHYTHMIAS – ATRIAL (PACS/OTHERS)
DYSRHYTHMIAS – VENTRICULAR (PVCS/OTHERS) [17] [25]
EPSILON WAVE [6] [25] [R PRECORD LEADS]
J WAVE (OSBORN WAVE)
QRS – LONG, NS [7]
QRS – RBBB/RBBB PATTERN
QRS, S WAVE UPSTROKE – LONG [V1-3; >55 MS]
T WAVE – INVER, ABN [25] [R PRECORDIAL LEADS]

Imaging [22]

EPICARD, FAT – INCR (MRI, EBCT)
LV, MYOCARD – LGE [23]
MYOCARD – FAT DEPOSITS (MRI, EBCT)
RV – PROMINENT MODERATOR BAND (RV ANGIO)
RV, CHAMBER, SIZE – INCR [SEG]
RV, CONTRACTION – DYSSYNCHRONY
RV, EF – DECR [25]
RV, INFUNDIBULUM – ANEURYSM [ECHO/RV ANGIO]
RV, MYOCARD – LGE [23]
RV, OUTFLOW TRACT, SIZE – INCR [25]
RV, TRABECULAE – THICK [RV ANGIO]
RV, WALL MOTION – DECR/AKINETIC [8][25]
RV, WALL MOTION – DYSKINETIC [25] [RV ANEURYSM]
TV, LEAFLET, MOTION – PROLAPSE [9]

Genomics [15]

FKBP12
GNB3 [17]
RYR2
DSC2 [28]
DSG2 [28]
DSP (DESMOPLAKIN) [25] [28]
JUP [28]
PKP2 [25] [28] [30–50 % OF CASES]
PLN
TGFBETA3
TMEM43

Other Tests

Ambulatory ECG monitoring [19]
Cardiac angiography [21]
Endomyocardial biopsy
EP testing: differentiate from Idiopathic RVOT Ventricular Tachycardia
Signal-averaged ECG

Treatment: Nonpharmacologic

Avoidance of strenuous physical activity

Treatment: Pharmacologic [10] [27]

Amiodarone
Beta-blockers
Sotalol

Treatment: Surgical/Invasive

ICD [11] [27]
RF Ablation
Cardiac transplantation [13]

Prevention

Family screening [23]

Course

Highly variable
Except for earlier onset in persons with mutations, course similar for persons with/without mutations
ICD decreases SCD

Notes

[1] May be clinical presentation
[2] Autosomal dominant with variable penetrance and incomplete expression in usual form; autosomal recessive in Naxos Disease
[3] With LBBB due to RV source
[4] Uncommon
[5] Especially RHF
[6] Characteristic of ARVD; due to delayed RV activation
[7] RV leads
[8] Hypokinetic and dyskinetic areas of RV may be localized
[9] Severe cases
[10] Should be undertaken only in centers highly experienced with this disease; inadequate data in asymptom-

atic persons or those without arrhythmias; class 1a, 1b, 1c antiarrhythmics less effective

[11] Indicated in persons with prior sustained VT/VF; role for primary prevention in ARVD patients with no prior episodes controversial; take precautions in implanting into RV, which may be penetrated; sensing may be abnormal

[12] Abnormal mainly in right precordial leads

[13] Consider when refractory to ICD

[14] Naxos Disease

[15] Reported frequency of mutation types varies among country populations studied

[16] Desmosomes: provide mechanical cell-cell adhesion, structural cardiac support, cardiac electrical integrity (sodium current/conduction velocity)

[17] Especially:

Frequent PVCs
Sustained/nonsustained VT with LBBB morphology/superior axis
Nonsustained VT of RVOT configuration with LBBB morph, inferior/unknown axis

[18] Typical:

RVOT VT: 1 morph (LBBB with inferior axis)
ARVD: multiple RVOT VT morphologies (LBBB with superior axis)

[19] ECG abnormalities (resting/ambulatory) highly sensitive for ARVD

[20] Mechanism: VF in young persons/athletes (often asymptomatic); VF rare in older patients

[21] >90 % sensitive for detecting akinesis/dyskinesis of RV infundibulum, apex, subtricuspid region

[22] Noninvasive imaging (echo/Doppler first-line) highly useful for both initial diagnosis and follow up for disease progression

[23] In patients with ARVD genotype: RV LGE highly specific/>50 % sensitive; LV LGE highly sensitive/>50 % specific

[24] Especially RHF in newborns/young children
[25] DSP mutation carriers: 4x LV dysfunction/HF incidence than PKP2 carriers in 1 study
[26] 1/3 of 1st degree relatives develop overt ARVD, with siblings at highest risk
[27] 2012 ACCF/AHA/HRS Focused Update Incorporated Into the ACCF/AHA/HRS 2008 Guidelines for Device-Based Therapy of Cardiac Rhythm Abnormalities (J Am Coll Cardiol 2013; 61:e6–75)

Arrhythmogenic Right Ventricular Dysplasia/ Cardiomyopathy

Arrhythmogenic RV dysplasia/cardiomyopathy (ARVD/C) is a genetic condition characterized by fibrofatty infiltration of the RV and less commonly the LV. It usually manifests clinically with sustained monomorphic VT with left bundle morphology in young individuals during exercise. There are no prospective randomized trials of pharmacological therapy versus ICD therapy in patients with ARVD/C for secondary prevention of SCD; however, observational reports from multiple centers consistently demonstrate a high frequency of appropriate ICD use for life-threatening ventricular arrhythmias and a very low rate of arrhythmic death in patients with ARVD/C treated with an ICD

[28] Increasingly found in pts with DCM as well

Guidelines

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Updates and More

https://clinicalguidecvd.com/arvd

Chapter 14
Athlete Heart

Management Keys

- Reassurance after excluding other causes of myocardial hypertrophy
- Careful evaluation of any athlete having syncopal episode during (not after) exercise, which is indicator of pathology
- Consider using ECG when screening athletes for CVD [18]

ICD-10 Code

I51.7

Alternate Names/Abbreviation

ATHLETIC HEART SYNDROME

Description/Etiology

Exercise training-induced changes in CV system causing increased LV SV at rest and during exercise

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 205
DOI 10.1007/978-1-4471-7293-2_14,

Comorbid Conditions

LEFT VENTRICULAR HYPERTROPHY

Demography

Competitive athletes

Pathophysiology

Isotonic (endurance) exercise: increased cardiac four chamber volume allowing sustained increase in cardiac output due to increased LVSV

Isometric (strength) exercise: increased peripheral vascular resistance with normal/near normal cardiac output and markedly increased systolic BP and LV afterload

Signs/Symptoms

CHEST – PALPITATIONS
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE) [10]
HEART, LSB, MID – MURMUR, SYS
HEART, LV, APEX – IMP, LAT DISPLACEMENT [1]
HEART, LV, APEX – SOUND, MID DIAS
HEART, LV, APEX, IMP – FORCEFUL/SUSTAINED [1]
HEART, LV, APEX, IMP – FORCEFUL/SUSTAINED [1]
HEART, RATE – SLOW (BRADYCARDIA) [3]
HEART, RHYTHM – IRREG
HEART, S3 LV
HEART, S4 LV

Differentiation

Arrhythmogenic Right Ventricular Dysplasia
Brugada Syndrome (Early Repolarization)

Cardiomyopathy – Hypertrophic [11]
Cardiomyopathy – Noncompaction (spongy myocardium)
Other causes of bradyarrhythmias
Other causes of LVH

Complications

Atrial Fibrillation [14]

Laboratory

NS

ECG [18]

AV COND – 1ST DEGREE BLOCK [3][4] [15]
AV COND – 2ND DEGREE BLOCK, MOBITZ I (WENCKEBACH)
DYSRHYTHMIAS – ATRIAL (PACS/OTHERS) [8]
DYSRHYTHMIAS – VENTRICULAR (PVCS/OTHERS) [8]
HEART,RATE–DECREASED (SINUSBRADYCARDIA) [3] [15]
J POINT – ELEV [17]
J WAVE (OSBORN WAVE) [12]
P WAVE, DUR – INCR, NOTCHED (P MITRALE) [16]
P WAVE, MORPH – VAR/ABN [2]
PR INTERVAL – LONG <1ST DEGREE BLOCK [3] [4]
PR INTERVAL – SHORT [16]
Q WAVE – ABN [16]
QRS – LBBB/LBBB PATTERN [16]
QRS – RBBB/RBBB PATTERN [16] [COMPLETE RBBB]
QRS – RBBB/RBBB PATTERN [15] [INCOMPLETE RBBB]

QRS – RVH PATTERN [16]
QRS, AMP – INCR [15]
QRS, AXIS – L [16]
QRS, AXIS – R [16]
QT INTERVAL – SHORT [16]
QT/QTC INTERVAL – LONG [16]
SINUS ARRHYTHMIA [3]
ST SEGMENT – DEPR [16]
ST SEGMENT – ELEV [5][9][17]
T WAVE – INVER, ABN [9] [16][17]
T WAVE – TALL/PEAKED [6]
VOLTAGE, GEN – INCR [9]

Imaging

CARDIOMEGALY [MILD]
LV, CHAMBER, SIZE – INCR [MILD]
LV, MYOCARD, WALL THICKNESS – INCR (HYPERTROPHY) [7][9]
MV, FLOW – REGURG [MILD]
TV, FLOW – REGURG [MILD]

Other Tests

Ambulatory ECG monitoring [8]

Treatment: Nonpharmacologic

NA

Treatment: Pharmacologic

NA

Treatment: Surgical/Invasive

NA

Course

Benign

Notes

[1] Associated more often with isometric conditioning
[2] Wandering atrial pacemaker
[3] Attributed to increased vagal tone
[4] Normalizes or shortens after exercise
[5] V2-4; normalizes with exercise; reciprocal changes in inferior leads
[6] Correlates with ST segment elevation
[7] <1.7 CM
[8] PACS/PVCS common
[9] More common/pronounced in African-American athletes compared to other ethnicities
[10] Immediately post-exercise only and attributed to neurocardiogenic mechanisms due to sudden decrease in venous return; syncope during exercise should be attributed to pathology
[11] In "gray zone" of LV thickness 13–15 mm, following are useful:

LV internal dimension: >54 mm in athlete
LA internal dimension: <40 mm in HCM; >39 mm in athlete
LV filling/relaxation: abnormal in HCM; normal in athlete
ECG diffuse T wave inversion: present in HCM; absent in athlete
Family history of HCM: positive in HCM; absent in athlete

[12] Common in highly trained athletes, usually associated with other ECG changes, and considered benign without increased risk for adverse events, including SCD/tachyarrhythmias

[13] RVH voltage criteria common in athletes and alone not associated with pathology

[14] High level athletic training associated with AF due to:

LA enlargement
Repetitive extreme hemodynamic loads
Vagal tone increase

[15] Occur in up to 80 % of athletes and deemed normal physiological variants, mostly due to increased vagal tone

[16] Occur in <5 % of athletes and warrant further investigation to exclude cardiac disease, specifically cardiomyopathy and ion channel disorders

[17] Repolarization changes:

>60 % of athletes
J-point elevation in most leads associated with concave ST segments commonly found in slender males
Particularly common in people of African/Afro-Caribbean origin; in latter group: ST segments in V1-V4 may be convex and associated with biphasic T waves or asymmetrical and occasionally deep T-wave inversion, seen in up to 13 % of African/Afro-Caribbean athletes, including women and adolescent athletes of both sexes

[18] ECG may be most effective screening strategy for CVD in athletes, with:

Five times more sensitive than history
Ten times more sensitive than physical exam
Higher positive likelihood ratio
Lower negative likelihood ratio
Lower false positive rate

Guidelines

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Updates and More

https://clinicalguidecvd.com/ath

Chapter 15
Atrial Fibrillation

Management Keys

Calculate CHA_2DS_2-VASc score for thromboembolic risk [28]

Diagnose underlying/precipitating/treatable causes, eg, Hyperthyridism/Graves Disease, electrolyte abnormalities, alcohol use, as part of initial evaluation

Follow Guideline-directed antithrombotic treatment for specific patients/clinical issues:

- General
- AF with prosthetic valves
- AF requiring coronary intervention
- Atrial Flutter
- Nonvalvular AF
- Warfarin
- Other anticoagulation

Follow Guideline- directed arrhythmia treatment for specific patient groups:

- AMI
- Familial AF
- HCM
- HF
- Hyperthyroidism
- Post-operative cardiac/thoracic surgery

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_15,

Pulmonary disease
WPW/Preexcitation syndromes

Make shared decisions with patient/family for antithrombotic therapy, including stroke/bleeding risks, patient preferences
Perform TEE as part of initial evaluation to detect LA/LAA thrombus, underlying structural heart disease, and to assess cardiac function/LA size
Select antithrombotic therapy based on thromboembolism risk rather than AF classification (paroxysmal, persistent, permanent)
Take special precaution when using antiarrhythmics (especially sotalol, dofetilide, disopyramide, quinidine) that may cause QT prolongation/torsade de pointes [30]
Life-long monitoring/risk reduction for stroke and other complications, regardless whether AF is idiopathic or caused by secondary precipitant
Pay attention to patient symptoms to optimize QOL [52]

ICD-10 Code

I48.0 Paroxysmal
I48.1 Persistent
I48.2 Permanent

Alternate Names/Abbreviation

AF
A Fib

Description/Etiology

Very common supraventricular tachyarrhythmia with uncoordinated atrial activation/ineffective atrial contraction due to structural/electrophysiological abnormalities of

atrial tissue promoting abnormal impulse formation/propagation
Often associated with rapid ventricular response rates
Causes:

Usually due to secondary precipitants (see COMORBIDS) [35]
Up to 30 % idiopathic
Familial form (s)

Classification:

Paroxysmal: spontaneous termination/conversion within 7 days of onset
Persistent: duration >7 days
Long-standing persistent: duration >12 months
Permanent: patient/clinician decide to cease efforts to restore/maintain sinus rhythm
Nonvalvular: absence of MS, mechanical/bioprosthetic valve, MV repair
Lone AF: refers to patients age <60 years with AF and no known heart disease

Clinical appearance often associated with exacerbation of underlying heart disease
Associated with increased risk of stroke/peripheral emboli from atrial thrombus, usually within LAA [26]
Risk of stroke determined by CHA_2DS_2-VASc score [28]
Often associated with other arrhythmias, including:

Atypical/typical atrial flutter
Focal Atrial Tachycardia
Multifocal Atrial Tachycardia

Comorbid Conditions

ACUTE INFECTION [ESP BRONCHITIS, PNEUMONIA]
ACUTE MYOCARDIAL INFARCTION
ACUTE PULMONARY EMBOLISM

ALCOHOL USE/EXCESS
ANEMIA
ANEURYSMS-OSTEOARTHRITIS SYNDROME
ANXIETY/ANXIETY DISORDER
AORTIC STENOSIS [52][54]
ARTHRITIS
ATHLETE HEART [38]
ATRIAL SEPTAL DEFECT – SECUNDUM [PRIMUM/ SECUNDUM]
ATRIAL SEPTAL INFARCTION
CANCER
CARDIAC INVASIVE PROCEDURES
CARDIAC TRAUMA
CARDIAC TUMOR (S)
CARDIOMYOPATHY – DILATED
CARDIOMYOPATHY – HYPERTROPHIC
CARDIOMYOPATHY – PERIPARTUM
CARDIOMYOPATHY – RESTRICTIVE [13]
CARDIOTHORACIC SURGERY
CATARACT
CHRONIC KIDNEY DISEASE
CHRONIC OBSTRUCTIVE PULMONARY DISEASE (EMPHYSEMA) [47][49]
COR TRIATRIATUM
CORONARY ARTERY DISEASE
DEMENTIA
DIABETES MELLITUS
DIPHTHERIA
DRUGS: CANCER THERAPY [21]
DRUGS: STIMULANTS [2]
DRUGS; CHOLINERGIC
DYSLIPIDEMIA
EMPYEMA
EXERCISE INTOLERANCE
FABRY DISEASE
FATIGUE – EXTREME
FEVER

FRAILTY
HEAD INJURY
HEART FAILURE [22]
HEART TRANSPLANT
HIV
HYPERTENSION – SYSTEMIC ARTERIAL [1]
HYPERTHYROIDISM
HYPOCALCEMIA
HYPOGLYCEMIA
HYPOKALEMIA
HYPOMAGNESEMIA
HYPOTHERMIA
HYPOXEMIA
INFLUENZA – SEASONAL [57]
LEFT VENTRICULAR HYPERTROPHY
METABOLIC SYNDROME
METABOLIC TOXICITY
MITRAL ANNULUS CALCIFICATION
MITRAL REGURGITATION
MITRAL STENOSIS – ACQUIRED
MITRAL STENOSIS – CONGENITAL
MITRAL VALVE PROLAPSE
MUSCULAR DYSTROPHY
MYOCARDIAL INFILTRATIVE DISEASES
MYOCARDITIS
MYXOMA – LEFT ATRIUM
NONTHORACIC SURGERY
OBESITY [51]
OBSTRUCTIVE SLEEP APNEA [37][49]
PAPILLARY FIBROELASTOMA
PATENT DUCTUS ARTERIOSUS
PERICARDITIS [16]
PHEOCHROMOCYTOMA
PNEUMONIA – COMMUNITY-ACQUIRED
POST-THORACOTOMY
PREEXCITATION SYNDROMES
PRIMARY ALDOSTERONISM

PULMONARY INFECTION
RENAL DIALYSIS
SECOND HAND SMOKE EXPOSURE [ESP IN UTERO/CHILDHOOD
SEVERE SEPSIS (SEPTIC SHOCK) [55]
SINUS NODE DYSFUNCTION
SPINAL CORD INJURY
STABLE ISCHEMIC HEART DISEASE
SUBSTANCE ABUSE – STIMULANTS [2]
SUPRAVENTRICULAR TACHYCARDIA
THORACIC SURGERY
TOBACCO USE
TRICUSPID REGURGITATION
TRICUSPID STENOSIS
TYPHOID FEVER

Demography

Aging: increased incidence
M > F
Ethnicity: more common in whites compared to blacks, Asians, Hispanics

Pathophysiology

Progressive disease process in most affected persons
Most often structural abnormalities of atrial architecture due to any disease process that causes:

- Altered wall stress
- Atrial dilatation
- Increased LA press
- Increased LA stiffness

Atrial histopathology includes:

- Fibrosis [45]
- Hypertrophy

- Inflammation
- Thrombus formation in LAA [36]

Pathophysiologic mechanisms (understanding incomplete and continues to evolve) involving initiation and maintenance including:

- Electrical/structural remodeling
- Fibrosis: atrial/ventricular [45] [53]
- Focal discharges, often from pulmonary veins
- Inflammation/oxidative stress
- RAAS stimulation
- Reentry
- Risk factors/associated heart disease
- Sympathetic nervous system

Atrial electrical wavelets with multiple reentry and patterns of rapid and irregular wave propagation

Altered secondary hemodynamics due to: [23]

- Beat-beat variation in ventricular filling
- Suboptimal ventricular rate control
  - Too rapid
  - Too slow
- Sympathetic activation
- Uncoordinated atrial contraction

Marked decreased in cardiac output may occur, especially when associated with:

- HCM
- Mitral Stenosis
- Restrictive Cardiomyopathy
- Systemic Arterial Hypertension

LA dilatation occurs in >70 % of cases

Thrombus formation due to sluggish blood flow in LA – especially LAA (>90 % localized in LAA) – as source of peripheral emboli; inflammatory/procoagulable state of AF/AF comorbidities may also contribute

LV systolic dysfunction common in AF [43] [53]

Signs/Symptoms [3] [20] [24] [50]

ARTERIAL PULSE–DEFICIT [12]
ARTERIAL PULSE, AMP–DECR/ABS
ARTERIAL PULSE, AMP–VAR
BREATHING–DIFF (DYSPNEA)
CHEST–PAIN
CHEST–PALPITATIONS [MOST COMMON SYMP]
COGNITION–DEFECT, NS [ESP HF]
CONSCIOUSNESS–LOSS, SUDDEN (SYNCOPE)
DIZZY/LIGHTHEADED/PRESYNCOPE
FATIGUE
HEART, LV, APEX–MURMUR, SYS
HEART, RATE–RAPID (TACHYCARDIA)
HEART, RHYTHM–IRREG [19]
HEART, S1, INTENSITY–VAR
HEART, S4–ABSENT
HYPOTENSION (BLOOD PRESSURE–DECREASED/LOW)
NECK, JVP, A WAVE–ABSENT
NECK, JVP, PULSATIONS–IRREG
URINATION–INCR (POLYURIA)

Differentiation

Atrial Flutter
ECG artifact [17]
Multifocal Atrial Tachycardia
Paroxysmal Atrial Tachycardia with Block
PACs (frequent)
PVCs (frequent)
VT

Complications [14]

Acute Pulmonary Embolism
Atrial Flutter
Cognitive impairment [11]
HF [6]
Ischemic Stroke [6] [24] [33] [39]
Mesenteric ischemia/bowel infarction
Myocardial ischemia
Peripheral embolization [9] [24] [33]
Renal dysfunction [46]
Tachycardia-Induced Cardiomyopathy
Ventricular arrhythmias [30]

Laboratory [24] [27]

BLOOD, CREATININE CLEARANCE (EST) (CCR) – DECR [46]
BLOOD, GALACTIN-3 – INCR [32] [HFpEF]
BLOOD, NT-PROBNP – INCR [25]
BLOOD, TROPONIN – INCR

ECG [4] [24]

FIBRILLATION WAVES, ATRIAL
P WAVE – ABSENT
QRS – LONG, NS [18]
RR INTERVAL – IRREG [19]
VENTRIC RESPONSE, RATE – VAR

Imaging [5] [20] [24]

ART, CORONARY – CALCIUM [31]
LA – FIBROSIS [45]

LA, CHAMBER, SIZE – INCR [CHRONIC A FIB] [40]
LA, EMPTYING FUNCTION – DECR [42]
LA, INTRACAVITY – MASS [THROMBUS] [26]
LAA – THROMBUS [26]
LV, DIAS – DYSF [44]
LV, SYS – DYSF [43]

Genomics

CAV1
KCNA5
KCNE2
KCNJ2
KCNQ1
NPPA
NUP155
SCN5A [SNP: rs6800541]

Other Tests

EP: for detecting trigger of AF, including;

SVT

AV node reentrant tachycardia
AV reentry with accessory pathway
Ectopic atrial tachycardia

Preexcitation (delta wave on ECG)

Also use EP for:

Detecting A Flutter
Differentiating AF from other dysrhythmias, especially AF with wide-complex from VT

Ambulatory ECG monitoring
Cognitive impairment testing (older patients) [11]
Stress test

Treatment: Nonpharmacologic

- Ablation of accessory or AV nodal pathways by RF/catheter ablation [29] [58]
- Direct current cardioversion [8] [59]
- CPAP for patients with OSA [47]

Treatment: Pharmacologic [73]

- Antithrombotic therapy: related only to risk, not AF classification [60]
- Prevention of thromboembolism: Calculate CHA_2DS_2-VASc score for thromboembolic risk; treat when score >1 or when prior stroke/TIA occurred with anticoagulation therapy [28]
- Guideline-directed patient-specific categories:
  - Nonvalvular AF: stroke risk assessment CHA_2DS_2-VASc [61]
  - Mechanical heart valve [62]
  - Patients treated with warfarin [63]
  - Bridging treatment for warfarin/new anticoagulants (interruption for procedure) [64]
  - Direct thrombin/factor Xa inhibitors: renal function [65]
  - Atrial Flutter [66]
  - Nonvalvular AF/CHA_2DS_2-VASc 0–1 [67]
  - Nonvalvular AF/CHA_2DS_2-VASc ≥ 2, or prior stroke, TIA [68]
  - Nonvalvular AF/CHA_2DS_2-VASc ≥ 2 and CKD/hemodialysis [69]
  - AF patients undergoing PCI [70]
  - Post-PCI [70]
- Antithrombotic options:
  - Antiplatelet agents
    - ASA
    - Clopidogrel

Direct thrombin inhibitors

- Bivalrudin
- Dabigatran

Heparin

Injectable direct factor Xa inhibitors

- Fondaparinux

Oral factor Xa inhibitors [15]

- Apixaban
- Edoxaban
- Rivaroxaban

Warfarin

Rate control: Guidelines [71]

- Amiodarone oral/IV
- Beta-blockers
- Digoxin [34]
- Non-dihydropyridine CCB [10]

Rhythm control [7] [73]:

- Pharmacological cardioversion [72]
- Maintenance
- Amiodarone
- Beta-blockers
- Disopyramide [30]
- Dofetilide (in hospital only) [30]
- Dronedarone
- Flecainide
- Ibutilide (IV)
- Propafenone
- Propafenone or flecainide + Beta-Blocker or non-dihydropyridine CCB
- Quinidine [30]
- Sotalol [30]

Thromboembolism prevention with electrical or pharmacological cardioversion

≥48 h/unkn duration and no anticoagulation preceding 3 weeks [74]
≥48 h/unkn duration: elective (warfarin) [75]
≥48 h/unkn duration: elective (dabigatran/rivaroxaban/apixaban) [76]
>48 h/unkn duration: immediate [77]
<48 h/unkn duration and low thromboembolic risk [78]
<48 h/high stroke risk [79]
Post cardioversion: long-term based on thromboembolic risk [80]

Treatment: Surgical/Invasive

LAA closure: no current guidelines [48]; possible candidates include:

Contraindication to NOACC/warfarin
Persistent noncompliance to NOACC/warfarin

Ablation [29] [41] [50] [58]

Catheter
Surgical (Maze procedures) [81]

Pacemakers/ICDs: treatment of symptomatic bradycardia, often related to Sick Sinus Syndrome (Sinus Node Dysfunction)

Prevention

Aerobic training [56]
Influenza vaccination [57]
Maintain/lose to optimal body weight [51]
Optimal cardiorespiratory fitness except competitive athletics [38]

Upstream treatment: [82]

HFrEF: ACE/ARB
Post-CABG: statin
Systemic Arterial Hypertension: ACE/ARB

Course

When untreated, 50 % of first episodes resolve spontaneously within 48 h of onset

Recurs in most patients, including those with secondary precipitants, with similar long-term risk for stroke and mortality in idiopathic/secondary precipitants

Notes

[1] Most common associated condition
[2] Especially alcohol
[3] Varies according to causes and complications
[4] May be normal if AF is paroxysmal
[5] Useful in search for underlying cardiac disease, especially MV disease and atrial thrombus
[6] Embolism from atrial thrombus or noncardiac site, eg carotid artery, aorta, cerebral vessels
[7] Other medications that have been used, but carry increased risk and not approved in USA for this indication, include quinidine, procainamide, disopyramide, flecainide, and propafenone
[8] Especially if hemodynamic instability present, eg HF, hypotension, myocardial ischemia
[9] Rarely occur after spontaneous/induced cardioversion
[10] Anti-tachycardia type
[11] Mild cognitive decline occurs in up to two-thirds of older pts hospitalized for chronic AF
[12] Ashman phenomenon: aberrancy of beat ending a short ventricular cycle that is immediately preceded by a long cycle
[13] EG, Amyloidosis, Sarcoidosis, Hemochromatosis

[14] Hospitalizations for AF as primary diagnosis are significantly increasing in USA and expected to continue to increase in coming decades
[15] Non-valve AF
[16] Especially chronic constrictive form
[17] EG, Parkinson tremor
[18] Due to aberrant conduction, which occurs frequently in AF
[19] Sometimes regular
[20] Other abnormalities may be present due to associated/causative condition (s)
[21] Include but not limited to anthracyclines, cisplatin, melphalan, interleukin-2
[22] AF is both a trigger for and predictor of HF progression; occurs in >50 % of patients with HF
[23] After restoration of normal sinus rhythm atrial mechanical function may not recover due to remodeling/underlying atrial disease
[24] Features of causative/associated diseases (eg, valvular heart disease, CAD, Hyperthyroidism) not included and should be sought in evaluation of patients with AF
[25] Increased in absence of HF and rapidly decreases after restoration of normal sinus rhythm
[26] TEE: most sensitive/specific modality for detecting LA/LAA thrombus, which is present in 5–15 % of patients prior to cardioversion
[27] Lab evaluation should also include tests for secondary causes (eg, electrolytes, thyroid/renal/hepatic function, blood count)
[28] CHA_2DS_2-VASc score

C Congestive HF/LV dysf	1 point
H Hx HTN	1
A Age 65-74 yrs	1
D Diabetes mellitus	1
S Stroke/TIA Hx	1
V Vasc dis (any territory)	1
A Age 65-74 yrs	2
Sc Sex - F	1

[29] AV node ablation usually reserved for older patients because causes pacemaker dependency; complications include:

Acute Myocardial Infarction
Air embolism
Atrial-esophageal fistula
Cardiac tamponade/perforation
Death
Iatrogenic atrial flutter
Gastric motility dis
MV injury
Pericarditis
Phrenic nerve injury/diaphragmatic paralysis
Pulmonary vein stenosis
Radiation injury
Stroke/TIA
Vascular access complications:
  A-V fistula
  Femoral pseudoaneurysm
  Hematoma

[30] High risk of QT prolongation with Torsades de Points; contraindicated in patients with:

Congenital/Acquired Long QT Syndrome
Baseline QT/QTc >440 msc
Severe renal impairment
Hypersensitivity to dofetilide

Other risk factors for Torsades:

Advanced age
Baseline long QT
Bradycardia
Female gender
HF
Hypokalemia
Hypomagnesemia
LVH (possible)
Other QT-prolonging drugs

[31] Independent association with risk for AF
[32] Galactin-3 serum level increase correlates with LA volume index in pts with AF and HFpEF
[33] In patients with HF, stroke/peripheral emboli unrelated to symptom status or LV dysfunction
[34] Digoxin should be used with caution, possibly only in select cases due to increased risk of mortality (also in HF)
[35] Most common secondary precipitants in Framingham Study:

Recent cardiothoracic surgery (30 %)
Acute infection (23 %)
Noncardiothoracic surgery (20 %)
Acute Myocardial Infarction (18 %)

[36] Thrombus formation in LAA:

Due to decreased LA blood flow
Found in 5–14 % of patients with AF lasting 2 or more days
Size up to 40 mm
75 % resolve after 1 month of anticoagulation therapy

[37] OSA association with AF is multifactorial, including:

Atrial stretch
Autonomic imbalance
Inflammation
Oxidative stress

[38] High level athletic training associated with AF due to:

LA enlargement
Repetitive/extreme hemodynamic loads
Vagal tone increase

[39] Strokes related to AF have poorer outcomes than non-AF related stroke, including worse functional impairment, recurrence, death
[40] LA dimension predictive of incident AF in Framingham Study; LA volume index strong predictor of stroke

[41] Ablation associated with transient effect on GI motility, probably due to interruption of vagal nerve function
[42] LA emptying function abnormalities may precede structural abnormality in pathophysiology of AF; may be measured by echo, but CT and CMR more accurate
[43] LV systolic dysfunction common and may cause or be secondary effect of AF
[44] LV diastolic dysfunction commonly associated with AF due to increased load on LA, but whether association is independent is unresolved
[45] Atrial fibrosis correlates with AF persistence/burden; may contribute to stroke risk; detected by delayed enhancement CMR
[46] Renal function often decreased, progressively with anticoagulation therapy, more so with warfarin than dabigatran
[47] CPAP: effective in preventing AF recurrence in patients with OSA
[48] LAA closure may be associated with improved rates of hemorrhagic stroke, CV/unexplained death, nonprocedural bleeding compared to warfarin, but exact role indefinite
[49] Reduced lung function associated with increased risk of AF
[50] Gastroparesis is reported complication, manifest as:

Abdominal/epigastric discomfort
Bloating
Nausea
Vomiting

[51] Sustained obesity: associated with increased propensity for AF via global biatrial endocardial remodeling with:

LA enlargement
Conduction abnormalities
Fractionated electrograms
Increased profibrotic TGF-ß1 expression
Interstitial atrial fibrosis

Every 5 unit increase in BMI associated with 10–29 % increased risk of incident, post-operative, post-ablation AF

[52] In ORBIT-AF:

61.8 % of pts with A Fib symptomatic
16.5 % of pts had severe/disabling symptoms
Symptoms associated with increased risk of hospitalization/borderline increased risk of major bleeding/not death
Most common symptoms:

Palpitations (32.7 %)
Dyspnea with exertion (27.6)
Fatigue (26.4)
Lightheadedness/dizziness (20.6)
Dyspnea at rest (10.3)
Exercise intolerance (10)
Chest tightness/discomfort (9.4)
Syncope/fainting (4.5)

[53] Ventricular fibrosis also often present and may play a role in frequent comorbidity of AF and HF

[54] AF: major predictor of mortality in both surgically/nonsurgically-treated patients with AS

[55] AF occurs in up to 25 % of patients with severe sepsis/septic shock and may be associated with worse short/long term outcomes

[56] Aerobic training: may shorten duration of AF episodes in patients with nonpermanent AF, improvement in AF symptoms, peak VO2, LA/LV function, lipid levels, QOL

[57] Influenza: 18 % increase risk of AF, which could be reduced through influenza vaccination

[58] **AF Catheter Ablation to Maintain Sinus Rhythm: Recommendations** ***
Sec. 6.3
Class I

1. AF catheter ablation is useful for symptomatic paroxysmal AF refractory or intolerant to at least 1 class I or III antiarrhythmic medication when a rhythm control strategy is desired. (Level of Evidence: A)

2. Prior to consideration of AF catheter ablation, assessment of the procedural risks and outcomes relevant to the individual patient is recommended. (Level of Evidence: C)

Class IIa

1. AF catheter ablation is reasonable for selected patients with symptomatic persistent AF refractory or intolerant to at least 1 class I or III antiarrhythmic medication. (Level of Evidence: A)
2. In patients with recurrent symptomatic paroxysmal AF, catheter ablation is a reasonable initial rhythm control strategy prior to therapeutic trials of antiarrhythmic drug therapy, after weighing risks and outcomes of drug and ablation therapy. (Level of Evidence: B)

Class IIb

1. AF catheter ablation may be considered for symptomatic long-standing (>12 months) persistent AF refractory or intolerant to at least 1 class I or III antiarrhythmic medication, when a rhythm control strategy is desired. (Level of Evidence: B)
2. AF catheter ablation may be considered prior to initiation of antiarrhythmic drug therapy with a class I or III antiarrhythmic medication for symptomatic persistent AF, when a rhythm control strategy is desired. (Level of Evidence: C)

Class III: Harm

1. AF catheter ablation should not be performed in patients who cannot be treated with anticoagulant therapy during and following the procedure. (Level of Evidence: C)
2. AF catheter ablation to restore sinus rhythm should not be performed with the sole intent of obviating the need for anticoagulation. (Level of Evidence: C)

[59] **Electrical and Pharmacological Cardioversion of AF and Atrial Flutter** ***
Sec. 6.1
Subsection 6.1.2 Direct-Current Cardioversion: Recommendations
Class I

1. In pursuing a rhythm-control strategy, cardioversion is recommended for patients with AF or atrial flutter as a method to restore sinus rhythm. If cardioversion is unsuccessful, repeated direct-current cardioversion attempts may be made after adjusting the location of the electrodes or applying pressure over the electrodes, or following administration of an antiarrhythmic medication. (Level of Evidence: B)
2. Cardioversion is recommended when a rapid ventricular response to AF or atrial flutter does not respond promptly to pharmacological therapies and contributes to ongoing myocardial ischemia, hypotension, or HF. (Level of Evidence: C)
3. Cardioversion is recommended for patients with AF or atrial flutter and pre-excitation when tachycardia is associated with hemodynamic instability. (Level of Evidence: C)

Class IIa

1. It is reasonable to perform repeated cardioversions in patients with persistent AF provided that sinus rhythm can be maintained for a clinically meaningful period between cardioversion procedures. Severity of AF symptoms and patient preference should be considered when embarking on a strategy requiring serial cardioversion procedures. (Level of Evidence: C)

[60] **Risk-Based Antithrombotic Therapy: Recommendations** ***
Sec 4.1

Class I

1. In patients with AF, antithrombotic therapy should be individualized based on shared decision making after discussion of the absolute and RRs of stroke and bleeding, and the patient's values and preferences. (Level of Evidence: C)
2. Selection of antithrombotic therapy should be based on the risk of thromboembolism irrespective of whether the AF pattern is paroxysmal, persistent, or permanent. (Level of Evidence: B)
8. Re-evaluation of the need for and choice of antithrombotic therapy at periodic intervals is recommended to reassess stroke and bleeding risks. (Level of Evidence: C)

[61] **Risk-Based Antithrombotic Therapy: Recommendations** ***

Sec 4.1 Class I

3. In patients with nonvalvular AF, the CHA2DS2-VASc score is recommended for assessment of stroke risk. (Level of Evidence: B)

[62] **Mechanical Heart Valve** ***

Sec 4.1 Risk-Based Antithrombotic Therapy: Recommendations

Class I

4. For patients with AF who have mechanical heart valves, warfarin is recommended and the target international normalized ratio (INR) intensity (2.0–3.0 or 2.5–3.5) should be based on the type and location of the prosthesis. (Level of Evidence: B)

Class III: Harm

1. The direct thrombin inhibitor, dabigatran, should not be used in patients with AF and a mechanical heart valve. (Level of Evidence: B)

[63] **Patients on warfarin** ***
Sec 4.1 Risk-Based Antithrombotic Therapy: Recommendations
Class I

6. Among patients treated with warfarin, the INR should be determined at least weekly during initiation of antithrombotic therapy and at least monthly when anticoagulation (INR in range) is stable. (Level of Evidence: A)

Class I

7. For patients with nonvalvular AF unable to maintain a therapeutic INR level with warfarin, use of a direct thrombin or factor Xa inhibitor (dabigatran, rivaroxaban, or apixaban) is recommended. (Level of Evidence: C)

[64] **Bridging with unfractionated heparin** ***
Sec 4.1 Risk-Based Antithrombotic Therapy: Recommendations
Class I

9. Bridging therapy with unfractionated heparin (UFH) or low-molecular-weight heparin (LMWH) is recommended for patients with AF and a mechanical heart valve undergoing procedures that require interruption of warfarin. Decisions regarding bridging therapy should balance the risks of stroke and bleeding. (Level of Evidence: C)
10. For patients with AF without mechanical heart valves who require interruption of warfarin or newer anticoagulants for procedures, decisions about bridging therapy (LMWH or UFH) should balance the risks of stroke and bleeding and the duration of time a patient will not be anticoagulated. (Level of Evidence: C)

[65] **Renal function** ***
Sec 4.1 Risk-Based Antithrombotic Therapy: Recommendations
Class I

11. Renal function should be evaluated prior to initiation of direct thrombin or factor Xa inhibitors and should be re-evaluated when clinically indicated and at least annually. (Level of Evidence: B)

[66] **Atrial Flutter** ***
Sec 4.1 Risk-Based Antithrombotic Therapy: Recommendations
Class I

12. For patients with atrial flutter, antithrombotic therapy is recommended according to the same risk profile used for AF. (Level of Evidence: C)

[67] **Nonvalvular A Fib/cha2ds2-vasc score** ***
Sec 4.1 Risk-Based Antithrombotic Therapy: Recommendations
Class IIa

1. For patients with nonvalvular AF and a CHA_2DS_2-VASc score of 0, it is reasonable to omit antithrombotic therapy. (Level of Evidence: B)

Class IIb

1. For patients with nonvalvular AF and a CHA_2DS_2-VASc score of 1, no antithrombotic therapy or treatment with an oral anticoagulant or aspirin may be considered. (Level of Evidence: C)

[68] **Prior stroke/TIA** ***
Sec 4.1 Risk-Based Antithrombotic Therapy: Recommendations
Class I

5. For patients with nonvalvular AF with prior stroke, transient ischemic attack (TIA), or a

CHA_2DS_2-VASc score of 2 or greater, oral anticoagulants are recommended. Options include: warfarin (INR 2.0–3.0) (Level of Evidence: A), or dabigatran, rivaroxaban, or apixaban (Level of Evidence: B).

[69] **End-stage CKD/Hemodialysis** ***

Sec 4.1 Risk-Based Antithrombotic Therapy: Recommendations

Class IIa

2. For patients with nonvalvular AF with a CHA_2DS_2-VASc score of 2 or greater and who have endstage CKD (creatinine clearance [CrCl] <15 mL/min) or are on hemodialysis, it is reasonable to prescribe warfarin (INR 2.0–3.0) for oral anticoagulation. (Level of Evidence: B)

Class III: No Benefit

1. The direct thrombin inhibitor, dabigatran, and the factor Xa inhibitor, rivaroxaban, are not recommended in patients with AF and end-stage CKD or on hemodialysis because of the lack of evidence from clinical trials regarding the balance of risks and benefits. (Level of Evidence: C)

Class IIb

2. For patients with nonvalvular AF and moderate-to-severe CKD with CHA2DS2-VASc scores of 2 or greater, treatment with reduced doses of direct thrombin or factor Xa inhibitors may be considered (e.g., dabigatran, rivaroxaban, or apixaban), but safety and efficacy have not been established. (Level of Evidence: C)

[70] **PCI** ***

Sec 4.1 Risk-Based Antithrombotic Therapy: Recommendations

Class IIb

3. In patients with AF undergoing percutaneous coronary intervention,* bare-metal stents may be considered to minimize the required duration of dual antiplatelet therapy. Anticoagulation may be interrupted at the time of the procedure to reduce the risk of bleeding at the site of peripheral arterial puncture. (Level of Evidence: C)

Class IIb

4. Following coronary revascularization (percutaneous or surgical) in patients with AF and a CHA_2DS_2-VASc score of 2 or greater, it may be reasonable to use clopidogrel (75 mg once daily) concurrently with oral anticoagulants but without aspirin. (Level of Evidence: B)

[71] **Rate control guidelines** ***
Sec 5
Class I

1. Control of the ventricular rate using a beta blocker or nondihydropyridine calcium channel antagonist is recommended for patients with paroxysmal, persistent, or permanent AF. (Level of Evidence: B)
2. Intravenous administration of a beta blocker or nondihydropyridine calcium channel blocker is recommended to slow the ventricular heart rate in the acute setting in patients without preexcitation. In hemodynamically unstable patients, electrical cardioversion is indicated. (Level of Evidence: B)
3. In patients who experience AF-related symptoms during activity, the adequacy of heart rate control should be assessed during exertion, adjusting pharmacological treatment as necessary to keep the ventricular rate within the physiological range. (Level of Evidence: C)

Class IIa

1. A heart rate control (resting heart rate <80 bpm) strategy is reasonable for symptomatic management of AF. (Level of Evidence: B)
2. Intravenous amiodarone can be useful for rate control in critically ill patients without preexcitation. (Level of Evidence: B)
3. AV nodal ablation with permanent ventricular pacing is reasonable to control the heart rate when pharmacological therapy is inadequate and rhythm control is not achievable. (Level of Evidence: B)

Class IIb

1. A lenient rate-control strategy (resting heart rate <110 bpm) may be reasonable as long as patients remain asymptomatic and LV systolic function is preserved. (Level of Evidence: B)
2. Oral amiodarone may be useful for ventricular rate control when other measures are unsuccessful or contraindicated. (Level of Evidence: C)

Class III: Harm

1. AV nodal ablation with permanent ventricular pacing should not be performed to improve rate control without prior attempts to achieve rate control with medications. (Level of Evidence: C)
2. Nondihydropyridine calcium channel antagonists should not be used in patients with decompensated HF as these may lead to further hemodynamic compromise. (Level of Evidence: C)
3. In patients with pre-excitation and AF, digoxin, nondihydropyridine calcium channel antagonists, or intravenous amiodarone should not be administered as they may increase the ventricular response and may result in ventricular fibrillation. (Level of Evidence: B)

4. Dronedarone should not be used to control the ventricular rate in patients with permanent AF as it increases the risk of the combined endpoint of stroke, MI, systemic embolism, or cardiovascular death. (Level of Evidence: B)

[72] **Pharmacological cardioversion** ***
Sec. 6.1.3
Class I

1. Flecainide, dofetilide, propafenone, and intravenous ibutilide are useful for pharmacological cardioversion of AF or atrial flutter provided contraindications to the selected drug are absent. (Level of Evidence: A)

Class IIa

1. Administration of oral amiodarone is a reasonable option for pharmacological cardioversion of AF. (Level of Evidence: A)
2. Propafenone or flecainide ("pill-in-the-pocket") in addition to a beta blocker or nondihydropyridine calcium channel antagonist is reasonable to terminate AF outside the hospital once this treatment has been observed to be safe in a monitored setting for selected patients. (Level of Evidence: B)

Class III: Harm

1. Dofetilide therapy should not be initiated out of hospital owing to the risk of excessive QT prolongation that can cause torsades de pointes. (Level of Evidence: B)

[73] **Pharmacological Maintenance** ***
Sec. 6.2.1
Class I

1. Before initiating antiarrhythmic drug therapy, treatment of precipitating or reversible causes of AF is recommended. (Level of Evidence: C)

2. The following antiarrhythmic drugs are recommended in patients with AF to maintain sinus rhythm, depending on underlying heart disease and comorbidities (Level of Evidence: A):

    a. Amiodarone
    b. Dofetilide
    c. Dronedarone
    d. Flecainide
    e. Propafenone
    f. Sotalol

3. The risks of the antiarrhythmic drug, including proarrhythmia, should be considered before initiating therapy with each drug. (Level of Evidence: C)
4. Owing to its potential toxicities, amiodarone should only be used after consideration of risks and when other agents have failed or are contraindicated. (Level of Evidence: C)

Class IIa

1. A rhythm-control strategy with pharmacological therapy can be useful in patients with AF for the treatment of tachycardia-induced cardiomyopathy. (Level of Evidence: C)

Class IIb

1. It may be reasonable to continue current antiarrhythmic drug therapy in the setting of infrequent, well-tolerated recurrences of AF, when the drug has reduced the frequency or symptoms of AF. (Level of Evidence: C)

Class III: Harm

1. Antiarrhythmic drugs for rhythm control should not be continued when AF becomes permanent (Level of Evidence: C) including dronedarone. (Level of Evidence: B)
2. Dronedarone should not be used for treatment of AF in patients with New York Heart Association

(NYHA) class III and IV HF or patients who have had an episode of decompensated HF in the past 4 weeks. (Level of Evidence: B)

[74] **Thromboembolism prevention** ***
Subsection 6.1.1 Thromboembolism Prevention: Recommendations
Class IIa

1. For patients with AF or atrial flutter of 48-h duration or longer or of unknown duration who have not been anticoagulated for the preceding 3 weeks, it is reasonable to perform a TEE prior to cardioversion and proceed with cardioversion if no LA thrombus is identified, including in the LAA, provided that anticoagulation is achieved before TEE and maintained after cardioversion for at least 4 weeks. (Level of Evidence: B)

[75] **Thromboembolism prevention** ***
Subsection 6.1.1 Thromboembolism Prevention: Recommendations
Class I

1. For patients with AF or atrial flutter of 48-h duration or longer, or when the duration of AF is unknown, anticoagulation with warfarin (INR 2.0–3.0) is recommended for at least 3 weeks prior to and 4 weeks after cardioversion, regardless of the CHA2DS2-VASc score and the method (electrical or pharmacological) used to restore sinus rhythm. (Level of Evidence: B)

[76] **Thromboembolism prevention** ***
Subsection 6.1.1 Thromboembolism Prevention: Recommendations
Class IIa

2. For patients with AF or atrial flutter of 48-h duration or longer, or when the duration of AF is unknown, anticoagulation with dabigatran, rivar-

oxaban, or apixaban is reasonable for at least 3 weeks prior to and 4 weeks after cardioversion. (Level of Evidence: C)

[77] **Thromboembolism prevention with immediate cardioversion** ***
Subsection 6.1.1 Thromboembolism Prevention: Recommendations
Class I

2. For patients with AF or atrial flutter of more than 48 h or unknown duration that requires immediate cardioversion for hemodynamic instability, anticoagulation should be initiated as soon as possible and continued for at least 4 weeks after cardioversion unless contraindicated. (Level of Evidence: C)

[78] **Thromboembolism prevention** ***
Subsection 6.1.1 Thromboembolism Prevention: Recommendations
Class IIb

1. For patients with AF or atrial flutter of less than 48-h duration who are at low thromboembolic risk, anticoagulation (intravenous heparin, LMWH, or a new oral anticoagulant) or no antithrombotic therapy may be considered for cardioversion, without the need for postcardioversion oral anticoagulation. (Level of Evidence: C)

[79] **Thromboembolism prevention** ***
Subsection 6.1.1 Thromboembolism Prevention: Recommendations
Class I

3. For patients with AF or atrial flutter of less than 48-h duration and with high risk of stroke, intravenous heparin or LMWH, or administration of a factor Xa or direct thrombin inhibitor, is recom-

mended as soon as possible before or immediately after cardioversion, followed by longterm anticoagulation therapy. (Level of Evidence: C)

[80] **Long term thromboembolism prevention**
Subsection 6.1.1 Thromboembolism Prevention: Recommendations
Class I

4. Following cardioversion for AF of any duration, the decision regarding long-term anticoagulation therapy should be based on the thromboembolic risk profile (Section 4). (Level of Evidence: C)

[81] **Maze procedure**
Sec. 6.5
Class IIa

1. An AF surgical ablation procedure is reasonable for selected patients with AF undergoing cardiac surgery for other indications. (Level of Evidence: C)

Class IIb

1. A stand-alone AF surgical ablation procedure may be reasonable for selected patients with highly symptomatic AF not well managed with other approaches. (Level of Evidence: B)

[82] **Upstream therapy** ***
Sec. 6.2.2
Class IIa

1. An ACE inhibitor or angiotensin-receptor blocker (ARB) is reasonable for primary prevention of new-onset AF in patients with HF with reduced LVEF. (Level of Evidence: B)

Class IIb

1. Therapy with an ACE inhibitor or ARB may be considered for primary prevention of new-onset AF in the setting of hypertension. (Level of Evidence: B)

2. Statin therapy may be reasonable for primary prevention of new-onset AF after coronary artery surgery. (Level of Evidence: A)

Class III: No Benefit

1. Therapy with an ACE inhibitor, ARB, or statin is not beneficial for primary prevention of AF in patients without cardiovascular disease. (Level of Evidence: B)

*** **Extracted from** 2012 ACCF/AHA/HRS Focused Update Incorporated Into the ACCF/AHA/HRS 2008 Guidelines for Device-Based Therapy of Cardiac Rhythm Abnormalities (J Am Coll Cardiol 2013;61: e6–75)

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Circulation. 2009;120:1768–74. http://circ.ahajournals.org/content/120/18/1768.full.

Obesity

J Am Coll Cardiol. 2015;66:1–11. http://content.onlinejacc.org/article.aspx?articleID=2375086.

Obesity

Am J Med. 2005;118:489–95. http://www.sciencedirect.com/science/article/pii/S0002934305000744.

Obesity: Risk for Atrial Fibrillation

J Am Coll Cardiol EP. 2015;1:139–52. http://electrophysiology.onlinejacc.org/article.aspx?articleID=2299812.

Obstructive Sleep Apnea

J Am Coll Cardiol. 2007;49:565–71. http://content.onlinejacc.org/article.aspx?articleID=1188673.

Obstructive Sleep Apnea/Stroke

Am J Cardiol. 2015;115:461–5. http://www.sciencedirect.com/science/article/pii/S0002914914021687.

Obstructive Sleep Apnea Treatment: AF Recurrence

JACCCEP. 2015;1:41–51. http://electrophysiology.onlinejacc.org/article.aspx?articleID=2277227.

Oral Antithrombotics (AHA Science Advisory)

Stroke. 2012;43:3442–53. http://stroke.ahajournals.org/content/43/12/3442.full.

Outcomes: Long-Term (Framingham)

Circulation. 2015;131:1648–55. http://circ.ahajournals.org/content/131/19/1648.full.

P Wave Duration: Risk

Heart Rhythm. 2016;12:1887–95. http://www.heartrhythmjournal.com/article/S1547-5271(15)00493-2/abstract.

Pathophysiology/Prognosis: Imaging

Circulation: Cardiovascular Imaging. 2015;8:e003020. http://circimaging.ahajournals.org/content/8/6/e00300.full.

Pericarditis: Acute

Herz. 2000;25:729–33. http://www.ncbi.nlm.nih.gov/pubmed/11200120.

Pericarditis: Constrictive

Ann Noninvasive Electrocardiol. 2009;14:258–61. http://www.ncbi.nlm.nih.gov/pubmed/19614637.

Perioperative Atrial Fibrillation

JAMA. 2014;312:616–22. http://jama.jamanetwork.com/article.aspx?articleid=1895248.

Peripheral Artery Disease

J Am Heart Assoc. 2014;3:e001270. http://jaha.ahajournals.org/content/3/6/e001270.long.

Pheochromocytoma

Rev Port Cardiol. 2014;33:245.e1–4. http://www.ncbi.nlm.nih.gov/pubmed/24780129.

Pneumonia

Clin Infect Dis. 2007;45:158–65. http://cid.oxfordjournals.org/content/45/2/158.long.

Predisposing Factors

Pacing Clin Electrophysiol. 1997;20:2670–4. http://onlinelibrary.wiley.com/doi/10.1111/j.1540-8159.1997.tb06115.x/abstract.

Preexcitation Syndromes

Am J Card. 1977;40:514–20. http://www.sciencedirect.com/science/article/pii/0002914977900650.

Prognosis

Circulation. 2012;126:e143–6. http://circ.ahajournals.org/content/126/10/e143.full.

QTC Duration/AF Risk

J Am Coll Cardiol. 2013;61:2557–64. http://content.onlinejacc.org/article.aspx?articleID=1679549.

Rate Control

N Engl J Med. 2010;362:1363–73. http://www.nejm.org/doi/full/10.1056/NEJMoa1001337.

Rate/Rhythm Control: Review

Ann Intern Med. 2014;160:760–73. http://annals.org/article.aspx?articleid=1877019.

Renal Dysfunction: Anticoagulants

J Am Coll Cardiol. 2015;65:2420–9. http://content.onlinejacc.org/article.aspx?articleID=2300733.

Rhythm Control: La Dysfunction

Chest. 2005;128:2551–2556. http://journal.publications.chestnet.org/article.aspx?articleID=1083870.

Risk Factors: Framingham Heart Study

JAMA. 1994;271:840–4. http://jama.jamanetwork.com/article.aspx?articleid=367563.

Risk: Long-Term

Am J Med. 2002;113:359–364. http://www.sciencedirect.com/science/article/pii/S0002934302012366.

Risk: Resting HR/Physical Activity

Heart. 2013;99:1755–60. http://heart.bmj.com/content/99/23/1755.abstract.

Risk for Stroke/Bleeding Associated with Antithrombotic Therapy

J Am Heart Assoc. 2015;4:e001921. http://jaha.ahajournals.org/content/4/7/e001921.abstract?etoc.

Rivaroxaban

N Engl J Med. 2011;365:883–91. http://www.nejm.org/doi/full/10.1056/NEJMoa1009638.

Second Hand Smoke Exposure

Heart Rhythm. 2016;13:3–9. http://www.heartrhythmjournal.com/article/S1547-5271(15)01018-8/abstract.

Sepsis

Chest. 2016;149:74–83. http://journal.publications.chestnet.org/article.aspx?articleID=2429069.

Sepsis

Chest. 2014;146:1187–95. http://journal.publications.chestnet.org/article.aspx?articleid=1860570.

Sick Sinus Syndrome

Europace. 2011;14:682–9. http://europace.oxfordjournals.org/content/14/5/682.

Silent Cerebral Ischemia

J Am Coll Cardiol. 2013;62:1990–7. http://content.onlinejacc.org/article.aspx?articleID=1711102.

Silent Stroke

Ann Intern Med. 2014;161:650–8. http://annals.org/article.aspx?articleid=1920505.

Spinal Cord Injury

Heart Rhythm. 2016;13:416–23. http://www.heartrhythmjournal.com/article/S1547-5271(15)01279-5/abstract.

Statins: Post-OP AF Prevention

J Thorac Cardiovasc Surg. 2010;139:803–4. http://www.sciencedirect.com/science/article/pii/S0022522309005121.

Stroke

N Engl J Med. 2014;370:2467–77. http://www.nejm.org/doi/full/10.1056/NEJMoa1311376.

Stroke: Prevention in Patients with CKD

Circulation. 2016; 133:1512–15. http://circ.ahajournals.org/content/133/15/1512.full.

Stroke: Subclinical AF

N Engl J Med. 2012;366:120–9. http://www.nejm.org/doi/full/10.1056/NEJMoa1105575.

Symptoms: Orbit-AF

Circ Outcomes. 2015;114.001303. http://circoutcomes.ahajournals.org/content/early/2015/06/08/CIRCOUTCOMES.114.001303.abstract.

Systemic Arterial Hypertension

Am J Cardiol. 2003;91:9G–14G. http://www.ncbi.nlm.nih.gov/pubmed/12781903.

Systemic Emboli

Circulation. 2015;132:796–803. http://circ.ahajournals.org/content/132/9/796.abstract.

Thoracic Surgery

J Am Coll Cardiol. 2009;54:2049–51. http://content.onlinejacc.org/article.aspx?articleid=1140222.

Thromboembolism Risk: CHA2DS2-VASc Score

Chest. 2010;137:263–72. http://journal.publications.chestnet.org/article.aspx?articleid=1086288.

Tobacco Use

Am Heart J. 2008;156:1163–9. http://www.sciencedirect.com/science/article/pii/S0002870308006601.

Vigorous Exercise

Am J Cardiol. 2009;103:1572–7. http://www.sciencedirect.com/science/article/pii/S0002914909005499.

Updates and More

https://clinicalguidecvd.com/afib

Chapter 16
Atrial Giant Cell Myocarditis

ICD-10 Code

I40.1

Alternate Names/Abbreviation

NS

Description/Etiology

Rare inflammatory disorder of atria consisting of giant cell and lymphocyte infiltrates of atrial wall [1]
Believed to be a distinct entity from giant cell myocarditis, which primarily involves ventricles, with much better prognosis and not associated with other inflammatory disorders
Often associated with MV/TV disease, especially MS
Etiology unknown

Comorbid Conditions

RHEUMATIC VALVE DISEASE [2]

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_16,

Demography

Too few cases reported but all middle age and older

Pathophysiology

Histology (atrial appendage):

- Cardiomyocyte hypertrophy
- Cardiomyocyte necrosis
- Eosinophils
- Giant cells
- Granulomas (poorly-formed granulomas)
- Interstitial fibrosis
- Interstitial lymphocytic inflammatory infiltrates
- Lymphocytic myocarditis-like foci

Atrial dilatation/hypokinesis with preserved ventricular function

Signs/Symptoms [3]

BREATHING – DIFF (DYSPNEA)
BREATHING – DIFF, RECLINING FLAT (ORTHOPNEA)
CHEST – PAIN
FATIGUE
HEART, RHYTHM – IRREG

Differentiation

Other causes of atrial dilatation
INFECTIOUS MYOCARDITIS [6]
SARCOIDOSIS [6]
WEGENER GRANULOMATOSIS [6]

Complications

Atrial Fibrillation
Heart Block
Peripheral Emboli

Laboratory

NS

ECG [3]

DYSRHYTHMIAS–ATRIAL (PACS/OTHERS) [4]

Imaging [3]

LA, CHAMBER, SIZE–INCR
LA, INTRACAVITY–MASS [MURAL THROMBUS]
LA, WALL, THICKNESS – INCR

Other Tests

NS

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic

Anticoagulation
Immunosuppression [5]

Treatment: Surgical/Invasive

Pacemaker

Prevention

NS

Course

Uncertain due to limited reporting but prognosis appears better than typical Giant Cell Myocarditis of ventricles

Notes

[1] Only small number of reports in literature; many detected only with microscopic study of atrial appendages removed at surgery
[2] Not all reported cases have this association; MS case report in 1968
[3] Does not include findings due to associated valve disease
[4] Especially AF
[5] Efficacy unknown
[6] Histology

Guidelines

NS

Patient Information

(NS for Atrial form; list is for Giant Cell Myocarditis in general)

Medlineplus

ENGLISH http://www.nlm.nih.gov/medlineplus/ency/article/000149.htm.
ESPANOL http://www.nlm.nih.gov/medlineplus/spanish/ency/article/000149.htm.

Myocarditis Foundation

http://www.myocarditisfoundation.org/about-myocarditis/.

Nord

https://rarediseases.org/rare-diseases/giant-cell-myocarditis/.

Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/myocarditis/basics/definition/con-20027303.

Professional Information

Editorial
Circulation. 2013;127:8–9. http://circ.ahajournals.org/content/127/1/8.extract.

Editorial

Circulation. 2013;127:8–9. http://circ.ahajournals.org/content/127/1/8.extract.

Case Series

Circulation. 2013;127:39–47. http://circ.ahajournals.org/content/127/1/39.full.

Case Report

Cardiovasc Pathol. 2010;19:e37–8. http://www.ncbi.nlm.nih.gov/pubmed/19026572.

Case Report

Pacing Clin Electrophysiol. 2006;29:1179–80. http://www.ncbi.nlm.nih.gov/pubmed/17038151.

Echo Features (Case Report)

J Am Soc Echocardiogr. 2004;17:1073–6. http://www.ncbi.nlm.nih.gov/pubmed/15452474.

Mitral Stenosis

J Clin Pathol. 1968;21:750–75. http://www.ncbi.nlm.nih.gov/pmc/articles/PMC473954/.

Updates and More

https://clinicalguidecvd.com/agcm

Chapter 17
Atrial Septal Defect: Secundum

ICD-10 Code

Q21.1

Alternate Names/Abbreviation

ASD
OSTIUM SECUNDUM DEFECT
PATENT FORAMEN OVALE

Description/Etiology

Persistent interatrial communication comprising several different types:

- Secundum ASD in region of fossa ovalis (75 % of cases)
- Primum ASD, inferior near crux of heart (15–20 %)
- Sinus venosus ASD, superior near SVC entry or inferiorly near IVC entry (5–10 %)
- Coronary sinus septal defect (<1 %)
- Patent foramen ovale [1]

Associated with other malformations in nearly 30 % of cases

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_17,

Others:

Familial
Iatrogenic [11]

Initial presentation in adults most often includes dyspnea and palpitations

Comorbid Conditions

ATRIOVENTRICULAR HEART BLOCK
CLEFT MITRAL VALVE
HOLT-ORAM SYNDROME
MIGRAINE [10]
MITRAL VALVE PROLAPSE
PARTIAL ANOMALOUS RIGHT PULMONARY VENOUS DRAINAGE
PULMONARY STENOSIS – VALVULAR
TREACHER COLLINS SYNDROME
TRISOMY 13 (PATAU SYNDROME)
TRISOMY 18 (EDWARDS SYNDROME)

Demography

F 2:1
All age groups [2]

Pathophysiology

RV volume overload and pulmonary over-circulation
Flow-related PAH occurs with large shunts
Pulmonary vascular obstructive disease may develop in adults

Signs/Symptoms [2] [5]

BODY, GROWTH – DECR
BREATHING – DIFF (DYSPNEA)
REATHING – DIFF, RECLINING FLAT (ORTHOPNEA)
BREATHING – DIFF, UPRIGHT (PLATYPNEA)
BREATHING – RAPID (TACHYPNEA)
CHEST – PAIN
CHEST – PALPITATIONS
CHEST, ANT, L – BULGE
CHEST, AXILLA – MURMUR, SYS
FATIGUE
HEART, LSB, LOWER – IMP, SYS
HEART, LSB, LOWER – MURMUR, DIAS
HEART, LSB, UPPER – IMP, SYS
HEART, LSB, UPPER – MURMUR, SYS
HEART, S2, SPLIT – FIXED
HEART, S2, SPLIT – WIDE
HEART, T1, INTENSITY – INCR
NECK, JVP, A WAVE – CREST EQUALS V WAVE
SKIN, COLOR – BLUE (CYANOSIS) [3]
SKIN, COLOR, EFFORT – BLUE (CYANOSIS)

Differentiation

Other causes of RV/pulmonary overload and pulmonary congestion

Complications

Atrial Fibrillation/Flutter [4]
Heart Block
Lower Respiratory infections (Recurrent)
Peripheral Embolism [8]
Paroxysmal Atrial Tachycardia

Pneumonia
Pulmonary Arterial Hypertension
Sick Sinus Syndrome
Stroke

Laboratory

NS

ECG

AV COND – 1ST DEGREE BLOCK
DYSRHYTHMIAS – ATRIAL (PACS/OTHERS)
P WAVE – TALL/PEAKED
P WAVE, INF LEADS – INVERTED
QRS – RBBB/RBBB PATTERN
QRS, AXIS – R

Imaging [12] [13]

AORTA, ROOT, SIZE – DECR [INCONSPICUOUS ON X-RAY]
ATRIA – FLOW – INTERATRIAL [DOPPLER]
ATRIA, SEPTUM – ANEURYSM
CARDIOMEGALY
IAS – ECHO-FREE SPACE
IVS, MOTION – PARADOX
LA, SIZE – N
PA, MAIN, SIZE – INCR
PA, TRUNK, SIZE – INCR
PUL, VASCULARITY – INCR
RA, CHAMBER, SIZE – INCR
RV, CHAMBER, SIZE – INCR
RV, WALL MOTION – INCR/HYPERDYNAMIC

Genomics

CSX
DTNA
MYH6
NKX2.5
TBX5

Other Tests

Exercise test for functional capacity [7]
Cardiac catheterization [6]

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic

Rate control for persistent AF, with anticoagulation

Treatment: Surgical/Invasive

Percutaneous closure [9]
Surgical closure [9]

Prevention

NA

Course

Often excellent but variable according to other factors, eg, magnitude of shunt and coexisting conditions, eg, CAD, hypertension
Often asymptomatic until middle age

Notes

[1] Flaplike communication involving septum primum covering fossa ovalis overlaps superior limbic band of septum secundum; may occur up to 30 months
[2] Initial clinical features often not present until adulthood
[3] When complicated by RV failure/increased PA pressures
[4] May precipitate initial symptoms in adults
[5] Features such as murmurs may become more pronounced or first detected during pregnancy
[6] Cardiac catheterization not required for uncomplicated ASDs in younger patients with adequate noninvasive imaging; usually for study of possible CAD in patients at increased risk and for whom surgical intervention is planned and to assess PVR and reactivity in patients with significant PAH

    May also be needed to evaluate ASD size, pulmonary venous return, and associated valvular disease if noninvasive methods inadequate
    Catheterization most often performed in conjunction with device closure of ASD

[7] Contraindicated in presence of severe PAH
[8] Paradoxical or from septal aneurysm
[9] Not indicated in presence of severe irreversible PAH and no evidence of L-R shunt
[10] Conflicting data whether correction has effect

[11] Persistent iatrogenic ASDs may occur after structural transseptal cardiac interventions, especially with use of larger sheaths

[12] TTE is primary diagnostic imaging modality for ASD:

- Include 2-dimensional imaging of atrial septum from parasternal, apical, and subcostal views with color Doppler demonstration of shunting
- Include subcostal views with deep inspiration
- Include high right parasternal views, which can be particularly helpful for imaging ASD in adults
- Visualize entire atrial septum from SVC orifice to IVC orifice to detect sinus venosus defects or extension of large secundum defects in these regions

[13] TEE may be necessary to determine connection of all pulmonary veins in patients with ASD; also provides exact localization and sizing of ASD and measurement of septal rims

Guidelines

ACC/AHA 2008 Guidelines for the management of adults with congenital heart disease

J Am Coll Cardiol. 2008;52:e143–263. http://content.onlinejacc.org/article.aspx?articleid=1188032#tab1.

ESC Guidelines for the management of grown-up congenital heart disease (new version 2010)

Eur Heart J. 2010; 31:2915–57. http://eurheartj.oxfordjournals.org/content/ehj/31/23/2915.full.pdf.

Patient Information

Images

http://www.nlm.nih.gov/medlineplus/ency/imagepages/18119.htm.

Medlineplus

ENGLISH

http://www.nlm.nih.gov/medlineplus/ency/article/000157.htm.

ESPANOL

http://www.nlm.nih.gov/medlineplus/spanish/ency/article/000157.htm.

Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/atrial-septal-defect/basics/definition/con-20027034.

AHA

http://www.heart.org/HEARTORG/Conditions/CongenitalHeartDefects/AboutCongenitalHeartDefects/Atrial-Septal-Defect-ASD-UCM-307021-Article.jsp.

CDC

http://www.cdc.gov/ncbddd/heartdefects/atrialseptaldefect.html.

Cleveland Clinic

http://my.clevelandclinic.org/services/heart/disorders/congenital-heart/atrial-septal-defect-asd.

Merck

http://www.merckmanuals.com/home/SearchResults?query=Atrial+Septal+Defect++(ASD).

Stanford Childrens

http://www.stanfordchildrens.org/en/topic/default?id=atrial-septal-defect-asd-90-P01766.

Professional Information

Review: Adults

Circulation. 2006;114:1645–53. http://circ.ahajournals.org/content/114/15/1645.full.

Review: Closure

Cardiol Clin. 2013;31:385–400. http://www.sciencedirect.com/science/article/pii/S0733865113000313.

Review: Transcatheter Closure Versus Medical Therapy

Eur Heart J. 2013;34:3342–52. http://eurheartj.oxfordjournals.org/content/34/43/3342.

Closure in Adults

Eur Heart J. 2011;32:553–60. http://eurheartj.oxfordjournals.org/content/32/5/553.long.

Closure: Patients with Cryptogenic Embolism

Eur Heart J. 2015;36:120–8. http://eurheartj.oxfordjournals.org/content/36/2/120.

Closure: Determinants of Improvement Post-procedure

J Am Coll Cardiol. 2004;43:1886–91. http://content.onlinejacc.org/article.aspx?articleid=1135607.

Closure in Adults: Long-Term Outcomes

J Am Coll Cardiol Intv. 2013;6:497–503. http://interventions.onlinejacc.org/article.aspx?articleid=1679520.

Familial ASD

Circulation. 1998;97:2043–8. http://circ.ahajournals.org/content/97/20/2043.full?sid=85d3d9c3-e0c8-43aa-8e8f-06010fa1b36c.

Iatrogenic ASD

Circulation Cardiovasc Interv. 2016;9:e003545. http://circinterventions.ahajournals.org/content/9/4/e003545.extract?etoc.

Migraine

Am J Med. 2014;127:411–20. http://www.sciencedirect.com/science/article/pii/S0002934313010747.

Natural HX

Circulation. 1968;37:805–15. http://circ.ahajournals.org/content/37/5/805.abstract?sid=85d3d9c3-e0c8-43aa-8e8f-06010fa1b36c.

Pacemakers/Stroke Risk

Circulation. 2013;128:1433–41. http://circ.ahajournals.org/content/128/13/1433.full.

Paradoxical Embolism

Catheter Cardiovasc Interv. 2009;74:1066–9. http://www.ncbi.nlm.nih.gov/pubmed/19670302.

Paradoxical Embolism

Circulation. 2014;129:1892–7. http://circ.ahajournals.org/content/129/18/1892.full.

Paradoxical Embolism: Images

J Am Coll Cardiol. 2010;56:e43. http://content.onlinejacc.org/article.aspx?articleid=1143932.

Post-closure Lv/Rv Function Improvement

J Am Coll Cardiol. 2005;45:499–504. http://content.onlinejacc.org/article.aspx?articleid=1136345.

Subclinical Cerebrovascular Disease/Stroke

J Am Coll Cardiol. 2013;62:35–41. http://content.onlinejacc.org/article.aspx?articleID=1686317.

Stroke

Stroke. 2015;46:e35–7. http://stroke.ahajournals.org/content/46/2/e35.extract?etoc.

Roke Prevention by Percutaneous Closure

STHeart. 2014;100:389–95. http://heart.bmj.com/content/100/5/389.abstract.

Unexplained Stroke

N Engl J Med. 2005;353:2361–72. http://www.nejm.org/doi/full/10.1056/NEJMcp043981.

Updates and More

https://clinicalguidecvd.com/asds

Chapter 18
Atrioventricular Heart Block

ICD-10 Code

I44.0 FIRST DEGREE
I44.1 SECOND DEGREE
I44.2 THIRD DEGREE

Alternate Names/Abbreviation

FIRST DEGREE HEART BLOCK
SECOND DEGREE HEART BLOCK
THIRD DEGREE HEART BLOCK
LENEGRE DISEASE
LEV DISEASE
VAGALLY-MEDIATED HEART BLOCK [17]

Description/Etiology

Slowed or absent cardiac impulse conduction at any site, or combination of sites, from sinus node to bundle branches from a variety of causes

Congenital:

Associated with other cardiac malformations
Cardiac ion channelopathies

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_18,

Complete AV block [1]

Acquired

Age-related fibrosis
Idiopathic: Paroxysmal atrioventricular block [14] [17]
Other acquired cardiac disorders (see COMORBID/ PREDISPOSING CONDTIONS) Vagal-mediated [17]
Vagal-mediated [17]

Comorbid/Predisposing Conditions

ALLERGIC ACUTE CORONARY SYNDROME (KOUNIS SYNDROME)
ACUTE ALCOHOL INGESTION
ACUTE MYOCARDIAL INFARCTION
ATRIOVENTRICULAR SEPTAL DEFECT
CARDIAC INVASIVE PROCEDURES [9]
CARDIOMYOPATHY – TAKOTSUBO
CORONARY ARTERY DISEASE [3]
DRUGS [2]
EBSTEIN ANOMALY
EHLERS-DANLOS SYNDROME
ELECTROLYTE ABNORMALITIES [11]
ENDOCRINOPATHIES [12]
FABRY DISEASE
HYPERURICEMIA
HYPOTHERMIA
INFECTION [8]
INFECTIVE ENDOCARDITIS
INFILTRATIVE [6]
KEARNS-SAYRE SYNDROME
LONG QT SYNDROME
MARFAN SYNDROME
MITRAL ANNULUS CALCIFICATION
NEUROPATHIES [7]
OBSTRUCTIVE SLEEP APNEA [15]
RADIATION
RHEUMATOID HEART DISEASE [5]

SARCOIDOSIS
SCLEROSIS/FIBROSIS OF CARDIAC SKELETON [4]
SCORPION STING
TRANSPOSITION OF GREAT ARTERIES – CORRECTED
TRICUSPID ATRESIA
VAGAL MEDIATION
VENTRICULAR SEPTAL DEFECT

Demography

Varies according to etiology

Pathophysiology

Morphology varies according to etiology
Physiologic effect: reduced cardiac output due to:

Slow HR
Atrioventricular dyssynchrony

Signs/Symptoms

BLOOD PRESSURE, ARTERIAL – INCREASED/ELEVATED [SYS]
BREATHING – DIFF (DYSPNEA)
CHEST – PAIN, EFFORT (ANGINA PECTORIS)
CHEST – PALPITATIONS
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)
DIZZY/LIGHTHEADED/PRESYNCOPE
FATIGUE
HEART, RATE – SLOW (BRADYCARDIA)
HEART, RHYTHM – IRREG
HEART, S1, INTENSITY – VAR
MENTATION – CONFUSION
MENTATION – WEAKNESS (MALAISE)
MOOD – LETHARGIC

SEIZURES

Differentiation

Other causes of syncope
Seizure disorder

Complications

Heart Failure
Stroke
Syncope
Sudden death

Laboratory

ASSESS FOR OTHER CAUSATIVE FACTORS, EG, ELECTROLYTES [11]

ECG

AV COND – 1ST DEGREE BLOCK [18]
AV COND – 2ND DEGREE BLOCK, MOBITZ I (WENCKEBACH)
AV COND – 2ND DEGREE BLOCK, MOBITZ II
AV COND – 3RD DEGREE BLOCK
AV CONDUCTION – AV DISSOCIATION, COMPLETE
DYSRHY – JUNCT
DYSRHYTHMIAS – VENTRICULAR (PVCS/ OTHERS)
P WAVE – ABSENT CONSTANT/INTERMITTENT
P WAVE – INDEPENDENT OF QRS
PR INTERVAL – VAR
QRS – LBBB/LBBB PATTERN

QRS – LONG, NS
QRS – RBBB/RBBB PATTERN [14]
RATE – DECREASED (SINUS BRADYCARDIA)
VAR DEPENDING ON TYPE/ETIOL

Imaging

FOR ETIOL, ESP ASSD CONGEN DEFECTS

Genomics

GATA 5
NKX2.5
SCN5A

Other Tests

Ambulatory ECG Monitoring
Exercise test
EP test

Treatment: Nonpharmacologic [19]

NS

Treatment: Pharmacologic [19]

DC drugs that may cause/exacerbate [2]
Antiinfectious agents when indicated [8]
Atropine [13]
Isoproterenol [13]

Treatment: Surgical/Invasive [19]

Pacemaker – temporary
Pacemaker – permanent

Prevention

NS

Course

Variable per cause

Notes

[1] Embryonic maldevelopment of AVN; >50 % may be due to maternal SLE
[2] EG, Digoxin, beta-blockers, CCBs, antiarrhythmics, clonidine, older antidepressants, chloroquine, marijuana, lithium, organophosphates, methyldopa
[3] With/without prior infarction; transient or permanent
[4] Progressive sclerosis/fibrosis of conduction system (LEV disease, Lenegre Disease); calcification of aortic/mitral annulus extending to conduction tracts
[5] EG, Rheumatoid arthritis, Scleroderma, SLE, Reiter Syndrome, Ankylosing Spondylitis, Wegener Granulomatosis, Polymyositis
[6] EG, Amyloidosis, Sarcoidosis, Hemochromatosis, tumors (benign, metastatic, leukemia)
[7] EG, Muscular Dystrophy (Myotonic, Becker, Peroneal Muscular Atrophy/Charcot-Marie-Tooth, ERB/limb-girdle), Kearns-Sayre Syndrome
[8] EG, Endocarditis, viral Myocarditis (e.g., Measles, Mumps, Varicella), bacterial Myocarditis (e.g. Lyme disease, Tuberculosis), Parasitic Myocarditis (e.g., Chagas disease, Toxoplasmosis)

[9] Especially AV replacement, cardiac catheterization, CABG, congenital repair, alcohol ablation
[10] Usually benign, with normal QRS; e.g., during swallowing, coughing, micturition, physical conditioning, carotid sinus massage
[11] EG, Hyperkalemia, Hypercalcemia, Hypermagnesemia
[12] EG, Hypothyroidism, Adrenal Insufficiency, Hypopituitarism
[13] Temporary only, until definitive treatment can be given
[14] Newly described entity with episodes of third-degree heart block manifest by recurrent syncope; RBBB reported in 1 series as common on resting ECG; non-progressive; adenosine infusion simulates block; treatable with pacing
[15] Especially during sleep
[16] Progressive cardiac conduction disease
[17] Paroxysmal AV block; localized to AV node; associated with sinus bradycardia; syncopal episodes
[18] Although generally regarded as an innocent finding, first degree block may be associated with adverse long-term outcomes, including significant increases in AF, HF, mortality
[19] First exclude all reversible/treatable underlying causes, which dictate specific treatment when present

Guidelines

2013 ESC Guidelines on cardiac pacing and cardiac resynchronization therapy
Eur Heart J. 2013;34:2281–329. http://eurheartj.oxfordjournals.org/content/34/29/2281.
2012 ACCF/AHA/HRS focused update incorporated into the ACCF/AHA/HRS 2008 guidelines for device-based therapy of cardiac rhythm abnormalities
JACC. 2013;61:e6–75. http://content.onlinejacc.org/article.aspx?articleid=1486116&resultClick=3.

Patient Information

AHA: Pacemakers

http://www.heart.org/HEARTORG/Conditions/Arrhythmia/PreventionTreatmentofArrhythmia/Living-With-Your-Pacemaker_UCM_305290_Article.jsp#.VzN9wfMUV1s.

HRS: Heart Block

http://www.hrsonline.org/Patient-Resources/Heart-Diseases-Disorders/Heart-Block.

Professional Information

Bradycardia: Evaluation and Management

N Engl J Med. 2000;342:703–9. http://www.nejm.org/doi/full/10.1056/NEJM200003093421006.

Cardiac Valve Surgery: Need For Pacing

J Am Coll Cardiol. 2003;41:795–801. http://www.sciencedirect.com/science/article/pii/S0735109702029261.

Drug-Induced Heart Block

J Am Coll Cardiol. 2004;44:105–8. http://www.sciencedirect.com/science/article/pii/S0735109704007818.

First Degree Block: Long-Term Outcomes

Heart.2016;102:672–680.http://heart.bmj.com/content/102/9/672?etoc.

Heart Failure During Pacing

Circulation. 2006;113:2082–8. http://circ.ahajournals.org/content/113/17/2082.

LV Function During/After RV Pacing

J Am Coll Cardiol. 2004;44:1883–8. http://www.sciencedirect.com/science/article/pii/S0735109704016043.

RV Pacing For Congenital Heart Block: Detrimental Ventricular Remodeling

Circulation. 2004;110:3766–72. http://circ.ahajournals.org/content/110/25/3766.

Sick Sinus Syndrome: Natural History

Am J Cardiol. 1998;82:1205–9. http://www.ajconline.org/article/S0002-9149(98)00605-5/abstract.

Sinus Node Dysfunction and Pacing

N Engl J Med. 2002;346:1854–62. http://www.nejm.org/doi/full/10.1056/NEJMoa013040.

Updates and More

https://clinicalguidecvd.com/avhb

Chapter 19
Atrioventricular Septal Defect

ICD-10 Code

Q21.1

Alternate Names/Abbreviation [8]

AVSD
AV CANAL DEFECT
ENDOCARDIAL CUSHION DEFECT
OSTIUM PRIMUM ATRIAL SEPTAL DEFECT
SEPTUM PRIMUM DEFECT

Description/Etiology

Large, central defect above AV valve (see Atrial Septal Defect) or may extend to variable degrees above and below AV valve; intercommunication may be small or large

Over 75 % of complete AVSDs are in Down syndrome patients; most partial AVSDs occur in non–Down syndrome patients (>90 %)

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_19,

Most adult patients with AVSD will have had surgical correction in childhood; unrepaired adults may be asymptomatic or may present with HF, exertional limitation, PAH and cyanosis, IE, or atrial flutter/AF

Patients with partial AVSD are more likely to become symptomatic at a younger age if significant left AV valve regurgitation is present

Predisposing/Comorbid Conditions

DOWN SYNDROME
NOONAN SYNDROME
TETRALOGY OF FALLOT

Demography

Gender equal

Pathophysiology

Morphology:

Large, central defect that may lie above AV valve or may extend to variable degrees above and below AV valve

Interventricular communication can be small or large

Common AV valve annulus stretches across both ventricles; may be a common superior leaflet, or superior leaflet may be separated at its distal margin into right and left components

AV valve may be misaligned with respect to the ventricles, in association with hypoplasia of right or left ventricle

Left AV valve is trileaflet with superior and inferior bridging leaflets separated by a mural leaflet

There may be abnormal lateral rotation of posteromedial papillary muscle

Right/left ventricles may be hypoplastic

Signs/Symptoms [2] [3]

BODY, GROWTH – DECR
BREATHING – DIFF (DYSPNEA)
BREATHING – DIFF, RECLINING FLAT (ORTHOPNEA)
BREATHING – DIFF, UPRIGHT (PLATYPNEA)
BREATHING – RAPID (TACHYPNEA)
CHEST – PAIN
CHEST, ANT, L – BULGE
FATIGUE
HEART, LSB, LOWER – IMP, SYS
HEART, LSB, LOWER – MURMUR, DIAS
HEART, LSB, UPPER – IMP, SYS
HEART, LSB, UPPER – MURMUR, SYS [1]
HEART, S2, SPLIT – FIXED
HEART, S2, SPLIT – WIDE
HEART, S3 LV
HEART, S4 LV
HEART, T1, INTENSITY – INCR
NECK, JVP, A WAVE – CREST EQUALS V WAVE
SKIN, COLOR – BLUE (CYANOSIS)
SKIN, COLOR, EFFORT – BLUE (CYANOSIS)

Differentiation

ATRIAL SEPTAL DEFECT
VENTRICULAR SEPTAL DEFECT
OTHER CAUSES OF MR

Complications

AF/Atrial Flutter [7]
Heart Failure
Infective Endocarditis
Pulmonary Arterial Hypertension

Laboratory

NS

ECG

AV COND – 1ST DEGREE BLOCK
DYSRHYTHMIAS – ATRIAL (PACS/OTHERS) [7]
P, DURATION – LONG
QRS – LVH PATTERN [4]
QRS – RVH PATTERN [5]
QRS, AXIS – L [6]
QRS, AXIS – R

Imaging [10]

CARDIOMEGALY
LV, OUTFLOW – OBS
PA, MAIN, SIZE – INCR [5]
PUL, VASCULARITY – INCR

Other Tests

Exercise test [for functional status]
Cardiac catheterization [9]

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic

For specific problems such as ACEI/diuretic for MR
IE Prophylaxis

Treatment: Surgical/Invasive

Primary repair if fixed PAH absent

Course

Highly variable depending on pathoanatomy

Notes

[1] Due to MR but may not radiate to left axilla as regurgitant jet is more medial than in usual forms of MR
[2] Usually clinically manifest in infancy, but may be detected in asymptomatic adults depending on magnitude of pathology
[3] For repaired patients, exam may be normal except for apical systolic murmur due to residual MR or SubAS
[4] Indicates significant AV regurgitation
[5] Indicates PAH
[6] With counterclockwise loop in frontal plane
[7] Especially in older patients
[8] Terms AVSD, AV canal defect, and endocardial cushion defect can be used interchangeably to describe this group of defects
[9] Cardiac catheterization has limited role in assessment of AVSD unless noninvasive findings equivocal; assessment of PAH and coronary anatomy may be needed when reoperation considered
[10] TTE is primary imaging modality:

Should include demonstration of borders of primum ASD, a VSD (if present), morphology and function of AV valve, ventricular size and shunting, and SubAS (if present)

In complete and unrepaired AVSD, study includes presence and size of septal defect, morphology and function of common AV valve, ventricular size and function; when ventricular portion of defect is large, ventricular septum may be deficient apically and inferiorly

Pulmonary artery pressures (generally very high in complete AVSD) should be evaluated by measuring TR and pulmonary regurgitation jet velocity with simultaneous systemic blood pressure measurement

Evidence of subaortic obstruction, caused by AV valve attachments to crest of the IVS, should be sought by imaging and Doppler

In post-repair patients, residua may include left AV valve dysfunction, SubAS, VSD patch leak, and PAH; may be difficult to distinguish residual LV-RA shunt from TR with RV hypertension; failure to distinguish these may result in erroneous diagnosis of PAH

Guidelines

ACC/AHA 2008 guidelines for the management of adults with congenital heart disease

J Am Coll Cardiol. 2008;52:e143–263. http://content.onlinejacc.org/article.aspx?articleid=1188032.

ESC Guidelines for the management of grown-up congenital heart disease (new version 2010)

Eur Heart J. 2010;31:2926–7. http://www.escardio.org/guidelines-surveys/esc-guidelines/Pages/grown-up-congenital-heart-disease.aspx.

Patient Information

Images

http://www.nlm.nih.gov/medlineplus/ency/imagepages/1056.htm.
http://www.nlm.nih.gov/medlineplus/ency/imagepages/1097.htm.
http://www.cdc.gov/ncbddd/heartdefects/AVSD-graphic.html.

Medlineplus

ENGLISH

http://www.nlm.nih.gov/medlineplus/ency/article/001114.htm.

ESPANOL

http://www.nlm.nih.gov/medlineplus/spanish/ency/article/001114.htm.

Genetics Home Reference

http://ghr.nlm.nih.gov/search/term=Atrioventricular+septal+defect+2.

Cleveland Clinic

http://my.clevelandclinic.org/heart/disorders/congenital/septal.aspx.

CDC

http://www.cdc.gov/ncbddd/heartdefects/avsd.html.
http://www.cdc.gov/ncbddd/heartdefects/living.html.

Texas Childrens

http://www.texaschildrens.calls.net/Ventricular-Septal-Defect/?-vsrefdom=p.4965.

Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/atrioventricular-canal-defect/basics/definition/con-20024932.

Professional Information

Review

Heart. 2006;92:1879–85. http://heart.bmj.com/content/92/12/1879.extract.

AV Valve Regurgurgitation

J Am Coll Cardiol. 1998;32:1449–53. http://content.onlinejacc.org/article.aspx?articleid=1125449.

Echo: Obstructive Lesions

J Am Coll Cardiol. 2001;38:253–61. http://content.onlinejacc.org/article.aspx?articleid=1127314.

Electroanatomical Relationships

J Am Coll Cardiol Img. 2009;2:1357–65. http://imaging.onlinejacc.org/article.aspx?articleid=1109507.

Fetal Diagnosis

J Am Coll Cardiol. 2000;36:593–601. http://content.onlinejacc.org/article.aspx?articleid=1126610.

Preoperative Cardiac Catheterization/ Angiography

J Am Coll Cardiol. 1988;11:373–8. http://content.onlinejacc.org/article.aspx?articleid=1113101.

Updates and More

https://clinicalguidecvd.com/avsd

Chapter 20
Brugada Syndrome

Management Keys

- Avoid drugs associated with triggering arrhythmias [21]
- Evaluation and long-term management (both drug and device) should be carried out only by professionals experienced in treating patients and families with Brugada
- Genetic testing and counseling of patients and families by qualified professionals and laboratories

ICD-10 Code

I45.8

Alternate Names/Abbreviation

BRS

Description/Etiology

- Inherited autosomal dominant channelopathy with ST/J wave elevation in right ECG precordial leads

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_20,

Affected persons at increased risk for SCD due to VF
Hallmark of diagnosis: 12 lead ECG features of coved type ST elevation/negative T wave in ECG right precordial leads with/without drug challenge
Other diagnostic features include:

- Agonal respirations during sleep
- Family history of coved-type ECG
- Family history of SCD
- History of VT/VF
- VT/VF inducible during EP study

Comorbid Conditions

ATRIAL FIBRILLATION
ATRIOVENTRICULAR HEART BLOCK
FAMILY HX: SUDDEN DEATH
MONOMORPHIC VENTRICULAR TACHYCARDIA
SINUS NODE DYSFUNCTION
SUPRAVENTRICULAR TACHYCARDIA
WOLFF-PARKINSON-WHITE SYNDROME [AND OTHER PREEXCITATION SYNDROMES]

Demography

Most common in SE Asia
Males 8:1 in phenotypic expression [1]
Most often initial clinical manifestations occur age 20–40 years but may occur at all ages from infancy to very aged
SCD/VF may be first manifestation

Pathophysiology

Increased collagen content in RVOT associated with

- Epicardial surface/intramyocardial fibrosis
- Decreased gap junction protein expression

Two mechanisms for arrhythmias proposed:

Depolarization abnormality
Repolarization abnormality

Signs/Symptoms

BREATHING – DIFF, NOCTURNAL (DYSPNEA, NOCT)
CARD ARREST
CHEST – PALPITATIONS
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)
MENTATION – CONFUSION [5]
MENTATION – NS CHANGES [5]
MENTATION, MEMORY – IMPAIRED (AMNESIA) [5]
SEIZURES [5]

Differentiation

Arrhythmogenic Right Ventricular Dysplasia
Catecholaminergic Polymorphic Ventricular Tachycardia
Drug (Especially antiarrhythmics) ECG Effects
Long QT Syndrome – Congenital
Long QT Syndrome – Acquired
Seizure Disorder [5]
Short QT Syndrome

Complications [2] [3] [4] [22]

Dysrhythmias [14]
Seizures
Sudden death (mean age 41 ± 15 years)
Syncope

Laboratory

NS

ECG [20]

AV COND – 1ST DEGREE BLOCK
AV CONDUCTION – AV DISSOCIATION, COMPLETE [11]
DYSRHY – PREEXCITATION
DYSRHYTHMIAS – ATRIAL (PACS/OTHERS) [16]
DYSRHYTHMIAS – VENTRICULAR (PVCS/ OTHERS) [14]
EPSILON WAVE
J WAVE (OSBORN WAVE)
P WAVE, DUR – INCR
QRS – FRAGMENTATION [8] [10]
QRS – LONG, NS
QRS – RBBB/RBBB PATTERN [17]
QT/QTC INTERVAL – LONG [8]
S WAVE, LEAD I – DEEP/WIDE [24]
ST SEGMENT – ELEV [6] [8] [ESP DURING FEVER IN CHILDREN]
T WAVE – INVER, ABN [7] [8]

Imaging

NS

Genomics

CACNA1C
GPD1L
HCN4
KCND3

KCNE3
KCNE5
MOG1
SCN10A
SCN1B
SCN3B
CACNB2b
SCN5A [19]

Other Tests

Ambulatory ECG for nighttime detection of characteristic ECG changes
TST [12]
Programmed ventricular stimulation [23]
Provocative drug testing [13]

Treatment: Nonpharmacologic

Fever control

Treatment: Pharmacologic

Antipyretics for fever
Antiarrhythmics [15]
Isoproterenol for acute VF
Long term oral treatment for VF suppression:

- Bepridil
- Cilostazol
- Denopamine
- Quinidine

Treatment: Surgical/Invasive

ICD
RF ablation

Prevention (of complications)

Avoid strenuous sports
Family screening

Notes

[1] Leading cause of SCD in males age <40 years
[2] Especially early morning hours and with bradycardia
[3] Triggered by fever, large meals, alcohol, cocaine
[4] Triggered by drugs, especially acetylcholine and certain:

Antiarrhythmics
Psychotropics
Anesthetics
For current drug list see: www.brugadadrugs.org

[5] May present with seizures and postictal state; some antiepileptic drugs may precipitate Brugada dysrhythmias, especially carbamazepine and phenytoin
[6] Coved downward or saddleback shape
[7] Little/no separation from ST segment
[8] Right precordial leads; changes may be more pronounced with lead placement cephalad from standard positions
[9] Characteristic ECG changes highly variable in individual patients; may be unmasked by many different stimuli in [3] and [4]
[10] Appears as multiple small spikes within QRS
[11] May occur in absence of QT or ST abnormalities

[12] May accentuate some ECG changes, eg, QRS widening, ST elevation

[13] Drug challenge with sodium channel blockers **(caution: must be performed only by trained personnel using continous ECG monitoring with full resuscitation equipment)**:

Ajmaline
Flecainide
Pilsicainide
Procainamide

[14] Especially Polymorphic VT

[15] Class 1a, 1c, and beta-blockers should be avoided; see www.brugadadrugs.org

[16] 20 % have AF

[17] RBBB may hide Brugada ECG features

[18] ICDs: efficacious but associated with high frequency of inappropriate shocks and complications

[19] SCN5A: most common genotype, inherited as autosomal dominant trait with incomplete penetrance; actual causal role of SCN5A not established

Also associated with other disorders, including:
Congenital atrial standstill
Dilated cardiomyopathy
Long QT syndrome type A
Progressive cardiac conduction disease
Sinus node dysfunction
Combinations of above

[20] Fever associated with ECG changes

[21] Listed at www.brugadadrugs.org

[22] Includes asymptomatic persons with Brugada phenotype

[23] Programmed ventricular stimulation: provoked arrhythmias associated with future ventricular arrhythmia risk; absence of arrhythmia provocation does not necessarily portend low ventricular arrhythmia risk, especially in patients with high-risk clinical features

[24] Wide/large lead I S-wave may be strong predictor of life-threatening ventricular arrhythmias in Brugada patients with no history of cardiac arrest at presentation; larger population studies needed for confirmation

Guidelines

HRS/EHRA/APHRS expert consensus statement on the diagnosis and management of patients with inherited primary arrhythmia syndromes

Heart Rhythm. 2013;10:1932–63. http://www.sciencedirect.com/science/article/pii/S1547527113005523.

Patient Information

Genetics Home Reference

http://ghr.nlm.nih.gov/condition/brugada-syndrome.

Mayo (English)

http://www.mayoclinic.org/diseases-conditions/brugada-syndrome/basics/definition/con-20034848.

SADS Foundation

http://www.sads.org/sads/media/sads-materials---brochures/sads-brugada-brochure-3-2011.pdf.

Texas Heart Institute

http://www.texasheartinstitute.org/HIC/Topics/Cond/brugada.cfm.

Professional Information

Early Description

J Am Coll Cardiol. 1992;20:1391–6. http://content.onlinejacc.org/article.aspx?articleID=1118420.

Heart Rhythm Society/European Heart Rhythm: Special Report

Circulation. 2005;111:659–70. http://circ.ahajournals.org/content/111/5/659.full.pdf+html.

Review/History

Eur Heart J. 2013;34:3610–5. http://eurheartj.oxfordjournals.org/content/34/47/3610.

Review

Circulation Arrhyth Electrophysiol. 2012;5:606–16. http://circep.ahajournals.org/content/5/3/606.full.

Review

J Am Coll Cardiol. 2008;51:1176–80. http://content.onlinejacc.org/article.aspx?articleid=1187781.

Review

Pacing Clin Electrophysiol. 2006;29:1130–59. http://onlinelibrary.wiley.com/doi/10.1111/j.1540-8159.2006.00507.x/abstract.

Review: Genetics of Sudden Cardiac Death

Circulation Research. 2015;116:1919–36. http://circres.ahajournals.org/content/116/12/1919.full.

Atrial Fibrillation

J Am Coll Cardiol. 2008;51:1149–53. http://content.onlinejacc.org/article.aspx?articleid=1187752.

Case Report

QJM. 2015;108:235–7. http://qjmed.oxfordjournals.org/content/108/3/235.extract?etoc.

Drugs Associated with Brugada

Brugada.org. http://www.brugadadrugs.org.

ECG/Fatal Events

J Am Coll Cardiol. 2014;63:2131–8. http://content.onlinejacc.org/article.aspx?articleID=1859511.

ECG: Lead 1 S Wave Marker of SCD

J Am Coll Cardiol. 2016;67:1427–40. http://content.onlinejacc.org/article.aspx?articleID=2505132.

Fever/Brugada ECG Changes

Circulation. 2013;127:2145–6. http://circ.ahajournals.org/content/127/21/2145.full.

Fibrosis/Connexin-43/Conduction Abnormality

J Am Coll Cardiol. 2015;66:1976–86. http://content.onlinejacc.org/article.aspx?articleID=2465341.

Genetic Basis

Heart Rhythm. 2007;4:756–7. http://www.sciencedirect.com/science/article/pii/S154752710700272X.

Genetic Testing (ED)

J Am Coll Cardiol. 2012;60(15):1419–20. http://content.onlinejacc.org/article.aspx?articleid=1224884.

ICD: 20-YR Experience

J Am Coll Cardiol. 2015;65:879–88. http://content.onlinejacc.org/article.aspx?articleID=2174617.

Monomorphic Ventricular Tachycardia

Heart Rhythm. 2016;13:669–82. http://www.heartrhythmjournal.com/article/S1547-5271(15)01368-5/fulltext.

Mutations: SCN10A

J Am Coll Cardiol. 2014;64:66–79. http://content.onlinejacc.org/article.aspx?articleID=1886832.

Outcomes Post-ICD Implantment

Circulation. 2013;128:1739–47. http://circ.ahajournals.org/content/128/16/1739.full.

Programmed Ventricular Stimulation: Risk Stratification

Circulation. 2016;133:622–30. http://circ.ahajournals.org/content/133/7/622.full.

RBBB: Hidden Brugada

Circulation. 2013;128:1048–54. http://circ.ahajournals.org/content/128/10/1048.full.

Risk Stratification

Heart Rhythm. 2016;13:299–310. http://www.heartrhythmjournal.com/article/S1547-5271(15)01128-5/abstract.

Risk Stratification: Asymptomatic Patients with Brugada Phenotype

Am J Cardiol. 2015;116:98–103. http://www.ajconline.org/article/S0002-9149(15)01050-4/abstract.

SCN10A: Common/Rare Variants

Cardiovasc Res. 2015;106:520–9. http://cardiovascres.oxfordjournals.org/content/106/3/520.full.

Syncope

Heart Rhythm. 2014. doi:10.1016/j.hrthm.2014.10.014. http://www.sciencedirect.com/science/article/pii/S1547527114011424#.

Ventricular Arrhythmias In Channelopathies: Management

Circulation Arrhyth Electrophysiol. 2015;8:221–31. http://circep.ahajournals.org/content/5/3/606.extract.

Updates and More

https://clinicalguidecvd.com/brugada

Chapter 21
Cantu Syndrome

ICD-10 Code

Q87.3

Alternate Names/Abbreviation

HYPERTRICHOTIC OSTEOCHONDRODYSPLASIA
CRANIOFACIAL-CARDIAC-SKELETAL SYNDROME

Description/Etiology

Multisystem (including CV, musculoskeletal, integumentary) congenital syndrome with autosomal dominant inheritance

Characterized by abnormalities of:

- Behavior
- CV system
- Facial features [18]
- Hair growth (hypertrichosis)
- Skeletal system [19]

Cardiovascular abnormalities include:

- AV Stenosis/bicuspid AV

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_21,

Cardiomegaly
Concentric ventricular hypertrophy (normal ventricular contractility)
MR
PDA (50 %)
Pericardial effusion (20 %)
Partial pulmonary venous obstruction
Pulmonary Hypertension
Increased vascular tortuosity

Intellect typically normal
Single gene disorder: ABCC9

Comorbid Conditions [11]

CARDIAC HYPERTROPHY [1]
IMMUNE DEFICIENCY
PATENT DUCTUS ARTERIOSUS [1] [13]
PERICARDIAL EFFUSION [1]
POLYHYDRAMNIOS
PULMONARY HYPERTENSION [1]

Demography

Initial clinical presentation: all ages from birth to adulthood

Pathophysiology

Varies according to system involvement

Signs/Symptoms [6]

ARTERIAL PULSE – DOUBLE (BISFERIENS) [4]
ARTERIAL PULSE PRESSURE – INCR [4]

ARTERIAL PULSE, FALL – RAPID [4]
ARTERIAL PULSE, RISE – RAPID [4]
BACK, CURV – LAT (SCOLIOSIS)
BODY, APPEARANCE – MUSCULAR
BREATHING – DIFF (DYSPNEA) [4]
BREATHING – STRIDOR [12]
CHEST – PAIN [4]
CHEST, ANT, L – BULGE [4]
EXTREM – LYMPHEDEMA
EXTREM, FINGERS, PADS – FETAL
EXTREM, LOWER, DIGITS – CLUBBING [4]
EXTREM, LOWER, DIGITS, COLOR – BLUE (CYANOSIS) [4]
EXTREM, PALMS/SOLES – CREASES, DEEP
EXTREM, UPPER, L, DIGITS – CLUBBING [4]
EXTREM, UPPER, L, DIGITS, COLOR – BLUE (CYANOSIS) [4]
EYES, PALPEBRAL FISS – LONG
EYES, SEPARATION – WIDE (HYPERTELORISM)
FACE, APPEARANCE – COARSE
FACE, NASAL BRIDGE – FLAT [2]
FACE, PHILTRUM – LONG
FATIGUE
HAIR – EXCESS (HYPERTRICHOSIS) [14]
HAIR, HEAD, ANT LINE – LOW
HEAD, SIZE – INCR (MACROCEPHALY)
HEART, LSB, UPPER – MURMUR, CONT [4]
HEART, LSB, UPPER – THRILL, CONT [4]
HEART, LV, APEX – IMP, TRIPLE [4]
HEART, LV, APEX – MURMUR, DIAS [4]
HEART, LV, APEX, IMP – FORCEFUL/SUSTAINED [4]
HEART, P2, INTENSITY – INCR [5]
HEART, S2, SPLIT – REVERSED (PARADOXICAL) [4]
JOINTS – LAX
JOINTS – SMALL
LIPS – THICK
LIVER – ENLARGED (HEPATOMEGALY) [8]

MENTATION, LEARNING, DEVELOPMENT – DECR [9] [10]
MOOD – ANXIOUS
MOUTH, GINGIVA, THICKNESS – INCR (GINGIVAL HYPERTROPHY)
SKIN – SOFT [3]
SKIN, COLOR – DARK (HYPERPIGMENTATION)
STERNUM, CURV – ANT (PECTUS CARINATUM) [4]
VOICE – HOARSE (LARYNGITIS)

Differentiation

Other causes of hypertrichosis, including:

Acquired hypertrichosis lanuginosa [17]
Porphyria cutanea tarda
Malnutrition, eg anorexia nervosa
Malignancy
Drugs, such as

Androgens
Cyclosporin
Minoxidil
Pheytoin

Complications [11]

GI Bleeding [7]
Obstructive Sleep Apnea
Recurrent respiratory infections

Laboratory

NS

ECG [6]

AV COND – 1ST DEGREE BLOCK [4]
P WAVE – FLAT [4]
P WAVE – TALL/PEAKED [4]
P WAVE, DUR – INCR [4]
QRS – BVH PATTERN [4]
QRS – LVH PATTERN [4]
QRS – RVH PATTERN [4]

Imaging [6]

CARDIOMEGALY [4]
DUCTUS ARTERIOSUS – CALCIUM [4]
LA, CHAMBER, SIZE – INCR [4]
LV, CHAMBER, SIZE – INCR [4]
PA, MAIN, SIZE – INCR [4]
PERICARD – FLUID [4]
PUL, VASCULARITY – INCR [4]

Genomics

ABCC9

Other Tests

NS

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic

NS [16]

Treatment: Surgical/Invasive

PDA Closure [15]

Percutaneous
Surgical

Course

Varies according to pathology

Notes

[1] CV manifestations of Cantu Syndrome
[2] Infants
[3] "Doughy"
[4] Due to PDA
[5] Due to PDA/PAH
[6] Does not include features of PAH, LVH, or pericardial effusion
[7] Due to duodenal ulcer/esophagitis
[8] Early childhood
[9] Usually mild, may include motor/speech delay
[10] IQ increased in some affected persons
[11] Numerous other abnormalities, including pituitary adenoma, hydrocephalus, renal changes, retinal artery occlusion, behavior disorders, have been reported in isolated cases
[12] Due to laryngomalacia
[13] May occur in >50 % of cases
[14] EG, bushy eyebrows, excess hair on extensor surfaces of limbs/back
[15] Not indicated for PAH and net R-L shunt

[16] ABCC9 antagonists (eg, Glibenclamide) suggested for possible treatment

[17] Occurs in persons people who may later develop some form of cancer; also known as "malignant down" often confined to facial and other sites that are normally hairless; mechanism of cancer relation unknown

[18] Craniofacial features include:

Anterior open bite
Anteverted nostrils
Broad nasal bridge
Coarse facial appearance
Epicanthal folds
Gingival hyperplasia
High or narrow palate
Long philtrum
Macroglossia
Wide mouth with full lips

[19] Skeletal abnormalities include:

Broad ribs
Coxa vara
Delayed bone age
Erlenmeyer flask-like long bones with metaphyseal flaring
Hypoplastic ischium and pubic bones
Narrow obturator foramen
Narrow shoulders/thorax
Osteopenia
Platyspondyly and ovoid vertebral bodies
Scoliosis
Thickened calvarium

Guidelines

NS

Patient Information

Treatment: Nonpharmacologic

http://ghr.nlm.nih.gov/condition/cantu-syndrome.

Patient Support

http://cantu-syndrome.org/index.html.

ESPANOL

http://rarediseases.info.nih.gov/resources/6/recursos-en-espanol.

Professional Information

ABCC9 Mutation/CANTU

Nature Genetics. 2012;44:793–6. http://www.nature.com/ng/journal/v44/n7/full/ng.2324.html.

ABCC9 Mutation: Wide Clinical Variability

Am J Med Genet A. 2013;161A:295–300. http://www.ncbi.nlm.nih.gov/pubmed/23307537.

Cardiac Pathology

Circulation. 2014;130:A19680. http://circ.ahajournals.org/content/130/Suppl-2/A19680.abstract?sid=f5a42990-dd9a-40ca-9d57-f204e30317ec.

Genomics

Am J Human Genet. 2012;90:1094–101. http://ac.els-cdn.com/S0002929712002170/1-s2.0-S0002929712002170-main.pdf?-tid=894ec2d6-845c-11e3-9954-00000aab0f26&acdnat=1390502052-c721c5ab83a9db8acf08964958db0999.

K_{ATP} Channels and Cardiovascular Disease

Circulation Research. 2013;112:1059–72. http://circres.ahajournals.org/content/112/7/1059.full.

Phenotype

Am J Med Genet. 2011;155:508–18. http://onlinelibrary.wiley.com/doi/10.1002/ajmg.a.33885/pdf.

Updates and More

https://clinicalguidecvd.com/cantu

Chapter 22
Carcinoid Heart Disease

Management Keys

- Use echo as primary diagnostic modality of cardiac lesions
- Consider cardiac valve surgery to relieve cardiac symptoms
- Engage multidisciplinary team experienced in dealing with Carcinoid Heart Disease to provide optimal management of this complex condition

ICD-10 Code

E34.0

Alternate Names/Abbreviation

NS

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_22,

Description/Etiology

- Carcinoid tumors: neuroendocrine cancer most often originating from GI tract enterochromaffin cells, usually growing slowly over a period of years in gastrointestinal tract; affected persons usually asymptomatic until tumors become large or metastasize
- Carcinoid tumors in midgut may secrete large amounts of vasoactive substances, including 5-hydroxytryptamine (5-HT), tachykinins, and prostaglandins, which are mostly inactivated by liver
- Carcinoid syndrome:
  - Characterized by flushing, diarrhea, bronchospasm
  - Occurs when tumor cells metastasize to liver as vasoactive substances reach systemic circulation via hepatic vein
- Fibrous tissue deposits form on endocardium of TV, PV, RA, and RV (MV and AV less often affected) due to effects of circulating vasoactive hormones [1]
- Carcinoid heart disease usually preceded by months-years of carcinoid syndrome, but up to 20 % of patients with carcinoid syndrome present with cardiac involvement at initial diagnosis [2]
- Tricuspid regurgitation is most common lesion seen in carcinoid heart disease (up to 90 %)
- Patients may be functional NYHA class I despite severe right heart lesions, but features of RHF, including dyspnea, LE edema, fatigue develop with disease progression
- Left heart lesions occur in about 15 % of cases

Comorbid Conditions

CARCINOID SYNDROME/TUMOR

Demography

Incidence: about 1 in 100,000 of general population

Pathophysiology

Vasoactive substances secreted by carcinoid metastatic tumor cells in liver reach right heart, causing fibrous tissue deposition on endocardial surfaces

Valve regurgitation/stenosis secondary to leaflet fibrosis, thickening, retraction, and fixation [10]

Left heart lesions: diffuse valve leaflet thickening, usually less severe than right heart valve lesions; serotonin believed to be inactivated when passing through lung parenchyma, so left heart valve lesions may be due to patent foramen ovale with R-L shunt, bronchial carcinoid, or high levels of circulating vasoactive substances

Signs/Symptoms

ABDOMEN – FLUID (ASCITES)
BOWEL MOVEMENTS – DIARRHEA [2]
BREATH SOUNDS – WHEEZES [2]
BREATHING – DIFF (DYSPNEA)
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE) [2]
DIZZY/LIGHTHEADED/PRESYNCOPE [2]
EXTREM, LOWER, BILAT – EDEMA
FATIGUE
HEART, LSB, LOWER – IMP, SYS
HEART, LSB, LOWER – MURMUR, DIAS
HEART, LSB, LOWER – MURMUR, SYS [3]
HEART, LSB, UPPER – MURMUR, DIAS
HEART, LSB, UPPER – MURMUR, SYS
HEART, RATE, VAR – DECR

HEART, RATE – INCR (TACHYCARDIA) [2]
HYPOTENSION (BLOOD PRESSURE – DECREASED/LOW) [2]
LIVER – ENLARGED (HEPATOMEGALY)
LIVER – PULSATION, SYS [3]
NAUSEA [2]
NECK, JVP – ELEV [3]
NECK, JVP, V WAVE – INCR/LARGE [3]
SKIN – FLUSHING [2]
SKIN – TELANGIECTASES
SKIN, COLOR – BLUE (CYANOSIS) [4]
VOMITING (EMESIS) [2]

Differentiation

Other causes of Tricuspid Regurgitation
Pulmonary Arterial Hypertension

Complications

AF
Constrictive pericarditis (rare)
HF

Laboratory

BLOOD, ANP – INCR [6]
BLOOD, NT-PROBNP – INCR [6]
URINE, 5-HIAA – INCR [5] [6]

ECG [7]

DYSRHYTHMIAS – ATRIAL (PACS/OTHERS)
P WAVE – TALL/PEAKED

QRS – RBBB/RBBB PATTERN
QRS, AMP – DECR
RATE – INCREASED (SINUS TACHYCARDIA)
ST-T WAVE – ABN, NS

Imaging

CARDIOMEGALY
LUNGS – NODULE(S) [9]
MV, LEAFLETS – THICK
PERICARD – FLUID
PLEURA – FLUID
PV, FLOW – REGURG
PV, LEAFLETS – THICK [8]
PV, PRESS – GRADIENT
RA, CHAMBER, SIZE – INCR
RV, CHAMBER, SIZE – INCR
TV, FLOW – OBS
TV, FLOW – REGURG
TV, LEAFLET, COAPTATION – INCOMPLETE [RETRACTED] [8]
TV, LEAFLETS – THICK

Other Tests

NS

Treatment: Nonpharmacologic [12]

HF regimen

Treatment: Pharmacologic [12]

HF regimen
Somatostatin analogues [11]

Treatment: Surgical/Invasive [12]

General:

Cancer resection when possible

Cardiac: [13]

Balloon valvuloplasty
Valve replacement

Course

Variable

Notes

[1] Tumors convert tryptophan to serotonin; most often located in GI tract, especially appendix and terminal ileum; other sites include bronchi and gonads
[2] Episodic symptoms due to release of serotonin and other vasodilating hormones, lasting minutes-hours, sometimes triggered by stress, exercise, alcohol ingestion
[3] Due to TR
[4] In presence of patent foramen ovale
[5] Breakdown product of serotonin; falsely elevated by foods high in tryptophan content, including:

Avocados
Bananas
Eggplant
Pineapples
Plums
Tomatoes
Walnuts

[6] High levels correlate with worse prognosis
[7] Normal in 50 % of cases

[8] Calcification rare and presence suggests another diagnosis
[9] With pulmonary metastases
[10] Almost all cases involve right heart, but MV/AV thickening/stenosis can occur in rare cases
[11] Inhibit release of serotonin and other biogenic amines and peptides; cause marked symptoms improvement but effect on survival not established
[12] Goals of treatment mainly symptom reduction, improved QOL, increased survival by tumor hormone inhibition or tumor load reduction
[13] Valve surgery may cause marked symptomatic improvement of patients in >1 New York Heart Association class

Guidelines

NS

Patient Information

Images

http://www.nlm.nih.gov/medlineplus/ency/imagepages/9877.htm.

Medlineplus

ENGLISH

http://www.nlm.nih.gov/medlineplus/ency/article/000347.htm.

ESPANOL

http://www.nlm.nih.gov/medlineplus/spanish/ency/article/000347.htm.

Mayoclinic

http://www.mayoclinic.org/diseases-conditions/carcinoid-syndrome/basics/definition/con-20027127.

Professional Information

Editorial

British Cardiovascular Society. https://www.bcs.com/pages/news_full.asp?NewsID=19792009.

Review

Circulation. 2007;116:2860–5. http://circ.ahajournals.org/content/116/24/2860.full?sid=9ae741f8-d724-487a-bc3f-4dbad479a8de.

Review

Heart. 2004;90:1224–8. http://heart.bmj.com/content/90/10/1224.full?sid=be2fef7e-a213-4e9a-b5d2-efe41706c9d6.

Cardiac Metastases (Echo)

J Am Coll Cardiol. 2002;40:1328–32. http://content.onlinejacc.org/article.aspx?articleid=1130376&resultClick=3.

Constrictive Pericarditis

Heart. 1999;82:641–3. http://heart.bmj.com/content/82/5/641.full?sid=e70fc97e-8ac6-47bd-9a8e-68ac5027a6d7.

Echo

Circulation. 1993;87:1188–96. http://circ.ahajournals.org/content/87/4/1188.abstract?sid=9ae741f8-d724-487a-bc3f-4dbad479a8de.

Images

J Am Coll Cardiol. 2010;55:1996–1996. http://content.onlinejacc.org/article.aspx?articleid=1142783&resultClick=3.

Prognosis

Circulation. 2005;112:3320–7. http://circ.ahajournals.org/content/112/21/3320.full?sid=9ae741f8-d724-487a-bc3f-4dbad479a8de.

Serotonin Levels and Cardiac Manifestations

Circulation. 1995;92:790–5. http://circ.ahajournals.org/content/92/4/790.full?sid=9ae741f8-d724-487a-bc3f-4dbad479a8de.

Surgery

J Am Coll Cardiol. 1995;25:410–6. http://content.onlinejacc.org/article.aspx?articleid=1119782&resultClick=3.

Surgery: Left Heart Valve Disease

Circulation. 2001;104:I-36–I-40. http://circ.ahajournals.org/content/104/suppl-1/I-36.full?sid=9ae741f8-d724-487a-bc3f-4dbad479a8de.

Surgery: PV Replacement

Circulation.2002;106:I-51–I-56.http://circ.ahajournals.org/content/106/12-suppl-1/I-51.full?sid=9ae741f8-d724-487a-bc3f-4dbad479a8de.

Updates and More

https://clinicalguidecvd.com/carcinoid

Chapter 23
Cardiac Allograft Vasculopathy

ICD-10 Code

T86.290

Alternate Names/Abbreviation

CAV
Transplant allograft vasculopathy

Description/Etiology

Diffuse cardiac vascular intimal hyperplasia and thickening post-cardiac transplantation, affecting 30–50 % of patients by 5 years post-transplant [5]
Involves epicardial and intramyocardial coronary arteries and veins
Angiographic definition: stenosis >50 % of intraluminal diameter
Early diagnosis of CAV difficult due to:

- Lack of ischemic symptoms in denervated allograft
- Insensitivity of coronary angiography (often underestimates disease extent/severity)

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_23,

Involvement of small intramyocardial vessels
Functional coronary alterations independent of morphological changes

After 1 year post-cardiac transplant: cancer and CAV are most common causes of death

Comorbid Conditions

DONOR

BRAIN DEATH DUE TO INTRACEREBRAL HEMORRHAGE [8]
HEPATITIS B
HEPATITIS C

RECIPIENT

CYTOMEGALOVIRUS INFECTION
DIABETES MELLITUS
DYSLIPIDEMIA [1]
HLA ANTIGEN MISMATCH
HYPERHOMOCYSTEINEMIA
HYPERTENSION – SYSTEMIC ARTERIAL
INFLAMMATION [2]
PERITRANSPLANT MYOCARDIAL ISCHEMIA
RECURRENT CELLULAR REJECTION

TOBACCO USE

Demography

Age >40 years

Pathophysiology

Mechanism (s) incompletely understood
Smooth muscle proliferation and extracellular matrix protein secretion leading to intimal hyperplasia and

thickening; advanced stages may include atherosclerotic processes [3]

Signs/Symptoms [4]

CHEST – PAIN, EFFORT (ANGINA PECTORIS)

Differentiation

Cardiac rejection
Coronary artery atherosclerotic disease

Complications

AMI
Dysrhythmias
HF
SCD

Laboratory

BLOOD, C-REACTIVE PROTEIN – INCR [2]
BLOOD, TROPONIN – INCR

ECG

N/NS ABN

Imaging

CORONARY ARTERY INTIMA, THICKNESS – INCR [INTRAVASCULAR ULTRASOUND]

Other Tests

Coronary angiography [7]
Dobutamine stress echo [10]

Treatment: Nonpharmacologic

Lifestyle for atherosclerotic risk factor modification and optimal exercise tolerance

Treatment: Pharmacologic

Immunosuppressants

Calcineurin inhibitors
Mycophenolate mofetil
Proliferation signal inhibitors

Serolimus/everolimus

Statins
Vasodilators

CCBs

Treatment: Surgical/Invasive

PTCA with stenting [9]
CABG [9]
Repeat transplantation

Prevention

Lifestyle optimization for atherosclerotic risk factor modification
Statins

Course

Highly variable and changing as new therapies and insights become available

Notes

[1] Highly important
[2] CRP: early marker of CAV
[3] Differences of CAV from typical atherosclerotic plaque include:

- Absence of calcium
- Concentric distribution
- Intact internal elastic lamina
- Involvement of intramyocardial arteries and veins
- Presence of inflammation

[4] Cardiac symptoms usually absent due to denervation, although reinnervation eventually occurs, and angina may by present after 5 years in presence of myocardial ischemia; other clinical manifestations include HF (eg, dyspnea, hepatomegaly) and dysrhythmias (eg, syncope)
[5] Noncardiac vasculature unaffected
[6] IVUS: The "gold standard" for detecting early CAV; intimal thickness ≥5 mm
[7] Much less sensitive than IVUS due to lesion concentricity
[8] "Explosive" brain death associated with catecholamine release, intensive vasoconstriction, and upregulation of circulating inflammatory cytokines
[9] Short-term benefit only in select cases; CABG has high periprocedural mortality
[10] Dobutamne stress echo:

- Sensitivity 80 % compared with coronary angiography
- Specificity up to 88 % when intimal thickening by intravascular ultrasound is used as "gold standard"
- May have predictive value for development of future CAV and outcome

Guidelines

NS

Patient Information

http://www.ncbi.nlm.nih.gov/pmc/articles/PMC3211528/table/t1-12/.

Professional Information

Review

Circulation. 1997;96:2069–77. http://circ.ahajournals.org/content/96/6/2069.full?sid=d34043c7-1dee-4473-b754-c3be7f009f24.

Review

Circulation. 2008;117:2131–41. http://circ.ahajournals.org/content/117/16/2131.full?sid=d34043c7-1dee-4473-b754-c3be7f009f24.

Review

World J Transplant. 2014;24:276–93. http://www.ncbi.nlm.nih.gov/pubmed/25540736.

CT Angiography

J Am Coll Cardiol. 2014;63:1992–2004. http://content.onlinejacc.org/article.aspx?articleID=1851425.

Donor-Specific Antibody

Am J Clin Pathol. 2014;142:809–15. http://www.ncbi.nlm.nih.gov/pubmed/25389335.

Imaging: Noninvasive DX

Heart. 2013;99:445–53. http://heart.bmj.com/content/99/7/445.abstract.

Imaging: Progression

J Am Coll Cardiol. 2013;61:e149–e149. http://content.onlinejacc.org/article.aspx?articleID=1656473.

Inflammatory Markers

PLoS One. 2014;9:e113260. doi: 10.1371. http://www.ncbi.nlm.nih.gov/pubmed/25490200.

Intracoronary Imaging

Circulation Cardiovasc Imaging. 2015;8:e002636. http://circimaging.ahajournals.org/content/8/1/e002636.extract?etoc.

MRI Assessment

J Am Coll Cardiol. 2014;63:799–808. http://content.onlinejacc.org/article.aspx?articleID=1792232.

Screening

J Heart Lung Transplant. 2014. http://www.sciencedirect.com/science/article/pii/S1053249814014399.

Sirolimus

J Heart Lung Transplant. 2013;32:784–91. http://www.ncbi.nlm.nih.gov/pmc/articles/PMC3727915/.

Updates and More

https://clinicalguidecvd.com/cav

Chapter 24
Cardiac Amyloidosis (Transthyretin-Associated Familial Amyloidosis)

Management Keys

- Consider this diagnosis in any patient with Heart Failure and preserved ejection fraction
- Refer to practitioners/centers with amyloid expertise when suspected
- Early diagnosis/typing of amyloid type for specific treatment
- Evaluate for associated plasma cell dyscrasia
- Caution using ACEIs or ARBs [30]
- Screen persons with family history of Cardiac Amyloidosis
- Differentiate from Hypertrophic Cardiomyopathy when LVOT obstruction detected by echo
- Consider medical treatmnet of TTR form with TTR stabilizers [32]

ICD-10 Code

E85.4

Alternate Names/Abbreviation

- AL Amyloid
- FAP Amyloid

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_24,

SSA Amyloid
TTR Transthyretin-associated familial amyloidosis (hereditary form)

Description/Etiology

Form of Restrictive/Infiltrative Cardiomyopathy caused by amyloid protein deposition in heart
Usually part of systemic disease, but may occur in isolation
Forms affecting heart:

- Isolated atrial (IAA) [25]
- Familial (FAP)
- Light chain (AL)
- Non-transthyretin variants
- Secondary (AA) (most common form in developing nations)
- Senile systemic (SSA)

WHO stages:

1. No symptoms/occult cardiac amyloid by biopsy/noninvasive test
2. Asymptomatic cardiac involvement by biopsy/noninvasive test or unexplained ECG low voltage [28]
3. Compensated symptomatic cardiac involvement
4. Uncompensated cardiomyopathy

Comorbid Conditions

CARPAL TUNNEL SYNDROME [33]
LYMPHOMA
MACROGLOBULINEMIA
MULTIPLE MYELOMA
NEUROPATHY – AUTONOMIC [35] [36]
NEUROPATHY – PERIPHERAL [35] [36] [>50 %]
SYSTEMIC AMYLOID DEPOSITION [1]

Demography

AA

Variable with associated condition

AL

Gender equal
Usually age >50 year

FAP

Mid/advanced age

IAA

Advanced age

SSA

Age >65 years
Males (almost 100 %)

Pathophysiology

Cardiac: amyloid protein deposition in myocardium, conduction paths, and coronary arteries causing LV/RV systolic/diastolic dysfunction
Associated with:

Systemic Amyloidosis (most cases)
Serum amyloid A
Transthyretin-related
Immunoglobin (AL)

Signs/Symptoms [17]

ABDOMEN – FLUID (ASCITES)
ABDOMEN – PAIN [2] [ESP RUQ]
APPETITE – DECR (ANOREXIA)

ARTERIAL PRESSURE, UPRIGHT – DECR (ORTHOSTATIC HYPOTENSION)
BOWEL MOVEMENTS – CONSTIPATION
BOWEL MOVEMENTS – DIARRHEA
BREATHING – DIFF (DYSPNEA)
CHEST – PAIN, EFFORT (ANGINA PECTORIS)
CHEST – PALPITATIONS
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE) [3]
CONSCIOUSNESS – LOSS, SUDDEN, EFFORT (EFFORT SYNCOPE) [4]
DIZZY/LIGHTHEADED/PRESYNCOPE
EXTREM – NUMB
EXTREM – PAIN, SHOOTING (PARESTHESIAS)
EXTREM, DIGITS, NAILS – DEGENERATION (DYSTROPHY) [AL]
EXTREM, LOWER, BILAT – EDEMA
EYES – PANDA/RACCOON [26]
EYES, PERIORBITAL – ECCHYMOSES
EYES, SCLERA – YELLOW (ICTERUS)
HEART, LSB, LOWER – MURMUR, SYS [20]
HEART, LSB, MID – MURMUR, SYS [22]
HEART, LV, APEX – MURMUR, SYS [21]
HEART, RSB, UPPER – MURMUR, SYS [22]
HYPOTENSION (BLOOD PRESSURE – DECREASED/LOW)
LIVER – ENLARGED (HEPATOMEGALY)
MOUTH – DRY (XEROSTOMIA)
NAUSEA
NECK, JVP – ELEV
SKIN – PETECHIAE/ECCHYMOSES/PURPURA
SYNDROME – CARPAL TUNNEL [33]
TONGUE, SIZE – INCR (MACROGLOSSIA) [AL]
URINE – FROTHY
VOICE – HOARSE (LARYNGITIS)
WEIGHT – LOSS [5]
WEIGHT – LOSS

Differentiation

Cancer cardiac infiltration
Cardiac Sarcoidosis
Cardiomyopathy – Hypertrophic
Cardiomyopathy – Restrictive
Cardiomyopathy – Iron Overload
Fabry Disease
Glycogen Storage Disease
Hypothyroidism
LVOT obstruction from other causes
LVH from other causes
Other infiltrative cardiomyopathies

Complications

AF
HF
High grade heart block
SCD
Thromboembolism

Laboratory [17]

BLOOD, ALK PHOS – INCR [14]
LOOD, HGB/HCT – DECR (ANEMIA)
BLOOD, NT-PROBNP – INCR [15]
BLOOD, TROPONIN – INCR [15]
BLOOD, URIC ACID – INCR [15]
URINE, PROTEIN – INCR (PROTEINURIA) [16]

ECG

AV COND – 1ST DEGREE BLOCK
AV CONDUCTION – AV DISSOCIATION, COMPLETE

DYSRHYTHMIAS – ATRIAL (PACS/OTHERS) [ESP A FIB] [23]
DYSRHYTHMIAS – VENTRICULAR (PVCS/OTHERS)
Q WAVE – ABN [7]
QRS – LBBB/LBBB PATTERN
QRS – LONG, NS
QRS – R PROGRESSION, POOR [8]
QRS, AXIS – L
QT/QTC INTERVAL – LONG
VOLTAGE, GEN – DECR [6]

Imaging

IVS, THICKNESS – INCR (SEPTAL HYPERTROPHY)
LA, CHAMBER, SIZE – INCR
LA, INTRACAVITY – MASS [THROMBUS]
LV, CHAMBER, SIZE – DECR [MAY BE N]
LV, DIAS – DYSF [DECR EARLY FILLING]
LV, EF – DECR [MAY BE N]
LV, FILLING – DECR/RESTRICTED
LV, MYOCARD – LGE [25] [31]
LV, MYOCARD, WALL THICKNESS – INCR (HYPERTROPHY) [24]
LV, PAP MUSCLES – THICK
LV, WALL MOTION – DECR [EARLY DIAS] [10]
MV, LEAFLETS – THICK
MYOCARD, APPEARANCE – GRANULAR (SPARKLING) [9]
PERICARD – FLUID [SMALL]
PLEURA – FLUID
RA, CHAMBER, SIZE – INCR
RV, WALL THICKNESS – INCR
TV, LEAFLETS – THICK

Genomics

TTR [34]

Other Tests

Bone marrow biopsy [12]
DNA analysis [18]
EMB [11]
Fat pad aspiration [13]
Gingival biopsy [13]

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic [19]

AL: high-dose IV melphalan chemotherapy
FAP: liver transplant [29]
SSA, IAA, AA: NS
TTR stabilizers: Tafamidis meglumine, Flunasil [32]
Corticosteroids
Diflunisal
HF regimen [30]

Treatment: Surgical/Invasive

HEART TRANSPLANT
ICD
LVAD

Course

Varies with form
Mean survival <6 months in patients with IVS thickness >15 mm, EF <40 %, HF symptoms

Notes

[1] Extra-cardiac multi-organ involvement including liver, spleen, lymphopoietic, mucocutaneous, peripheral nervous system, kidneys, gastrointestinal tract
[2] Due to congestion and/or amyloid infiltration
[3] Due to orthostatic hypotension, electromechanical dissociation, dysrhythmias
[4] Due to decreased cardiac output combined with inability to increase HR/maintain vascular tone
[5] Can occur despite fluid retention
[6] Due to replacement of normal cardiac tissue with electrically inert amyloid
[7] Pseudoinfarction pattern
[8] Especially V1-3
[9] NS; also seen in other forms of LVH
[10] Prior to HF
[11] Definitive: tissue shows Congo red apple-green birefringence under polarized light microscopy
[12] For AL
[13] Screening for systemic Amyloidosis
[14] Liver infiltration
[15] In AL, highly sensitive and marker of poor prognosis
[16] Renal involvement
[17] Signs/symptoms of HF often initial presentation; because amyloidosis is usually multi-system disease, many clinical features other than those listed may be present
[18] Helps differentiate between acquired and hereditary forms
[19] For HF, standard regimen, especially diuretics; precaution when using RAAS inhibitors as patients may depend on angiotensin for BP maintenance
[20] TR
[21] MR
[22] Due to amyloid involvement of AV
[23] Onset of AF may cause rapid clinical deterioration and increased thromboembolic risk
[24] Early asymmetric IVS, becoming concentric later in course

[25] Common in advanced age with no systemic involvement and of little clinical significance in these patients except possible relation to AF
[26] Discolored skin around eyes due to bleeding
[27] Highly sensitive/specific for AL
[28] EG, LV wall thickness >1.1 cm in absence of prior HTN/valvular heart disease
[29] To remove source of mutant protein
[30] Caution using ACEIs/ARBs, which may cause hypotension and under-filling of stiff hearts
[31] Global, subendocardial, or segmental LGE and highly specific pattern of myocardial/blood pool gadolinium kinetics
[32] TTR tetramer stabilizers with favorable effects on cardiac and neurological function
[33] Carpal Tunnel Syndrome: occurs in 50 % of patients with wild-type TTR; caused by compression of median nerve and manifest by:

Intermittent numbness/pain/tingling of:

Thumb
Index finger
Middle finger
Ring finger (radial side)

Pain worse at night
Pain extending upward to arm/shoulder

[34] V122I mutation of TTR mutation: common in black men and associated with significantly increased incidence of HF
[35] All patients with amyloid peripheral/autonomic neuropathy should be screened for cardiac involvement
[36] Characteristics of Familial Amyloid Polyneuropathy:

Symmetrical/distal begins in LEs/progressing to UEs
Affects all nerve functions

Autonomic
Motor
Sensory

Onset typically late teens/later with rapid progression in next 2 years

Family history of similar neuropathy usually but not always present
Concurrent involvement of visceral organs (eg, kidneys, heart) common

Guidelines

NS

Patient Information

IMAGES

http://www.nlm.nih.gov/medlineplus/ency/imagepages/1056.htm.
http://www.nlm.nih.gov/medlineplus/ency/imagepages/18136.htm.
http://www.nlm.nih.gov/medlineplus/ency/imagepages/8904.htm.

Medline Plus

ENGLISH

http://www.nlm.nih.gov/medlineplus/ency/article/000193.htm.

ESPANOL

http://www.nlm.nih.gov/medlineplus/spanish/ency/article/000193.htm.

Genetics Home Reference

http://ghr.nlm.nih.gov/condition/transthyretin-amyloidosis.

Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/amyloidosis/basics/complications/con-20024354.

Professional Information

Early Description

Virchows Arch Pathol Anat. 1854;6:135–8. http://link.springer.com/article/10.1007/BF01930815.

Review

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Review

Heart. 2011;97:75–84. http://heart.bmj.com/content/97/1/75.extract.

Review

J Am Coll Cardiol. 2007;50:2101–10. http://content.onlinejacc.org/article.aspx?articleid=1138688.

Review

J Am Heart Assoc. 2012;1:e000364. http://jaha.ahajournals.org/content/1/2/e000364.full.

Review: TTR

Circulation. 2012;126:1286–1300. http://circ.ahajournals.org/content/126/10/1286.extract.

AL LV Function

Circulation. 2014;129:1840–49. http://circ.ahajournals.org/content/129/18/1840.abstract.

Amyloidogenic V122I Transthyretin Variant in African Americans

N Engl J Med. 2015; 372:21–9. http://www.nejm.org/doi/full/10.1056/NEJMoa1404852#t=abstract.

Conduction Abnormalities/Atrial Arrhythmias

Heart Rhythm. 2016;13:383–90. http://www.heartrhythmjournal.com/article/S1547-5271(15)01186-8/abstract.

High-Sensitivity Troponin T

Heart. 2014;100:383–88. http://heart.bmj.com/content/100/5/383.abstract.

Intracardiac Thrombus/Anticoagulation

Circulation. 2009;119:2490–97. http://circ.ahajournals.org/content/119/18/2490.abstract.

LGE

J Am Coll Cardiol Img. 2010;3:155–64. http://imaging.onlinejacc.org/article.aspx?articleid=1813145.

LVOT Obstruction: Differentiation from HCM

Am J Cardiol. 2008;101:674–76. http://www.sciencedirect.com/science/article/pii/S000291490702139X.

MRI Differentiation: AL vs TTR

J Am Coll Cardiol Img. 2014;7:133–42. http://imaging.onlinejacc.org/article.aspx?articleid=1813145.

Tafamadis Meglumine

Proc Natl Acad Sci USA. 2012;109:9629–34. http://www.pnas.org/content/109/24/9629.long.

TTR Transthyretin-Associated Familial Amyloidosis: Exclusive Card Phenotype

Eur Heart J. 2013;34:520–28. http://eurheartj.oxfordjournals.org/content/34/7/520.

Updates and More

https://clinicalguidecvd.com/cardamy

Chapter 25
Cardiac Angiosarcoma

ICD-10 Code

NS

Alternate Names/Abbreviation

NS

Description/Etiology

Extremely rare, most common primary tumor of heart, usually involving RA/pericardium
Familial form reported

Comorbid Conditions

NS

Demography

M > F
Average age first presentation: 40 years

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_25,

Pathophysiology

Endothelial cell tumor appearing as multicentric mass
Primary cardiac sites:

- RA (90 %), affecting surrounding structures including invasion of IVC and TV
- Pericardium
- RCA

Signs/Symptoms

ARTERIAL PULSE – PARADOXICAL (PARADOXICAL PULSE)
ARTERIAL PULSE PRESSURE – DECR
BREATHING – DIFF (DYSPNEA) [MOST COMMON SYMP]
CHEST – PAIN [PLEURITIC/NON-PLEURITIC] [3]
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)
COUGH
EXTREM, LOWER, BILAT – EDEMA
FATIGUE
FEVER
HEART, RATE – RAPID (TACHYCARDIA)
HEART, SOUNDS, INTENSITY – DECR
LIVER – ENLARGED (HEPATOMEGALY)
NECK, JVP – ELEV
SKIN – SWELLING, EDEMA (ANASARCA) [1]
SKIN, COLOR – BLUE (CYANOSIS) [1]
SPUTUM – BLOOD (HEMOPTYSIS)
VEINS, SUPERFICIAL – DIL [1]

Differentiation

Right Atrial Myxoma

Complications

Cardiac tamponade
Chest wall invasion
Heart failure
Metastasis (lungs most often)
Myocardial rupture
RA rupture
Supraventricular arrhythmias

Laboratory

BLOOD, HGB/HCT – DECR (ANEMIA)

ECG

N/NS ABN

Imaging

PERICARD – FLUID

Other Tests

Pleural/pericardial fluid cytology: unreliable even with tumor invasion
EMB: 50 % positive

Treatment: Nonpharmacologic [2]

Radiation

Treatment: Pharmacologic [2]

Chemotherapy
Immunotherapy

Treatment: Surgical/Invasive [2]

Surgical resection

Course

Poor prognosis due to highly aggressive nature of tumor and usual advanced state at time of initial diagnosis
Mean survival without surgical resection: 4 months
With aggressive combination treatment, survival up to 30 months reported

Notes

[1] Especially head, upper trunk; described as Superior Vena Cava Syndrome in some cases
[2] No standard protocols
[3] Chest pain: chief complaint in 40 % of cases

Guidelines

NS.

Patient Information

NS

Professional Information

Early Description

Am Heart J. 1934;9:480–91. http://www.sciencedirect.com/science/article/pii/S000287033490096X.

Review

Med Sci Monit. 2014;23:20:103–9. http://www.ncbi.nlm.nih.gov/pubmed/24452054.

Cardiac Sarcoma: Cleveland Clinic Series

Am J Clin Oncol. 2014. http://www.ncbi.nlm.nih.gov/pubmed/25036471.

Cardiac Sarcoma: Mayo Series

Cancer. 2008;112:2440–6. doi: 10.1002/cncr.23459. http://www.ncbi.nlm.nih.gov/pubmed/18428209.

Echo

J Am Coll Cardiol. 2013;61. doi:10.1016/S0735-1097(13)61085-2. http://content.onlinejacc.org/article.aspx?articleid=1665367.

Familial Angiosarcoma

J Thor Cardiovasc Surg. 2002;124:392–4. http://www.sciencedirect.com/science/article/pii/S0022522302000922.

RA Rupture

J Am Coll Cardiol. 2013;61:e175. http://content.onlinejacc.org/article.aspx?articleid=1675800.

Updates and More

https://clinicalguidecvd.com/cardang

Chapter 26
Cardiac Arrest

Management Keys

Resuscitation duration should continue for at least 40 min in all adults with bystander-witnessed out-of-hospital cardiac arrest
Early access to cardiac catheterization laboratory [7]

ICD-10 Code

I46.9

Description/Etiology

Acute circulatory failure due to absent/ineffective cardiac systolic function associated with:

- Asystole
- Pulseless electrical activity
- Pulseless ventricular tachycardia
- Ventricular fibrillation

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_26,

Comorbid Conditions

ACUTE MYOCARDIAL INFARCTION
ACUTE PULMONARY EMBOLISM
ANOREXIA NERVOSA
ATRIOVENTRICULAR HEART BLOCK
BRUGADA SYNDROME
CARDIAC TAMPONADE
CARDIOMYOPATHY [ALL FORMS]
CHEST TRAUMA
COCAINE
CORONARY ARTERY DISEASE
DIABETES MELLITUS
DRUG OVERDOSE
DYSRHYTHMIAS – VENTRICULAR
HEART FAILURE
HYPERGLYCEMIA – ACUTE
HYPERKALEMIA
HYPERTENSION – SYSTEMIC ARTERIAL
HYPOGLYCEMIA
HYPOKALEMIA
HYPOMAGNESEMIA
HYPOTHERMIA
HYPOVOLEMIA
HYPOXEMIA
LONG QT SYNDROME – ACQUIRED
LONG QT SYNDROME – CONGENITAL
METABOLIC ACIDOSIS
PULMONARY HYPERTENSION
RESPIRATORY ACIDOSIS
SEVERE SEPSIS (SEPTIC SHOCK)
TENSION PNEUMOTHORAX
TOXINS
VALVULAR HEART DISEASE

Signs/Symptoms

CHEST – PAIN
CHEST – PALPITATIONS
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)

Complications

Acute Kidney Injury [6]
Ischemic brain injury [4]
Ischemic liver injury (shock liver) [5]
Rib and sternal fractures [3]
Recurrent cardiac arrest
Death

Laboratory [1]

ECG [1]
DYSRHYTHMIAS – VENTRICULAR (PVCS/OTHERS)
ELECTRICAL ACTIVITY – ABSENT
IMAGING [1]
GENOMICS [1]

Treatment: Nonpharmacologic

CPR [8]
Defibrillation [10]
Induced hypothermia

Treatment: Pharmacologic

Antiarrhythmic:

Amiodarone: 300 mg IV in single dose initially, followed by 150 mg in single dose if required

Lidocaine: 1–1.5 mg/kg IV in single dose initially, followed by 0.5–0.75 mg/kg q 5–10 min (max 3 mg/kg total dose)

Atropine: for pulseless electrical activity/bradycardia: 0.5–1 mg IV q 3–5 min (max 3 mg total dose) [2]

Vasopessors

Epinephrine: 1 mg IV q 3–5 min [9]
Vasopressin: 40 units IV in single dose

For torsades: MgSO4: 2 g IV in single dose over 5–10 min

Treatment: Surgical/Invasive

Pacemaker

Procedures to correct underlying cause, such as PCI for Acute Myocardial Infarction, embolectomy for severe Acute Pulmonary Embolism when indicated and appropriate [7]

Notes

[1] Variable with etiology

[2] Efficacy of atropine in setting of cardiac arrest unproven

[3] Traumatic injury including rib/sternal fractures occurs in about 1/3 of patients; very low rate of related serious complications; follow-up care should include evaluation for penetrating trauma to internal organs

[4] Up to 50 % of cardiac arrest survivors have subsequent neurologic abnormalities, due to:

Increased cerebral oxygen consumption
Increased post-arrest intracranial pressure
Inflammation
Oxygen free radicals generation/accumulation

[5] Caused by decreased/loss of splanchnic circulation during cardiac arrest, propagated by inflammatory response

of hepatocytes, Kupffer cells, endothelium; also due to reperfusion injury caused by resuscitation/restoration of oxygen/generation of reactive oxygen species

[6] Due to Acute Tubular Necrosis, comprising:

Endothelial damage
Inflammatory mediators
Procoagulant state
Vascular dysregulation

[7] Early access to cardiac cath lab after cardiac arrest due to shockable rhythm associated with 65 % survival rate to hospital discharge with good neurological outcome

[8] According to 1 study, resuscitation duration should be for at least 40 min in all adults with bystander-witnessed out-of-hospital cardiac arrest

[9] AHA Guidelines: "reasonable to consider" epinephrine q 3–5 min for patients with a non-shockable rhythm and q 3–5 min after 2nd defibrillation in patients with a shockable rhythm

[10] Current recommendations are for single shock protocols with 2 min of chest compressions between defibrillation attempts for persistent VT/VF

Guidelines

2015 international consensus on cardiopulmonary resuscitation and emergency cardiovascular care science with treatment recommendations
Circulation. 2015;132:S2–S311. https://circ.ahajournals.org/content/132/16_suppl_1.toc.

2015 ESC guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death
Eur Heart J. 2015;36;2793–867. http://eurheartj.oxfordjournals.org/content/36/41/2793.

Cardiac arrest and cardiopulmonary resuscitation outcome reports: update of the utstein resuscitation registry templates for out-of-hospital cardiac arrest
Circulation. 2015;132:1286–1300. http://circ.ahajournals.org/content/132/13/1286.full?sid=f4f6f125-85fd-4bf3-916a-2bcda1b7200c.

Professional Information

Best Practices Adoption by EMS.

J Am Heart Assoc. 2016;5:e002892. http://jaha.ahajournals.org/content/5/1/e002892.full.

Coronary Vasospasm (Case Study)

Circulation. 2016;133:756–63. http://circ.ahajournals.org/content/133/8/756.full.

Defibrillation: Time Intervals/Outcomes

BMJ. 2016;353:i1653. http://www.bmj.com/content/353/bmj.i1653.

Early Access to Cardiac Cath Lab

J Am Heart Assoc. 2016;5:e002670. http://jaha.ahajournals.org/content/5/1/e002670.full.

Epinephrine Misuse

BMJ. 2016;353:i1577. http://www.bmj.com/content/353/bmj.i1577.

Hypomagnesemia

J Am Heart Assoc. 2016;5:e002707. http://jaha.ahajournals.org/content/5/1/e002707.full.

ICD for SCD: Outcomes

Circ Arrhythmia Electrophysiol. 2016;9:e003283. doi:10.1161/CIRCEP.115.003283. http://circep.ahajournals.org/content/9/3/e003283.abstract.

Mitral Valve Prolapse

Heart Rhythm. 2016;13:498–503. http://www.heartrhythmjournal.com/article/S1547-5271(15)01196-0/abstract

Noncardiac Causes: Outside Hospital

Eur Heart J. 1997;18:1122–8. http://eurheartj.oxfordjournals.org/content/18/7/1122.abstract?ijkey=fa1dd075929478401e5e62a8db4161f7faf5937a&keytype2=tf-ipsecsha.

Resuscitation Duration

Circulation. 2016;133:1386–96. http://circ.ahajournals.org/content/133/14/1386.full.

Risk Factors (USA)

Epidemiology. 1997;8:175–180. http://journals.lww.com/epidem/pages/articleviewer.aspx?year=1997&issue=03000&article=00013&type=abstract.

Sudden Death in Young

Circulation. 2016;133:1006–26. http://circ.ahajournals.org/content/133/10/1006.abstract.

Survival

Resuscitation. 2010;81:1479–87. http://www.resuscitationjournal.com/article/S0300-9572(10)00432-6/abstract.

Therapeutic Hyperthermia

J Clin Diagn Res. 2015;9:OD01–2. http://www.ncbi.nlm.nih.gov/pmc/articles/PMC4606266/.

Utstein Style Templates for Reporting

Resuscitation. 2008;79:193–7. http://www.ncbi.nlm.nih.gov/pubmed/18805620?dopt=Abstract.

Updates and More

https://clinicalguidecvd.com/cardar

Chapter 27
Cardiac Contusion

ICD-10 Code

S26.11XA

Alternate Names/Abbreviation

BLUNT CARDIAC CONTUSION
MYOCARDIAL CONTUSION

Description/Etiology

Cardiac damage secondary to blunt chest trauma (non-penetrating chest injury) [1]
Most often due to:

- Abdominal and LE trauma [2]
- Auto accidents
- Crush injuries
- Cardiac compression (resuscitation)
- Rapid change in atmospheric pressure [3]
- Sports

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_27,

Comorbid Conditions

BLUNT CHEST TRAUMA
OTHER SITES OF TRAUMA

Demography

Most common among younger aged persons engaged in sports

Pathophysiology

Local cardiac muscle damage (necrosis, edema, hemorrhage) due to direct trauma or coronary insufficiency
RV most vulnerable due to anterior chest location

Signs/Symptoms [6]

CHEST – PALPITATIONS
CHEST – PAIN [4]
CHEST, WALL – BRUISES [5]
OFTEN ASYMP

Differentiation

Acute Myocardial Infarction
Cardiomyopathy – Takotsubo

Complications [21]

Acute Myocardial Infarction
Cardiac Dysrhythmias

Cardiac Tamponade
Commotio Cordis [15]
Coronary Artery Dissection
Heart Failure [20]
Hypotension/Shock
Mitral Regurgitation – Acute
Rupture of IVS
Tricuspid Regurgitation
Ventricular Aneurysm [20]

Laboratory

BLOOD, CKMB – INCR [7]
BLOOD, TROPONIN – INCR [8] [9]

ECG [10]

ATRIAL TACHYCARDIA
AV COND – 1ST DEGREE BLOCK
AV COND – 2ND DEGREE BLOCK
AV COND – 3RD DEGREE BLOCK
DYSRHYTHMIAS – ATRIAL (PACS/OTHERS) [14] [INCL A FIB]
DYSRHYTHMIAS – VENTRICULAR (PVCS/OTHERS) [14]
Q WAVE – ABN [INCL NEW Q WAVE]
QRS – RBBB/RBBB PATTERN
QRS, AXIS – L [FASCICULAR BLOCK]
QRS, AXIS – R [FASCICULAR BLOCK]
QT/QTC INTERVAL – LONG
RATE – DECREASED (SINUS BRADYCARDIA)
RATE – INCREASED (SINUS TACHYCARDIA)
ST SEGMENT – DEPR
ST SEGMENT – ELEV [12]
ST-T WAVE – ABN, NS [11]

Imaging

LV, CHAMBER, SIZE – INCR
LV, INTRACAVITY – MASS [THROMBUS]
LV, WALL MOTION, SEG – DECR/AKINETIC [17]
MV, FLOW – REGURG [18]
PERICARD – FLUID [19]
RV, CHAMBER, SIZE – INCR
RV, INTRACAVITY – MASS [THROMBUS]
RV, WALL MOTION, SEG – DECR/AKINETIC
TV, FLOW – REGURG [18]

Other Tests

ECG telemetry

Treatment: Nonpharmacologic

NS [16]

Treatment: Pharmacologic

NS [16]

Treatment: Surgical/Invasive

NS [16]

Prevention

Trauma precautions

Course

Variable with anatomic location, magnitude of injury, associated trauma

Notes

[1] Occurs in up to 70 % of cases of blunt chest trauma; blunt (nonpenetrating) chest trauma can also cause:

Aneurysm formation
Coronary artery thrombosis
Endocardial damage
Hemopericardium/tamponade
Myocardial laceration/rupture
Valve rupture

[2] "Hydraulic ram effect" due to upward displacement of viscera
[3] Example: explosion
[4] May resemble pain secondary to coronary insufficiency/AMI
[5] Usually absent
[6] Signs/symptoms due to other traumatic injuries may predominate, suggesting possible Cardiac Contusion, including:

Fractures of sternum, clavicle, ribs
New heart murmur
Pericardial friction rub

[7] Non-specific: also increased by trauma to skeletal muscle, liver, intestines, diaphragm
[8] Troponin T: highly specific with low sensitivity
[9] Troponin I: highly specific with varying reports of sensitivity
[10] May be normal because contusion mainly occurs in RV; changes may be due to pericardial damage; abnormalities

due to other causes related to trauma may be present, such as electrolyte changes, lung contusion, hypoxia, head injury
[11] Most common change, usually transient and resolving in 24–48 h
[12] Pericarditis
[13] Abnormal in >50 % of cases studied; TEE often necessary due to limiting circumstances (eg, chest tubes) surrounding trauma care
[14] Occur in up to 70 % of cases in blunt chest trauma; onset may be delayed by up to 48 h after incident
[15] Blunt trauma, which can be mild, usually young athletes
[16] No specific guidelines exist except treatment of comorbidities/complications, such as dysrhythmias, tamponade
[17] Especially anteroseptal and inferoposterior hypokinesis
[18] MR/TR rarely occur; due to papillary muscle injury
[19] Pericardial fluid, even associated with sternal fracture, does not necessarily indicate myocardial injury or tamponade
[20] Long term, due to myocardial necrosis
[21] Life-threatening dysrhythmias, such as severe valve regurgitation, almost all occur within 48 h of trauma

Guidelines

NS.

Patient Information

Images

http://www.nlm.nih.gov/medlineplus/ency/imagepages/1056.htm.
http://www.nlm.nih.gov/medlineplus/ency/imagepages/1097.htm.

Medlineplus

ENGLISH

http://www.nlm.nih.gov/medlineplus/ency/article/000202.htm.

ESPANOL

http://www.nlm.nih.gov/medlineplus/spanish/ency/article/000202.htm.

Merck

http://www.merckmanuals.com/home/SearchResults?query=Blunt+Cardiac+Injury&icd9=861.01.

Professional Information

Review

Cardiol Clin. 2012;30:545–55. http://www.ncbi.nlm.nih.gov/pubmed/23102031.

Review

Heart. 2003;89:485–9. http://heart.bmj.com/content/89/5/485.full.

Cardiac Troponin

Am Heart J. 1996;131:308–12. http://www.sciencedirect.com/science/article/pii/S0002870396903592.

Case: 8 Years Old Kicked by Horse

Circ Cardiovasc Imaging. 2015;8:e002857. http://circimaging.ahajournals.org/content/8/3/e002857.full.

Intramyocardial Hemorrhage

Circulation. 2004;109:277. http://circ.ahajournals.org/content/109/2/277.full?sid=f64aeddd-332d-4cd5-98df-f6d7e2da4a5f.

TEE

Chest. 1996;109:73–7. http://journal.publications.chestnet.org/article.aspx?articleID=1069358.

Soccer/MRI

Circulation. 2010;121:2456–61. http://circ.ahajournals.org/content/121/22/2456.full?sid=f64aeddd-332d-4cd5-98df-f6d7e2da4a5f.

Sudden Death: RA Contusion

Injury. 2005;36:213–7. http://www.ncbi.nlm.nih.gov/pubmed/15589944.

Updates and More

https://clinicalguidecvd.com/cardcon

Chapter 28
Cardiac Sarcoidosis

Management Keys

Aggressive use of prophylactic ICD when ventricular dysrhythmias occur, due to high incidence of subsequent SCD

Consider this diagnosis in young patients with otherwise unexplained heart block

ICD-10 Code

D86.85

Alternate Names/Abbreviation

Cardiac Sarcoid

Description/Etiology

Etiology unknown

Multisystem disease consisting of granulomas affecting all organ systems, most often lungs; 20–25 % of patients have

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_28,

cardiac involvement at autopsy, often without clinical manifestations, and sometimes as sole form of disease

Cardiac involvement may present as asymptomatic LV dysfunction, AV block, HF, atrial/ventricular dysrhythmia, SCD

Most common noncardiac sites of involvement:

Eyes
Liver/GI tract
Lungs (hilar lymph nodes, parenchyma)
Nervous system
Skin [15]

Comorbid Conditions

NS

Demography

Increased incidence in Norway, Ireland, Japan

In USA, African-Americans 3:1 compared to Caucasians

F > M

Pathophysiology

Noncaseating granulomas in myocardium, pericardium, conduction pathways; any/all 4 cardiac chambers can be involved

Involvement of LV myocardium (order of frequency):

LV free wall/papillary muscles
Basal IV septum
RV free wall
Atrial walls

Signs/Symptoms [1]

BREATHING – DIFF (DYSPNEA)

CHEST – PAIN
CHEST – PAIN, EFFORT (ANGINA PECTORIS)
CHEST – PALPITATIONS
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)
FATIGUE [2]
FEVER [2]
HEART, LV, APEX – MURMUR, SYS
WEIGHT – LOSS [2]

Differentiation

Arrhythmogenic Right Ventricular Dysplasia/ Cardiomyopathy
Coronary Artery Disease [6] [20]
Other causes of dysrhythmias/conduction abnormalities
Other causes of HF
Other causes of Myocarditis
Other causes of Chronic Pericarditis
Other causes of Restrictive Cardiomyopathy

Complications (Cardiac)

Acute Sarcoid Myocarditis [4]
Atrioventricular Heart Block (High Grade)
Cardiac Tamponade
Cardiomyopathy – Dilated
Cardiomyopathy – Hypertrophic
Cardiomyopathy – Restrictive
Coronary Artery Vasculitis
Heart Failure [18]
Pericarditis – Constrictive
Sudden Death

Laboratory

BLOOD, ACE – INCR [5]
BLOOD, CALCIUM – INCR [5]

BLOOD, GLOBULINS – INCR [5]
BLOOD, HGB/HCT – DECR (ANEMIA) [5]
BLOOD, TROPONIN – INCR
BLOOD, URIC ACID – INCR [5]
BLOOD, WBC – DECR (LEUKOPENIA) [5]

ECG [3]

AV COND – 1ST DEGREE BLOCK [16]
AV COND – 2ND DEGREE BLOCK [16]
AV COND – 3RD DEGREE BLOCK [9] [16]
DYSRHYTHMIAS – ATRIAL (PACS/OTHERS) [7]
DYSRHYTHMIAS – VENTRICULAR (PVCS/OTHERS) [8] [17]
QRS – LBBB/LBBB PATTERN [16]
QRS – LONG, NS
QRS – RBBB/RBBB PATTERN [16]
ST-T WAVE – ABN, NS

Imaging [3]

CARDIOMEGALY [10]
IVS, THICKNESS – DECR
IVS, THICKNESS – INCR (SEPTAL HYPERTROPHY) [12]
LUNGS – INFILTRATES
LUNGS, HILUM – ADENOPATHY
LV, CHAMBER, SIZE – INCR
LV, DIAS – DYSF
LV, EF – DECR
LV, MYOCARD – EDEMA (MRI) [22]
LV, MYOCARD – LGE [23]
LV, MYOCARD, THALLIUM UPTAKE – DECR [20]
LV, WALL MOTION – DECR SEG/GEN
LV, WALL MOTION, SEG – DECR/AKINETIC
LV, WALL THICKNESS, SEG – DECR [11]
LV, WALL THICKNESS, SEG – INCR
MYOCARD – EDEMA/INFLAMMATION [MRI] [21]
MYOCARD – FIBROSIS

MYOCARD – SCAR (S) [MRI]
PERICARD – FLUID

Other Tests

Ambulatory ECG
EMB [13]

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic

Corticosteroids [14]
HF regimen

Treatment: Surgical/Invasive

CRT [24]
ICD [24]
Pacemaker [24]
Heart transplant [24]

Course

Variable

Notes

[1] Vary according to location; patients with sarcoid lesion in heart often have no clinical manifestations, depending on location and size; clinical features often dominated by

extra-cardiac involvement, especially of lungs, which are not listed here; initial presentation may be HF or SCD
[2] Signs/symptoms of systemic Sarcoidosis
[3] ECG/echo findings may be only clinical manifestations
[4] High-degree HB, ventricular dysrhythmias, HF
[5] Non-specific abnormalities present in systemic Sarcoidosis
[6] Chest pain may resemble angina pectoris/AMI
[7] Especially AF/atrial flutter
[8] Especially NSVT
[9] Most common clinical finding, in up to 30 % of patients with cardiac sarcoid; occurs at younger age than usual causes of complete HB
[10] Pericardial effusion, cardiomyopathy
[11] Especially IVS; when basal anterior septum width is thin, highly suggestive of Sarcoidosis in young person with DCM
[12] Rare, resembles HCM
[13] Noncaseating granuloma: diagnostic, but may be normal due to patchy involvement
[14] Use with caution to avoid HF
[15] Erythema Nodosum, granulomas in scars/tattoos
[16] Heart block due to scar/granuloma of basal septum or ischemia of conduction system due to involvement of nodal artery
[17] VT most common, caused by increased automaticity and reentrant tachycardia at site of granuloma
[18] 2nd most common cause of death (after ventricular dysrhythmia) due to myocardial infiltration, ventricular aneurysm, rhythm abnormalities, Cor Pulmonale secondary to PAH, valve regurgitation, and combinations of these abnormalities
[19] Tamponade rare
[20] Decreased thallium uptake: NS for Sarcoidosis; uptake increases with exercise, termed “reverse distribution”, which may help differentiate from CAD
[21] Mainly mid-myocardium/epicardium but rarely endocardium
[22] Resolves with steroid treatment
[23] Sarcoidosis with LGE associated with significant increased risk for death/VT, even with preserved LVEF
[24] See current Heart Failure/Cardiomyopathy Guidelines for indications/appropriate use

Guidelines

2013 ACCF/AHA guideline for the management of heart failure
J Am Coll Cardiol. 2013;62:e147–239. http://content.onlinejacc.org/article.aspx?articleid=1695825.
ESC guidelines for the diagnosis and treatment of acute and chronic heart failure 2012
Eur Heart J. 2012;33:1787–847. http://eurheartj.oxfordjournals.org/content/ehj/33/14/1787.full.pdf.

Patient Information

IMAGES

http://www.nlm.nih.gov/medlineplus/ency/imagepages/1613.htm.
http://www.nlm.nih.gov/medlineplus/ency/imagepages/1614.htm.
http://www.nlm.nih.gov/medlineplus/ency/imagepages/1615.htm.
http://www.nlm.nih.gov/medlineplus/ency/imagepages/9248.htm.

AAFP

http://familydoctor.org/familydoctor/en/diseases-conditions/sarcoidosis.printerview.all.html.

Cleveland Clinic

http://my.clevelandclinic.org/lungs-breathing-allergy/departments-centers/sarcoidosis-center.aspx.

Medline Plus

ENGLISH

http://www.nlm.nih.gov/medlineplus/ency/article/000076.htm.

ESPANOL

http://www.nlm.nih.gov/medlineplus/spanish/ency/article/000076.htm.

NIH

http://www.nhlbi.nih.gov/health/health-topics/topics/sarc/.

Professional Information

History

WASOG. http://www.wasog.org/pdfs/sharma.pdf.

Review

Heart.2016;102:184–90.http://heart.bmj.com/content/102/3/184?etoc.

Review

Heart. 2006;92:282–8. http://heart.bmj.com/content/92/2/282.

Review

N Engl J Med. 2007;357:2153–65. http://www.nejm.org/doi/full/10.1056/NEJMra071714#t=article.

Review

Prog Cardiovascular Dis. 2010;52:336–46. https://www.deepdyve.com/lp/elsevier/diagnosis-and-management-of-cardiac-sarcoidosis-rfCaWyeN10.

Acute Sarcoidosis: Resemblance to AMI

Am J Med. 2013;126:21–4. http://www.sciencedirect.com/science/article/pii/S0002934312007930.

Atrial Arrhythmias

Am J Cardiol. 2014;113:1556–60. http://www.ajconline.org/article/S0002-9149(14)00636-5/abstract.

Atrioventricular Heart Block: Initial Manifestation

Am J Cardiol. 2015;115:505–9. http://www.sciencedirect.com/science/article/pii/S0002914914021699.

CMR/PET Images

Am Coll Cardiol. 2011;58:438. http://content.onlinejacc.org/article.aspx?articleid=1146665&resultClick=3.

Coronary Artery Vasculitis

Circulation. 2012;125:e344–6. http://circ.ahajournals.org/content/125/6/e344.full?sid=de4bce44-ce54-47c7-9df4-4d485f6f0a86.

Imaging

Circ Cardiovasc Imaging. 2016;9:e000867. http://circimaging.ahajournals.org/content/9/3/e000867.abstract.

Imaging

Eur Heart J. 2013;34:1278. http://eurheartj.oxfordjournals.org/content/34/17/1278.

Imaging

J Am Coll Cardiol. 2013;61:1548. http://content.onlinejacc.org/article.aspx?articleID=1673460.

Imaging: Integrated MRI/PET

Circulation. 2013;127:e639–41. http://circ.ahajournals.org/content/127/22/e639.full.

Imaging: Integrated MRI/PET

Heart. 2014;100:89–90. http://heart.bmj.com/content/100/1/89.full.

Imaging and Outcomes

Circulation. 2009;120:1969–77. http://circ.ahajournals.org/content/120/20/1969.full?sid=d63e1631-debd-4cdc-af62-33068c8e0041.

Imaging: LGE (Case Report)

Eur Heart J. 2013;34:2411. http://eurheartj.oxfordjournals.org/content/34/30/2411.

Imaging: LGE/Prognosis

Eur Heart J Cardiovasc Imaging. 2015;16:634–41. http://ehjcimaging.oxfordjournals.org/content/16/6/634.abstract?.

Imaging: Presentation as RV Mass (Case Report)

Eur Heart J. 2016;37:859. http://eurheartj.oxfordjournals.org/content/37/11/859.full.

LGE: Increased Risk

Circ Cardiovasc Imaging. 2016;9:e003738. http://circimaging.ahajournals.org/content/9/1/e003738.abstract?etoc.

LGE: Predictor of Adverse Outcomes

Heart. 2014;100:1165–72. http://heart.bmj.com/content/100/15/1165.abstract.

PET: Prognosis Assessmenyt

J Am Coll Cardiol. 2014;63:329–36. http://content.onlinejacc.org/article.aspx?articleID=1757083.

Rhythm Abnormalities

Heart Lung Circ. 2014;23:1100–9. http://www.sciencedirect.com/science/article/pii/S1443950614006234.

Ventricular Tachycardia

Circ Arrhythmia Electrophysiol. 2015;8:87–93. http://circep.ahajournals.org/content/8/1/87.abstract?etoc.

Updates and More

https://clinicalguidecvd.com/cardsarc

Chapter 29
Cardiac Tamponade

Management Keys

Treat as emergency condition in hospital if possible
Drain pericardial fluid ASAP
Administer IV fluids to normalize BP until pericardial fluid removed
Administer oxygen
Determine cause as soon as patient is stabilized, and begin appropriate treatment of cause as appropriate

ICD-10 Code

I31.4

Description/Etiology

Accumulation of fluid in pericardial space raising intrapericardial pressure with progressive impairment of ventricular diastolic filling and subsequent decreased cardiac output

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_29,

Comorbid Conditions

ACUTE MYOCARDIAL INFARCTION
AORTIC DISSECTION
AUTOIMMUNE/CONNECTIVE TISSUE DISEASE
BLUNT CHEST TRAUMA
CANCER [12]
CARDIAC SURGERY [RECENT]
CARDIAC TUMOR [PRIMARY]
DRUGS
HYPOTHYROIDISM
INDWELLING INSTRUMENTATION
INFECTION – BACTERIAL
INFECTION – FUNGAL
INFECTION – VIRAL [11]
RADIATION
RENAL FAILURE (UREMIA)
SEPTICEMIA [5]
TUBERCULOSIS

Demography

Varies with underlying cause

Pathophysiology

Increased intrapericardial pressure compresses all four cardiac chambers, resulting in:

- Decreased diastolic compliance
- Decreased ventricular inflow
- Equalization of mean diastolic pericardial and intracardiac pressures

Cardiac output initially maintained with increased HR and systolic vascular resistance; eventually cardiac output falls leading to cardiogenic shock

Only small amount of effusion necessary for adverse hemodynamic changes if accumulation is rapid; with slow accumulation, pericardial space may accommodate large effusion with little/no adverse hemodynamic effect due to increased myocardial compliance

Signs/Symptoms

ABDOMEN – PAIN
APPETITE – DECR (ANOREXIA) [2]
ARTERIAL PULSE – DICROTIC [15]
ARTERIAL PULSE – PARADOXICAL (PARADOXICAL PULSE)
ARTERIAL PULSE PRESSURE – DECR
ARTERIAL PULSE, AMP – ALTERNATING (PULSUS ALTERNANS)
BACK – PAIN [3] [ESP THORAX]
BREATHING – DIFF (DYSPNEA)
BREATHING – DIFF, RECLINING FLAT (ORTHOPNEA)
BREATHING – RAPID (TACHYPNEA)
CHEST – PAIN
HEART – FRICTION RUB, PERICARD
HEART, LV, APEX, IMP – DECR/ABSENT
HEART, RATE – RAPID (TACHYCARDIA)
HEART, RATE – SLOW (BRADYCARDIA) [4]
HEART, S3 LV
HEART, SOUNDS, INTENSITY – DECR
HYPOTENSION (BLOOD PRESSURE – DECREASED/LOW) [6]
LIVER – ENLARGED (HEPATOMEGALY)
MENTATION – CONFUSION
MENTATION – WEAKNESS (MALAISE)
MOOD – ANXIOUS
MOOD – LETHARGIC
MOOD – RESTLESS/IRRITABLE/COMBATIVE
NECK, JVP – ELEV

NECK, JVP, X DESCENT – PROMINENT/RAPID
NECK, JVP, Y DESCENT – ABSENT
SKIN, TEMP – DECR
SWEATING – INCR (DIAPHORESIS/ HYPERHIDROSIS)
URINE, VOL – DECR/ABSENT (OLIGURIA/ANURIA)
WEIGHT – LOSS [2]

Differentiation

Cardiomyopathy – Restrictive
Pericardial Effusion without Tamponade
Pericarditis – Constrictive
Septic Shock [5]

Complications

Shock
SCD

Laboratory

NS

ECG

AV CONDUCTION – AV DISSOCIATION, COMPLETE [8]
QRS – ELECTRICAL ALTERNANS, QRS ONLY
RATE – DECREASED (SINUS BRADYCARDIA) [4]
RATE – INCREASED (SINUS TACHYCARDIA)
ST-T WAVE – ABN, NS

Imaging

CARDIOMEGALY
IVC – ABSENT COLLAPSE
IVC, SIZE – INCR (PLETHORA)
IVS, MOTION, DIAS – ABN [16]
LA, CHAMBER – INVERSION
LV, MYOCARD, WALL THICKNESS – INCR (HYPERTROPHY)
MV, FLOW, DURING INSP – DECR [>25 %]
PERICARD – FLUID
RA, CHAMBER – INVERSION
RV, CHAMBER – INVERSION
TV, FLOW, INSP – DECR
VEINS, SYSTEMIC, FLOW, REVERSAL – INCR

Other Tests

Cardiac Catheterization [14]
Pericardiocentesis

Treatment: Nonpharmacologic [9]

NS
Avoid mechanical ventilation

Treatment: Pharmacologic [9] [13]

Volume expansion
Inotropes for hypotension

Treatment: Surgical/Invasive [9] [10]

Pericardiocentesis/pericardial catheter drainage
Percutaneous balloon pericardiotomy
Pericardiectomy

Prevention

Variable per etiology

Course

Variable per etiology

Notes

[1] See also Acute Pericarditis chapter
[2] Chronic form
[3] Interscapular
[4] Severe form; otherwise tachycardia typical
[5] Septic shock may be misdiagnosed for tamponade secondary to septicemia
[6] May be normal or increased in patients with prior Systemic Arterial Hypertension
[8] Severe
[9] Treatment should be supportive and primarily aimed at underlying cause until definitive treatment/drainage
[10] See current guidelines for details
[11] Include (but not limited to) Coxsackievirus, Adenovirus, Influenza, Mumps, HIV, Infectious Mononucleosis, Echovirus
[12] Especially breast, lung, melanoma, lymphoma
[13] Avoid diuretics and vasodilators
[14] Only to confirm in unclear cases; may delay definitive treatment and not recommended as routine

[15] Abnormal carotid pulse with 2 palpable pulsations, the second being diastolic; associated with decrease cardiac output
[16] Termed septal bounce "dip-plateau" phenomenon: abnormal early outward/inward IVS diastolic motion; also occurs in Constrictive Pericarditis

Guidelines

2015 ESC guidelines for the diagnosis and management of pericardial diseases
Eur Heart J. 2015;36:2921–64. http://eurheartj.oxfordjournals.org/content/36/42/2921.

Patient Information

Images

http://www.nlm.nih.gov/medlineplus/ency/imagepages/1097.htm.
http://www.nlm.nih.gov/medlineplus/ency/imagepages/18081.htm.
http://www.nlm.nih.gov/medlineplus/ency/imagepages/18123.htm.

Medlineplus

ENGLISH

http://www.nlm.nih.gov/medlineplus/ency/article/000194.htm.

ESPANOL

http://www.nlm.nih.gov/medlineplus/spanish/ency/article/000194.htm.

Cleveland Clinic

http://www.clevelandclinicmeded.com/medicalpubs/diseasemanagement/cardiology/pericardial-disease/#cesec48.

Merck

http://www.merckmanuals.com/home/SearchResults?query=Cardiac+Tamponade&icd9=861.01.

Professional Information

Review

Circulation. 2006;113:1622–32. http://circ.ahajournals.org/content/113/12/1622.full.pdf.

Review

Cleve Clin J Med. 2013;80(2):109–16. http://www.ccjm.org/content/80/2/109.full.pdf.

Diagnosis

JAMA. 2007;297:1810–18. http://jama.jamanetwork.com/article.aspx?articleid=206803.

Cardiac Hemangioma

Circulation. 2015;131:e21–3. http://circ.ahajournals.org/content/131/3/e21.extract?etoc.

Imaging Guidelines

J Am Soc Echocardiogr. 2013;26:965–1012.e15. http://ac.els-cdn.com/S0894731713005336/1-s2.0-S0894731713005336-main.pdf?-tid=d4bed034-f244-11e3-af0e-00000aab0f6c&acdnat=1402586499-ba4d064f354d0346278f6f823a0323d6

Management of Pericardial Effusion

Eur Heart J. 2013;34:1186–97. http://eurheartj.oxfordjournals.org/content/34/16/1186.full.pdf.

Septal Bounce

Heart. 2013;99:1376. http://heart.bmj.com/content/99/18/1376.extract.

Updates and More

https://clinicalguidecvd.com/cardtam

Chapter 30
Cardiomyopathy: Danon Disease (Lamp2 Cardiomyopathy)

Management Keys

- Consider heart transplant at earliest sign of HF due to rapid demise in patients with this disease
- Consider Danon Disease in all young male patients with HCM and WPW Syndrome
- Use multidisciplinary team approach for management, due to multisystem involvement, including:
  - Cardiac
  - Genetic counseling patient/family
  - Ophthalmological
  - Physical therapy
  - Psychosocial support
- Monitor closely/regularly cardiac function after initial diagnostic/baseline measures (eg, rest/ambulatory ECG, echo, MRI, BNP) for life-threatening dysrhythmias/LV dysfunction
- Meticulous management of volume/fluid status, especially avoidance of dehydration

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_30,

ICD-10 Code

E74.0

Alternate Names/Abbreviation

LAMP2 Cardiomyopathy

Description/Etiology

X-linked autosomal dominant multisystem disease, most often associated with abnormalities of heart, skeletal muscle, mental development, eyes; less often liver, lungs [1]
Deficiency of lysosome-associated membrane protein 2 (LAMP2)
Main cardiac abnormality: hypertrophic cardiomyopathy sometimes with WPW/AV conduction defects; progresses to Dilated Cardiomyopathy in up to 12 % of affected males

Comorbid Conditions

ATRIOVENTRICULAR HEART BLOCK
ATTENTION DEFICIT HYPERACTIVITY DISORDER
FAMILY HX: HEART FAILURE
FAMILY HX: SUDDEN DEATH
MENTAL RETARDATION
PREEXCITATION SYNDROMES [9]
VISION IMPAIRED

Demography

M > F [1]
All ethnicities
Prevalence unknown

Pathophysiology

Decreased LAMP2 causes cardiac and skeletal myopathy with intracytoplasmic vacuoles containing autophagic material and glycogen
Myocardial fibrosis prominent in explanted female hearts
Postmortem cardiac findings: extensive muscle fibrosis/necrosis

Signs/Symptoms

ABDOMEN – PAIN
ARTERIAL PULSE – DOUBLE (BISFERIENS)
ARTERIAL PULSE, RISE – RAPID
BOWEL MOVEMENTS – CONSTIPATION
BOWEL MOVEMENTS – DIARRHEA
BREATH SOUNDS – WHEEZES
BREATHING – DIFF (DYSPNEA)
BREATHING – DIFF, NOCTURNAL (DYSPNEA, NOCT)
CHEST – PAIN [16]
CHEST – PALPITATIONS [16]
COGNITION – DEFECT, NS
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)
DIZZY/LIGHTHEADED/PRESYNCOPE
EXTREM, FEET, ARCH – HIGH (PES CAVUS)
EYES, LENS – ABN, NS [12]
EYES, RETINA, EPITHELIUM, PIGMENTATION – DECR/ABSENT [12]
EYES, VISION – DECR/LOSS [12]
FATIGUE
HEART, LSB, LOWER – THRILL, SYS
HEART, LSB, MID – MURMUR, SYS
HEART, LV, APEX – MURMUR, DIAS
HEART, LV, APEX – MURMUR, SYS
HEART, LV, APEX, IMP – DOUBLE
HEART, LV, APEX, IMP – FORCEFUL/SUSTAINED
HEART, LV, APEX, IMP – DISPLACED, LAT

HEART, LV, APEX, IMP – TRIPLE
HEART, S2, SPLIT – REVERSED (PARADOXICAL)
HEART, S3 LV
LIVER – ENLARGED (HEPATOMEGALY)
MENTATION, LEARNING, DEVELOPMENT – DECR [15]
MUSCLES – CRAMPS [MORE COMMON IN F]
MUSCLES, EXTREM, PROX – ATROPHY [13]
MUSCLES, EXTREM, PROX – WEAKNESS [13]
NECK, MUSCLES – WEAK [13]
SPLEEN, SIZE – INCR (SPLENOMEGALY) [RARE]

Differentiation

Cardiomyopathy – Peripartum [11]
Other causes of Cardiomyopathy – Dilated [11]
Other causes of Cardiomyopathy – Hypertrophic [11]

Complications

Atrial Fibrillation
Atrioventricular Heart Block
Cardiomyopathy – Dilated
Heart Failure
Sudden Death

Laboratory

BLOOD, ALDOLASE – INCR
BLOOD, CHOLESTEROL, TOTAL – INCR
BLOOD, CK – INCR
BLOOD, LDH – INCR
BLOOD, LIVER ENZYMES – INCREASED
BLOOD, NT-PROBNP – INCR

ECG [5]

AV COND – 3RD DEGREE BLOCK
DELTA WAVE [2] [9]
DYSRHYTHMIAS – ATRIAL (PACS/OTHERS)
DYSRHYTHMIAS – VENTRICULAR (PVCS/OTHERS)
P WAVE – FLAT
P WAVE, DUR – INCR
PR INTERVAL – SHORT [2] [9]
Q WAVE – ABN
QRS – LVH PATTERN [10]
QRS, AXIS – L
QRS, R WAVE – TALL
QT/QTC INTERVAL – LONG
RATE – DECREASED (SINUS BRADYCARDIA)
ST SEGMENT – DEPR [2]
T WAVE – INVER, ABN

Imaging

CARDIOMEGALY
IVS, THICKNESS – INCR (SEPTAL HYPERTROPHY) [3]
LA, CHAMBER, SIZE – INCR
LV, CHAMBER, SIZE – DECR
LV, MYOCARD – LGE
LV, MYOCARD, WALL THICKNESS – INCR (HYPERTROPHY)
LV, OUTFLOW – OBS
LV, WALL THICKNESS, SEG – INCR
MV, FLOW – REGURG
MV, LEAFLETS, MOTION, SYS – ANT

Other Tests

Ambulatory ECG monitoring
EEG [7]
EMB
EMG [6]
Skeletal muscle biopsy [14]

Genomics

LAMP2

Treatment: Nonpharmacologic

Careful avoidance of hypovolemia/dehydration

Treatment: Pharmacologic

NS

Treatment: Surgical/Invasive

ICD
Pacemaker
Heart transplant [8]

Course

Without transplant:
Males: most die of HF by age 30 year
Females: milder course than males, but cardiac death common

Notes

[1] Phenotypic expression much greater in males; HCM occurs in males by age 20 year and after age 30 years in females; females less often have mental retardation and skeletal myopathy; females less studied than males
[2] WPW pattern; about 70 % of males, 27 % of females; when associated with HCM in males, WPW strongly suggests Danon Disease
[3] Can be massive: largest thicknesses recorded to date
[4] Usually absent; mild when present
[5] ECG abnormalities by early teens in males; 100 % of patients have abnormal ECG
[6] Myogenic in all males
[7] Mild abnormalities in some males
[8] May be considered at earliest signs of HF due to rapid demise; needed by most males by age 30 years
[9] WPW may be sole marker and manifestation in females
[10] LVH and WPW may be first clinical manifestation in young males
[11] Females with Danon Disease often initially misdiagnosed
[12] Ophthalmologic abnormalities:

Include:

Central scotoma
Abnormal color vision
Serious cone-rod dystrophy

May be first indicators of Danon, especially in females
Visual abnormalities occur in about 70 % of males
Females more often have peripheral pigmentary retinopathy; males usually have diffuse/near complete retinal pigment loss

[13] Progressive proximal muscle weakness occurs in 80–90 % of males; seldom debilitating; much less common and normal/less severe in females

[14] Intracytoplasmic vacuoles with autophagic material and glycogen; absent LAMP2 protein expression in males
[15] Usually mild, able to function including hold job, read, enter relationships, live independently
[16] Palpitations (about 70 %) and chest pain (about 40 %) most common symptoms in both males and females

Guidelines

2011 ACCF/AHA guideline for the diagnosis and treatment of hypertrophic cardiomyopathy
J Am Coll Cardiol. 2011;58:e212–60. http://content.onlinejacc.org/article.aspx?articleid=1147838.
2014 ESC Guidelines on diagnosis and management of hypertrophic cardiomyopathy
Eur Heart J. doi:10.1093/eurheartj/ehu284. http://eurheartj.oxfordjournals.org/content/early/2014/08/28/eurheartj.ehu284.

Patient Information

Genetics Home Reference

http://ghr.nlm.nih.gov/condition/danon-disease.

Nord

http://www.rarediseases.org/rare-disease-information/rare-diseases/byID/1179/printFullReport.

ESPANOL

http://ww.nlm.nih.gov/medlineplus/spanish/cardiomyopathy.html.

Professional Information

Early Description

Neurology. 1981;31:51–7. http://www.neurology.org/content/31/1/51.short.

Review

Circ Heart Fail. 2014;7:843–9. http://circheartfailure.ahajournals.org/content/7/5/843.full?sid=385d56bb-ec97-4bbc-be90-abfb1a18cb40.

Children

Circulation. 2005;112:1612–17. http://circ.ahajournals.org/content/112/11/1612.full.

Concentric LVH

Eur Heart J. 2012;33:649–56. http://eurheartj.oxfordjournals.org/search?fulltext=DANON&submit=yes&x=16&y=11.

General Features

Neurology. 2002;58:1773–8. http://www.neurology.org/content/58/12/1773.short.

Genetics Home Reference

http://ghr.nlm.nih.gov/condition/danon-disease.

Natural History

Genet Med. 2011;13:563–8. http://www.nature.com/gim/journal/v13/n6/full/gim9201190a.html.

Ophthalmic Features

Ophthalmology. 2006;113:1010–13. http://www.aaojournal.org/article/S0161-6420(06)00291-0/abstract.

Outcome/Phenotypic Expression

JAMA. 2009;301:1253–9. http://jama.jamanetwork.com/article.aspx?articleid=183623.

Sudden Death in Women

Am J Cardiol. 2012;109:406–11. http://www.sciencedirect.com/science/article/pii/S0002914911029389.

Young Women

J Am Coll Cardiol. 2010;55:408–10. http://content.onlinejacc.org/article.aspx?articleID=1140391.

Updates and More

https://clinicalguidecvd.com/danon

Chapter 31
Cardiomyopathy: Dilated

Management Keys

Rule out ischemic etiology
Rule out reversible causes, including:

- Cardiomyopathy – Peripartum
- Cardiomyopathy – Tachycardia-Induced
- Cardiomyopathy – Takotsubo
- Cobalt toxicity [28]
- Pheochromocytoma

Diagnose underlying disease, including detailed investigation for familial cause [26]
Treat according to evidence-based guidelines: prompt HF medical and device therapy can improve functional class
Manage/prevent associated risks, especially stroke, serious dysrhythmia, end-organ failure
Alcohol abstinence [22]
Caution using beta-blockers when cocaine may be etiologic factor [25]

ICD-10 Code

I42.0

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_31,

Alternate Names/Abbreviation

DCM
FAMILIAL DILATED CARDIOMYOPATHY (FDC) [26]
IDIOPATHIC CARDIOMYOPATHY [12]
IDIOPATHIC DILATED CARDIOMYOPATHY [12]
NONISCHEMIC CARDIOMYOPATHY (NICM)

Description/Etiology

Four chamber cardiac dilatation and LV systolic dysfunction (sometimes with diastolic component) with normal cardiac wall thickness
20–35 % familial (autosomal dominant, recessive or sex-linked)
Depending on cause, disease either confined to heart or part of general systemic disease, often leading to HF/complications/SCD
Causes:

- Alcohol abuse [1]
- Electrolyte imbalance, including:
  - Hypocalcemia
  - Hypophosphatemia
- Endocrine, including:
  - ACROMEGALY
  - DIABETES MELLITUS
  - PHEOCHROMOCYTOMA
- Genetic/mixed genetic/non-genetic, including: [26]
  - BARTH SYNDROME
  - CARDIOMYOPATHY – NONCOMPACTION
  - DUCHENNE/BECK MUSCULAR DYSTROPHY
  - EMERY-DREIFUSS MUSCULAR DYSTROPHY
  - TUBEROUS SCLEROSIS [29]

MYOCARDITIS – bacterial infection, including:

LEGIONELLOSIS
LEPTOSPIROSIS
LYME DISEASE
ROCKY MOUNTAIN SPOTTED FEVER
SHIGELLOSIS
TRICHINELLOSIS
YERSINIOSIS

MYOCARDITIS – protozoal infection, including:

CHAGAS DISEASE

MYOCARDITIS – rickettsial infection, including:

Q FEVER
ROCKY MOUNTIAN SPOTTED FEVER
TYPHUS – SCRUB

MYOCARDITIS – viral infection, including:

ADENOVIRUS
CHICKENPOX
COXSACKIEVIRUS
DENGUE
ENTEROVIRUS
EBOLA VIRUS DISEASE
INFECTIOUS MONONUCLEOSIS
HERPES VIRUS 6
HUMAN IMMUNODEFICIENCY VIRUS
INFLUENZA
LASSA FEVER
LYMPHOCYTIC CHORIOMENINGITIS
MARBURG HEMORRHAGIC FEVER
MENINGOCOCCEMIA
MUMPS
PARVOVIRUS B19
PSITTACOSIS
RUBEOLA
SMALLPOX
WEST NILE FEVER

MYOCARDITIS – inflammation, incuding:

CARDIAC SARCOIDOSIS
DRUG HYPERSENSITIVITY (EOSINOPHILIC MYOCARDITIS)
GIANT CELL MYOCARDITIS [3]
SYSTEMIC LUPUS ERYTHEMATOSIS

METABOLIC, including:

KWASHIORKOR
PELLAGRA
THIAMINE DEFICIENCY

PHYSIOLOGIC, including:

HEAT STROKE
TACHYCARDIA

TOXIC, including:

ANTICANCER RX (ESP ANTHRACYCLINES)
COBALT [28]
COCAINE
ETHANOL

Comorbid Conditions

ALCOHOL USE/EXCESS
DYSRHYTHMIAS – VENTRICULAR [30]
FAMILY HISTORY: SUDDEN DEATH
VARIES WITH CAUSE

Demography

Highly variable according to etiology
Global
All ethnicities
Clinical onset all ages, most often 30–50 year
Males > females (adults)

Pathophysiology

Variable according to etiology
Histology: myocyte hypertrophy/interstitial fibrosis

LV:

Spherical dilatation
Increased wall stress
Functional MR

RV:

Dysfunction in 35 %

Contributory:

Conduction blocks
Dysrhythmias

Signs/Symptoms [9] [16]

ABDOMEN – DISTENSION
ABDOMEN – FLUID (ASCITES)
ABDOMEN – FULLNESS
ABDOMEN – PAIN [ESP RUQ]
APPETITE – DECR (ANOREXIA)
ARTERIAL PRESSURE, DIAS – DECR
ARTERIAL PULSE, AMP – ALTERNATING (PULSUS ALTERNANS)
BODY, APPEARANCE – WASTING (CACHEXIA)
BREATH SOUNDS – CRACKLES (RALES)
BREATH SOUNDS – DECR
BREATH SOUNDS – WHEEZES
BREATHING – DIFF (DYSPNEA)
BREATHING – DIFF, NOCTURNAL (DYSPNEA, NOCT)
BREATHING – DIFF, RECLINING FLAT (ORTHOPNEA)

BREATHING – RAPID (TACHYPNEA)
BREATHING – RHYTHMIC CHANGES (CHEYNE-STOKES)
CHEST – PAIN [4]
CHEST – PALPITATIONS
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)
COUGH
COUGH – NOCT
EXTREM, LOWER, BILAT – EDEMA [5]
EYES – PROMINENT (EXOPHTHALMOS/ PROPTOSIS) [2]
EYES, SCLERA – YELLOW (ICTERUS)
EYES/SKIN – YELLOW (JAUNDICE)
FATIGUE [6]
HEADACHE
HEART, LV, APEX, IMP – DISPLACED, INF
HEART, LV, APEX, IMP – DISPLACED, LAT
HEART, P2, INTENSITY – INCR
HEART, RATE – RAPID (TACHYCARDIA)
HEART, RHYTHM – IRREG [3]
HEART, S3 LV
HEART, S3 RV
HEART, S4 LV
HEART, S4 RV
HEART, SOUNDS, INTENSITY – DECR
HYPOTENSION (BLOOD PRESSURE – DECREASED/LOW) [6]
LIVER – ENLARGED (HEPATOMEGALY)
LIVER – TENDER
MENTATION – CONFUSION
MUSCLES – ATROPHY
MUSCLES – WEAK
NAUSEA
NECK, JVP – ABDOMINOJUGULAR REFLUX
NECK, JVP – ELEV
SKIN – ITCHING (PRURITUS)
SKIN, COLOR – BLUE (CYANOSIS)

SLEEP – DISTURBED (INSOMNIA)
SPLEEN, SIZE – INCR (SPLENOMEGALY)
SPUTUM – BLOOD (HEMOPTYSIS)
URINATION – NIGHTTIME (NOCTURIA)
VOMITING (EMESIS)

Differentiation

CAD
Identifiable causes of cardiomyopathy

Complications

Dysrhythmias – Atrial [13]
Dysrhythmias – Ventricular
Heart Failure
Stroke
Sudden Death
Systemic Emboli

Laboratory [9]

BLOOD, NT-PROBNP – INCR
BLOOD, ST2 – INCR

ECG [9]

AV COND – 1ST DEGREE BLOCK
AV COND – 2ND DEGREE BLOCK
AV COND – 3RD DEGREE BLOCK
DYSRHYTHMIAS – ATRIAL (PACS/OTHERS) [13]
DYSRHYTHMIAS – VENTRICULAR (PVCS/OTHERS) [30]

P WAVE, DUR – INCR, NOTCHED (P MITRALE)
Q WAVE – ABN
QRS – LBBB/LBBB PATTERN
QRS – LONG, NS
QRS – LVH PATTERN
QRS – R PROGRESSION, POOR
QRS – RBBB/RBBB PATTERN
RATE – INCREASED (SINUS TACHYCARDIA)
ST-T WAVE – ABN, NS

Imaging [9]

CARDIOMEGALY
LA, CHAMBER, SIZE – INCR
LV, CHAMBER, SIZE – INCR
LV, DIAS – DYSF [7]
LV, EF – DECR
LV, INTRACAVITY – MASS [THROMBUS]
LV, SYS – DYSF [7]
LV, WALL MOTION – DECR
MV, FLOW – REGURG
MYOCARD – FIBROSIS [14]
PERICARD – FLUID
PUL, VASCULARITY – INCR
RA, CHAMBER, SIZE – INCR [15]
RV, CHAMBER, SIZE – INCR [15]
TV, FLOW – REGURG [15]

Genomics [23]

ACTC
DMD
FKBP12
LMNA
MYH7

RBM20
TAZ-G4.5
TCAP
TNNT2
Z-BAND
6P21 [?]
DSC2 [31]
DSG2 [31]
DSP (DESMOPLAKIN) [31]
JUP [31]
PKP2 [31]
TTN [27]

Other Tests [17]

Card cath [21]
EMB

Treatment: Nonpharmacologic [10] [17]

Alcohol abstinence [22]
Fluid restriction
Low salt diet

Treatment: Pharmacologic [10] [17]

Aldosterone antagonists [19]
Angiotensin-ii receptor blockers
Angiotensin-converting enzyme inhibitors
Beta-blockers
Digoxin [18]
Diuretics

- Furosemide
- Chlorothiazides

Treatment: Surgical/Invasive [10] [17]

CRT [20]
ICD
LVAD
Heart transplant [8]

Prevention

Treat specific cause, when identified, to help prevent progression to more advanced stages

Course

Highly variable depending on

- Complicating features, eg, dysrhythmia, renal dysfunction, thromboembolism
- Functional capacity
- Severity of associated HF
- Specific etiology

Medical treatment and CRT (if eligible) may improve LVEF and functional status

Notes

[1] Most common cause, due to heavy intake; mechanism likely multifactorial, including toxic, secondary neurohormonal, secondary hypertension, nutritional deficiency, genetic vulnerability
[2] Due to chronically increased venous pressure
[3] Suspect with rapidly progressive HF, ventricular dysrhythmias, HB
[4] Chest pain may occur in absence of demonstrable CAD
[5] May be absent even in presence of volume overload, esp younger persons

[6] May be due to low cardiac output
[7] Some patients have combined sys and dias dysfunction
[8] Most common cause of heart transplantation
[9] Usually initial clinical manifestations HF-related, but may be first detected as incidental finding of LVH/cardiomegaly on imaging/ECG for other indications
[10] HF treatment indicated once clinically manifest; prior to HF, treatment dictated by cause if identified, such as by EMB
[11] From LV mural thrombus
[12] Term "idiopathic" refers to all genetic and non-genetic forms after all identifiable causes excluded
[13] Especially AF
[14] Mid-wall; may be associated with adverse prognosis
[15] With HF or primary RV dysfunction
[16] Includes clinical manifestations of RV failure
[17] See Heart Failure chapter and other cardiomyopathies for further information
[18] Some evidence that digoxin may reduce HF admissions
[19] Reduces mortality
[20] Class I indication for NYHA Class 2–4 with LBBB and QRS >150 msc; Class 2a if LBBB and QRS 120–149 msc
[21] To rule out obstructive CAD as cause of cardiomyopathy
[22] LV dysfunction may revert when alcohol is discontinued and is contributory
[23] >30 genetic mutations discovered
[24] Approximately 50 % have identifiable cause; remainder classified as idiopathic
[25] Due to unopposed alpha adrenergic stimulation
[26] Clinical screening of asymptomatic 1st degree relatives; DCM in >1 related family member suggests familial disease
[27] TTN mutations produce truncated forms of titan; appear in about 20 % of patients with end-stage DCM and 2 % of general population
[28] Cobalt toxicity should be considered in patients with prior prosthetic (metal-on-metal) hip replacements; other manifestations include:

Hypothyroidism
Neuropathy
Polycythemia

[29] Tuberous sclerosis usually associated with Rhabdomyoma, but DCM reported in rare cases
[30] Frequent PVCs associated with decreased LVEF, increased incidence of HF, and increased mortality
[31] Originally described in association with ARVD but increasingly found in patients with DCM as well

Guidelines

2013 ACCF/AHA guideline for the management of heart failure
J Am Coll Cardiol. 2013;62:e147–239. http://content.onlinejacc.org/article.aspx?articleid=1695825.

ESC guidelines for the diagnosis and treatment of acute and chronic heart failure 2012
Eur Heart J. 2012;33:1787–47. http://eurheartj.oxfordjournals.org/content/ehj/33/14/1787.full.pdf.

Patient Information

Images

http://www.nlm.nih.gov/medlineplus/ency/imagepages/18136.htm.

Genetics Home REF

http://ghr.nlm.nih.gov/condition/dmd-associated-dilated-cardiomyopathy.

Medlineplus

ENGLISH

http://www.nlm.nih.gov/medlineplus/ency/article/000168.htm.

ESPANOL

http://www.nlm.nih.gov/medlineplus/spanish/ency/article/000168.htm.

Texas Heart Institute

http://www.texasheart.org/HIC/Topics/Cond/dilated.cfm.

Professional Information

Review

Lancet. 2010;375:752–2. http://www.sciencedirect.com/science/article/pii/S0140673609620237.

Review: Genetics of Sudden Cardiac Death

Circ Res. 2015;116:1919–36. http://circres.ahajournals.org/content/116/12/1919.full.

Alcohol and HF

Curr Atheroscler Rep. 2008;10:117–120. http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2365733/.

Cardiac Steatosis

Heart. 2014;100:1107–12. http://heart.bmj.com/content/100/14/1107.abstract.

Cobalt Toxicity/HIP Prosthesis

Circ Cardiovasc Imaging. 2015;8:e003352. http://circimaging.ahajournals.org/content/8/6/e003352.extract?.

Chemical Cardiomyopathies

Am J Med. 2011;124:480–8. http://www.amjmed.com/article/S0002-9343(11)00187-2/abstract.

Childhood Forms

Circulation. 2006;114:2671–78. http://circ.ahajournals.org/content/114/24/2671.full?sid=935a0a43-e90f-4e1d-b314-e459f91528bd.

Childhood Forms: Long-Term

Circulation. 2013;128:2039–46. http://circ.ahajournals.org/content/128/18/2039.full?sid=935a0a43-e90f-4e1d-b314-e459f91528bd.

Children: Recovery of Function (ECHO)

J Am Coll Cardiol. 2014;63:1405–13. http://content.onlinejacc.org/article.aspx?articleID=1833385.

CMRI

Eur Heart J. 2005;26:1461–74. http://eurheartj.oxfordjournals.org/content/26/15/1461.long.

Classification of Cardiomyopathies: AHA Scientific Statement

Circulation. 2006;113:1807–16. http://circ.ahajournals.org/content/113/14/1807.full.

Classification of Cardiomyopathies: ESC

Eur Heart J. 2008;29:270–6. http://eurheartj.oxfordjournals.org/content/29/2/270.full.

EMB: AHA Scientific Statement

J Am Coll Cardiol. 2007;50:1914–31. http://content.onlinejacc.org/article.aspx?articleID=1138635.

Familial Dilated Cardiomyopathy: Genetic Testing Complexities

Circ Cardiovasc Genetics. 2016;9:95–9. http://circgenetics.ahajournals.org/content/9/1/95.full.

Familial Dilated Cardiomyopathy

J Am Coll Cardiol. 1999;34:181–91. http://content.onlinejacc.org/article.aspx?articleid=1125868.

Familial TPN Cardiomyopathy

J Am Coll Cardiol. 2010;55:320–9. http://content.onlinejacc.org/article.aspx?articleid=1140389.

Genetic Forms and HF

Circ Res. 2013;113:660–75. http://circres.ahajournals.org/content/113/6/660.abstract.

Genetics

Eur Heart J. 2015;36:2327–37. http://eurheartj.oxfordjournals.org/content/36/34/2327.

Genetics

J Am Coll Cardiol. 2011;57:1641–49. http://www.ncbi.nlm.nih.gov/pmc/articles/PMC3088091/pdf/nihms-288703.pdf.

Genetics

GeneReviews. 2013. http://www.ncbi.nlm.nih.gov/books/NBK1309/.

Genetics

J Clin Invest. 2013;123:19–26. http://www.jci.org/articles/view/62862.

Genetics: 6P21

Eur Heart J. 2014;35:1069–77. http://eurheartj.oxfordjournals.org/content/35/16/1069.

Genetic Testing: Complexities

Circ Cardiovasc Genetics. 2016;9:95–9. http://circgenetics.ahajournals.org/content/9/1/95.extract.

HIV

Circulation. 2014;129:1781–89. http://circ.ahajournals.org/content/129/17/1781.full.

Mitral Valve Repair

Semin Thorac Cardiovasc Surg. 2012;24:51–8. http://www.ncbi.nlm.nih.gov/pubmed/22643662.

Myocardial Fibrosis

JAMA. 2013;309:896–908. http://jama.jamanetwork.com/article.aspx?articleid=1660382.

Outcomes: Idiopathic Dilated Cardiomyopathy

Am J Cardiol. 2015;116:952–9. http://www.ajconline.org/article/S0002-9149(15)01539-8/abstract.

Pheochromocytoma

Acta Cardiol. 1998;53:227–9. http://www.ncbi.nlm.nih.gov/pubmed/9842409.

Pheochromocytoma

Int J Clin Pract. 2003;57:547–58. http://www.ncbi.nlm.nih.gov/pubmed/12918898?dopt=Abstract.

PVCS: LVEF/HF/Mortality

J Am Coll Cardiol. 2015;66:101–9. http://content.onlinejacc.org/article.aspx?articleID=2383167.

RV Systolic Dysfunction

Circulation. 2013;128:1623–33. http://circ.ahajournals.org/content/128/15/1623.full.

Sudden Death: Stratification

J Am Coll Cardiol. 2014;63:1879–89. http://content.onlinejacc.org/article.aspx?articleID=1827541.

TTN Mutations

N Engl J Med. 2012;366:619–28. http://www.nejm.org/doi/full/10.1056/NEJMoa1110186.

Tuberous Sclerosis (Case Report)

Eur Heart J. 2015;36:2338. http://eurheartj.oxfordjournals.org/content/36/34/2338.extract?etoc.

X-Linked DCM

Circulation. 1993;87:1854–65. http://circ.ahajournals.org/content/87/6/1854.abstract.

Updates and More

https://clinicalguidecvd.com/dcm

Chapter 32
Cardiomyopathy: Hypertrophic (Yamaguchi Disease)

Management Keys

- Aggressive treatment of atherosclerotic risk factors
- Anticoagulation for both persistent/intermittent AF due to high risk of thromboemboli
- Avoid peripheral vasodilators in presence of resting or provocable LVOT obstruction (dehydration, high-dose diuretics, vasodilating drugs including dihydropyridine CCBs) [44]
- Careful evaluation of unexplained syncope, which is associated with increased risk of SCD, especially in young patients [42] [43]
- Counsel regarding participation in competitive sports
- Diagnose/quantify LVOT obstruction [30]
- Differentiate from Athlete Heart in highly trained athletes [47]
- Individualize treatment due to diverse pathology
- Pregnancy management by obstetricians/cardiologists trained in high-risk obstetrics
- Regular follow up every 1–3 years depending on individual patient disease characteristics
- Risk stratification for SCD regardless of presence/absence of symptoms
- Rule out other causes of LVOT obstruction, especially prior to intervention, including subaortic membranes, other MV leaflet abnormalities, mid-cavity obstruction, Cardiac Amyloidosis

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_32,

Screen all 1st degree relatives
Septal reduction (myectomy, alcohol ablation) for LVOT obstruction only in centers experienced in these techniques
Use CMRI when echo inconclusive/inadequate to detect myocardial hypertrophy in nonvisualized areas, especially LV apex

ICD-10 Code

142.1

Alternate Names/Abbreviation [21]

HCM
HYPERTROPHIC OBSTRUCTIVE CARDIOMYOPATHY (HOCM)
YAMAGUCHI DISEASE (APICAL HCM)

Description/Etiology

Cardiomyopathy caused by mutations in cardiac sarcomere protein genes [3] associated with wall thickness >14 mm (adults) or >2 standard deviations > predicted mean (children) (by any imaging mode)
Clinical diagnosis by increased LV wall thickness (≥13 mm) in presence of nondilated ventricular chambers, unexplained by other cardiac or systemic disease
Diagnosis with wall thickness of 13–14 mm requires other features, including positive FH, ECG abnormalities, other tests
Familial form: autosomal dominant

Comorbid Conditions

CARDIOMYOPATHY – NONCOMPACTION
FAMILY HX: SUDDEN DEATH
LEOPARD SYNDROME
NOONAN SYNDROME
PREEXCITATION SYNDROMES
PRKAG2 SYNDROME
VON WILLEBRAND SYNDROME [48] [ACQUIRED]
WOLFF-PARKINSON-WHITE SYNDROME

Demography

Global
Gender equal
All ethnicities
Clinical onset: any age; most often LVH apparent in adolescence/young adults
Prevalence: 1/500
Many affected persons undiagnosed

Pathophysiology

Among patients with HCM:

One-third: constant resting gradient of ≥30 mmHg [30]
One-third: labile/dynamic gradient of <30 mm at rest/≥30 mmHg with physiologic provocation [12] [30]
One-third: nonobstructive form of <30 mmHg at rest and with physiologic provocation

Anatomy of LV hypertrophy in HCM:

Age-dependent penetrance, usually around puberty, but ranges from infancy to advanced age

Any segment of LV myocardium can be affected, with basal anterior IVS most common
>95 % asymmetric but may be concentric, including familial forms
Increased wall thickness confined to only one or two segments, especially apical, in 10 %
Increased wall thickness may be noncontiguous, with normal areas of thickness separated by areas of increased thickness

Histology:

Abnormal intracoronary arterioles with decreased luminal size/decreased vasodilatory capacity [27]
Myocyte hypertrophy and disarray
Increased interstitial fibrosis

MV:

Anomalous papillary muscle insertion into anterior leaflet
Elongated leaflets/accessory tissue
MR usually present with systolic SAM [35]
Papillary muscle anterior displacement/hypertrophy
SAM usually present, but not required for diagnosis [30]

Other features:

Autonomic dysfunction
Diastolic dysfunction [1]
Dysrhythmias
MV inflow obstruction
Myocardial fibrosis
Myocardial ischemia/infarction [2]
LV Apical Aneurysm [26]

Signs/Symptoms

ARTERIAL PULSE – DOUBLE (BISFERIENS)
ARTERIAL PULSE, RISE – RAPID
BREATHING – DIFF (DYSPNEA) [ESP WITH EXERTION]

BREATHING – DIFF, NOCTURNAL (DYSPNEA, NOCT)
CHEST – PAIN [2]
CHEST – PALPITATIONS
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE) [42] [43]
DIZZY/LIGHTHEADED/PRESYNCOPE [42]
FATIGUE
HEART, LSB, LOWER – THRILL, SYS
HEART, LSB, MID – MURMUR, SYS [33]
HEART, LV, APEX – MURMUR, DIAS
HEART, LV, APEX – MURMUR, SYS [34] [35]
HEART, LV, APEX, IMP – FORCEFUL/SUSTAINED
HEART, LV, APEX, IMP – FORCEFUL/SUSTAINED
HEART, LV, APEX, IMP – DISPLACED, LAT
HEART, LV, APEX, IMP – DOUBLE
HEART, LV, APEX, IMP – TRIPLE
HEART, S2, SPLIT – REVERSED (PARADOXICAL)
HEART, S3 LV
NECK, JVP, A WAVE – INCR/LARGE (CANNON WAVE)

Differentiation [15]

Aortic Stenosis – Subvalvular
Aortic Stenosis – Supravalvular
Aortic Stenosis – Valvular
Athlete Heart
Cardiac Amyloidosis (Al, TTR, ATTR)
Cardiomyopathy – Danon Disease
Cardiomyopathy – Takotsubo
Fabry Disease
Friedreich Ataxia
Glycogen Storage Disease [51]
Hypertension – Systemic Arterial (with LVH)
Leopard Syndrome
Mitochondrial Disease

Noonan Syndrome
Pheochromocytoma-related LVH

Complications

Acute hypotension [44]
AF
Bleeding (Heyde Syndrome) [48]
Cardiac Arrest
Dysrrhthmias – Atrial (ESP AF) [29] [52]
Dysrhythmias – ventricular [43]
HF – acute, uncommon, triggered by:

- AMI/myocardial ischemia [44]
- Comorbidities (eg, Anemia, Hyperthyroidism)
- Dysrhythmias
- MR – Acute (due to chordal rupture, infective endocarditis)

HF – chronic [32]
Myocardial ischemia [16]
Peripheral emboli [52]
Pulmonary Hypertension
Stroke – Ischemic
Sudden death [23] [28] [53]
Syncope [42] [43]

Laboratory

NS [40] [41]

ECG [4]

DELTA WAVE [5]
DYSRHYTHMIAS – ATRIAL (PACS/OTHERS)
DYSRHYTHMIAS – VENTRICULAR (PVCS/OTHERS) [43]

P WAVE – FLAT
P WAVE, DUR – INCR
PR INTERVAL – SHORT [5]
Q WAVE – ABN [6]
QRS – LVH PATTERN
QRS, AXIS – L
QRS, R WAVE – TALL
QT/QTC INTERVAL – LONG
ST SEGMENT – DEPR [2]
T WAVE – INVER, ABN

Imaging [9] [14]

CARDIOMEGALY [8]
IVS, THICKNESS – INCR (SEPTAL HYPERTROPHY)
LA, CHAMBER, SIZE – INCR [13] [37]
LV, DIAS – DYSF [50]
LV, EF – INCR [OR N] [38]
LV, MYOCARD – LGE [53]
LV, MYOCARD, WALL THICKNESS – INCR (HYPERTROPHY) [10]
LV, OUTFLOW – OBS [12]
LV, WALL THICKNESS, SEG – INCR [10]
MV, FLOW – REGURG [35] [36]
MYOCARD – FIBROSIS

Genomics [25]

ACTC1
ACTN2
ANKRD1
CAV3
CSRP3
DES
GLA
ILK

JPH2
LAMP2
LDB3
MTTG
MTTI
MTTK
MTTQ
MYBPC3
MYBPC3
MYH6
MYH7
MYL2
MYL3
MYLK2
MYOZ2
NEXN
PDLIM3
PLN
PRKAG2
TCAP
TNNC1
TNNT2
TPM1
TTR
VCL
MRPL3 [22]
TNNI3

Other Tests

Ambulatory 24-h ECG monitor [7]
Exercise test

- Hypotensive BP response
- With echo: provoke dynamic LVOT obstruction [30]
- Functional status

Coronary angiography for CAD

Coronary angiography/intracoronary contrast echo prior to alcohol ablation [16]
Cardiac MRI
EMB [39]
EP

Treatment: Nonpharmacologic

Avoid:

- Afterload-reducing drugs
- Burst activities (sprinting)
- Competitive endurance training
- Dehydration
- Intense isometric exercise

Comorbidity management
Fluid administration for acute hypotension
Low intensity aerobic exercise program

Treatment: Pharmacologic [19] [44] [45]

Aggressive Rx of usual CAD risk factors
Anticoagulation for recurrent/persistent AF [52]
Bacterial IE prophylaxis: case by case basis
Beta-blockers [17]
CCBs (esp Verapamil) [17]
IV Phenylephrine (or other vasoconstrictor) for Acute Hypotension [19]
Oral diuretic for dyspnea without LVOT obstruction [18]
Relative/absolute contraindication with LVOT obstruction:

- Pure vasodilators
- High dose diuretics
- ACEIs and ARBs

Treatment of LV systolic dysfunction

Treatment: Surgical/Invasive [20] [45]

Arrhythmia modification
Dual chamber pacing ICD [43]
MV repair/replacement (indicated in up to 20 % of patients undergoing septal reduction)
Septal reduction

- Surgical myectomy (Morrow procedure)
- Alcohol ablation [31]

Cardiac transplantation

Prevention

SCD risk stratification
Genetic testing/counseling of family members
Preconception/pregnancy counseling, planning, management
Counseling to avoid competitive sports participation
Occupational choice/counseling

Course

Variable per severity/genetics/type
Many affected persons live normal lifespans with no HCM clinical manifestations

Notes

[1] With exercise: systolic BP falls or fails to rise >20 mmHg in 25 % of patients
[2] Due to myocardial supply–demand mismatch/myocardial bridging; frequency unrelated to CAD
[3] Autosomal dominant with high penetration: >95 % lifetime risk for developing clinical/phenotypic evidence of HCM

[4] Abnormal in 75–95 % of patients with HCM; may precede LV wall thickening and does not correlate with echo severity or pattern of LVH
[5] WPW pattern
[6] Mimics Acute Myocardial Infarction
[7] As part of initial evaluation and when symptoms such as palpitations and lightheadedness occur; NSVT occurs in 25 % of adults; PSVT occurs in >35 %
[8] Variable; cardiomegaly due to enlarged LV/LA; does not correlate with severity of obstruction
[9] Doppler/echo used for initial evaluation, serial monitoring, and family member screening; LV wall thickness should be measured at MV, mid-LV, and apex; LV wall motion abnormalities with exercise echo may correlate with outcomes
[10] A variety of morphological patterns have been described, including asymmetric hypertrophy of IVS
[11] Features of SAM (also found in normal persons and other conditions, eg, TGA, Discrete Subaortic Stenosis, COA, Pompe Disease, Friedreich Ataxia) include:

Abnormal coaptation
Annulus calcification
Chordal elongation
Leaflet prolapse
Leaflet thickening

[12] Dynamic obstruction correlates with risk for complications including SCD; may be provoked by amyl nitrate with echo/Doppler; also occurs in other disorders, including calcification of posterior MV annulus, HTN, hypovolemia, hyper-contractile states
[13] Increased LA volume index ($\geq$34 ml/m^2) associated with increased incidence of abnormal LV diastolic filling and less favorable outcomes
[14] Also used for preoperative myectomy evaluation for length of myectomy, MR quantification
[15] Contrast CMRI especially useful when echo nondefinitive

[16] To define anatomy of septal perforators, exclude CAD, ensure correct localization of alcohol
[17] Initial treatment of symptomatic patients; improve exertional dyspnea by decreasing HR/O2 consumption; efficacy in patients without symptoms uncertain
[18] Use with caution in patients with LVOT obstruction
[19] IV positive inotropes, eg dopamine, norepinephrine contraindicated with LVOT obstruction
[20] Imperative: operator and institutional experience
[21] >70 names of this condition have been used in the literature, initially popularly termed Idiopathic Hypertrophic Subaortic Stenosis, now seldom used because 30 % of affected patients have no LV outflow obstructive component
[22] Monogenic: abnormal assembly of mitochondrial respiratory chain
[23] Most common cause of SCD in young/competitive athletes
[24] Fibrosis is progressive and may be indicator for increased risk of ventricular dysrhythmias and future systolic dysfunction
[25] HCM genetics highly heterogeneous; most mutations private and not recurring; MYH7/MYBPC3 comprise >75 % of cases; additional genetic mutations likely involved
[26] Subgroup of HCM patients at increased risk of SCD, embolic stroke, HF
[27] Cause decreased myocardial blood flow during stress; over time may cause recurrent myocardial ischemia, fibrosis, cell death
[28] Risk factors for SCD in patients with HCM:

Decreased BP response to exercise
Extreme LVH
FH premature HCM-related death in close/multiple relatives
LGE
NSVT on serial ambulatory ECGs
Unexplained syncope

[29] AF/flutter occurs in as many as 25 % of HCM patients at some time and is one cause of HF and peripheral emboli
[30] LVOT obstruction at rest/effort is most important determinant of HF symptoms, stroke, death; highly variable even in same patient, necessitating provocative testing when no/low gradient at rest, either TST (preferred) or other means (pharmacological, Valsalva)
[31] Absolute alcohol infusion into 1st/2nd major perforator coronary artery; improved functional status/mortality similar to surgical myectomy; AV block most common complication of procedure
[32] HF symptoms occur in about 5 % of HCM patients; marked remodeling of LV myocardium to hypertrophy regression/LV dilatation with systolic dysfunction; LVOT obstruction (rest/provoked) is strong HF predictor
[33] Radiates to upper RSB and apex; increased intensity with decreased ventricular preload/afterload, eg, rising from squatting to standing position/Valsalva
[34] Murmur due to radiation of LVOT murmur/MR
[35] MR also dynamic and degree correlates with magnitude of LVOT obstruction
[36] MR jet: inferior/lateral oriented; when central/anterior directed, intrinsic MV abnormality may be present and should be sought
[37] Multifactorial cause, mainly due to MR/increased LV filling pressure
[38] LVEF is unreliable measure of LV systolic function in presence of LVH
[39] Not part of routine evaluation; may be used to assess for myocardial infiltrative/storage disease
[40] Laboratory tests should be performed in all HCM patients for target organ dysfunction (eg, renal disease) and in evaluation for other causes of HCM (eg, liver function for Danon Cardiomyopathy, blood IGM for Cardiac Amyloidosis, blood sugar for some forms of mitochondrial DNA disease)
[41] Increased BNP, NT-PROBNP, Troponin T associated with CV events, HF, SCD

[42] Causes of presyncope/syncope in HCM: Hypovolemia, AV Heart Block, Sinus Node Dysfunction, Sustained VT, LVOT obstruction, atrial dysrhythmia with rapid ventricular response (especially AF), abnormal vascular reflexes; patients with HCM may also have comorbidities that cause syncope (eg, seizure disorder); neural-related syncope (vasovagal) is not associated with increased risk of SCD risk in patients with HCM

[43] Unexplained syncope near time of initial diagnosis may warrant ICD implant

[44] Rarely, HCM patients with LVOT obstruction can present with hypotension/pulmonary edema, resembling AMI: use of vasodilators/positive inotropes can be life-threatening in this setting and treatment should instead consist of oral/IV beta-blockers or vasoconstrictors (eg, phenylephrine, metaraminol, norepinephrine)

[45] LV mid-cavity obstruction: often also have apical aneurysm; patients often very symptomatic; may be treated with high-dose beta-blockers/diltiazem/verapamil but response often suboptimal; surgical myectomy may be considered; apical aneurysms rarely need treatment when occur alone

[46] Select patients with SVT, ventricular pre-excitation, monomorphic sustained VT; not for SCD risk assessment

[47] In "gray zone" of LV thickness 13–15 mm, the following findings are useful:

LV internal dimension: >54 mm in athlete
LA internal dimension: <40 mm in HCM; >39 mm in athlete
LV filling/relaxation: abnormal in HCM; normal in athlete
ECG diffuse T wave inversion: present in HCM; absent in athlete
FH HCM: positive in HCM; absent in athlete

[48] Heyde Syndrome: acquired von Willebrand syndrome related to bleeding from GI angiodysplasia in patients

with AS and HOCM; caused by destruction of circulating von Willebrand factor multimers by sheer stress of blood flow through stenotic orifice

[49] Thin-filament type gene mutation associated with increased incidence of LV dysfunction/HF

[50] LV diastolic dysfunction can appear preclinically/without associated LVH in some patients with genetic mutation encoding a component of sarcomeres; suggests that diastolic dysfunction is early HCM phenotype rather than secondary to LVH

[51] Glycogen Storage Disease due to gene mutation of AMP-activated protein Kinase (PRKAG2); differentiated by EP abnormalities, especially ventricular preexcitation

[52] High risk of thromboembolism associated with AF requires anticoagulation (both persistent/intermittent types)

[53] LGE in patients with low risk HCM (by other current standards) associated with increased risk of SCD

[54] Eg, phosphodiesterase 5 inhibitor use for sexual dysfunction, which is common in patients with HCM

Guidelines

2011 ACCF/AHA guideline for the diagnosis and treatment of hypertrophic cardiomyopathy

Circulation. 2011;124:e783–831. http://circ.ahajournals.org/content/124/24/e783.

2014 ESC guidelines on diagnosis and management of hypertrophic cardiomyopathy

Eur Heart J. 2014. http://eurheartj.oxfordjournals.org/content/35/39/2733.

Patient Information

Cardiomyopathy Association (CMA)

http://www.cardiomyopathy.org.

Childrens Cardiomyopathy Foundation (CCF)

http://www.childrenscardiomyopathy.org.

Cleveland Clinic

http://my.clevelandclinic.org/heart/disorders/hcm/default.aspx.

Genetics Home Reference

http://ghr.nlm.nih.gov/condition/familial-hypertrophic-cardiomyopathy.

Hypertrophic Cardiomyopathy Association (HCMA)

http://www.4hcm.org.

Images

http://www.nlm.nih.gov/medlineplus/ency/imagepages/1056.htm.
http://www.nlm.nih.gov/medlineplus/ency/imagepages/1097.htm.
http://www.nlm.nih.gov/medlineplus/ency/imagepages/18141.htm.

Medline Plus

ENGLISH

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ESPANOL

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Circ Res 2015;116:1919–36. http://circres.ahajournals.org/content/116/12/1919.full.

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Alcohol Septal Ablation

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Alcohol Septal Ablation: Long-Term Outcomes

Eur Heart J. 2014;35:2040–45. http://eurheartj.oxfordjournals.org/content/35/30/2040

Alcohol Septal Ablation: Outcome Predictors

Circ Cardiovasc Interv. 2016;9:e002675. http://circinterventions.ahajournals.org/content/9/3/e002675.abstract

Apical HCM: ECG

Am J Med. 2014;127:31–3. http://www.sciencedirect.com/science/article/pii/S0002934313008681

Athlete Heart Differentiation

Am J Cardiol. 2014;114:1383–89. http://www.sciencedirect.com/science/article/pii/S0002914914015914

Atrial Fibrillation

Heart. 2014;100:465–72. http://heart.bmj.com/content/100/6/465.abstract

Atrial Fibrillation: Outcomes After Nonpharm RX

Heart Rhythm. 2015;12:1438–47. http://www.heartrhythmjournal.com/article/S1547-5271(15)00341-0/abstract

Atrial Fibrillation (Paroxysmal)/Exercise Capacity

Heart. 2014;100:624–30. http://heart.bmj.com/content/100/8/624.abstract.

BNP: Outcomes

Eur Heart J. 2013;34:2529–37. http://eurheartj.oxfordjournals.org/content/34/32/2529.abstract.

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Heyde Syndrome

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Circ Heart Fail. 2016;9:e002764. doi: 10.1161/CIRCHEARTFAILURE.115.002764. http://circheartfailure.ahajournals.org/content/9/3/e002764.abstract.

Myocardial Fibrosis/LGE: Risk for SCD

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LV Dias DYSF Preclinical/Independent of LVH

Circulation. 2002;105:2992–97. http://circ.ahajournals.org/content/105/25/2992.full.

LV Outflow Obstruction

J Am Coll Cardiol. 2009;54:191–200. http://content.onlinejacc.org/article.aspx?articleid=1139879.

Morrow Procedure

Circulation. 1975;52:88–102. http://circ.ahajournals.org/content/52/1/88.abstract.

Mortality in Young Persons with Current Treatment Strategies

Circulation. 2016;133:62–73. http://circ.ahajournals.org/content/133/1/62.abstract.

Mortality/Thromboembolism

Heart. 2014;100:465–72. http://heart.bmj.com/content/100/6/465.abstract.

MV Inflow OBS

Circulation. 2013;128:870–72. http://circ.ahajournals.org/content/128/8/870.full.

Myocardial Fibrosis

J Am Coll Cardiol. 2012;60:922–29. http://content.onlinejacc.org/article.aspx?articleid=1352834.

Myocardial Fibrosis

Heart. 2014;100:631–38. http://heart.bmj.com/content/100/8/631.abstract.

Obesity

J Am Coll Cardiol. 2013;62:449–57. http://content.onlinejacc.org/article.aspx?articleID=1686303.

Outcomes: Obstructive vs Nonobstructive HCM

Am J Cardiol. 2015;116:938–44. http://www.ajconline.org/article/S0002-9149(15)01534-9/abstract.

Paradox Response to Exercise

J Am Coll Cardiol. 2013;62:842–50. http://content.onlinejacc.org/article.aspx?articleID=1709473.

Pharmacological Treatment

Eur Heart J. 2012;33:1724–33. http://eurheartj.oxfordjournals.org/content/33/14/1724.full?sid=591bc7a4-51ae-4173-9504-e34e7cac981d.

Phosphodiester 5 Use in Patients with HCM

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Circulation. 2013;127:585–93. http://circ.ahajournals.org/content/127/5/585.full.

Risk Stratification/Outcomes: Validation of Guidelines

Heart. 2013;99:534–41. http://heart.bmj.com/content/99/7/509.extract.

Septal Myectomy: USA

J Am Coll Cardiol. 2005;46:470–76. http://content.onlinejacc.org/article.aspx?articleid=1136777.

Septal Myectomy: Europe

Eur Heart J. 2012;33: 2080–87. http://eurheartj.oxfordjournals.org/content/33/16/2080.full?sid=591bc7a4-51ae-4173-9504-e34e7cac981d.

Septal Myectomy: Effect on PAH

Eur Heart J. 2014;35:2032–39. http://eurheartj.oxfordjournals.org/content/35/30/2032.

Septal Myectomy vs Alcohol Ablation

Circulation. 2014;130:1617–24. http://circ.ahajournals.org/content/130/18/1617.full.

Sudden Death

Circulation. 2010;121:445–56. http://circ.ahajournals.org/content/121/3/445.full?sid=baa0fe51-fe9a-44a3-b361-ef1356cdfbec.

Sudden Death: Prevention

Heart. 2014;100:254–60. http://heart.bmj.com/content/100/3/254.extract.

Sudden Death: ICD Prevention

J Am Coll Cardiol. 2013;61:1527–35. http://content.onlinejacc.org/article.aspx?articleID=1673454.

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Eur Heart J. 2014;35:2010–20. http://eurheartj.oxfordjournals.org/content/35/30/2010.

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Circulation. 2013;128:209–16. http://circ.ahajournals.org/content/128/3/209.full.

Syncope/Sudden Death

Circulation. 2009;119:1703–10. http://circ.ahajournals.org/content/119/13/1703.full.

Thin Filament Mutations: Phenotype/Outcomes

J Am Coll Cardiol. 2014;64:2589–600. http://content.onlinejacc.org/article.aspx?articleID=2042965.

Wall Motion Abnormalities by Echo/CMR as Outcome Predictors

Eur Heart J Cardiovasc Imaging. 2015;16:423–32. http://ehjcimaging.oxfordjournals.org/content/16/4/423.full?etoc.

Updates and More

https://clinicalguidecvd.com/hcm

Chapter 33
Cardiomyopathy: Iron Overload (Hemochromatosis)

Management Keys

Close monitoring/early intervention (phlebotomy/chelation therapy) among patients with IOC/at increased risk, especially disorders requiring multiple blood transfusions, stem cell treatment, bone marrow transplant, as progression to overt HF can be prevented

Recognize stigmata of iron overload in patients receiving chronic blood transfusions and with diastolic dysfunction and Restrictive Cardiomyopathy

Screen first degree relatives of patients with Hereditary Hemochromatosis

ICD-10 Code

E83.110 (HEREDITARY HEMOCHROMATOSIS)

Alternate Names/Abbreviation

IRON OVERLOAD CARDIOMYOPATHY (IOC)
HEMOCHROMATOSIS

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_33,

Description/Etiology [28]

Myocardial dysfunction (primarily diastolic) due to excess cardiac iron deposition, progressing to Dilated Cardiomyopathy

Excess body iron accumulation occurs with:

- Blood transfusions
- Congenital increased GI absorption (Hereditary Hemochromatosis)
- Increased GI absorption assd with other conditions
- Increased dietary intake

2 phenotypes:

- Dilated Cardiomyopathy (most common)
- Restrictive Cardiomyopathy

Excess iron overload associated with hyperadrenergic state (reversible with iron removal)

Hereditary Hemochromatosis: autosomal dominant with 4 subtypes, all causing increased GI iron absorption in presence of normal diet

Comorbid Conditions

ACERULOPLASMINEMIA
APLASTIC ANEMIA
ATRIAL FIBRILLATION [25]
CHRONIC KIDNEY DISEASE
CHRONIC LIVER DISEASE
CONGENITAL ATRANSFERRINEMIA
CONGENITAL DYSERYTHROPOIETIC ANEMIA
DIABETES MELLITUS
DIAMOND-BLACKFAN ANEMIA
FRIEDREICH ATAXIA
HEMOCHROMATOSIS [15]
HEPATITIS B
HYPOGONADISM

HYPOTHYROIDISM
LEUKEMIA
LIVER CIRRHOSIS
MYELODYSPLASTIC SYNDROMES
MYELOFIBROSIS
MYELOPROLIFERATIVE DISORDERS
PARENTERAL IRON THERAPY
PORPHYRIA CUTANEA TARDA
SICKLE CELL DISEASE/TRAIT
SIDEROBLASTIC ANEMIA
STEM CELL TRANSPLANT
THALASSEMIA [14] [18]

Demography

Variable according to etiology
Global increase due to increased uses of bone marrow transplant, stem cell therapy, and greater longevity of patients with congenital hemoglobinopathies
Increased incidence in Sub-Saharan Africans

Pathophysiology

(Normal iron metabolism: absorbed primarily in duodenum; transported via ferroportin, which is inhibited by hepcidin; stored in tissue as ferritin)
Excess deposition of iron in myocytes, beginning in ventricular myocardium and progressing to involve atrial myocardium and conduction system; epicardial iron concentration generally higher than in endocardium
Typically progresses from Restrictive Cardiomyopathy to remodeling and Dilated Cardiomyopathy
LV diastolic dysfunction may be masked by high-output state due to anemia associated with hematologic disorders (e.g., hemoglobinopathies)

Signs/Symptoms [7]

ABDOMEN – FLUID (ASCITES) [4]
ABDOMEN – PAIN [3] [ESP RUQ]
ARTERIAL PULSE, AMP – ALTERNATING (PULSUS ALTERNANS) [4]
ARTERIAL PULSE, AMP – DECR/ABS [4]
BREATH SOUNDS – CRACKLES (RALES) [4]
BREATHING – DIFF (DYSPNEA) [4] [5]
BREATHING – DIFF, RECLINING FLAT (ORTHOPNEA) [4] [5]
CHEST – PAIN [4]
CHEST – PALPITATIONS
CHEST, ANT, RSB – PULSATION [4]
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)
EXTREM, LOWER, BILAT – EDEMA [4]
EYES – PROMINENT (EXOPHTHALMOS/PROPTOSIS) [4]
FATIGUE
GENITALS, TESTICLES, BILAT – ATROPHY [3]
HEART, LSB, LOWER – MURMUR, SYS [4]
HEART, LV, APEX – MURMUR, SYS [4]
HEART, P2, INTENSITY – INCR
HEART, RATE – RAPID (TACHYCARDIA) [4]
HEART, RATE – SLOW (BRADYCARDIA)
HEART, S3 LV [4]
HEART, S3 RV [4]
HEART, S4 LV [4]
HEART, S4 RV [4]
HEART, SOUNDS, INTENSITY – DECR [4]
HYPOTENSION (BLOOD PRESSURE – DECREASED/LOW) [4]
JOINTS – PAIN (ARTHRALGIA) [3] [6]
LIVER – ENLARGED (HEPATOMEGALY)
LIVER – PULSATION, SYS [4]
MUSCLES – ATROPHY [4]
MUSCLES – WEAK [4]

NECK, JVP – ELEV [4]
NECK, JVP – INSP RISE (KUSSMAUL SIGN) [4]
NECK, JVP, A WAVE – INCR/LARGE (CANNON WAVE) [4]
NECK, JVP, V WAVE – INCR/LARGE [4]
NECK, JVP, X DESCENT – PROMINENT/RAPID [4]
NECK, JVP, Y DESCENT – RAPID [4]
SEX, FUNCTION – DECR/ABSENT (IMPOTENCE) [3]
SKIN, PIGMENTATION – ABN [3] [12]
THROAT – PAIN/TIGHTNESS [4]

Differentiation [2]

Other Causes of Cardiomyopathy – Restrictive
Other Causes of Cardiomyopathy – Dilated
Other Causes of Dysrhythmias
Other Causes of Heart Block

Complications [2]

Advanced Heart Block [26]
Dysrhythmias
Heart Failure
Pulmonary Hypertension
Sudden Death

Laboratory [2]

BLOOD, FERRITIN – INCR [8]
BLOOD, LIVER ENZYMES – INCREASED
BLOOD, TRANSFERRIN SATURATION – INCR [8]
GENETIC TESTING [1]
ALSO TEST FOR ASSD DIS/COMPS [19]

ECG [10]

AV COND – 1ST DEGREE BLOCK [26]
AV COND – 2ND DEGREE BLOCK [26]
AV COND – 3RD DEGREE BLOCK [26]
DYSRHYTHMIAS – ATRIAL (PACS/OTHERS) [25]
DYSRHYTHMIAS – VENTRICULAR (PVCS/OTHERS) [25]
P WAVE – BIFID [4]
QRS – LONG, NS [4]
QRS – LVH PATTERN
ST-T WAVE – ABN, NS [4] [20]

Imaging [27]

ART, PUL, PRESS – INCR
CARDIOMEGALY
LA, CHAMBER, SIZE – INCR [4]
LUNGS, INTERSTITIUM – EDEMA/INFILTRATES [4]
LV, CHAMBER, SIZE – INCR [9]
LV, DIAS – DYSF [4]
LV, SYS – DYSF [9]
LV, WALL THICKNESS, SEG – INCR [24]
MRI ESP USEFUL FOR DETECTING MYOCARD IRON OVERLOAD [21]
MV, FLOW – REGURG [4]
MYOCARD – FIBROSIS [MRI] [16]
MYOCARD – SCAR [4]
PLEURA – FLUID
PUL, VASCULARITY – INCR [4]
RA, CHAMBER, SIZE – INCR [4]
RV, CHAMBER, SIZE – INCR [9]
RV, DIAS – DYSF
RV, SYS – DYSF
TV, FLOW – REGURG [4]

Genomics [13]

HAMP
HEMOJUVELIN [HJV]
HFE
SLC40A1
TfR2

Other Tests

Ambulatory ECG monitoring
EMB [11]
Liver biopsy/ultrasound [17]

Treatment: Nonpharmacologic

Avoid iron containing supplements, Vitamin C, red meat intake
Alcohol restriction
Phlebotomy [22]

Treatment: Pharmacologic

Chelation [23]
Guideline-directed HF protocol

Treatment: Surgical/Invasive

Pacemaker
Heart transplant

Prevention

Dietary iron restrictions

Course

Variable depending on stage/treatment when first detected

Notes

[1] For hereditary form and hemoglobinopathies
[2] Does not include non-cardiac features of Hemochromatosis, especially DM-related
[3] Hemochromatosis
[4] Due to Restrictive Cardiomyopathy
[5] Pulmonary signs and symptoms also occur due to PAH in patients with Hemolytic Anemia (group 5 PAH)
[6] Most often hands/fingers
[7] Signs/symptoms due to LV/RV HF; list includes select key features of Hereditary Hemochromatosis
[8] Useful for screening but poorly correlates with excess cardiac iron deposits
[9] Advanced, Dilated Cardiomyopathy
[10] Unresolved whether ECG changes are mainly due to LV dysfunction or iron deposition in conduction paths per se
[11] Limited value due to patchy deposition in heart; no stainable iron is present in normal heart
[12] Bronze skin
[13] All autosomal recessive except Hemochromatosis Type 4, which is autosomal dominant
[14] Also an increase in iron absorption due to inappropriate hepcidin suppression caused by ineffective erythropoiesis
[15] Typically seen in CKD and anemias such as Aplastic Anemia, Sickle Cell Disease, Sideroblastic Anemia, Thalassemia
[16] Iron overload on MRI
[17] Evaluation for Cirrhosis

[18] IOC: leading cause of death in Thalassemia Major
[19] Eg, DM, Hypothyroidism, Hypogonadism
[20] Nonspecific repolarization changes also occur with chelation therapy
[21] CMRI-derived T2 relaxation time provides quantification of cardiac iron deposition and risk prediction for HF:

T2* >20 ms: low HF risk
T2* 10–20 ms: intermediate HF risk
T2* <10 ms: high HF risk/may need increased chelation Rx

[22] Especially effective for hereditary forms; prevents but does not reverse iron deposition complications
[23] Especially secondary iron overload; make IOC almost 100 % preventable
[24] Iron deposition in myocardium is heterogeneous and iron itself is pro-arrhythmic
[25] AF is most common dysrhythmia in IOC and usually associated with myocardial damage
[26] Due to iron deposition in conduction pathways
[27] Imaging abnormalities associated with high output states may also occur, such as increased LVEF
[28] ACCF/AHA statement (2013 ACCF/AHA Guideline for the Management of Heart Failure: J Am Coll Cardiol 2013;62:e147-e239): "Iron overload cardiomyopathy manifests itself as systolic or diastolic dysfunction secondary to increased deposition of iron in the heart and occurs with common genetic disorders such as primary hemochromatosis or with lifetime transfusion requirements as seen in beta-thalassemia major. Hereditary hemochromatosis, an autosomal recessive disorder, is the most common hereditary disease of Northern Europeans, with a prevalence of approximately 5 per 1000. The actuarial survival rates of persons who are homozygous for the mutation of the hemochromatosis gene C282Y have been reported to be 95, 93, and 66 %, at 5, 10, and 20 years, respectively. Similarly, in patients with thalassemia major, cardiac failure is one of the most frequent causes of death. Chelation therapy, including newer forms of oral chelators, such as deferoxamine, and phlebotomy,

have dramatically improved the outcome of hemochromatosis, and the roles of gene therapy, hepcidin, and calcium channel blockers are being actively investigated."

Guidelines

2013 ACCF/AHA guideline for the management of heart failure
J Am Coll Cardiol. 2013;62:e147–239. http://content.onlinejacc.org/article.aspx?articleid=1695825&_ga=1.181011681.794932871.1444049701.

Patient Information

Medline Plus

ENGLISH

http://www.nlm.nih.gov/medlineplus/hemochromatosis.html.

ESPANOL

http://www.nlm.nih.gov/medlineplus/spanish/hemochromatosis.html.

Genetics Home Reference

http://ghr.nlm.nih.gov/condition/hemochromatosis.

CDC

http://www.cdc.gov/ncbddd/hemochromatosis/links.html.

Professional Information

Early Description

Clinique medicale de l'Hotel-Dieu de Paris. 1865;2:663–98. https://archive.org/details/cliniquemdicaled01trou.

Review

Circulation. 2011;124:2253–63. http://circ.ahajournals.org/content/124/20/2253.full.pdf.

Review

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Review

J Cardiac Fail. 2010;16:888–900. http://www.onlinejcf.com/article/S1071-9164(10)00215-0/abstract.

Review: Hereditary Hemochromatosis

Gastroenterol. 2010;139:393–408. http://www.ncbi.nlm.nih.gov/pubmed/20542038.

Arrhythmias

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Hereditry Hemochromatosis: Screening

Ann Intern Med. 2005;143:517–21. http://annals.org/article.aspx?articleid=718758.

Hyperadrenergic State/Iron Overload

Eur Heart J. 2016;37:988–95. http://eurheartj.oxfordjournals.org/content/37/12/988.abstract?etoc.

Presenting as Restrictive Cardiomyopathy

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RV Cardiomyopathy and B-Thalassemia Major

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Thalassemia

Eur Heart J. 2002;23:102–5. http://eurheartj.oxfordjournals.org/content/23/2/102.full.pdf+html.

Updates and More

https://clinicalguidecvd.com/iocm

Chapter 34
Cardiomyopathy: Noncompaction (Spongy Myocardium)

Management Keys

Highly individualize diagnostic evaluation and subsequent treatment as management of this disorder can be complex and is controversial

ICD-10 Code

I42.8

Alternate Names/Abbreviation

LVNC
ISOLATED LEFT VENTRICULAR NONCOMPACTION
LV HYPERTRABECULATION
SPONGY MYOCARDIUM

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_34,

Description/Etiology

Caused by intrauterine arrest of compaction (normal compaction occurs at 5–8 weeks gestation) of loose meshwork of fetal myocardial primordium with subsequent persistence of myocardial deep trabecular recesses
Most often clinically manifest as HF, dysrhythmias, systemic/pulmonary emboli [14] [15]
May be first detected on echo/screening of affected family members
Genetically heterogeneous with two forms:

Familial
Isolated with autosomal dominant, autosomal recessive, x-linked inheritance

Often associated with other congenital cardiac abnormalities
May first appear as Restrictive Cardiomyopathy in children

Comorbid Conditions

ADRENAL HYPERPLASIA
ATRIAL FIBRILLATION
BARTH SYNDROME
CARDIOMYOPATHY – HYPERTROPHIC
COMPLEX CYANOTIC CONGENITAL HEART DISEASE
CORONARY ARTERY ANOMALIES
DIGEORGE SYNDROME
EMERY-DREIFUSS MUSCULAR DYSTROPHY
MYOTUBULAR MYOPATHY
SINUS NODE DYSFUNCTION [12]
WOLFF-PARKINSON-WHITE SYNDROME

Demography

M > F
All ages [10]

Pathophysiology

Always involves LV; <50 % also RV
Diastolic and systolic dysfunction sometimes progressing to HF
Congenital form: trabeculations communicate with coronary arteries/ventricular cavity
Isolated form: trabeculations communicate only with ventricular cavity
Abnormal subendocardial perfusion may occur due to isometric contraction of endocardium and myocardium within deep intertrabecular recesses

Signs/Symptoms [1]

BREATHING – DIFF (DYSPNEA)
CHEST – PAIN
CHEST – PALPITATIONS
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)
EARS, POSITION – LOW SET [16]
EYES, MOTION – WANDERING (STRABISMUS) [16]
FACE, CHIN – SMALL (MICROGNATHIA) [16]
FACE, FOREHEAD – BROAD/PROMINENT [16]
HEART – MURMUR, NS
HEART, RATE – SLOW (BRADYCARDIA) [12]
MOUTH, PALATE – HIGH/ARCHED

Differentiation

Arrhythmogenic Right Ventricular Dysplasia

Endomyocardial Fibrosis
Cardiac Fibroma
Cardiac Metastases
Cardiomyopathy – Dilated
Cardiomyopathy – Hypertensive
Cardiomyopathy – Hypertrophic (With Apical Hypertrophy)
Eosinophilic Myocarditis
False LV tendons/aberrant chordae tendinae (Echo)
Intracardiac Tumor
LV Apical Thrombus
Other causes Of HF

Complications [1]

Cardiogenic shock
Heart Block
Heart Failure
Pulmonary Embolism [19]
Stroke
Sudden Death
Supraventricular Tachycardia [2]
Systemic emboli [19]
Ventricular Tachycardia – Nonsustained
Ventricular Tachycardia – Sustained

Laboratory

NS

ECG [9] [17]

AV COND – 3RD DEGREE BLOCK
DELTA WAVE [2]
DYSRHY – PREEXCITATION [2]

DYSRHYTHMIAS – ATRIAL (PACS/OTHERS) [ESP A FIB]
DYSRHYTHMIAS – VENTRICULAR (PVCS/ OTHERS)
PR INTERVAL – SHORT [2]
QRS – LBBB/LBBB PATTERN
QRS – LONG, NS
QRS – LVH PATTERN
QRS – RBBB/RBBB PATTERN
QRS – RVH PATTERN
QRS, AMP – INCR
RATE – DECREASED (SINUS BRADYCARDIA) [12]

Imaging [5] [9] [18]

CARDIOMEGALY
LA, CHAMBER, SIZE – INCR
LV, DIAS – DYSF
LV, INTRACAVITY – MASS [THROMBUS]
LV, SYS – DYSF [8]
LV, WALL THICKNESS, SEG – INCR [6]
MV, FLOW – DECR [4]
MYOCARD, MORPH – NONCOMPACTION [3]

Genomics

ACTC [22] [23] [25]
CSX [20] [24] [25] [26] [27]
DTNA [20]
LMNA [22] [25]
MYH7 [22] [23] [25]
NKX2.5 [20] [24] [25] [26] [27]
SCN5A [24] [25] [28] [29]
TNNT2 [22] [23] [25]
Z-BAND [22] [24]
FKBP12 [21] [22] [24]

HCN4 [25]
TAZ-G4.5 [22] [24]
TBX5 [20] [24] [25] [26] [27]

Other Tests [9]

Ambulatory ECG monitoring [for ventricular dysrhythmias]
Echo screening
EP
Neurological screening
Skeletal muscle biopsy [7]

Treatment: Nonpharmacologic [9]

Consult current HF guidelines

Treatment: Pharmacologic [9]

ASA/Anticoagulants may be especially important for this condition
Consult current HF guidelines

Treatment: Surgical/Invasive [9]

ICD [11]
CRT [11]
Heart transplant
Consult current HF guidelines

Prevention

Echo screening of 1st degree relatives

Course

Similar to Dilated Cardiomyopathy

Highly variable but as many as 60 % of symptomatic patients may die or require cardiac transplant within 6 years; incidental discovery by echo/family members detected by screening often have indolent course

Poor prognostic signs: [11]

AF
BBB
Dilated LV at time of initial diagnosis
NYHA class III-IV

Notes

[1] Complications, especially HF, sometimes first clinical manifestation
[2] WPW reported in children
[3] 2 layered myocardial appearance: outer thin compact layer and inner noncompact thick trabeculated endocardial layer; usually near cardiac apex below papillary muscles; usually LV, but RV also sometimes involved
[4] Pattern of Restricted Cardiomyopathy
[5] In advanced stage, findings are those of Dilated Cardiomyopathy/HF
[6] Noncompacted local areas of wall thickness may remain unchanged from diastole to systole
[7] Children: mitochondrial proliferation and myopathic changes may be present
[8] In children, undulating pattern of improvement followed by deteriorating LV systolic function may occur
[9] Diagnosis and treatment are complex and controversial and must be highly individualized
[10] Newborn to age 90+ years
[11] Treatment with ICD/CRT may be especially important for patients at high risk

[12] HCN4: isolated reports in families along with sinus node dysfunction; associated with sinus bradycardia
[13] Compaction normally progresses from epicardium to endocardium and from cardiac base to apex
[14] Ventricular dysfunction/dysrhythmias may be due to endocardial/myocardial hypoperfusion
[15] Adults: ventricular dysrhythmias occur in >40 % (uncommon/rare in children); AF in 25 %
[16] Head/facial dysmorphisms occur in about 1/3 of affected children; unreported/rare in adult-onset form
[17] ECG abnormal in about 90 %
[18] Echo procedure of choice but MRI preferable when apex not visualized by echo as apex most common area of noncompaction

Proposed (some believe too sensitive/lack adequate specificity) echo criteria:

Absence of coexisting cardiac abnormalities
Segmental LV wall thickening, with:

Thin compacted epicardial layer
Thick endocardial layer with prominent trabeculations/deep recesses
Noncompaction to compaction ratio >1 at end-systole

Predominant localization of pathology in LV regions:

Lateral
Apical
Midinferior

Color Doppler provides evidence of deep, perfused intertrabecular recesses

Proposed MRI criteria:

Noncompacted LV myocardial mass >25 %
Total noncompacted LV myocardial mass index >15 g/m2

Noncompacted/compacted myocardial ratio equal to or >3:1 in at least 1 segment, excluding apical segment
Trabeculation in segments 4–6 equal to or >2:1 (noncompacted to compacted)

[19] Increased risk with AF/LV systolic dysfunction
[20] Shared mutation with ASD and VSD
[21] Shared mutation with ARVD
[22] Shared mutation with DCM
[23] Shared mutation with HCM
[24] Shared mutation with other cardiomyopathies, including:

X-Linked infantile cardiomyopathy
X-Linked Endocardial Fibroelastosis
Hypoplastic Left Heart Syndrome

[25] Shared mutation with conduction abnormalities, including:

Atrioventricular nodal blocks
Bradyarrhythmias
Bundle branch blocks
Tachyarrhythmias

[26] Shared mutation with Tetralogy of Fallot
[27] Shared mutation with Ebstein's Anomaly
[28] Shared mutation with Brugada Syndrome
[29] Shared mutation with Congenital Long QT Syndrome

Guidelines

2013 ACCF/AHA guideline for the management of heart failure
J Am Coll Cardiol. 2013;62:e147–239. http://content.onlinejacc.org/article.aspx?articleid=1695825.

ESC guidelines for the diagnosis and treatment of acute and chronic heart failure 2012
Eur Heart J. 2012;33:1787–847. http://eurheartj.oxfordjournals.org/content/ehj/33/14/1787.full.pdf.

Patient Information

Medline Plus

ENGLISH

http://www.nlm.nih.gov/medlineplus/cardiomyopathy.html.

ESPANOL

http://www.nlm.nih.gov/medlineplus/spanish/cardiomyopathy.html.

Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/cardiomyopathy/basics/definition/con-20026819.

Professional Information

Review

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Review

ESC Council for Cardiol Pract E-J. 2012;(10). http://www.escardio.org/communities/councils/ccp/e-journal/volume10/Pages/Echocardiographiccriteria.aspx#.UycT82dOW8E.

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Children: Clinical Characteristics

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Echo

J Am Soc Echocardiogr. 2012;25:363–75. http://www.sciencedirect.com/science/article/pii/S0894731711009758.

Echo

Heart. 2001;86:666–71. http://heart.bmj.com/content/86/6/666.full.

Images: Thromboembolic Events, Ventric Arrhythmias, HF

J Am Coll Cardiol. 2014;63:e39–9. http://content.onlinejacc.org/article.aspx?articleID=1841607.

Mortality/Sudden Death

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MRI

Eur Radiol. 2012;22:2699–709. http://link.springer.com/article/10.1007/s00330-012-2554-7.

MRI

Eur J Radiol. 2010;74:147–51. http://www.sciencedirect.com/science/article/pii/S0720048X09000084.

Mutations: HCN4

J Am Coll Cardiol. 2014;64:745–56. http://content.onlinejacc.org/article.aspx?articleID=1898532.

Mutations: SCN4/Familial Sinus Node DYSF

J Am Coll Cardiol. 2014;64:757–67. http://content.onlinejacc.org/article.aspx?articleID=1898539.

Sinus Bradycardia

J Am Coll Cardiol. 2014;64:745–56. http://content.onlinejacc.org/article.aspx?articleID=1898532.

Updates and More

https://clinicalguidecvd.com/ncm

Chapter 35
Cardiomyopathy: Peripartum (Pregnancy-Associated Cardiomyopathy)

Management Keys

- Recognize/manage as a reversible form of HF
- Prompt diagnosis
- Management by expert multidisciplinary care team including cardiology, perinatology, hematology, obstetrics
- Aggressive treatment of mother during pregnancy as maternal health critical to fetal well-being
- Vaginal delivery with effective pain control
- Use diuretics with caution in patients with coexisting preeclampsia
- Do not prescribe ACEIs or ARBs until after delivery
- Breastfeeding should not be discouraged; may be beneficial for recovery
- Expert counseling regarding future pregnancies

ICD-10 Code

O90.3

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_35,

Alternate Names/Abbreviation

PPCM
CARDIOMYOPATHY – POSTPARTUM
PREGNANCY-ASSOCIATED CARDIOMYOPATHY

Description/Etiology [18]

Left ventricular systolic dysfunction in 3rd trimester and up to 5–6 months post-delivery in absence of another cause [12]
45 % occur in first week post-delivery, 75 % in 1st month
Phenotypic variations in different geographical areas
Cause unknown [2]

Risk factors:

Advanced maternal age
African descent
Hypertension
Multiple gestations (but 1/3 primigravida)
Multifetal pregnancy
Prior toxin exposure (e.g., cocaine)

Comorbid Conditions

ANEMIA
AUTOIMMUNE/CONNECTIVE TISSUE DISEASE
DIABETES MELLITUS
ECLAMPSIA
HYPERTENSION – SYSTEMIC ARTERIAL [GESTATIONAL]
HYPERTENSION – SYSTEMIC ARTERIAL
OBESITY
PREECLAMPSIA
SUBSTANCE ABUSE
TOBACCO USE

Demography

In USA: more common in African-Americans, least frequent in Hispanics, intermediate in Caucasians/Asians; incidence is increasing
Age >30 years [>60 %]
High incidence in Haiti

Pathophysiology

Mechanisn(s) uncertain, but an underlying inflammatory process is suggested by high concentrations of tumor necrosis factor-α (TNFα), interferon-γ, interleukin-6, C-reactive protein (CRP), and Fas/apoptosis antigen 1 (Apo-1)
Patients with peripartum cardiomyopathy also have increased levels of oxidative stress

Signs/Symptoms [3]

BREATH SOUNDS – CRACKLES (RALES) [14]
ABDOMEN – PAIN [1]
ARTERIAL PRESSURE, UPRIGHT – DECR (ORTHOSTATIC HYPOTENSION)
ARTERIAL PULSE PRESSURE – DECR
BREATHING – DIFF (DYSPNEA)
BREATHING – DIFF, NOCTURNAL (DYSPNEA, NOCT)
BREATHING – DIFF, RECLINING FLAT (ORTHOPNEA)
CHEST – PAIN
CHEST – PALPITATIONS
COUGH
EXTREM, LOWER, BILAT – EDEMA
HEART, LSB, LOWER – MURMUR, SYS
HEART, LV, APEX – MURMUR, SYS
HEART, LV, APEX, IMP – DISPLACED, LAT

HEART, RATE – RAPID (TACHYCARDIA) [14]
HEART, S3 LV [14]
HEART, S4 LV
HEART, SOUNDS, INTENSITY – DECR
LIVER – ENLARGED (HEPATOMEGALY)
NECK, JVP – ELEV [14]

Differentiation

Acute Myocardial Infarction
Normal Pregnancy [3]
Preeclampsia
Other forms of HD, e.g. cardiac valve disease
Other forms of cardiomyopathy
Pulmonary Arterial Hypertension

Complications

Cardiac Dysrhythmias
HF
Lower Extremity DVT
Peripheral Arterial Embolism
Pulmonary Embolism
Stillbirth
SCD

Laboratory [16]

BLOOD, NT-PROBNP – INCR [15]
BLOOD, TROPONIN – INCR [10]

ECG

DYSRHYTHMIAS – ATRIAL (PACS/OTHERS) [INCL A FIB]

DYSRHYTHMIAS – VENTRICULAR (PVCS/OTHERS) [INCL V TACH]
QRS – LONG, NS [11]
QRS – LVH PATTERN
RATE – INCREASED (SINUS TACHYCARDIA)
ST-T WAVE – ABN, NS

Imaging

CARDIOMEGALY
LA, CHAMBER, SIZE – INCR [5]
LV, CHAMBER, SIZE – INCR
LV, EF – DECR
LV, MYOCARD – LGE
PLEURA – FLUID
PUL, VASCULARITY – INCR
RA, CHAMBER, SIZE – INCR [5]
RV, CHAMBER, SIZE – INCR [5]

Other Tests

EMB [6]
Intense fetal/maternal monitoring during delivery

Treatment: Nonpharmacologic

Decrease sodium intake
Breastfeeding should not be discouraged, and is associated with increased rate of recovery of LV function

Treatment: Pharmacologic

Same as treatment for other forms of nonischemic cardiomyopathy except avoidance of drugs unsafe during pregnancy and lactation

ACEIs and ARBs contraindicated during pregnancy but safe/effective post-delivery
Most HF medications are safe during breastfeeding
Digoxin
Beta-blockers
Diuretics
Nitrates
Hydralazine
Anticoagulation [7]

Treatment: Surgical/Invasive

Cardiac transplantation [8]
VAD [8]
ICD [9]

Prevention

Affected patients may consider avoidance of future pregnancy
Close monitoring of pregnant patients with prior PPCM

Course

5 year survival >90 %
Full restoration to normal LV function usually within 2–6 months in about 50 % of PPCM cases
Predictors of LV recovery:

Blood troponin not elevated or at lower level
Breastfeeding
Diagnosis made post-delivery
LV diastolic dimension <5.5–6.0 cm
LVEF >30–35 %
LV thrombus absent
Non-African American

Notes

[1] Due to liver congestion
[2] Suggested: myocarditis, viral, hormonal, abnormal immune response, malnutrition, abnormal response to pregnancy hemodynamics, inflammation, apoptosis, decreased cardiac microcirculation
[3] Several signs and symptoms are often present in normal pregnancy, such as peripheral edema and dyspnea, which may delay diagnosis of PPCM
[4] Sudden onset of signs and symptoms should raise possibility of PPCM rather than normal pregnancy
[5] Severe forms
[6] Not recommended routinely; unproven clinical value except when used to exclude other specific forms of cardiomyopathy; may show myocarditis, but this finding is NS
[7] In event of thromboembolism; warfarin may carry risk for fetal complications, especially fetal cerebral hemorrhage in 2nd–3rd trimester; unfractionated heparin preferable but current guidelines should be consulted; should be continued until LVEF >35 % and especially important during first 6–8 weeks after delivery
[8] Severe, medically-refractory cases
[9] For life-threatening ventricular dysrhythmias
[10] Low sensitivity; correlates with duration of LV dysfunction
[11] Poor prognostic sign
[12] Some cases present in 2nd trimester; PPCM occurs in patients with other forms of heart disease so this diagnosis must be considered in those patients when HF develops
[13] High recurrence rate, especially if systolic function not fully recovered to normal
[14] Abnormal findings in pregnancy and suggest possible HF when present
[15] Not increased in normal pregnancy but may be markedly increased in PPCM

[16] In addition to routine tests, such as for anemia, electrolyte abnormalities, kidney dysfunction, liver dysfunction, thyroid dysfunction, inflammation, sepsis, viral serology

[17] GNB3 TT genotype associated with lower LVEF at 6–12 months in women with PPCM, especially in blacks

[18] Statement from ACCF/AHA statement (2013 ACCF/AHA Guideline for the Management of Heart Failure: *J Am Coll Cardiol* 2013;62:e147–e239): "Peripartum cardiomyopathy is a disease of unknown cause in which LV dysfunction occurs during the last trimester of pregnancy or the early puerperium. It is reported in 1:1300–1:4000 live births. Risk factors for peripartum cardiomyopathy include advanced maternal age, multiparity, African descent, and long-term tocolysis. Although its etiology remains unknown, most theories have focused on hemodynamic and immunologic causes. The prognosis of peripartum cardiomyopathy is related to the recovery of ventricular function. Significant improvement in myocardial function is seen in 30–50 % of patients in the first 6 months after presentation. However, for those patients who do not recover to normal or near-normal function, the prognosis is similar to other forms of DCM. Cardiomegaly that persists for >4–6 months after diagnosis indicates a poor prognosis, with a 50 % mortality rate at 6 years. Subsequent pregnancy in women with a history of peripartum cardiomyopathy may be associated with a further decrease in LV function and can result in clinical deterioration, including death. However, if ventricular function has normalized in women with a history of peripartum cardiomyopathy, the risk may be less. There is an increased risk of venous thromboembolism, and anticoagulation is recommended, especially if ventricular dysfunction is persistent."

Guidelines

2013 ACCF/AHA Guideline for the Management of Heart Failure

J Am Coll Cardiol. 2013;62:e147–239. http://content.onlinejacc.org/article.aspx?articleid=1695825&_ga=1.181011681.794932871.1444049701.

ESC guidelines on the management of cardiovascular diseases during pregnancy

Eur Heart J. 2011;32:3147–97. http://eurheartj.oxfordjournals.org/content/32/24/3147.full.

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Eur Heart J. 2011;32:3174–6. http://eurheartj.oxfordjournals.org/content/32/24/3147.full.

Patient Information

AHA

http://circ.ahajournals.org/content/127/20/e622.full.

Images

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Medlineplus

ENGLISH

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Professional Information

Early Description

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Review

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Review

J Perinat Neonatal Nurs. 2006;20:324–32. http://journals.lww.com/jpnnjournal/pages/articleviewer.aspx?year=2006&issue=10000&article=00011&type=abstract.

Review: Cardiovascular Disease in Pregnancy

Cardiovasc Res. 2014;101(4):554–60. http://cardiovascres.oxfordjournals.org/content/101/4/554.abstract.

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J Am Coll Cardiol HF. 2013;409–16. http://heartfailure.onlinejacc.org/article.aspx?articleID=1737285.

Cardiovascular Physiology of Pregnancy

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African Americans

J Card Fail. 2013;19:214–8. http://www.onlinejcf.com/article/S1071-9164(13)00072-9/abstract.

African Americans: Risk

J Am Coll Cardiol. 2010;55:654–9. http://content.onlinejacc.org/article.aspx?articleID=1140457.

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JAMA. 2000;283(9):1183–88. http://jama.jamanetwork.com/article.aspx?articleid=192436.

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Outcomes: South African Study

Heart. 2013;99:308–13. http://heart.bmj.com/content/99/5/308.abstract.

Preeclampsia Relationship

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Presenting as AMI

Mayo Clin Proc. 2002;77:500–1. http://www.mayoclinicproceedings.org/article/S0025-6196(11)62221-0/abstract.

Predictors of Recovery/ICD Use

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Cardiomyopathy: Peripartum

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Updates and More

https://clinicalguidecvd.com/ppcm

Chapter 36
Cardiomyopathy: Restrictive

Management Keys

Rule out Constrictive Pericarditis
Diagnose/treat secondary forms and provide appropriate/specific treatment [14]

ICD-10 Code

I42.5

Alternate Names/Abbreviation

RCM
INFILTRATIVE CARDIOMYOPATHY [1]

Description/Etiology

Heterogeneous group of adult disorders with wide range of underlying causes and characterized by rapid early diastolic filling, with minimal/no further filling in late diastole, due to compromised ventricular expansion; ventricular contractility often maintained

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_36,

Usual course is progressive HF, increased mortality, and poor prognosis

Myocardial noninfiltrative cardiomyopathies include:

- Diabetic
- Familial
- Hypertrophic
- Idiopathic
- Pseudoxanthoma Elasticum
- Scleroderma

Myocardial infiltrative cardiomyopathies include:

- Amyloidosis
- Fatty infiltration
- Gaucher Disease
- Hurler Syndrome
- Primary Hyperoxaluria
- Sarcoidosis

Storage diseases include:

- Fabry Disease
- Glycogen Storage Disease
- Hemochromatosis

Endomyocardial diseases include:

- Anthracycline toxicity
- Carcinoid heart disease
- Drug – Induced Fibrous Endocarditis [13]
- Endomyocardial Fibrosis
- Hypereosinophilic Syndrome
- Metastatic cancers
- Radiation

Comorbid Conditions

AMYLOIDOSIS
BEHCET DISEASE
CARCINOID SYNDROME/TUMOR
CARDIOMYOPATHY – ANTHRACYCLINE

CARDIOMYOPATHY – DIABETIC
CARDIOMYOPATHY – FAMILIAL
CARDIOMYOPATHY – HYPERTROPHIC
DRUG-RELATED ENDOCARDITIS [13]
ENDOMYOCARDIAL FIBROSIS
FABRY DISEASE
FATTY INFILTRATION
GAUCHER DISEASE
HEMOCHROMATOSIS
HURLER SYNDROME
HYPEREOSINOPHILIC SYNDROME
METASTATIC CANCER
PSEUDOXANTHOMA ELASTICUM
RADIATION [12]
SARCOIDOSIS
SCLERODERMA

Demography

Variable according to cause

Pathophysiology

Pathology varies according to etiology

Abnormal ventricular diastolic filling with:

Increased LVEDP
Dilated atria
Variable systolic function [1]

Signs/Symptoms

ABDOMEN – FLUID (ASCITES)
ARTERIAL PULSE, AMP – ALTERNATING (PULSUS ALTERNANS)

ARTERIAL PULSE, AMP – DECR/ABS
BREATH SOUNDS – CRACKLES (RALES)
BREATHING – DIFF (DYSPNEA) [2]
BREATHING – DIFF, RECLINING FLAT (ORTHOPNEA)
CHEST – PAIN
CHEST, ANT, RSB – PULSATION [3]
EXTREM, LOWER, BILAT – EDEMA
EYES – PROMINENT (EXOPHTHALMOS/ PROPTOSIS)
FACE, SHAPE – ROUND [4]
FATIGUE
HEART, LSB, LOWER – MURMUR, SYS [5]
HEART, LV, APEX – MURMUR, SYS [6]
HEART, RATE – RAPID (TACHYCARDIA)
HEART, S3 LV
HEART, S3 RV
HEART, S4 LV
HEART, S4 RV
HEART, SOUNDS, INTENSITY – DECR
HYPOTENSION (BLOOD PRESSURE – DECREASED/LOW)
LIVER – ENLARGED (HEPATOMEGALY)
LIVER – PULSATION, SYS
MUSCLES – ATROPHY
MUSCLES – WEAK
NECK, JVP – ELEV
NECK, JVP – INSP RISE (KUSSMAUL SIGN)
NECK, JVP, A WAVE – INCR/LARGE (CANNON WAVE)
NECK, JVP, V WAVE – INCR/LARGE
NECK, JVP, X DESCENT – PROMINENT/RAPID
NECK, JVP, Y DESCENT – RAPID
THROAT – PAIN/TIGHTNESS

Differentiation

Constrictive Pericarditis

Complications

Heart block [15]
HF
Systemic emboli

Laboratory [7]

BLOOD, NT-PROBNP – INCR

ECG [10]

DYSRHYTHMIAS – ATRIAL (PACS/OTHERS) [8]
DYSRHYTHMIAS – VENTRICULAR (PVCS/OTHERS) [9]
P WAVE – BIFID
QRS – LONG, NS
ST-T WAVE – ABN, NS

Imaging [1]

LA, CHAMBER, SIZE – INCR
LUNGS, INTERSTITIUM – EDEMA/INFILTRATES
MV, FLOW – REGURG
PLEURA – FLUID
PUL, VASCULARITY – INCR
RA, CHAMBER, SIZE – INCR
TV, FLOW – REGURG

Genomics

MYH7 [16]
TTN [16]

Other Tests

Card catheterization:

- Increased LVEDP
- Normal/decreased LVEF
- Increased PCWP
- Increased RA pressure
- Increased RVEDP

EMB

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic

NS

Treatment: Surgical/Invasive

Cardiac transplantation
Pacemaker

Course

Variable according to etiology

Notes

[1] LV/RV chamber size may be normal; wall thickness may be normal or increased

[2] May be paroxysmal
[3] Due to enlarged RA
[4] "Moon" face
[5] TR; increased intensity with inspiration
[6] MR
[7] Does not include abnormal clinical features specific to various forms, e.g., myopathy in Familial RCM, anemia in Loeffler Fibrosis
[8] Most often AF (<70 %)
[9] Mainly PVCS
[10] Findings differ when disease confined to either left or right heart, which more often occurs in secondary forms
[11] Additional clinical features may be present in secondary forms
[12] Pericardial damage and secondary Constrictive Pericarditis more common from radiation
[13] Include serotonin, methysergide, ergotamine, mercurials, busulfan
[14] 50 % idiopathic in USA
[15] Heart block due to fibrosis of sinus/AV nodes
[16] Familial RCM

Guidelines

NS

Patient Information

Images

http://www.nlm.nih.gov/medlineplus/ency/imagepages/1056.htm.
http://www.nlm.nih.gov/medlineplus/ency/imagepages/1097.htm.

Medlineplus

ENGLISH

https://www.nlm.nih.gov/medlineplus/ency/article/000189.htm.

ESPANOL

http://www.nlm.nih.gov/medlineplus/spanish/ency/article/000189.htm.

Genetics Home Reference

http://ghr.nlm.nih.gov/condition/familial-restrictive-cardiomyopathy.

CMA

http://www.cardiomyopathy.org/Restrictive-cardiomyopathy.html.

ESPANOL

http://www.nlm.nih.gov/medlineplus/spanish/ency/article/000189.htm.

Professional Information

Early Description: Idiopathic RCM

Circulation. 1980;61:1206–12. http://circ.ahajournals.org/content/61/6/1206.abstract.

Review

N Engl J Med. 1997;336:267–76. http://www.nejm.org/doi/full/10.1056/NEJM199701233360407.

Differential Diagnosis: Invasive Hemodynamics

Cardiol Clin. 2011;29:191–9. http://www.cardiology.theclinics.com/article/S0733-8651(11)00010-5/abstract.

Familial RCM: MYH7 Mutation

Eur Heart J. 2015;36:178. http://eurheartj.oxfordjournals.org/content/36/3/178.extract?etoc.

Familial RCM: TTN Mutation

Int J Cardiol. 2014;171:24–30. http://www.sciencedirect.com/science/article/pii/S016752731302055X.

Genetics: Secondary RCM

Heart Failure Clin. 2010;6:179–86. http://www.heartfailure.theclinics.com/article/S1551-7136(09)00107-X/abstract.

LV Mechanics; RCM Versus Constrictive Pericarditis

J Am Coll Cardiol Imaging. 2008;1:29–38. http://imaging.onlinejacc.org/article.aspx?articleID=1096954.

Outcomes: Children

Circulation. 2012;126:1237–44. http://circ.ahajournals.org/content/126/10/1237.full.

Outcomes: Idiopathic RCM

Circulation. 2000;101:2490–96. http://circ.ahajournals.org/content/101/21/2490.full.

MRI

Clin Radiol. 2012;67:95–105. http://www.clinicalradiologyonline.net/article/S0009-9260(11)00356-4/abstract.

Pathology

J Am Coll Cardiol. 1991;18:1230–5. http://content.onlinejacc.org/article.aspx?articleID=1117799.

Primary Hyperoxaluria (Case Report)

J Am Coll Cardiol. 2013;62(24):e525–5.doi:10.1016/j.jacc.2013.05.104 http://content.onlinejacc.org/article.aspx?articleid=1757154.

Sarcomeric Protein Mutations

Heart. 2014;100:1916–23. http://heart.bmj.com/content/100/24/1916.long.

Updates and More

https://clinicalguidecvd.com/rcm

Chapter 37
Cardiomyopathy: Tachycardia-Induced

Management Keys

Recognize/manage as a reversible form of HF
Diagnose specific causative tachyarrhythmia
Diagnose associated structural cardiac abnormalities, if any
Reverse/control causative tachyarrhythmia
Close follow-up after tachyarrhythmia termination for recurrence

ICD-10 Code

NS

Alternate Names/Abbreviation

TIC
TACHYCARDIA MEDIATED CARDIOMYOPATHY

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 523
DOI 10.1007/978-1-4471-7293-2_37,

Description/Etiology

Systolic/diastolic ventricular dilatation/dysfunction caused by prolonged persistent/very frequent intermittent tachyarrhythmia, including:

- Atrial tachyarrhythmias (especially AF) [1]
- Ventricular tachyarrhythmias (including frequent PVCs)
- Rapid external pacing

Reversible by control of arrhythmia/heart rate by:

- Anti-tachycardia pacing
- Cardioversion
- Catheter ablation
- Negative inotropic agents

Diagnosis often made when ventricular function normalizes after tachyarrhythmia rate/rhythm correction

Occurs in patients:

- Without structural heart disease
- With underlying heart disease in whom tachycardia aggravates ventricular function

Comorbid Conditions

ALL CARD DIS CAUSING TACHYARRHYTHMIAS
ATRIAL FIBRILLATION [1]
DYSRHYTHMIAS – ATRIAL
DYSRHYTHMIAS – VENTRICULAR
HYPERTHYROIDISM [INCL GRAVES DIS]

Demography

All ages, including fetuses

Pathophysiology

Animal studies:

- Hemodynamics: increased ventricular filling pressure, severe biventricular systolic dysfunction, decreased cardiac output, increased systemic vascular resistance
- Neurohormonal: marked increased plasma atrial natriuretic peptides, epinephrine/norepinephrine, plasma renin, aldosterone
- Blunted response to beta-adrenergic stimulation

Cardiac morphology:

- 4 chamber dilatation
- Normal/decreased ventricular wall thickness
- Normal/unchanged cardiac mass

Cellular: myocyte loss, cell elongation, myofibril misalignment, loss of sarcomere register, calcium handling, oxidative stress

Myocardial Ischemia

Mitral Regurgitation

Propensity may vary with type of tachycardia (up to 50 % of patients have permanent junctional reciprocating tachycardia)

Signs/Symptoms

ABDOMEN – DISTENSION
ABDOMEN – FLUID (ASCITES)
ABDOMEN – FULLNESS
ABDOMEN – PAIN
APPETITE – DECR (ANOREXIA)
ARTERIAL PRESSURE, DIAS – DECR
ARTERIAL PULSE, AMP – ALTERNATING (PULSUS ALTERNANS)
BEHAVIOR – BIZARRE/CHANGED
BREATH SOUNDS – CRACKLES (RALES)
BREATH SOUNDS – DECR

BREATH SOUNDS – WHEEZES
BREATHING – DIFF (DYSPNEA)
BREATHING – DIFF, RECLINING FLAT (ORTHOPNEA)
BREATHING – RHYTHMIC CHANGES (CHEYNE-STOKES)
CHEST – PAIN
CHEST – PALPITATIONS
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)
COUGH
COUGH – NOCT
EXTREM, LOWER, BILAT – EDEMA
EYES–PROMINENT (EXOPHTHALMOS/PROPTOSIS)
FATIGUE
HEADACHE
HEART, LV, APEX, IMP – DISPLACED, INF
HEART, LV, APEX, IMP – DISPLACED, LAT
HEART, P2, INTENSITY – INCR
HEART, RATE – RAPID (TACHYCARDIA) [3]
HEART, RHYTHM – IRREG [2]
HEART, S3 LV
HEART, S3 RV
HEART, S4 LV
HEART, S4 RV
HEART, SOUNDS, INTENSITY – DECR
HYPOTENSION (BLOOD PRESSURE – DECREASED/LOW)
LIVER – ENLARGED (HEPATOMEGALY)
LIVER – TENDER
MENTATION – CONFUSION
MUSCLES – WEAK
NAUSEA
NECK, JVP – ABDOMINOJUGULAR REFLUX
NECK, JVP – ELEV
SKIN, COLOR – BLUE (CYANOSIS)
SLEEP – DISTURBED (INSOMNIA)
SPLEEN, SIZE – INCR (SPLENOMEGALY)
SPUTUM – BLOOD (HEMOPTYSIS)
URINATION – NIGHTTIME (NOCTURIA)
VOMITING (EMESIS)

Differentiation

Other causes of DCM
Other causes of HF

Complications

Late diffuse LV fibrosis [6]
Recurrence after initial treatment [5]
SCD
Thromboembolism

Laboratory

BLOOD, NT-PROBNP – INCR
NS [4]

ECG

[FEATURES OF DYSRHY MAY ALSO BE PRESENT]
HEART, RATE – INCR [3]

Imaging

[FEATURES OF OTHER FORMS OF CARD DIS MAY BE PRESENT]
CARDIOMEGALY
LA, CHAMBER, SIZE – INCR
LV, CHAMBER, SIZE – INCR
LV, EF – DECR
LV, SYS – DYSF
LV, WALL MOTION – DECR
PLEURA – FLUID
PUL, VASCULARITY – INCR
RA, CHAMBER, SIZE – INCR
RV, CHAMBER, SIZE – INCR

Other Tests

Ambulatory ECG monitoring
EP

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic

Rhythm/rate control per causative dysrhythmia

Treatment: Surgical/Invasive

Rhythm/rate control per causative dysrhythmia

Prevention

Prevent causative dysrhythmia

Course

Ventricular function fully restored about 4 weeks post-rate/rhythm control [6]

Notes

[1] In patients with AF, 25–50 % of episodes of associated ventricular dysfunction at least in part attributed to TIC
[2] Especially AF, frequent PVCS
[3] Prolonged at >100 bpm

[4] Include increased BUN, creatinine, liver enzymes, troponin I/T, uric acid, glucose; decreased total cholesterol, HGB/HCT, sodium
[5] May occur suddenly before LV function has returned to normal after treatment
[6] Late long-term fibrosis may occur after apparent cure; may be associated with SCD

Guidelines

2013 ACCF/AHA guideline for the management of heart failure
J Am Coll Cardiol. 2013;62:e147–239. http://content.onlinejacc.org/article.aspx?articleid=1695825.

Patient Information

NS

Professional Information

Early Description

QJM. 1913;6:435–40. http://qjmed.oxfordjournals.org/content/os6/4/435.

Review

Am J Med. 2003;114:51–5. http://www.sciencedirect.com/science/article/pii/S0002934302014729.

Review

Int J Cardiol. 2014;40–6. http://www.sciencedirect.com/science/article/pii/S0167527314000400.

Hyperthyroidism

Curr Heart Fail Rep. 2008;5:170–6. http://link.springer.com/article/10.1007/s11897-008-0026-9.

Hyperthyroidism

Heart. 2007;93:483–7. http://heart.bmj.com/content/93/4/483.abstract.

Mitral Regurgitation

Circulation. 2001;104:47–53. http://circ.ahajournals.org/content/104/suppl_1/I-47.full.

NT-PROBNP

Clin Res Cardiol. 2011;100:887–96. http://link.springer.com/article/10.1007/s00392-011-0319-y.

PVCS/Recovery

Pacing Clin Electrophysiol. 2013;36:612–7. http://www.readcube.com/articles/10.1111%2Fpace.12087.

Treatment

J Interv Card Electrophysiol. 2013;36(1):27–32. http://link.springer.com/article/10.1007%2Fs10840-012-9727-9.

Treatment

Heart Fail Rev. 2011;16(5):467–76. http://link.springer.com/article/10.1007%2Fs10741-011-9240-8.

Updates and More

https://clinicalguidecvd.com/tcm

Chapter 38
Cardiomyopathy: Takotsubo (Stress Cardiomyopathy)

Management Keys

Recognize/manage as a reversible form of HF
Close follow up for HF recurrence/persistence [18]

ICD-10 Code

I51.81

Alternate Names/Abbreviation

STRESS CARDIOMYOPATHY
TAKOTSUBO SYNDROME
TRANSIENT LEFT VENTRICULAR BALLOONING SYNDROME

Description/Etiology

Acute cardiac event manifest by:

Resting chest pain
Dyspnea
Increased cardiac enzymes (e.g., troponin)

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_38,

ECG: ST elevation/other ischemic changes
Reversible abnormal contraction of LV apex and mid-portion with sparing of basal segments

Etiology/mechanism unknown
Triggers include:

Emotional stress, including:

Death of close friend/relative
Quarrel
Financial loss (e.g., business, gambling)
Frightening event (e.g., volcano eruption)

Iatrogenic – drugs, including:

Adrenergics (e.g., oxymetazoline)
Alpha-adrenergic vasoconstrictors (e.g., ergonovine)
Anticholinergics/parasympatholytics (e.g., atropine)
Chemotherapy
Norepinephrine reuptake inhibitors (e.g., nortriptyline)
Serotonin reuptake inhibitors
Sympathomimetic Inotropes (e.g., Dobutamine)

Iatrogenic – invasive/procedural, including:

Cardiac surgery
Noncardiac surgery

Neurological, including:

Cerebrovascular accidents [1]
Seizures [15]

Physical stress, including:

Extreme exercise
Falls
Near-drowning

Physiological stress, including:
Alcohol withdrawal
Envenomation

Insect bite
Jellyfish bite
Snake bite

GI bleed
Opioid withdrawal
Pheochromocytoma

Proposed Mayo criteria:

1. Transient LV apical/mid-segment akinesis/dyskinesis with regional wall motion abn extending beyond a single epicard vasc distribution
2. Absence of obstructive coronary disease/angiographic evidence of acute plaque rupture
3. New ECG abnormalities (either ST elevation/T Wave inversion)
4. Absence of:

   Hypertrophic Cardiomyopathy
   Intracranial bleeding
   Myocarditis
   Obstructive epicardial CAD
   Pheochromocytoma
   Recent significant head trauma

Comorbid Conditions

ANXIETY/ANXIETY DISORDER [16]
CANCER
CHRONIC OBSTRUCTIVE PULMONARY DISEASE (EMPHYSEMA)
CORONARY ARTERY DISEASE
DEPRESSION [16]
DRUG ABUSE
DYSLIPIDEMIA
FAMILY HX: CORONARY ARTERY DISEASE
HYPERTENSION – SYSTEMIC ARTERIAL
MOOD DISORDERS [16]
PERICARDITIS – ACUTE

PHEOCHROMOCYTOMA
SEPSIS
STROKE
TOBACCO USE

Demography

>80 % female
Age usually >60 years

Pathophysiology [18]

Hypothesis: sympathetic hyperactivity causes myocardial stunning/hypocontractility and associated coronary artery vasospasm; decreased estrogen may be factor
Systolic dysfunction of LV apical and mid-LV myocardium with hyperkinesis of basal LV myocardium
LVOT obstruction due to LV basal hypercontractility (25 % of pts)
Cases isolated to RV reported

Signs/Symptoms [2]

ABDOMEN – PAIN
BREATHING – DIFF (DYSPNEA)
CHEST – PAIN
CHEST – PALPITATIONS
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)
DIZZY/LIGHTHEADED/PRESYNCOPE
HEART, LV, APEX – MURMUR, SYS [3]
HYPOTENSION (BLOOD PRESSURE – DECREASED/LOW)
MOOD – ANXIOUS
NAUSEA
VOMITING (EMESIS)

Differentiation

Acute Myocardial Infarction
Cardiomyopathy – Hypertrophic [10]
Cardiomyopathy – Sepsis-Induced
Head Trauma
Heart Failure – Other Causes
Myocarditis
Pheochromocytoma
Syndrome X
Variant Angina

Complications

Atrioventricular Heart Block
Cardiogenic Shock [17]
Death
Dynamic intraventricular obstruction
Extension to RV [6]
Heart Failure
Left Ventricular free wall rupture
LV thrombus
Mitral Regurgitation – Acute
Pericarditis – Acute
Ventricular arrhythmias [17]

Laboratory

BLOOD, TROPONIN – INCR [7]

ECG

AV COND – 1ST DEGREE BLOCK
AV COND – 3RD DEGREE BLOCK [4]
DYSRHYTHMIAS – ATRIAL (PACS/OTHERS)
DYSRHYTHMIAS – VENTRICULAR (PVCS/OTHERS)

Q WAVE – ABN
QRS – R PROGRESSION, POOR
QT/QTC INTERVAL – LONG
RATE – DECREASED (SINUS BRADYCARDIA)
ST SEGMENT – ELEV
T WAVE – INVER, ABN [5]

Imaging

[COR ANGIO: NONOBS LESIONS OR N]
LV, EF – DECR [14]
LV, WALL MOTION, SEG – DECR/AKINETIC [BALLOONING] [8]
LV, WALL MOTION, SEG – INCR/HYPERDYNAMIC [9]

Other Tests

NS

Treatment: Nonpharmacologic [11]

NS

Treatment: Pharmacologic [11]

Anticoagulation – short-term [13]
Beta-blockers [12]
Vasopressors (for Hypotension)

Treatment: Surgical/Invasive

NA

Course [18]

>90 % fully recover
10 % recurrence
Small number develop persistent HF

Notes

[1] Associated with 10x increased incidence
[2] Cardiac arrest due to VF may be initial clinical manifestation
[3] Transient
[4] Reported but uncommon
[5] Symmetric, most leads
[6] Possible poorer prognosis
[7] Cardiac biomarkers generally mildly increased (not all cases); peak at initial presentation
[8] Apex and mid-LV, extends beyond distribution of 1 coronary artery, reverses to normal within days on serial exams
[9] Posterior/basilar LV
[10] LV outflow gradient rarely reported
[11] Acute management with cardiac monitoring, O_2, aspirin, etc, same as AMI protocols until this syndrome diagnosed
[12] Unless epicardial coronary artery vasospasm present
[13] Consider to prevent LV thrombus formation
[14] Transient, 20–40 %; normal within weeks
[15] Seizure-related: more often males, younger age, chest pain less frequent, increased rate of cardiogenic shock/recurrence
[16] Up to 40 % have depression/anxiety
[17] VF/cardiogenic shock 1–4 % in reported cases
[18] Autonomic dysfunction (increased sympathetic/decreased parasympathetic modulation of HR) may persist for several years after acute episode

Guidelines

2013 ACCF/AHA guideline for the management of heart failure
J Am Coll Cardiol. 2013;62:e147–239. http://content.onlinejacc.org/article.aspx?articleid=1695825.
ESC Guidelines for the diagnosis and treatment of acute and chronic heart failure 2012
Eur Heart J. 2012;33:1787–847.http://eurheartj.oxfordjournals.org/content/ehj/33/14/1787.full.pdf.

Patient Information

Medline Plus

ENGLISH

http://www.nlm.nih.gov/medlineplus/cardiomyopathy.html.

ESPANOL

https://www.nlm.nih.gov/medlineplus/spanish/cardiomyopathy.html.

Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/broken-heart-syndrome/basics/definition/con-20034635.

Johns Hopkins-FAQS

http://www.hopkinsmedicine.org/asc/faqs.html.

Harvard

http://www.health.harvard.edu/heart-health/takotsubo-cardiomyopathy-broken-heart-syndrome.

Professional Information

Mayo Criteria

Ann Intern Med. 2004;141:858–65. http://annals.org/article.aspx?articleid=717989.

Early Description (Japanese)

J Cardiol. 1991;21:203–14. http://www.ncbi.nlm.nih.gov/pubmed/1841907.

Review

Am J Med. 2014;127:699–704. http://www.sciencedirect.com/science/article/pii/S000293431400309X.

Review

Heart Failure Clin. 2013;9:249–66. http://www.sciencedirect.com/science/article/pii/S1551713612001286.

Review

World J Cardiol. 2014;6:602–9. http://www.wjgnet.com/1949-8462/full/v6/i7/602.htm.

Review

J Nurse Pract. 2009;10:767–72. http://www.sciencedirect.com/science/article/pii/S1555415508006041.

Review

Am Heart J. 2008;155:408–17. http://www.ncbi.nlm.nih.gov/pubmed/18294473?dopt=Abstract.

Review

Circulation. 2008;118:2754–62. http://circ.ahajournals.org/content/118/25/2754.full#ref-9.

Review

Eur Heart J. 2006;27:1523–29. http://eurheartj.oxfordjournals.org/content/27/13/1523.full?sid=f5a37f78-d848-4de0-88dc-f749782cef20.

Autonomic Changes

Am J Cardiol. 2016;117:206–13. http://www.ajconline.org/article/S0002-9149(15)02192-X/abstract.

Biomarkers: MicroRNAs

Eur Heart J. 2014;35:999–1006. http://eurheartj.oxfordjournals.org/content/35/15/999.

Catecholamine Hyperactivity

Am J Cardiol. 2015;115:1580–6. http://www.ajconline.org/article/S0002-9149(15)00908-X/abstract.

Clinical Profile of High Risk PTS

Am J Cardiol. 2015;116:765–72. http://www.ajconline.org/article/S0002-9149(15)01438-1/abstract.

Coronary Microvascular Dysfunction: Reversible

Eur Heart J. 2010;31:1319–27. http://eurheartj.oxfordjournals.org/content/31/11/1319.

Echocardiography

J Am Soc Echocardiogr. 2015;28:57–74. http://www.ncbi.nlm.nih.gov/pubmed/25282664.

Envenomation: Jellyfish

Eur Heart J. 2011;32(1):18. doi:10.1093/eurheartj/ehq349. http://eurheartj.oxfordjournals.org/content/32/1/18.long.

Envenomation: Snake

Pan Afr Med J. 2012;13:51. http://www.ncbi.nlm.nih.gov/pubmed/23330042.

Envenomation: Spider

Medicine. 2015;94:e457. http://journals.lww.com/md-journal/Fulltext/2015/02010/Transient-Reverse-Takotsubo-Cardiomyopathy.12.aspx.

Heart Failure

Circulation. 2008;118:2754–62. http://circ.ahajournals.org/content/118/25/2754.full?sid=b34e3a21-488c-43b7-808e-408fa6bb9c47.

Heart Transplant Recipient

Circulation. 2013;128:e62–3. http://circ.ahajournals.org/content/128/5/e62.full.

Images

J Am Coll Cardiol. 2014;63:e53–3. http://content.onlinejacc.org/article.aspx?articleID=1859513.

Images

Eur Heart J. 2013;34:397. http://eurheartj.oxfordjournals.org/content/34/5/397.

LVOT Obstruction

Am Heart J. 2008;156:543–8. http://www.ahjonline.com/article/S0002-8703(08)00369-4/abstract.

LV Systolic/Diastolic Mechanics

Circulation. 2014;129:1659–67. http://circ.ahajournals.org/content/129/16/1659.full.

Outcomes Predictors

Am J Cardiol. 2015;116:1586–90. http://www.ajconline.org/article/S0002-9149(15)01851-2/abstract.

Pericarditis

Am J Emerg Med. 2012;30:382–3. http://www.sciencedirect.com/science/article/pii/S0735675711005031.

Pheochromocytoma

Eur Heart J. http://dx.doi.org/10.1093/eurheartj/ehm449830. http://eurheartj.oxfordjournals.org/content/29/6/830.

Reverse Takotsubo

Congest Heart Fail. 2010;16:284–6. http://www.ncbi.nlm.nih.gov/pubmed/21091614/.

RV Involvement (Case Report)

http://ehjcimaging.oxfordjournals.org/content/16/3/285.extract?etoc. Eur Heart J Cardiovasc Imaging. 2015;16:285.

RV Involvement

Eur Heart J. 2006;27:2433–9. http://eurheartj.oxfordjournals.org/content/27/20/2433.

Seizures

Epilepsia. 2011;52:e160–7. http://onlinelibrary.wiley.com/doi/10.1111/j.1528-1167.2011.03185.x/full.

Sepsis-Induced Cardiomyopathy

Heart Fail Rev. 2010;15:605–11. http://link.springer.com/article/10.1007%2Fs10741-010-9176-4.

Stemi Mimic

J Electrocardiol. 2007;40:e1–6. http://www.ncbi.nlm.nih.gov/pubmed/17067626?dopt=Abstract.

Stress

N Engl J Med. 2005;352:539–48. http://www.nejm.org/doi/full/10.1056/NEJMoa043046.

Treatment

Heart Fail Clin. 2013;9:177–86. http://www.ncbi.nlm.nih.gov/pubmed/23562118.

Volcano Eruptions

JAMA. 2005;294:305–7. http://jama.jamanetwork.com/article.aspx?articleid=201243.

Updates and More

https://clinicalguidecvd.com/takotsubo

Chapter 39
Carotid Artery Stenosis

Management Keys

- Carotid Endarterectomy within 2 weeks of TIA reduces risk of stroke
- Atherosclerotic risk factor modification for both asymptomatic patients and patients with TIA

ICD-10 Code

I65.2

Abbreviation

CAS

Description/Etiology

- A form of extracranial cerebrovascular disease encompassing several disorders involving arteries that supply the brain
- Important cause of stroke and transient cerebral ischemic attack

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_39,

Most frequent cause is atherosclerosis [other causes include Fibromuscular Dysplasia, Cystic Medial Necrosis, Arteritis, Dissection]
Patients with CAS have an increased risk for other CV events, including AMI, PAD, and SCD
Obstruction of cerebral blood flow due to CAS typically occurs at bifurcation of common carotid artery and internal carotid artery

Comorbid Conditions

ABDOMINAL AORTIC ANEURYSM
AORTIC DISSECTION
ATHEROSCLEROSIS IN OTHER CV AREAS [1][2]
CORONARY ARTERY DISEASE [12]
DIABETES MELLITUS
DYSLIPIDEMIA
ELEVATED LIPOPROTEIN (A)
FIBROMUSCULAR DYSPLASIA
GOUT
HYPERTENSION – SYSTEMIC ARTERIAL
PERIPHERAL ARTERY DISEASE
RADIATION
TOBACCO USE
VASCULITIS
WILLIAMS SYNDROME

Demography

Advanced age
M > F age <75 years; reverse for age >75 years
African American and Hispanic ethnicity: higher incidence

Pathophysiology

Pathobiology of carotid (and vertebral artery) atherosclerosis is similar to process in other arteries, including:

- Early lesion development initiated by intimal accumulation of lipoprotein particles
- Particles undergo oxidative modification and elaborate cytokines that cause expression of adhesion molecules and chemoattractants that facilitate uptake and migration of monocytes into artery wall
- Monocytes become lipid-laden macrophages (foam cells) due to accumulation of modified lipoproteins and subsequently release additional cytokines, oxidants, and matrix metalloproteinases
- Smooth muscle cells migrate from the media to intima, proliferate, and elaborate extracellular matrix as extracellular lipid accumulates in central core surrounded by layer of connective tissue
- Initially atherosclerotic lesion grows in an outward direction ("arterial remodeling")
- With plaque growth, encroaches on lumen and causes stenosis
- Plaque disruption and thrombus formation contribute to progressive narrowing of lumen and to clinical events (TIAs, Ischemic Stroke)

- Due to predilection for atherosclerotic plaque formation at flow dividers and branch points, where there is both turbulence and shifts in shear stress, CAS most often forms at bifurcation of common carotid artery into internal and external carotid arteries
- Stroke and TIAs caused by several mechanisms, including:
  - Artery-to-artery embolism of thrombus formed on an atherosclerotic plaque
  - Atheroembolism of cholesterol crystals or other atheromatous debris (e.g., Hollenhorst plaque)
  - Acute thrombotic occlusion of extracranial artery resulting from plaque rupture
  - Structural disintegration of arterial wall due to dissection or subintimal hematoma,
  - Decreased cerebral perfusion due to critical stenosis or occlusion caused by progressive plaque growth

Signs/Symptoms [3]

ARTERIAL PULSE, CAROTID – DECR/ABSENT
ARTERY, CAROTID – BRUIT [13]
EARS, BILAT, EARLOBE CREASE, DIAGONAL
EXTREM, UNILAT – SHAKING/FLAPPING [5]
EXTREM, UNILAT – WEAKNESS
EXTREM, UNILAT, SENSORY – DECR/ABSENT
EYES, RETINA – PLAQUES, CHOLESTEROL (HOLLENHORST PLAQUES) [9]
EYES, VISION – DECR/LOSS [8]
EYES, VISION, MONOCULAR – DECR/TOTAL LOSS, TRANSIENT (AMAUROSIS FUGAX) [8]
FACE, MUSCLES, UNILAT – WEAK
FACE, SENSATION, UNILAT – DECR
SPEECH – DISTURBED (DYSPHASIA) [4]

Differentiation

AORTIC VALVE STENOSIS
AORTIC DISSECTION
CARDIAC EMBOLI
CAROTID DISSECTION
MOYAMOYA DISEASE
SEIZURES
VERTEBROBASILAR INSUFFICIENCY

Complications

Stroke

Laboratory

NS

ECG

NS/VAR PER COMORBIDITY(S) [ESP CAD]
N/NS ABN [6]

Imaging [7]

ARTERY, CAROTID – PLAQUE
ARTERY, CAROTID, FLOW – OBS
ARTERY, CAROTID, IMT – INCR
COR ANGIO MAY BE INDICATED IN ADDITION TO CAROTID IMAGING
CT/MRI OF AORTIC ARCH FOR MORPH MAY ALSO BE INDICATED FOR DIS
CTA AND MRA ARE COMPARABLE TO CAROTID ANGIO FOR DEFINITIVE DX

Other Tests

Stress test – especially preoperatively [12]

Treatment: Nonpharmacologic

Atherosclerotic risk factor modification [11]
Tobacco cessation
Exercise

Treatment: Pharmacologic

Antiplatelet agents [11] [14]
Hypertension control [11] [15]
Statins [10] [11]

Treatment: Surgical/Invasive [1]

Endarterectomy [16]
Stent [17]

Notes

[1] Often asymptomatic; in such patients, medical versus surgical therapy controversial and current guidelines should be consulted
[2] Large majority; increased incidence of atherosclerotic CAD and PAD
[3] Most persons with CAS are asymptomatic; listed symptoms are those of TIA when lasting 1–24 h; when lasting >24 h, considered a completed stroke
[4] Expressive aphasia (Broca's Aphasia) – word finding difficult; receptive aphasia (Wernicke's Aphasia) – inability to follow commands
[5] Global ischemia of cerebral hemisphere; usually severe Carotid Stenosis and usually bilateral; may resemble seizure
[6] AF and changes due to CAD, LVH may be present
[7] Duplex ultrasound usually first choice but MRI, CT comparable in detecting significant CAS alone; gold standard is contrast angiography with DSA
[8] Loss of vision can occur secondary to retinal emboli (ipsilateral to stenosis vision loss or ophthalmic cortex (contralateral)
[9] On ophthalmoscopy seen as yellow fleck at retinal artery bifurcation; likely due to embolus from carotid plaque
[10] Regardless of baseline cholesterol, ACC/AHA guidelines CV risk calculator include stroke as outcome, so high dose statin therapy recommended for all patients for secondary prevention and in specific high-risk patients for primary prevention
[11] Also reduce risk of other forms of atherosclerotic disease, especially CAD

[12] Patients with CAS have high risk of death due to CAD
[13] Bruit is insensitive sign of CAS
[14] For stroke secondary prevention, guidelines recommend either ASA 50–100 mg daily or ASA 25 mg daily plus extended release dipyridamole 200 mg bid or clopidogrel 75 mg daily
[15] Thiazide diuretics and ACEI/ARB are first-line treatment in patients with cerebrovascular disease
[16] Symptomatic patients with TIA or Stroke symptoms within 6 months, with perioperative stroke or mortality risk <6 %, should undergo endarterectomy if diameter of ipsilateral internal carotid artery is reduced by >70 % by non-invasive imaging or >50 % by catheter angiography; in asymptomatic patients no strong evidence to support invasive procedures vs medical therapy; however, in asymptomatic patients with >70 % stenosis of internal carotid artery, reasonable to perform endarterectomy if perioperative risks low
[17] Stenting: indicated alternative to endarterectomy for symptomatic patients meeting criteria in [16]; sometimes preferred when neck anatomy unfavorable for surgery; stenting controversial in asymptomatic patients

Guidelines

Guidelines for the prevention of stroke in patients with stroke and transient ischemic attack
Stroke. 2014;45:2160–236. http://stroke.ahajournals.org/content/45/7/2160.long.

Update of the European Stroke Initiative (EUSI) recommendations for stroke management
European handbook of neurological management, vol. 1. 2nd ed. https://www.eaneurology.org/fileadmin/user_upload/guidline_papers/EFNS_guideline_2011_Ischaemic_stroke_and_transient_ischaemic_attack.pdf.

Patient Information

AHA

http://circ.ahajournals.org/content/114/7/e244.full.

Medlineplus

ENGLISH

http://www.nlm.nih.gov/medlineplus/carotidarterydisease.html.

ESPANOL

http://www.nlm.nih.gov/medlineplus/spanish/carotidarterydisease.html.

Cleveland Clinic

http://my.clevelandclinic.org/services/heart/disorders/hic-Carotid-Artery-Disease.

Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/carotid-artery-disease/basics/symptoms/con-20030206.

Professional Information

Review

J Am Coll Cardiol. 2015;65:1134–43. http://content.onlinejacc.org/article.aspx?articleID=2199422.

Review

Cochrane Database Syst Rev. 2012;(9):CD000515. http://www.ncbi.nlm.nih.gov/pubmed/22972047.

Asymptomatic

Circulation. 2013;127:739–42. http://circ.ahajournals.org/content/127/6/739.long.

Asymptomatic

J Am Coll Cardiol. 2013;61:2510–13. http://content.onlinejacc.org/article.aspx?articleID=1677758.

GOUT: Increased Risk for TIA/Stroke

Ann Rheum Dis. 2015;74:642–7. http://ard.bmj.com/content/74/4/642.full.

Medical Treatment

Stroke. 2009;40:e573–83. http://stroke.ahajournals.org/content/40/10/e573.full.

Plaque Histopathology

Neurosurgery. 2006;59(5 Suppl 3):S219–27. http://www.ncbi.nlm.nih.gov/pubmed/17053606.

Screening

U.S. Preventive Services Task Force. http://www.uspreventiveservicestaskforce.org/uspstf/uspsacas.htm.

Stenting

J Am Coll Cardiol. 2014;64:722–31. http://content.onlinejacc.org/article.aspx?articleID=1895460.

Stenting

Eur Heart J. 2015;36:13–21. http://eurheartj.oxfordjournals.org/content/36/1/13.

Stenting/Endarterectomy

N Engl J Med. 2016;374:1011–20. http://www.nejm.org/doi/full/10.1056/NEJMoa1515706.

Stenting/Endarterectomy

N Engl J Med. 2010; 363:11–23. http://www.nejm.org/doi/full/10.1056/NEJMoa0912321#t=article.

Stenting/Endarterectomy

Stroke. 2011;42:2019–25. http://stroke.ahajournals.org/content/42/7/2019.abstract?ijkey=11dcad9b1c257491aa0a951e2bb73409df09281a&keytype2=tf-ipsecsha.

Stenting/Endarterectomy (Elderly)

JAMA. 2014;311:1244–5. http://jama.jamanetwork.com/article.aspx?articleid=1849967.

Updates and More

https://clinicalguidecvd.com/cas

Chapter 40
Catecholaminergic Polymorphic Ventricular Tachycardia

Management Keys

- Consider this diagnosis in any young person with unexplained syncope
- Refer to practitioners/centers with CPVT expertise
- Genetic counseling/screen family members
- Treat with beta-blockers for life
- Consider left sympathectomy in patients with drug failure
- Consider use of ICD in select patients [8]
- Educate patient/family members about symptoms, trigger (exercise, emotional) avoidance

ICD-10 Code

I47.2

Alternate Names/Abbreviation

CPVT
FAMILIAL POLYMORPHIC VT

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_40,

Description/Etiology

Exercise and stress-induced polymorphic or bi-directional VT in absence of anatomical heart disease or Long QT on resting ECG
Often misdiagnosed as seizure disorder or Benign Fainting Syndrome
SCD often first manifestation

Comorbid Conditions

FAMILY HX: SUDDEN DEATH

Demography

F: 60 %
All ages but initial presentation usually childhood-young adults

Pathophysiology

Rapid VT with reduced cardiac output; may self-terminate or result in VF/SCD

Signs/Symptoms [1]

CARD ARREST
CHEST – PALPITATIONS
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)
DIZZY/LIGHTHEADED/PRESYNCOPE
HEART, RATE – SLOW (BRADYCARDIA) [2]

Differentiation

Andersen-Tawil Syndrome [3]
Ankyrin-B Syndrome
Arrhythmogenic Right Ventricular Dysplasia
Brugada Syndrome
CAD and coronary artery anomalies
Cardiomyopathy – Hypertrophic
Long QT Syndrome
Non-CV causes of syncope
Other types of underlying heart disease
Seizure disorders

Complications

Syncope
SCD

Laboratory

GENETIC TESTING

ECG [2]

QTC – N [6]
RATE – DECREASED (SINUS BRADYCARDIA) [2]

Imaging

NS/VAR WITH COMORBID

Genomics

CASQ2 [AUTOSOMAL RECESSIVE]
RYR2 [AUTOSOMAL DOMINANT] [7]
TRDN [AUTOSOMAL RECESSIVE]

Other Tests

Exercise test to provoke: [5]

- PVCs
- Polymorphic VT
- Supraventricular tachyarrhythmias
- VF

Ambulatory ECG monitoring [5]
Provocative testing (isoproterenol)

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic

Beta-blockers [4]

Treatment: Surgical/Invasive

ICD [8]
Catheter ablation
Sympathetic denervation [9]

Prevention

Exercise limitation/avoidance
Family screening

Course

Untreated mortality: 30–50 % by age 40 years

Notes

[1] Induced by exercise/emotion
[2] Usually normal; some patients have bradycardia
[3] Long QT, periodic muscle weakness, unusual facial features, clinodactyly, scoliosis, short stature
[4] CCBS and flecainide have been proposed as second-line agents
[5] Dysrhythmias cannot be provoked in all patients
[6] May be borderline long in some patients
[7] RYR2 mutation found in >50 % of pts with CPVT
[8] ICD may be required in as many as 30 % of patients
[9] Consider when syncope/repeat ICD shocks occur despite optimal drug treatment

Guidelines

2011 HRS/EHRA expert consensus statement on genetic testing for channelopathies and cardiomyopathies
Heart Rhythm. 2011;8:1308–39. http://www.heartrhythmjournal.com/article/S1547-5271(11)00607-2/abstract.

2013 HRS/EHRA/APHRS expert consensus statement on the diagnosis and management of patients with inherited primary arrhythmia syndromes
Heart Rhythm. 2013;10:1932–63. http://www.hrsonline.org/content/download/14025/629471/file/IA-Final%20Published-Dec%202013%20(2).pdf.

Patient Information

Genetics Home Reference

http://ghr.nlm.nih.gov/condition/catecholaminergic-polymorphic-ventricular-tachycardia

NYU Langone

http://cvgenetics.med.nyu.edu/genetic-conditions/catecholaminergic-polymorphic-ventricular-tachycardia

SADS Foundation

http://www.sads.org/library/cpvt#.VLewhGfwvds

Professional Information

Editorial

Circulation. 2015;131:2169–71 http://circ.ahajournals.org/content/131/25/2169.full.

Review

Heart. 2013;99:497–504. http://heart.bmj.com/content/99/7/497.extract.

Review: Children

Circulation. 1995;91:1512–19. http://circ.ahajournals.org/content/91/5/1512.full.

Review: Genetics of Sudden Cardiac Death

Circ Res. 2015;116:1919–36. http://circres.ahajournals.org/content/116/12/1919.full.

Review: NSVT

J Am Coll Cardiol. 2012;60:1993–2004. http://content.onlinejacc.org/article.aspx?articleid=1378223.

CLIN/Molecular Characterization

Circulation. 2002;106:69–74. http://circ.ahajournals.org/content/106/1/69.full.

Protocol: Treatment/Follow Up

J Am Coll Cardiol. 2013;61(10-S). doi:10.1016/S0735-1097(13)60346-0.http://content.onlinejacc.org/article.aspx?articleid=1664493.

Sympathetic Denervation

Circulation. 2015;131:2185–93. http://circ.ahajournals.org/content/131/25/2185.full.

Updates and More

https://clinicalguidecvd.com/cpvt

Chapter 41
Coarctation of Aorta

ICD-10 Code

Q25.1

Management Keys

- Evaluate for associated anomalies, especially bicuspid AV [7]
- Abstain from high-intensity exercise with severe COA
- Surgical correction indicated for children with peak-to-peak gradient >20 mmHg, imaging showing collateral flow, or history of HF
- Surgical correction indicated for adults with peak-to-peak gradient ≥20 mmHg, or imaging showing significant coarctation with collateral flow
- Close follow-up/monitoring after correction for hypertension, re-coarctation, or aneurysm formation after intervention [14]

Alternate Names/Abbreviation

COA

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_41,

Description/Etiology

Narrowed aorta in region of ligamentum arteriosum adjacent to origin of left subclavian artery
Presentation varies based on degree of obstruction; generally asymptomatic
Primarily congenital; can be isolated or occur with other forms of congenital HD
Acquired cases may occur secondary to aortic inflammatory syndromes

Predisposing/Comorbid Conditions

AORTIC ARCH HYPOPLASIA
AORTIC STENOSIS – SUBVALVULAR
ATRIAL SEPTAL DEFECT
BICUSPID AORTIC VALVE [7]
CIRCLE OF WILLIS ANEURYSM
DESCENDING AORTIC ANEURYSM
DOUBLE OUTLET RIGHT VENTRICLE
HYPERTENSION – SYSTEMIC ARTERIAL
MARFAN SYNDROME
MITRAL VALVE ATRESIA
MITRAL VALVE PROLAPSE
NOONAN SYNDROME
PATENT DUCTUS ARTERIOSUS
PERSISTENT LEFTSUPERIOR VENA CAVA
PULMONARY STENOSIS
TRANSPOSITION OF GREAT ARTERIES
TREACHER COLLINS SYNDROME
TURNER SYNDROME
VENTRICULAR SEPTAL DEFECT

Demography

Males 3:1
Often familial

Pathophysiology

Direct consequences mainly due to Systemic Arterial Hypertension/impaired renal perfusion
Hemodynamic abnormalities begin only after birth, due to closure of ductus arteriosus
Acts as an obstruction to LVOT, leading to increased LV afterload
LVH and collaterals develop as compensatory mechanisms [9]

Signs/Symptoms [3] [10]

ARTERIAL PRESSURE, SYS, LE – DECR
ARTERIAL PRESSURE, SYS, UE – INCR, R > L
ARTERIAL PULSE PRESSURE – INCR [10]
ARTERIAL PULSE, BRACHIAL, L – DECR/ABSENT
ARTERIAL PULSE, CAROTID – HYPERDYNAMIC
ARTERIAL PULSE, FEMORAL – DECR/ABSENT
ARTERIAL PULSE, FEMORAL – DELAYED
ARTERIAL PULSE, ILIAC – DECR/ABSENT
ARTERIAL PULSE, UE – ASYMMETRIC
BACK, INTERSCAPULA – MURMUR/BRUIT
BLOOD PRESSURE, ARTERIAL – INCREASED/ELEVATED
CHEST, ANT, INFRACLAV, L – MURMUR, SYS
EXTREM, LOWER, BILAT – PAIN, EFFORT (CLAUDICATION)
EXTREM, LOWER, DIGITS, TEMP – DECR
EXTREM, UPPER, L – FATIGUE
EXTREM, UPPER, L – THIN
HEADACHE
HEART, A2, INTENSITY – INCR
HEART, LSB, MID – MURMUR, SYS
HEART, LV, APEX – MURMUR, DIAS
HEART, LV, APEX, IMP – FORCEFUL/SUSTAINED

HEART, LV, APEX, IMP – SUSTAINED
HEART, RSB, MID – IMP, SYS
HEART, RSB, UPPER – IMP, SYS
HEART, S4 RV
MUSCLE, EXTREM, LOWER, BILAT – WEAK
NECK, SENSATION – PULSATIONS
NECK, SUPRASTERNAL NOTCH – MURMUR, SYS
NECK, SUPRASTERNAL NOTCH – THRILL, SYS
NOSE – BLOOD (EPISTAXIS)
SKIN, SUBCUT, INTRASCAPULA – ART PULSE
SKIN, SUBCUT, LSB – ART PULSE
SKIN, SUBCUT, RSB – ART PULSE
SWALLOWING – DIFFICULT (DYSPHAGIA)

Differentiation

Aortic Dissection
AS
Essential/other causes Systemic Arterial Hypertension
HCM
Peripheral artery disease
Supravalvular AS
Thoracic aorta aneurysm

Complications

Aortic Aneurysm [8]
Aortic Dissection/Rupture [8]
Cerebrovascular Accident
Complications associated with severe hypertension
HF
Infective Endocarditis
Intercostal Artery Aneurysm
Intracerebral aneurysm/hemorrhage
Resistant arterial hypertension [4]
SCD

Laboratory

NS

ECG

P WAVE – FLAT
P WAVE, DUR – INCR
QRS – LVH PATTERN
QRS – RBBB/RBBB PATTERN
QRS, AXIS – L

Imaging [1] [2] [11]

[ASSESS FOR OTHER CARD STRUCTURAL ABN, ESP AV]
[MRI/CT ANGIO: LOCATE EXACT SITE OF COA]
3 SIGN [1]
AORTA, ASCEND, FLOW – TURBULENT [PROXIMAL]
AORTA, ASCEND, SIZE – INCR
RIBS – NOTCHING

Other Tests

Stress test and stress-echo
Cardiac Catheterization/coronary angiography: for suspected CAD when surgery planned

Treatment: Nonpharmacologic

Hypertension diet

Treatment: Pharmacologic

Prostaglandin E1 [5]
Antihypertensives [12]
Endocarditis prophylaxis not indicated except for repaired or uncomplicated COA unless prior history of IE

Treatment: Surgical/Invasive [13]

Percutaneous angioplasty/stent
Surgical repair

Course

Unrepaired: 75 % mortality by age 45 years
After surgical correction, patients still at increased risk for recurrent hypertension, accelerated CAD, CVA, SCD [14]
5–10 % risk of re-coarctation after intervention
Postop survival:

10 years: about 92 %
20 years: about 85 %
30 years: about 73 %

Notes

[1] CXR pattern due to combination of descending aorta convexity to left with dilated left subclavian artery
[2] Echo: demonstrate COA via suprasternal notch view
[3] Isolated COA often asymptomatic until features of severe hypertension develop
[4] Can be seen even after intervention; appears to correlate with duration of pre-correction hypertension
[5] To dilate ductus arteriosus in neonates with critical COA
[6] Surgical correction generally preferred in infants due to risk of reintervention with angioplasty; angioplasty generally preferred in adults and patients with isolated COA
[7] Bicuspid AV occurs in >40 % of patients with COA

[8] Associated intrinsic abnormality in aortic wall predisposes to dissection or rupture in ascending aorta or area of coarctation

[9] Abundant collaterals may reduce COA gradient and mask obstruction severity

[10] ACC/AHA 2008 Guidelines recommendation:

"Every patient with systemic arterial hypertension should have the brachial and femoral pulses palpated simultaneously to assess timing and amplitude evaluation to search for the "brachial-femoral delay" of significant aortic coarctation. Supine bilateral arm (brachial artery) blood pressures and prone right or left supine leg (popliteal artery) blood pressures should be measured to search for differential pressure. (*Level of Evidence*: *C*)
"

[11] ACC/AHA 2008 Guidelines recommendation:

"Every patient with coarctation (repaired or not) should have at least 1 cardiovascular MRI or CT scan for complete evaluation of the thoracic aorta and intracranial vessels. (*Level of Evidence*: *B*)"

[12] ACC/AHA 2008 Guidelines recommendation:

"Hypertension should be controlled by beta blockers, ACE inhibitors, or angiotensin-receptor blockers as first-line medications. The choice of beta blockers or vasodilators may be influenced in part by the aortic root size, the presence of AR, or both."

[13] ACC/AHA 2008 Guidelines Recommendations for Interventional and Surgical Treatment of Coarctation of the Aorta in Adults

"Class I

1. Intervention for coarctation is recommended in the following circumstances:

   A. Peak-to-peak coarctation gradient greater than or equal to 20 mmHg. (*Level of Evidence*: *C*)
   B. Peak-to-peak coarctation gradient less than 20 mmHg in the presence of anatomic imaging evidence of significant coarctation with radiological evidence of significant collateral flow. (*Level of Evidence*: *C*)

2. Choice of percutaneous catheter intervention versus surgical repair of native discrete coarctation should be determined by consultation with a team of ACHD cardiologists, interventionalists, and surgeons at an ACHD center. (*Level of Evidence*: *C*)
3. Percutaneous catheter intervention is indicated for recurrent, discrete coarctation and a peak-to-peak gradient of at least 20 mmHg. (*Level of Evidence*: *B*)
4. Surgeons with training and expertise in CHD should perform operations for previously repaired coarctation and the following indications:)

    A. Long recoarctation segment. (*Level of Evidence*: *B*)
    B. Concomitant hypoplasia of the aortic arch. (*Level of Evidence*: *B*)

Class IIb

1. Stent placement for long-segment coarctation may be considered, but the usefulness is not well established, and the long-term efficacy and safety are unknown. (*Level of Evidence*: *C*)"

[14] ACC/AHA 2008 Recommendations for Key Issues to Evaluate and Follow-Up

"Class I

1. Lifelong cardiology follow-up is recommended for all patients with aortic coarctation (repaired or not), including an evaluation by or consultation with a cardiologist with expertise in ACHD. (*Level of Evidence*: *C*)
2. Patients who have had surgical repair of coarctation at the aorta or percutaneous intervention for coarctation of the aorta should have at least yearly follow-up. (*Level of Evidence*: *C*)
3. Even if the coarctation repair appears to be satisfactory, late postoperative thoracic aortic imaging should be performed to assess for aortic dilatation or aneurysm formation. (*Level of Evidence*: *B*)

4. Patients should be observed closely for the appearance or reappearance of resting or exercise-induced systemic arterial hypertension, which should be treated aggressively after recoarctation is excluded. (*Level of Evidence*: *B*)

Evaluation of the coarctation repair site by MRI/CT should be performed at intervals of 5 years or less, depending on the specific anatomic findings before and after repair. (*Level of Evidence*: *C*)"

Guidelines

ACC/AHA 2008 guidelines for the management of adults with congenital heart disease

J Am Coll Cardiol. 2008;52:e143–263. http://content.onlinejacc.org/article.aspx?articleid=1188032#tab1.

ESC Guidelines for the management of grown-up congenital heart disease (new version 2010)

Eur Heart J. 2010;31:2915–57. http://eurheartj.oxfordjournals.org/content/ehj/31/23/2915.full.pdf.

Patient Information

Images

http://www.nlm.nih.gov/medlineplus/ency/imagepages/18128.htm.
http://www.cdc.gov/ncbddd/heartdefects/CoarctationOfAorta-graphic.html.

Medlineplus

ENGLISH

http://www.nlm.nih.gov/medlineplus/ency/article/000191.htm.

ESPANOL

http://www.nlm.nih.gov/medlineplus/spanish/ency/article/000191.htm.

AHA

http://circ.ahajournals.org/content/131/9/e363.full.

CDC

http://www.cdc.gov/ncbddd/heartdefects/coarctationofaorta.html.

Professional Information

Review

J Cardiovasc Med (Hagerstown). 2007;8:123–8. http://journals.lww.com/jcardiovascularmedicine/Abstract/2007/02000/Aortic-coarctation--an-overview.8.aspx.

Comorbid Anomalies

Circulation. 1970;41:1067–75. http://circ.ahajournals.org/content/41/6/1067.abstract?sid=d9281a83-249e-4f61-bcd3-e0bcbac8836c.

Coronary Artery Disease

Circulation. 2012;126:16–21. http://circ.ahajournals.org/content/126/1/16.full?sid=d9281a83-249e-4f61-bcd3-e0bcbac8836c.

Image (3-D MR Angio)

ACC Cardiosource. http://www.acc.org/education-and-meetings/image-and-slide-gallery/media-detail?id=5185914e76bc4c25a9016ca693dc11cf.

Intracranial Aneurysms

Am J Cardiol. 2015;116:630–33. http://www.ajconline.org/article/S0002-9149(15)01354-5/abstract.

Long-Term Follow-Up

J Am Coll Cardiol. 2013;62:1020–25. http://content.onlinejacc.org/article.aspx?articleID=1711132.

Long-Term Follow-Up

Am J Cardiol. 2002;89:541–7. http://www.sciencedirect.com/science/article/pii/S0002914901022937.

Long-Term Outcomes

Heart. 2015;101:1190–95. http://heart.bmj.com/content/101/15/1190?etoc.

Long-Term Outcomes

Circulation. 1989;80:840–5. http://www.ncbi.nlm.nih.gov/pubmed/2791247.

Post-stent Blood Pressure

Am J Cardiol. 2013;111:731–6. http://www.sciencedirect.com/science/article/pii/S0002914912024575.

Pregnancy: Hypertension Complications

Am J Cardiol. 2011;107:1529–34. http://www.sciencedirect.com/science/article/pii/S0002914911003456.

Stenting

Arch Cardiovas Dis. 2011;104:627–35. http://www.sciencedirect.com/science/article/pii/S1875213611002828.

Stenting: Self-expanding

Circ Cardiovasc Intervent. 2015;8:e001799. http://circinterventions.ahajournals.org/content/8/1/e001799.abstract?etoc.

Updates and More

https://clinicalguidecvd.com/coa

Chapter 42
Coronary Arteriovenous Fistula

ICD-10 Code

Q24.5

Alternate Names/Abbreviation

CAVF

Description/Etiology

Rare congenital anomaly comprising abnormal communication between coronary artery and a cardiac chamber or vessel adjacent to heart

Present in 0.002 % of general population and visualized in about 0.25 % of patients having cardiac catheterization

Comorbid Conditions

NON-CONGENITAL:

PRIOR CARDIAC/CORONARY ARTERY INTERVENTIONS

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_42,

PRIOR MYOCARDIAL BIOPSY
TRAUMA

Demography

Gender equal

Pathophysiology

Congenital: arise from:

RCA (50 %)
LCA (42 %)
Both LCA/RCA (8 %)

Congenital: drain into:

Coronary sinus
RA
RV
PA
Left heart (rare)

Signs/Symptoms [9]

ARTERIAL PRESSURE, DIAS – DECR [1]
ARTERIAL PULSE PRESSURE – INCR [1]
BLOOD PRESSURE, ARTERIAL – INCREASED/ELEVATED [1] [SYS]
BREATHING – DIFF (DYSPNEA)
CHEST – PAIN, EFFORT (ANGINA PECTORIS)
FATIGUE
HEART, LSB, LOWER – IMP, SYS [3] [5]
HEART, LSB, LOWER – MURMUR, CONT [4] [5]
HEART, LSB, MID – MURMUR, CONT [5] [6]
HEART, LSB, MID – MURMUR, SYS [8]
HEART, LSB, UPPER – MURMUR, CONT [6]

HEART, LV, APEX – MURMUR, DIAS [7]
HEART, LV, APEX, IMP – FORCEFUL/SUSTAINED [2]
HEART, RSB, LOWER – MURMUR, CONT [4]
HEART, RSB, UPPER – MURMUR, SYS [8]
HEART, SUBXIPHOID – MURMUR, CONT [5]

Differentiation

All causes of continuous murmurs, especially PDA
Atherosclerotic CAD

Complications

AMI
HF
Infective Endocarditis
SCD

Laboratory

NS

ECG [10]

DYSRHYTHMIAS – ATRIAL (PACS/OTHERS)
P WAVE – TALL/PEAKED [11]
P WAVE, DUR – INCR [12]
QRS – LVH PATTERN [13]
QRS – RVH PATTERN [13]
ST SEGMENT – DEPR [14]

Imaging [15]

ECHO/DOPPLER MAY DETECT ORIGIN, COURSE, SITE OF DRAINAGE

Other Tests

Cardiac catheterization with coronary angiography

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic

NS

Treatment: Surgical/Invasive [17]

Transcatheter closure [16]
Surgical closure [16]

Course

Unrepaired: insufficient data

Notes

[1] Large fistula, with drainage into either RH or LH
[2] With large fistula; hyperdynamic LV impulse if drainage into LV or RV
[3] With large fistula; hyperdynamic RV impulse if drainage into RV

[4] Drainage into RA or coronary sinus; may extend over sternum and lower LSB
[5] Drainage into body of RV
[6] Drainage into PA trunk or LA
[7] Decrescendo, drainage into LV
[8] Systolic ejection murmur due to increased flow across AV
[9] First clinical manifestation may be complication
[10] Often normal
[11] RA drainage
[12] RV, PA trunk, LA drainage
[13] With large volume, biventricular hypertrophy; except when LV only, LVH alone occurs
[14] If myocardial ischemia occurs
[15] CXR often normal
[16] All large fistulae and all fistulae with complications, myocardial ischemia, or ventricular dysfunction
[17] ACC/AHA 2008 Recommendations:

"Class I

1. Surgeons with training and expertise in CHD should perform operations for management of patients with CAVF. (*Level of Evidence*: *C*)
2. Transcatheter closure of CAVF should be performed only in centers with expertise in such procedures. (*Level of Evidence*: *C*)
3. Transcatheter delineation of CAVF course and access to distal drainage should be performed in all patients with audible continuous murmur and recognition of CAVF. (*Level of Evidence*: *C*)"

Guidelines

ACC/AHA 2008 guidelines for the management of adults with congenital heart disease
J Am Coll Cardiol. 2008;52:e143–263http://content.onlinejacc.org/article.aspx?articleid=1188032#tab1.

ESC Guidelines for the management of grown-up congenital heart disease (new version 2010)
Eur Heart J. 2010;31:2915–57. http://eurheartj.oxfordjournals.org/content/ehj/31/23/2915.full.pdf.

Patient Information

Images

http://www.ncbi.nlm.nih.gov/pmc/articles/PMC555824/figure/f1-14/.

Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/arteriovenous-fistula/basics/causes/con-20034876.

Professional Information

Review

Circulation. 1979;59:849–54. http://circ.ahajournals.org/content/59/5/849.abstract.

Clinically Silent

Am J Cardiol. 1999;83:407–11. http://www.sciencedirect.com/science/article/pii/S0002914998008789.

Echo-Guided Device Closure

J Am Coll Cardiol. 2013;61:1458–68. http://content.onlinejacc.org/article.aspx?articleid=1671092.

IMAGE

J Am Coll Cardiol. 2009;53:523. http://content.onlinejacc.org/article.aspx?articleid=1139407.

Image: Coronary Steal

Eur Heart J Cardiovasc Imaging. 2015;16:176. http://ehjcimaging.oxfordjournals.org/content/16/2/176.

Natural History/Management

J Cardiothorac Surg. 2009;4:62. http://cardiothoracicsurgery.biomedcentral.com/articles/10.1186/1749-8090-4-62.

Transcatheter Closure

J Am Coll Cardiol. 2002;39:1026–32. http://content.onlinejacc.org/article.aspx?articleid=1127840.

Updates and More

https://clinicalguidecvd.com/cav

Chapter 43
Deep Vein Thrombosis: Lower Extremity (Venous Thromboembolism/VTE)

ICD-10 Code

I80.2

Alternate Names/Abbreviation

DVT
THROMBOPHLEBITIS
VENOUS THROMBOEMBOLISM (VTE) [20]

Description/Etiology

Blood clot formation usually within deep vein of lower leg or proximal iliofemoral vein [1]

Caused by combination of pathophysiologic states, including:

- Endothelial injury
- Hypercoagulability
- Stasis

Thrombi may embolize through IVC and right heart, obstructing pulmonary arterial tree, with resultant

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_43,

hemodynamic and gas-exchange abnormalities. DVT can also cause paradoxical embolism to arterial circulation [18]

Occurs spontaneously or associated with other disease; >50 % of patients have >2 risk factors/comorbid conditions [See Appendix A]

May-Thurner Syndrome: obstruction/entrapment of common iliac vein by overlying common iliac artery

Associated with non-O blood groups [24]

Comorbid Conditions

[See Appendix A and B]

ACUTE INFECTION
ACUTE MYOCARDIAL INFARCTION
ACUTE RHEUMATOLOGIC DISORDER
AIR TRAVEL [EXTENDED]
CANCER [21]
CHRONIC KIDNEY DISEASE
DEHYDRATION [2]
DISEASE-MODIFYING BIOLOGIC DMARDS
DRUGS: CANCER THERAPY [17]
FAMILY HX: THIS CONDITION
HEART FAILURE
HORMONE USE
HOSPITALIZATION
HX: THIS CONDITION
HYPERCOAGULATION STATES
HYPOTENSION
IMMOBILITY [ESP LIMB] [2]
INFLAMMATORY BOWEL DISEASE
MAY-THURNER SYNDROME [25]
NEPHROTIC SYNDROME
OBESITY
ORAL CONTRACEPTIVES
ORTHOSTATIC HYPOTENSION
POSTPARTUM [19]

PREGNANCY
RESPIRATORY FAILURE
RHEUMATOID ARTHRITIS
SICKLE CELL DISEASE/TRAIT [16]
STROKE
SURGERY [RECENT] [10]
THROMBOPHILIA [22]
TOBACCO USE
TRAUMA [RECENT]
VENOUS STASIS
VESSEL WALL ABNORMALITIES, ESP TRAUMA

Demography

All populations

Pathophysiology

Obstruction of venous return, local inflammation, thromboembolism [20]

Signs/Symptoms [8]

EXTREM, CALF – PAIN
EXTREM, CALF – PAIN, REST [6]
EXTREM, CALF – TENDER
EXTREM, POP SPACE – PAIN [7]
EXTREM, UNILAT – EDEMA
FEVER [3]
HEART, RATE – RAPID (TACHYCARDIA) [4]
SKIN, COLOR – RED (ERYTHEMA)
VEINS, POP – TENDER [5]
VEINS, SUPERFICIAL – DIL
VENOUS CORD – PALPABLE

Differentiation [9]

Achilles Tendonitis
Acute Arterial Ischemia
Advential Cystic Disease of common femoral vein
Baker Cyst rupture
Calf hematoma
Cellulitis
Erysipelas
Fracture
Lymphedema
Muscle Vein Thrombosis
Pelvic Vein Obstruction
Superficial Vein Thrombosis
Torn gastrocnemius
Torn meniscus

Complications

Acute Pulmonary Embolism
Paradoxical Embolism [18]
Phlegmasia Cerulea Dolans [13]
Postthrombotic Syndrome [15]
Venous ulcer

Laboratory

BLOOD, D-DIMERS – INCR [11]

ECG

N/NS ABN [12]

Imaging

[DUPLEX ULTRASOUND, CT, MR VENOGRAPHY]

Other Tests

Contrast venography

Treatment: Nonpharmacologic

Intermittent pneumatic compression [23]
Leg elevation

Treatment: Pharmacologic

Antithrombotic options:

- Antiplatelet agents
  - ASA
  - Clopidogrel
- Direct thrombin inhibitors
  - Bivalrudin
  - Dabigatran
- Heparin
  - LMWH (preferable to UFH)
  - UFH
- Injectable direct factor Xa inhibitors
  - Fondaparinux
- Oral factor Xa inhibitors
  - Apixaban

Edoxaban
Rivaroxaban

Warfarin

Treatment: Surgical/Invasive

Thrombolysis [14]

Prevention [See Appendix A and B]

ASA
Ambulation
Compression hosiery
Intermittent pneumatic compression [23]
Leg elevation

Notes

[1] DVT can also occur in other veins, including subclavian and visceral veins and IVC
[2] Possible contributors to DVT associated with long-duration plane travel
[3] Low grade: Michaeli's Sign
[4] Mahler Sign
[5] Pratt Sign
[6] May be precipitated by foot dorsiflexion
[7] May be precipitated by knee extension (Sigg sign)
[8] Clinical features of DVT highly insensitive and often subsequently found not due to DVT; may be more reliable in outpatient settings
[9] Note: many conditions in differential occur as comorbid conditions
[10] Especially orthopedic; knee/hip replacement particularly problematical for DVT due to thromboplastins release from dissected soft tissue/reamed bone, com-

bined with venous stasis during surgery/postop immobility

[11] Highly sensitive but NS

[12] Abnormal with comorbid conditions or pulmonary embolism

[13] Secondary arterial insufficiency due to extension of thrombus into venules and capillaries, manifest by cyanosis and often gangrene

[14] Mainly iliofemoral and upper extremity DVT

[15] Long-term, chronic pain, edema, erythema, varicosities, paresthesias; venous ulceration in severe cases

[16] Blacks with Sickle Cell Disease/Trait at particularly high risk for DVT during pregnancy

[17] Including, but not limited to Bevacizumab, Sorafenib, Sunitinib, Erlotinib, Thalidomide, Lenalidomide, Tamoxifen, Cisplatin, Vorinostat

[18] Requires comorbid intracardiac or pulmonary shunt for entry into systemic arterial circulation

[19] Low baby birth weight associated with 3x risk for postpartum thromboembolism

[20] 2/3 of patients with VTE present with DVT; 1/3 with Acute Pulmonary Embolism

[21] Especially lung, pancreas, colorectal, kidney, prostate

[22] Including:

Antiphospholipid Syndrome
Antithrombin defects
Factor V Leiden defect
Protein C/S defects

[23] Intermittent pneumatic compression preferable to heparin in patients with increased bleeding risk in medical/nonorthopedic surgery

[24] Non-O blood groups associated with >30 % of venous thromboembolic events, but clinical utility of this observation unresolved

[25] May–Thurner syndrome: common anatomical anomaly in which right common iliac artery extrinsically compresses left common iliac vein

Appendix A

Caprini Risk Assessment Model

(Wells PS, Anderson DR, Roger M, Forgie M, Kearon C, Dreyer J, Kovacs G, Mitchell M, Lewandowski B, Kovacsb MJ. *N Engl J Med* 2003;349:1227–1235)

DVT unlikely: 0–1
DVT likely: 2/greater

1 Point

Acute Myocardial Infarction
Age 41–60 year
BMI >25 kg/m2
COPD
Heart Failure
Hx major surg (<1 month)
Lung dis (serious)
Medical pt on bed rest
Minor surg
Sepsis (<1 month)
Swollen legs
Varicose veins

2 Points

Age 61–74 years
Arthroscopic surg
Cancer (past/present)
Central venous access
Immobilizing plaster cast
Laparoscopic surg
Major open surg

3 Points

Age
Elev serum homocysteine
FH VTE
Heparin-induced thrombocytopenia
Hx VTE
Pos Factor V Leiden
Pos lupus anticoagulant
Pos prothrombin 20210A
Other cong/acq thrombophilia

4 Points

Acute spinal cord injury (<1 month)
Elective arthroplasty
Fracture
Hip
Leg
Pelvis
Multiple trauma (<1 month)
Stroke (<1 month)

Appendix B

Primary Care Rule

(Toll DB, Oudega R, Vergouwe Y, Moons KG. *Fam Pract* 2008;25:3–8)

Very low risk: 0–3
Increased risk: 4/greater
Absence of leg trauma 1
Active cancer past 6 months 1
Collateral leg vein distention 1
Male gender 1

Surgery prev mo 1
Use hormone contraceptive 1
Calf circumference difference
>2 cm^2
Abnormal D-dimer assay 6

Guidelines

Antithrombotic therapy for VTE disease: CHEST guideline and expert panel report
Chest. 2016. doi:10.1016/j.chest.2015.11.026. http://journal.publications.chestnet.org/article.aspx?articleid=2479255.

Patient Information

AHA: Guide to Recovery

Circulation. 2014;129:e477–9. http://circ.ahajournals.org/content/129/17/e477.full.

AHA (Prevention)

http://circ.ahajournals.org/content/110/16/e445.full?sid=081dcf54-2735-4e1c-bac6-0b9c5f2445f4.

AMA: AIR Travel Prevention

http://jama.jamanetwork.com/article.aspx?articleid=1486833.

CDC

http://wwwnc.cdc.gov/travel/page/dvt.
http://www.cdc.gov/features/thrombosis/.

NHLBI

https://www.nhlbi.nih.gov/health/health-topics/topics/dvt/.

Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/deep-vein-thrombosis/basics/definition/con-20031922.

Medline Plus

ENGLISH

http://www.nlm.nih.gov/medlineplus/deepveinthrombosis.html.

ESPANOL

http://www.nlm.nih.gov/medlineplus/spanish/deepveinthrombosis.html.

Merck

http://www.merckmanuals.com/home/heart-and-blood-vessel-disorders/venous-disorders/deep-vein-thrombosis-dvt.

Cleveland Clinic

http://my.clevelandclinic.org/services/heart/disorders/hypercoagstate/hic-Deep-Vein-Thrombosis-DVT.

Professional Information

AHA Scientific Statement

Circulation. 2011;123:1788–830. http://circ.ahajournals.org/content/123/16/1788.full?sid=d8df4ed1-c165-4899-a78-ce4e8942fac0.

Review (CME)

Ann Int Med. 2015;162:ITC1–15. http://annals.org/article.aspx?articleid=2288552.

Review

Surgery (Oxford). 2013;31:206–10. http://www.sciencedirect.com/science/article/pii/S0263931913000331.

Review: Treatment

JAMA. 2014;311:717–28. http://jama.jamanetwork.com/article.aspx?articleid=1829996.

Adventitial Cystic Disease of Common Femoral Vein

Ann Intern Med. 2016. http://annals.org/article.aspx?articleid=2499477.

ASA: Recurrence Prevention

Circulation. 2014;130:1062–71. http://circ.ahajournals.org/content/130/13/1062.full.

Cancer

Circulation. 2003;107:I-17–21. http://circ.ahajournals.org/content/107/23-suppl-1/I-17.full?sid=081dcf54-2735-4e1c-bac6-0b9c5f2445f4.

Caprini Risk Assessment Model

Am J Surg. 2010;199:S3–10. http://www.americanjournalofsurgery.com/article/S0002-9610(09)00638-2/abstract.

DVT and Pulmonary Embolism

Arch Intern Med. 2008;168:425–30. http://archinte.jamanetwork.com/article.aspx?articleid=413988&resultClick=3.

Endovascular Treatment

Interven Cardiol Clin. 2014;3:607–17. http://www.sciencedirect.com/science/article/pii/S2211745814000698.

Heart Failure

Circulation. 2014;130:410–8. http://circ.ahajournals.org/content/130/5/410.full.

Home Treatment/Comorbid Conditions

Am J Med. 2016;129:392–7. http://www.sciencedirect.com/science/article/pii/S000293431501027X.

Incidence/In-hospital Mortality Trends

Circulation. 2013;128:115–21. http://circ.ahajournals.org/content/128/2/115.full.

Knee/Hip Replacement Surg

Circulation. 2012;125:2151–5. http://circ.ahajournals.org/content/125/17/2151.full#ref-2.

Long-Term Outcomes

Arch Intern Med. 1995;155:1031–7. http://archinte.jamanetwork.com/article.aspx?articleid=620531&resultClick=3.

Marathon Athletes: INCR Risk

Circulation. 2013;128:e469–71. http://circ.ahajournals.org/content/128/25/e469.full.

May-Thurner Syndrome

Ann Vasc Surg. 2013;27:984–95. http://www.sciencedirect.com/science/article/pii/S0890509613002094.

Non-O Blood Group ASSN with VTE

Circulation. 2016;133:1449–57 http://circ.ahajournals.org/content/133/15/1449.abstract.

Oral Anticoagulants

Eur Heart J. 2014;35:1836–43. http://eurheartj.oxfordjournals.org/content/35/28/1836.

Orthostatic Hypotension

Am J Hypertens. 2016;29:634–40. http://ajh.oxfordjournals.org/content/29/5/634.abstract?etoc.

Paradoxical Embolism

J Am Coll Cardiol. 2014;64:403–15. http://content.onlinejacc.org/article.aspx?articleID=1889485.

Postpartum Risk: Low Birth Weight

Circulation. 2015;131:1471–6. http://circ.ahajournals.org/content/131/17/1471.abstract.

Postthrombotic Syndrome (AHA Statement)

Circulation. 2014;130:1636–61. http://circ.ahajournals.org/content/130/18/1636.full.

Prevention: Critically Ill Patients

JAMA. 2014;312:2135–45. http://jama.jamanetwork.com/article.aspx?articleid=1921813.

Prognostic Score for Low-Risk Outputs

Am J Med. 2015;128:90.e9–90.e15. http://www.sciencedirect.com/science/article/pii/S000293431400789X.

Rheumatoid Arthritis/DMARDS

Am J Med. 2015;128:539e7–17. http://www.sciencedirect.com/science/article/pii/S0002934314012066.

Risk Factors

Circulation. 2003;107:I9–16. http://circ.ahajournals.org/content/107/23-suppl-1/I-9.long.

Risk/Prophylaxis (Endorse Study)

Lancet. 2008;371:387–94. http://www.sciencedirect.com/science/article/pii/S0140673608602020.

Rivaroxaban

Br J Cardiol. 2015;22:78. http://bjcardio.co.uk/2015/06/treatment-of-vte-in-primary-care-building-a-new-approach-to-patient-management-with-rivaroxaban/.

Sickle Cell Disease

Am J Med. 2013;126:443–9. http://www.sciencedirect.com/science/article/pii/S0002934313001071.

Smoking: Risk

Circulation. 2013;127:1276–82. http://circ.ahajournals.org/content/127/12/1276.full.

Spinal Cord Injury

Thromb Res. 2014;133:579–84. http://www.sciencedirect.com/science/article/pii/S0049384814000322.

Thrombolysis: Short/Long Term Results

J Am Coll Cardiol. 2000;36:1336–43. http://content.onlinejacc.org/article.aspx?articleid=1126726.

Vitamin K Antagonists: Long-Term Verses Short-Term Use

JAMA. 2015;314:72–3. http://jama.jamanetwork.com/article.aspx?articleid=2382956.

Updates and More

https://clinicalguidecvd.com/dvtle

Chapter 44
Deep Vein Thrombosis: Upper Extremity

ICD-10 Code

I80.2

Alternate Names/Abbreviation

PAGET-SCHROETTER SYNDROME (EFFORT-RELATED) [1]
THROMBOPHLEBITIS

Description/Etiology

Blood clot formation, in 1 or multiple deep veins:

Axillary
Brachial
Internal jugular
Radial
Subclavian
Superior vena cava
Ulnar

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_44,

Primary causes:

Idiopathic
Effort-related [1]

Secondary causes:

Central venous catheter
Central venous pacemaker lead
Mass lesion

Comorbid Conditions

Mass lesion
Thrombophilia (acquired/inherited]

Demography

Variable according to etiology
No gender or ethnic predilection except when effort-related, which is more common in males

Pathophysiology

Obstruction of UE venous return, local inflammation, thromboembolism

Signs/Symptoms [5]

BREATHING – DIFF (DYSPNEA) [4]
DIZZY/LIGHTHEADED/PRESYNCOPE [4]
EXTREM, HAND – SWOLLEN
EXTREM, HAND – WEAKNESS (PARESIS) [2]
EXTREM, UPPER – PAIN
EXTREM, UPPER – PARESTHESIA
EXTREM, UPPER – SWELLING
EXTREM, UPPER, SHOULDER – PAIN

EYES, VISION – BLURRED [4]
FACE – SWELLING [4]
HEAD, SENSATION – FULLNESS [4]
MUSCLES, ARM – WEAK [2]
MUSCLES, EXTREM, UPPER DISTAL – WEAK (HAND DROP) [2]
MUSCLES, HAND – ATROPHY [2]
NECK – PAIN
NECK, JVP – ELEV
NECK, SUPRACLAV FOSSA – TENDER [2] [3]
SKIN, UE, COLOR – BLUE (CYANOSIS)
VEINS, SUPERFICIAL – DIL
VENOUS CORD – PALPABLE

Differentiation

Lymphedema
Mass compression of blood vessels
Muscle injury
Superficial vein thrombosis

Complications

Acute Pulmonary Embolism
Postthrombotic Syndrome [6] [7]

Laboratory

BLOOD, D-DIMERS – INCR [8]

ECG

N/NS ABN

Imaging

[COMPRESSION/DOPPLER ULTRASOUND, CT CONTRAST, MR CONTRAST]

Other Tests

Venography

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic

Anticoagulation

Treatment: Surgical/Invasive

Catheter/ultrasound-directed thrombolysis
Thrombectomy [9]
Venous bypass [9]

Prevention

In-line venous catheter care

Notes

[1] Paget-Schroetter Syndrome: axillary/subclavian thrombosis, associated with repetitive and strenuous use of UE, usually dominant arm; especially involves young athletes; usual presentation is pain and arm swelling; anterior scalene muscles usually overdeveloped

[2] Thoracic Outlet Syndrome; symptoms may be positional and worsened by hyperabduction of shoulder or lifting
[3] Brachiocephalic nerves
[4] SVC obstruction
[5] In patients with central venous catheter, only sign may be inability to access
[6] Occurs in up to 33 % of cases of upper extremity DVT
[7] Long term, including:

Chronic pain
Edema
Erythema
Paresthesias
Varicosities
Venous ulceration in severe cases

[8] Highly limited value as other conditions/procedures usually present that also increase D-dimers
[9] Refractory cases

Guidelines

NS

Patient Information

Medlineplus

ENGLISH

http://www.nlm.nih.gov/medlineplus/deepveinthrombosis.html.

ESPANOL

http://www.nlm.nih.gov/medlineplus/spanish/deepveinthrombosis.html.

Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/deep-vein-thrombosis/basics/definition/con-20031922.

CDC

http://wwwnc.cdc.gov/travel/page/dvt.
http://www.cdc.gov/features/thrombosis/.

Professional Information

Review

Circulation. 2002;106:1874–80. http://circ.ahajournals.org/content/106/14/1874.full.

Effort-Related

Acta Haematol. 2006;115:214–20. http://www.ncbi.nlm.nih.gov/pubmed/16549899.

Line-Associated

Am J Surg. 2014;208:45–9. http://www.ncbi.nlm.nih.gov/pubmed/24530041.

Thrombophilia

Thromb Haemost. 2008;100:440–6. http://www.ncbi.nlm.nih.gov/pubmed/18766260.

Updates and More

https://clinicalguidecvd.com/dvtue

Chapter 45
Early Repolarization Syndrome

ICD-10 Code

NS

Alternate Names/Abbreviation

ERS

Description/Etiology

Definition of early repolarization (all criteria required):

1. End-QRS notch or slur on downslope of a prominent R-wave; if there is a notch, it should lie entirely above the baseline; onset of a slur must also be above baseline
2. Jp = 0.1 mV in 2 or more contiguous leads of 12-lead ECG, excluding leads V1-3
3. QRS duration <120 ms

AHA definition: occurring in patients with early repolarization pattern who have survived idiopathic VF with clinical evaluation unrevealing for other explanations

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_45,

ECG positive S wave deflection in at least 2 consecutive inferior or lateral leads and at least 1 mm ST elevation above baseline

Malignant form:

Associated with idiopathic VF and increased mortality
ECG characteristic pattern: horizontal ST segment
Autosomal dominant transmission
Family history of SCD

Benign form:

Often observed in healthy persons, especially:

Young age
Male
Athletic
African-American

ECG characteristic pattern: rapidly upsloping ST segment

Comorbid Conditions

VENTRICULAR FIBRILLATION/CARDIAC ARREST
DYSRHYTHMIAS – VENTRICULAR

Demography

NS

Pathophysiology

Undefined

Signs/Symptoms (Malignant Form)

CHEST – PALPITATIONS
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)

Differentiation (Malignant Form) (ECG)

AMI [3]
ARVD
Athlete Heart
Brain injury
Brugada Syndrome
Long QT Syndrome
Hypercalcemia
Hypervagotonia
Hypothermia
Vasospastic Angina

Complications

Idiopathic VF
SCD
VF during AMI
VF during vasospastic angina

Laboratory

NS

ECG

J WAVE (OSBORN WAVE) [4] [5]
ST SEGMENT – ELEV [1] [4]

Imaging

NS/VAR WITH COMORBID

Genomics

KCNJ8

Other Tests

Ambulatory ECG monitoring
Exercise test [6]
Isoproterenol infusion [6]

Treatment: Nonpharmacologic

NS [7]

Treatment: Pharmacologic

NS [7]

Treatment: Surgical/Invasive

NS [7]

Prevention

NS [7]

Course

Variable

Notes

[1] Horizontal slope, as opposed to ST elevation in Athlete Heart, which has downward slope
[2] Other causes of J Wave
[3] Post-AMI with occlusion of LCA
[4] Vary with fluctuations in HR and autonomic tone; may appear primarily in either inferior or lateral leads
[5] Increase in amplitude immediately prior to onset of VF
[6] Decreases early repolarization
[7] Treatment recommendations specific for patients with familial ERS not developed

Guidelines

AHA scientific statement: electrocardiographic early repolarization
Circulation. 2016;133:1520–9. http://circ.ahajournals.org/content/133/15/1520.full.
The early repolarization pattern
J Am Coll Cardiol. 2015;66:470–7. http://content.onlinejacc.org/article.aspx?articleID=2398001.

Patient Information

NS

Professional Information

Review

Circulation. 2013;127:1620–9. http://circ.ahajournals.org/content/127/15/1620.full.

Review

J Am Coll Cardiol. 2013;62:863–8. http://content.onlinejacc.org/article.aspx?articleID=1699350.

Review: Genetics of Sudden Cardiac Death

Circ Res. 2015;116:1919–36. http://circres.ahajournals.org/content/116/12/1919.full.

ECG: Athlete Heart Versus Brugada Syndrome

Am J Cardiol. 2015;115:529–32. http://www.sciencedirect.com/science/article/pii/S0002914914021766.

ECG: Benign Versus Malignant

Heart Rhythm. 2012;9:225–9. http://www.ncbi.nlm.nih.gov/pubmed/21914497?dopt=Abstract.

ECG Phenotypes with Favorable Long-Term Outcome

Circulation. 2011;123:2666–73. http://circ.ahajournals.org/content/123/23/2666.abstract?ijkey=dca455f38e695f65591ac697bc026b62d84e5c13&keytype2=tf-ipsecsha.

Family Investigation

J Am Coll Cardiol. 2013;61:164–72. http://content.onlinejacc.org/article.aspx?articleID=1555160.

Incidence/Prognostic Value of ERS Pattern on ECG

Circulation.2011;123:2931–7. http://circ.ahajournals.org/content/123/25/2931.full.

J Wave Syndromes

Heart Rhythm. 2010;7:549–58. http://www.ncbi.nlm.nih.gov/pubmed/20153265?dopt=Abstract.

Large Families

J Am Coll Cardiol. 2013;61:164–72. http://content.onlinejacc.org/article.aspx?articleid=1555160&resultClick=3.

Long-Term Outcomes

Eur Heart J. 2011;32:3098–106. http://www.ncbi.nlm.nih.gov/pubmed/21785106.

Long-Term Outcomes

N Engl J Med. 2009;361:2529–37. http://www.nejm.org/doi/full/10.1056/NEJMoa0907589

Risk: Arrhythmia Death

J Am Coll Cardiol. 2013;61:645–50 http://content.onlinejacc.org/article.aspx?articleid=1555243&resultClick=3.

Sudden Death

N Engl J Med. 2008;358:2016–23. http://www.nejm.org/doi/full/10.1056/NEJMoa071968.

Updates and More

https://clinicalguidecvd.com/ers

Chapter 46
Ebstein Anomaly

ICD-10 Code

746.2

Alternate Names/Abbreviation

Ebstein's Anomaly

Description/Etiology

Wide variety of TV malformations, mainly inferior displacement of proximal attachments of TV leaflets from AV valve ring and RV malformations

Key features include:

- Adherence of TV leaflets to underlying myocardium (failure of delamination)
- Apical displacement of TV septal and posterior leaflets to below AV junction in RV
- Atrialization/dilatation of RV inflow
- Anterior TV leaflet redundancy, tethering, fenestrations
- Varying degrees of TR, which may be severe
- RA dilatation
- Varying degrees of cyanosis

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_46,

Atrial Septal Defect, other congenital abnormalities, and accessory conduction pathways often present.

Adults with mild Ebstein anomaly may be asymptomatic with no functional limitation and survival as long as ninth decade.

Symptoms in persons age >10 years more often due to electrophysiological rather than hemodynamic abnormalities.

Comorbid Conditions

ATRIAL SEPTAL DEFECT – SECUNDUM
COARCTATION OF AORTA
MITRAL VALVE PROLAPSE
PATENT DUCTUS ARTERIOSUS
PATENT FORAMEN OVALE
PREEXCITATION SYNDROMES
PULMONARY ATRESIA
PULMONARY STENOSIS
RIGHT VENTRICULAR HYPOPLASIA
VENTRICULAR SEPTAL DEFECT

Demography

Gender equal

Pathophysiology

Primary hemodynamic abnormality: tricuspid regurgitation (which may be severe) due to valve apical displacement and associated TV abnormalities

Signs/Symptoms

ARTERIAL PULSE – DECR/ABSENT
BREATHING – DIFF (DYSPNEA)
CHEST – PAIN

CHEST – PALPITATIONS
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)
EXTREM, LOWER, DIGITS – CLUBBING
EXTREM, UPPER, BILAT, DIGITS – CLUBBING
HEART, LSB, LOWER – MURMUR, DIAS
HEART, LSB, LOWER – MURMUR, SYS
HEART, LSB, LOWER – THRILL, SYS
HEART, LSB, MID – IMP, SYS
HEART, P2, INTENSITY – DECR/ABSENT
HEART, RSB, LOWER – PERCUSSION, DULLNESS
HEART, S1, SPLIT – WIDE
HEART, S2 – SINGLE
HEART, S2, SPLIT – FIXED
HEART, S2, SPLIT – WIDE
HEART, S3 RV
HEART, S4 RV
HEART, T1, INTENSITY – INCR
LIVER – ENLARGED (HEPATOMEGALY)
NECK, JVP – ELEV [3]
SKIN, COLOR – BLUE (CYANOSIS)

Differentiation

ASD
PAH [4]

Complications

Dysrhythmias [2]
HF
Peripheral embolus
SCD

Laboratory

NS

ECG

AV COND – 1ST DEGREE BLOCK
DYSRHY – PREEXCITATION [1]
DYSRHYTHMIAS – ATRIAL (PACS/OTHERS)
P WAVE – TALL/PEAKED
QR V1-4
QRS – RBBB/RBBB PATTERN
T WAVE – INVER, ABN

Imaging

AORTA, ASCEND, SIZE – DECR
PA, MAIN, SIZE – DECR
RA, CHAMBER, SIZE – INCR
RV, CHAMBER, SIZE – INCR
TV, FLOW – REGURG

Genomics

CSX
NKX2.5
TBX5

Other Tests

Ambulatory ECG monitoring
EP testing
Exercise testing

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic

Anticoagulation

- AF
- History of paradoxical embolus

IE prophylaxis: cyanotic and post-valve replacement patients

Treatment: Surgical/Invasive

Catheter ablation of accessory conduction pathways
Surgery:

- Closure of intra-atrial communications
- Division of accessory conduction pathways
- TV repair
- TV replacement

Prevention

NA

Course

Highly variable according to abnormalities but persons with mild forms generally have excellent long-term prognosis

Notes

[1] Type B
[2] Especially AF/Flutter
[3] Even with severe TR may be normal due to large and compliant RA
[4] Rare in Ebstein, but may be erroneously diagnosed in Ebstein patients with cyanosis

Guidelines

ACC/AHA 2008 guidelines for the management of adults with congenital heart disease
J Am Coll Cardiol. 2008;52:e143–263. http://content.onlinejacc.org/article.aspx?articleid=1188032#tab1.

ESC guidelines for the management of grown-up congenital heart disease (new version 2010)
Eur Heart J. 2010; 31:2915–57. http://eurheartj.oxfordjournals.org/content/ehj/31/23/2915.full.pdf.

Patient Information

Images

http://www.nlm.nih.gov/medlineplus/ency/imagepages/19884.htm.

Medlineplus

ENGLISH

http://www.nlm.nih.gov/medlineplus/ency/article/007321.htm.

ESPANOL

http://www.nlm.nih.gov/medlineplus/spanish/ency/article/007321.htm.

Genetics Home Reference

http://ghr.nlm.nih.gov/glossary=ebsteinanomaly.

Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/ebsteins-anomaly/basics/definition/con-20033605.

Texas Heart Institute

http://www.texasheart.org/HIC/Topics/Cond/ebstein.cfm.

Cincinnati Childrens-Ebsteins in Children

http://www.cincinnatichildrens.org/health/e/ebstein/.

AHA

http://www.heart.org/HEARTORG/Conditions/CongenitalHeartDefects/AboutCongenitalHeartDefects/Ebsteins-Anomaly-UCM-307025-Article.jsp.

Boston Childrens

http://www.childrenshospital.org/conditions-and-treatments/conditions/ebsteins-anomaly.

Cleveland Clinic

http://my.clevelandclinic.org/services/heart/disorders/congenital-heart/Ebsteins-Anomaly.

Memorial Hermann Childrens

http://childrens.memorialhermann.org/services/ebstein-s-anomaly/?gclid=CjwKEAjw1f6vBRC7tLqO-aih5WISJAAE0CYwj0kSP2edF4DwJTBjl2aLdCWbu2zNG-LCUPEswuuOMhoCN17w-wcB.

Ebsteins Anamoly Foundation

http://www.ebsteinsanomaly.org/what-is-ea.html.

Professional Information

Review (Adult)

Circulation. 2007;115:277–85. http://circ.ahajournals.org/content/115/2/277.full.

ECG/Echo: Case Report

Am J Med. 2015;128:367–68. http://www.amjmed.com/article/S0002-9343(14)01085-7/fulltext.

Presentation/Outcome

J Am Coll Cardiol. 1994;23:170–6. http://content.onlinejacc.org/article.aspx?articleid=1119154.

Exercise Tolerance

J Am Coll Cardiol. 1997;29:1615–22. http://content.onlinejacc.org/article.aspx?articleid=1121770.

Image

N Engl J Med. 2005;353:e1. http://www.nejm.org/doi/full/10.1056/NEJMicm031104.

Mortality: Risk Factors

Eur Heart J. 2007;28:2661–6. http://eurheartj.oxfordjournals.org/content/28/21/2661.

LV Noncompaction/Bicuspid AV

J Am Coll Cardiol. 2010;56:899–9. http://content.onlinejacc.org/article.aspx?articleid=1143125.

Updates and More

https://clinicalguidecvd.com/ebstein

Chapter 47
Endomyocardial Fibrosis (Davies Disease)

Management Keys

Differentiate from potentially treatable disease (e.g., Constrictive Pericarditis, Rheumatic Valve Disease)

ICD-10 Code

I42.3

Alternate Names/Abbreviation

EMF
DAVIES DISEASE

Description/Etiology

Etiology unproven
Idiopathic form of Restrictive Cardiomyopathy
Most common form of Restrictive Cardiomyopathy in world
Familial occurrence (undetermined whether genetic/environmental)

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 623
DOI 10.1007/978-1-4471-7293-2_47,

Resembles Loefflers Disease outside tropics
Occurs as rare complication of Behcet Disease

Comorbid Conditions

BEHCET DISEASE

Demography

Common in equatorial (Sub-Saharan) Africa, South America, India, China; rare outside tropics [1]
All ages; mostly impoverished children and young adults with bimodal peaks at ages 10/30 years, especially females in reproductive years
Gender equal in children; F 2:1 in adults in some reports

Pathophysiology

Subendocardial/inner 1/3 of myocardium dense fibrosis of LV/RV apices/inflow tracts and fibrosis of AV valves
Involvement may be biventricular (usual) or predominantly in either ventricle (usually RV), with corresponding atrial, valvular, hemodynamic changes, especially restriction of ventricular filling
MR/TR caused by scar tissue binding of leaflet(s)

Signs/Symptoms

ABDOMEN – FLUID (ASCITES) [MAY BE MASSIVE]
ARTERIAL PULSE, AMP – ALTERNATING (PULSUS ALTERNANS)
ARTERIAL PULSE, AMP – DECR/ABS
BREATH SOUNDS – CRACKLES (RALES)
BREATHING – DIFF (DYSPNEA)
BREATHING–DIFF,RECLININGFLAT(ORTHOPNEA)
CHEST – PAIN

CHEST, ANT, RSB – PULSATION [DUE TO ENLARGED RA]
EXTREM, DIGITS – CLUBBED
EXTREM, LOWER, BILAT – EDEMA [OFTEN ABSENT]
EYES – PROMINENT (EXOPHTHALMOS/PROPTOSIS)
FACE, SHAPE – ROUND (MOON) [FACIAL EDEMA]
FATIGUE
HEART, LSB, LOWER – MURMUR, SYS [3] [7] [TR, INCR WITH RESP]
HEART, LV, APEX – MURMUR, SYS [3] [7] [MR]
HEART, LV, APEX – OPENING SNAP [6] [DELAYED]
HEART, RATE – RAPID (TACHYCARDIA)
HEART, S3 LV
HEART, S3 RV
HEART, S4 LV
HEART, S4 RV
HEART, SOUNDS, INTENSITY – DECR
HYPOTENSION (BLOOD PRESSURE – DECREASED/LOW)
LIVER – ENLARGED (HEPATOMEGALY)
LIVER – PULSATION, SYS
MUSCLES – ATROPHY
MUSCLES – WEAK
NECK, JVP – ELEV
NECK, JVP – INSP RISE (KUSSMAUL SIGN)
NECK, JVP, A WAVE – INCR/LARGE (CANNON WAVE)
NECK, JVP, V WAVE – INCR/LARGE
NECK, JVP, X DESCENT – PROMINENT/RAPID
NECK, JVP, Y DESCENT – RAPID
THROAT – PAIN/TIGHTNESS

Differentiation

Other forms of Restrictive Cardiomyopathy [10]
Cancer infiltration of heart
Constrictive Pericarditis [11]

Ebstein Anomaly
Loeffler Endocarditis
Rheumatic Valve Disease

Complications

HF
Sudden Death

Laboratory

BLOOD, EOSINOPHILES – INCR [30 % OF CASES]

ECG

AV COND – 1ST DEGREE BLOCK [8]
DYSRHYTHMIAS – ATRIAL (PACS/OTHERS) [2]
DYSRHYTHMIAS – VENTRICULAR (PVCS/OTHERS) [USUALLY ONLY PVCS]
P WAVE – BIFID
P WAVE – TALL/PEAKED
QRS – LONG, NS
QRS – LVH PATTERN
QRS – RVH PATTERN
QRS, AXIS – L
QRS, AXIS – R
ST SEGMENT – DEPR [LAT LEADS]
ST-T WAVE – ABN, NS
T WAVE – INVER, ABN [LAT LEADS]
VOLTAGE, GEN – DECR [DUE TO PERICARD EFFUSION]

Imaging [9]

[ECHO MOST WIDE-USED DX TEST]
ABDOM – FLUID (ASCITES)
CARDIOMEGALY

IVS, MOTION – PARADOX
LA, CHAMBER, SIZE – INCR
LV, CHAMBER, APEX – OBLITERATED [4]
LV, INTRACAVITY – MASS [5]
LV, WALL MOTION, BASAL – HYPERKINETIC (MERLON SIGN)
MV, FLOW – DECR
MV, FLOW – REGURG [3]
MV, LEAFLETS – THICK
MV, POST LEAFLET – DECR/ABSENT
PERICARD – FLUID
PLEURA – FLUID
PUL, VASCULARITY – INCR
PV – OPENING, DIAS
RA, CHAMBER, SIZE – INCR
RV, APEX – RETRACTION
RV, CHAMBER, APEX – OBLITERATED [4]
RV, CHAMBER, SIZE – DECR
RV, INTRACAVITY – MASS [5]
RV, OUTFLOW TRACT, SIZE – INCR
TV – ADHERENT TO ENDOCARD
TV, FLOW – REGURG [3]
TV, LEAFLETS – THICK
VENA CAVA, INF, SIZE – INCR

Other Tests

EMB: usually not necessary and often technically difficult
Card Catheterization: not routine as noninvasive tests usually adequate

RV findings:

Equal pressures in RA, RV, PA
Flat RV apex
Smooth trabeculae
Free TV reflux
Large RA
Dilated SVC
Active infundibulum

LV findings:

LVEDP increased (marked) with dip/plateau pattern
Variable degrees of pulmonary hypertension
LV apex obliterated
MR
Normal to severely abnormal LV wall motion depending on site/severity of EMF

Treatment: Nonpharmacologic

Heart Failure protocol

Treatment: Pharmacologic

Heart Failure protocol

Treatment: Surgical/Invasive

Pericardiocentesis
Pleurocentesis
Paracentesis
Endocardectomy with associated AV replacement [12]

Course

Unknown prior to advanced stage due to lack of data; high mortality in patients with HF

Notes

[1] Countries with highest incidence: Uganda, Brazil, Columbia, Cote D'ivoire, Nigeria (Southern), Mozambique (costal), India (Kerala State); China

(Guangxi Province); recent marked decreased incidence in Nigeria, especially Southwest region
[2] Especially AF, often with slow ventricular response; other dysrhythmias include junctional rhythm
[3] MR/TR due to adhesion of valve apparatus to ventricular wall
[4] Fibrotic obliteration/negative contrast of either/both ventricular apex highly characteristic of EMB
[5] Due to thrombus; preserved normal wall motion characteristic in EMF
[6] Due to abnormal anterior MV leaflet movement
[7] Murmurs typically soft/absent
[8] Advanced heart block rare
[9] MRI: gold standard for diagnosis but not generally available in endemic areas; CT also useful when available
[10] Especially apical hypertrophic form of HCM
[11] By echo, in EMF: normal pericardial thickness, markedly dilated atria, large A waves, early diastolic dip, reversed diastolic pressure gradient across PV
[12] Technically complex, associated with high morbidity/mortality

Guidelines

NS

Patient Information

NORD

https://www.rarediseases.org/rare-disease-information/rare-diseases/byID/232/viewAbstract.

Professional Information

Early Description

East Afr Med J. 1948;25:10. (NO INTERNET ACCESS).

Review

Heart. 2013;99:1481–7. http://heart.bmj.com/content/99/20/1481?etoc.

Case Reports

J Am Coll Cardiol. 1985;5:983–8. http://content.onlinejacc.org/article.aspx?articleid=1111141&resultClick=3.

Diagnostic Pitfalls

Chest. 2005;28:3985–93. http://journal.publications.chestnet.org/article.aspx?articleID=1084084.

ECG

Br Heart J. 1960;22:311–5. http://www.ncbi.nlm.nih.gov/pmc/articles/PMC1017661/.

Epidemiology/Etiology

PLoS Negl Trop Dis. 2008;2:E97. http://www.plosntds.org/article/info%3Adoi%2F10.1371%2Fjournal.pntd.0000097.

Images

J Am Coll Cardiol. 2010;55:e137. http://content.onlinejacc.org/article.aspx?articleid=1142833&resultClick=3.

India

Indian J Med Res. 2012;136:729–38. http://www.ncbi.nlm.nih.gov/pmc/articles/PMC3573593/.

Mozambique

N Engl J Med. 2008;359:43–9. http://www.nejm.org/doi/full/10.1056/NEJMoa0708629.

Surgery

J Am Coll Cardiol. 1990;16:11245–251. http://content.onlinejacc.org/article.aspx?articleid=1115731&resultClick=3.

Ebstein Anomaly/Behcet

Mod Rheumatol. 2014;24:532–36. http://informahealthcare.com/doi/abs/10.3109/14397595.2013.874731.

Updates and More

https://clinicalguidecvd.com/emf

Chapter 48
Erdheim-Chester Disease

ICD-10 Code

E88.89

Alternate Names/Abbreviation

ECD

Description/Etiology

Non-Langerhans histiocytic multisystem disorder primarily involving skeleton and also CV system, CNS, lungs, glands, retroperitoneum, kidneys, other tissue
Cardiac manifestations include:

- AR
- Coronary artery perifibrosis
- Heart block
- MR
- Pericardial fibrosis
- RA pseudotumors

Cause unknown

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_48,

Comorbid Conditions

LANGERHANS HISTIOCYTOSES [2]
ROSAI-DORFMAN DISEASE [2]

Demography

Onset age 40–60 years
M 3:1
Global

Pathophysiology

Multiorgan tissue infiltration by lipid-containing histiocytes; effects determined by location and extent of involvement

Signs/Symptoms [1]

ABDOMEN – FLUID (ASCITES)
ABDOMEN – PAIN, AFTER MEALS (ABDOMINAL ANGINA)
BLOOD PRESSURE, ARTERIAL – INCREASED/ELEVATED [9]
BONE, LOCAL – PAIN [3]
BREATH SOUNDS – CRACKLES (RALES)
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)
COUGH
EXTREM, LOWER, BILAT – EDEMA
EYES – PROMINENT (EXOPHTHALMOS/PROPTOSIS) [6]
EYES – XANTHELASMAS
EYES, RETINA – PAPILLEDEMA
EYES, VISION – DECR/LOSS
FEVER [10]
HEADACHE

HEART, LSB, MID – MURMUR, DIAS
HEART, LV, APEX – MURMUR, SYS
HEART, SOUND – EARLY DIAS
HEART, SOUNDS, INTENSITY – DECR [5]
LIVER – ENLARGED (HEPATOMEGALY)
NECK, JVP – ELEV
NERVOUS SYSTEM – MULTIPLE SIGNS/SYMPTOMS [14]
SEIZURES
SKIN – RASH, MACULAR/PAPULAR [8]
THIRST – INCR [7]
URINATION – INCR (POLYURIA) [7]
WEIGHT – LOSS

Differentiation

Other causes of HF
Other causes of Osteosclerosis
Other causes of Pericardial Effusion
Other causes of right heart masses/tumors
Other causes of Vasculitis

Complications

Acute pulmonary edema [4]
AMI
Cardiac Tamponade
Diabetes Insipidus
DVT
Heart Block
HF
Hydronephrosis
Pulmonary Embolism
Pulmonary Hypertension
RAS
Stroke
Sudden Death

Laboratory

BLOOD, ALK PHOS – INCR
BLOOD, C-REACTIVE PROTEIN (CRP) – INCR
BLOOD, ESR – INCR
BLOOD, IL-6 – INCR
BLOOD, SODIUM – INCR

ECG

PR INTERVAL – SHORT
Q WAVE – ABN
QRS – LVH PATTERN
RATE – DECREASED (SINUS BRADYCARDIA)
SINOAURICULAR BLOCK
ST SEGMENT – ELEV [10]
ST-T WAVE – ABN, NS

Imaging [13]

AORTA, ARCH, SIZE – INCR
AORTA, PERIAORTA, TISS – FIBROSIS [11]
AORTA, ROOT, SIZE – INCR
ART, PERIVASC, TISSUE – FIBROSIS [12]
AV SULCUS, R – INFILTRATE
CARDIOMEGALY
MV, FLOW – REGURG
PERICARD – FLUID
PERICARD – THICK
RA, CHAMBER, SIZE – INCR
RA, INTRACAVITY – MASS

Other Tests

Tissue biopsy

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic [13]

Chemotherapy
Corticosteroids
Interferon alpha
Radiation

Treatment: Surgical/Invasive [13]

Variable

Prevention

NS

Notes

[1] Only CV and most common non-cardiac features listed; due to extreme heterogeneity, many other features have been reported
[2] Other forms of Histiocytoses occur more often than chance and should be sought in patients with ECD
[3] Most common symptom; especially distal LE; often mild; onset any time in course of disorder
[4] Due to MR
[5] Due to pericardial effusion/thickening
[6] Unilateral or bilateral
[7] Due to Diabetes Insipidus
[8] NS; may be pruritic
[9] Often resistant when due to RAS secondary to retroperitoneal fibrosis/direct renal infiltration

[10] Uncommonly reported
[11] In some patients described as "coated aorta"; may involve all or some segments of abdominal/thoracic aorta
[12] Involved arteries include carotids, basilar, vertebral, subclavian, main PA and branches, coronary, mesenteric, renal
[13] Highly variable depending on sites/tissue involvement
[14] Especially cerebral, cerebellar, and spinal cord involvement

Guidelines

NS

Patient Information

ECD.ORG

http://www.erdheim-chester.org/.

Histiocyte Society

http://www.histiocytesociety.org/.

Professional Information

Histiocyte Society

http://www.histiocytesociety.org/.

Imaging Brain/Heart

J Am Coll Cardiol. 2014;63:1121–1. http://content.onlinejacc.org/article.aspx?articleid=1827544&resultClick=24.

Interferon Alpha and Cardiac Involvement

J Am Coll Cardiol. 2011;58:2695–5. http://content.onlinejacc.org/article.aspx?articleid=1147836&resultClick=24.

Pericarditis

Circulation. 2008;118:e511–2. http://circ.ahajournals.org/content/118/14/e511.full.pdf+html?sid=5755f16d-a18b-4c38-be74-b37c089810d2.

Images

J Am Coll Cardiol. 2014;63:1121–1 http://content.onlinejacc.org/article.aspx?articleID=1827544.

Imaging Series

Circulation. 2009;119:e597–8. http://circ.ahajournals.org/content/119/25/e597.full?sid=5755f16d-a18b-4c38-be74-b37c089810d2.

Imaging/Pacemaker Insertion

Circulation. 2007;115:e412–4. http://circ.ahajournals.org/content/115/16/e412.full?sid=5755f16d-a18b-4c38-be74-b37c089810d2.

Pericarotid Fibrosis

Circulation. 2004;110:e443–4. http://circ.ahajournals.org/content/110/15/e443.full?sid=5755f16d-a18b-4c38-be74-b37c089810d2.

Imaging Atrial Pseudo-Mass

Eur Heart J. 2009;30:3063. http://eurheartj.oxfordjournals.org/content/30/24/3063.full?sid=bb1abc0e-f6fd-4bbb-9395-2105288c26e9.

Imaging Cerebral and Cardiac Mass

Eur Heart J. 2008;29:1929. http://eurheartj.oxfordjournals.org/content/29/16/1929.full?sid=bb1abc0e-f6fd-4bbb-9395-2105288c26e9.

Updates and More

https://clinicalguidecvd.com/ecd

Chapter 49
Fabry Disease (Alpha-Galactosidase A Deficiency)

ICD-10 Code

E75.21

Alternate Names/Abbreviation

ALPHA-GALACTOSIDASE A DEFICIENCY
ANDERSON-FABRY DISEASE
ANGIOKERATOMA CORPORIS DIFFUSUM
ANGIOKERATOMA DIFFUSE
CERAMIDE TRIHEXOSIDASE DEFICIENCY
GLA DEFICIENCY
HEREDITARY DYSTOPIC LIPIDOSIS

Description/Etiology

X-Chromosome linked genetic Lysosomal Storage Disorder involving heart and other systems including:

Dermatological
Gastrointestinal
Neurological
Renal

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_49,

Comorbid Conditions

NS

Demography

Both genders, affecting males earlier, but with comparable degrees of severity over time

Pathophysiology

Multiorgan cell damage due to accumulation of globotriaosylceramide (GL-3) caused by inadequate breakdown by enzyme alpha-galactoside

Auditory:

- Cochlear vessel narrowing/occlusion
- Ischemic Neuropathy

Cardiovascular:

- Coronary artery atherosclerosis/stenosis/vasospasm
- LVH/progressive myocardial fibrosis (strong predictor of outcome) [23]
- Thrombosis/Thromboembolism

Dermatologic:

- Epidermal capillary wall weakening/vascular ectasia
- Small vessel narrowing around eccrine sweat glands

Gastrointestinal:

- Mesenteric small vessel narrowing

Neurologic: ischemic injury (prothrombotic/occlusive) and metabolic abnormalities causing: [2]

- Ischemic small vessel multifocal leukoencephalopathy
- Large vessel ectasia

Neuronal cell functional disruption
Small myelinated/unmyelinated fiber loss

Ophthalmologic:

Central retinal artery occlusion
Conjunctival/retinal vasculopathy
Corneal epithelial streaks
Lacrimal hyposecretion

Pulmonary:

Airway narrowing
Capillary blockage

Renal:

Glomerular Sclerosis
Interstitial Fibrosis
Tubular atrophy

Signs/Symptoms [1]

ABDOMEN – PAIN, AFTER MEALS (ABDOMINAL ANGINA)
APPETITE, SATIETY – EARLY
BLOOD PRESSURE, ARTERIAL – INCREASED/ELEVATED
BODY, SENSATION, COLD – INTOLERANCE
BODY, SENSATION, HEAT – INTOLERANCE
BOWEL MOVEMENTS – DIARRHEA [19]
BREATH SOUNDS – WHEEZES
BREATHING – DIFF (DYSPNEA) [9]
CHEST – PAIN, EFFORT (ANGINA PECTORIS)
CHEST – PALPITATIONS
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE) [8]
COUGH
DIZZY/SPINNING (TRUE VERTIGO)
EARS – RINGING (TINNITUS)
EARS, HEARING – LOSS (DEAFNESS)

EXERCISE, TOLERANCE – DECR
EXTREM, ANKLES – SWOLLEN
EXTREM, FEET – PAIN [2][3]
EXTREM, HANDS – PAIN [2][3]
EXTREM, HANDS/FEET – NUMB [2]
EXTREM, HANDS/FEET – TINGLING [2]
EYES, CORNEA – STREAKS (VERTICILLATA) [7]
EYES, VISION, LIGHT SENSITIVITY – INCR (PHOTOPHOBIA)
FACE – DYSMORPHIC
FATIGUE
HEADACHE
HEART, LSB, MID – MURMUR, DIAS [11]
HEART, LV, APEX – MURMUR, SYS [12]
HEART, RATE – SLOW (BRADYCARDIA) [10]
HEART, RHYTHM – IRREG
HEART, S1, INTENSITY – DECR [13]
HEART, S1, INTENSITY – INCR [13]
HEART, S1, INTENSITY – VAR [13]
HEART, S2, SPLIT – REVERSED (PARADOXICAL) [13]
HEART, S3 LV
HEART, S4 LV
JOINTS – PAIN (ARTHRALGIA)
MUSCLES – WEAK
NAUSEA
SKIN – RASH, RED-PURPLE (ANGIOKERATOMA) [6]
SWEATING – DECR (ANHIDROSIS) [5]
VOMITING (EMESIS)

Differentiation

Atherosclerotic CAD [14]
Cerebrovascular disease
Collagen vascular disease
Familial Globotriaosylceramide-associated Cardiomyopathy [25]

Hypertrophic Cardiomyopathy
Other causes of multi-system disease

Complications

AF
AMI
Body Overheating [4]
Cataracts [7]
Heart Block
HF
Malignant Ventricular Arrhythmias
Osteoporosis/Osteopenia
Renal Failure
Stroke/TIA [may occur at young age]

Laboratory [18]

BLOOD GB3 – INCR
BLOOD, GALACTOSIDASE – DECR [MALES]
URINE, GB3 – INCR
URINE, PROTEIN – INCR (PROTEINURIA)

ECG

AV COND – 1ST DEGREE BLOCK
AV COND – 2ND DEGREE BLOCK, MOBITZ II
AV COND – 3RD DEGREE BLOCK
DYSRHY – PREEXCITATION
DYSRHYTHMIAS – ATRIAL (PACS/OTHERS)
DYSRHYTHMIAS – VENTRICULAR (PVCS/OTHERS)
PQ INTERVAL – SHORT [22]
PR INTERVAL – SHORT [22]
QRS – LBBB/LBBB PATTERN
QRS – LVH PATTERN

QRS, AMP – INCR
RATE – DECREASED (SINUS BRADYCARDIA)
ST SEGMENT – DEPR
T WAVE – INVER, ABN

Imaging

AV, LEAFLETS – THICK
CARDIOMEGALY
IVS, THICKNESS – INCR (SEPTAL HYPERTROPHY) [15]
LA, CHAMBER, SIZE – INCR
LV, CHAMBER, SIZE – INCR
LV, MYOCARD – LGE [ESP POST LAT]
LV, MYOCARD, WALL THICKNESS – INCR (HYPERTROPHY) [15]
MV, FLOW – REGURG
MV, LEAFLETS – THICK
MV, LEAFLETS, MOTION – PROLAPSE
RA, CHAMBER, SIZE – INCR

Genomics

GLA

Other Tests

Ambulatory ECG monitoring [23]
Cardiac EP
EEG
Mutational analysis [21]
Slit lamp: corneal whorling
Others (as indicated):

Audiometry
Bone mineral density
Endoscopy
Renal function
Retinal function
Spirometry
Tear secretions
Visual

Treatment: Nonpharmacologic [24]

Hearing aid
Tobacco cessation

Treatment: Pharmacologic [17] [24]

Specific: enzyme replacement therapy
Antianginals [16]
Anticoagulation
Antihypertensives
RAAS blockade
Statins

Treatment: Surgical/Invasive [17] [24]

Pacemaker
Septal alcohol ablation

Prevention

Genetic counseling for family planning

Notes

[1] Symptoms highly variable in individuals and from person to person
[2] Neuropathic pain: hallmark of this disease, likely due to dorsal nerve root nerve fiber degeneration in dorsal root ganglion cells associated with axonal degeneration of small fibers in pain pathways ; may be triggered by heat, weather change, stress, exercise, fatigue
[3] May spread to other parts of body, lasting minutes-days, termed "Fabry crises"
[4] Due to anhidrosis
[5] Due to nerve/sweat gland damage
[6] Highly characteristic of this disease; most often between navel and knees and on elbows and knees; usually initially appear in adolescence and may enlarge with age; Fabry diagnosis should be considered in patients with diffuse appearance of angiokeratomas
[7] Yellow, gray, brown; vision not affected
[8] Due to heart block or less often LVOT obstruction
[9] Due to LV diastolic dysfunction
[10] Due to involvement of sinus node/AV conduction
[11] AR
[12] MR
[13] Variable according to whether preexcitation/heart block present
[14] Conditions may coexist, but myocardial insufficiency can occur in absence of epicardial coronary artery obstruction
[15] LVH may be symmetrical or asymmetrical
[16] Beta-blockers should be used with caution due to conduction abnormalities
[17] As indicated in individual patients
[18] Secondary abnormalities highly variable depending on specific organ involvement
[19] Most common GI complaint
[20] Unreliable in females
[21] Confirmatory for diagnosis of Fabry Disease
[22] Short PR/PQ in absence of preexcitation: characteristic of Fabry; mechanism uncertain

[23] Use regular ambulatory ECG monitoring to detect malignant ventricular arrhythmias, which correlate with myocardial fibrosis/LGE
[24] In addition to other adjunctive treatments for Fabry-related comorbidities
[25] Rare Gb3-assoociated cardiomyopathy; autosomal recessive inheritance likely; genetic and metabolic causes unknown

Guidelines

American College of Medical Genetics and Genomics

Genet Med. 2006;8:539–48. http://www.nature.com/gim/journal/v8/n9/full/gim200691a.html.

Patient Information

Medline Plus-Genetic Brain Disorders

ENGLISH

https://www.nlm.nih.gov/medlineplus/geneticbraindisorders.html.

ESPANOL

https://www.nlm.nih.gov/medlineplus/spanish/geneticbraindisorders.html.

Fabry Disease Foundation

http://www.fabrydisease.org/.

Genetics Home Reference

http://ghr.nlm.nih.gov/condition/fabry-disease.
Professional Information

Review

Circulation. 2014;130:1081–90 http://circ.ahajournals.org/content/130/13/1081.full?sid=b3b35701-fcf8-4814-ada1-0d218feb5436.

Review

Prog Cardiovasc Dis. 2010;52:326–35. http://dx.doi.org/10.1016/j.pcad.2009.11.002.

Angiokeratoma

http://dermatology.cdlib.org/114/NYU/NYUtexts/0419054.html.

Angiokeratoma

Br J Dermatol. 2012;166:712–20. http://www.ncbi.nlm.nih.gov/pubmed/22452439?dopt=Abstract.

Angina Pectoris/Coronary Small Vessel Disease

Circulation Heart Failure. 2008;1:161–9. http://circheartfailure.ahajournals.org/content/1/3/161.full.

Angina Pectoris/Coronary Small Vessel Disease

Circulation. 2014;130:1530–1. http://circ.ahajournals.org/content/130/17/1530.full.

Cardiac Troponin I: Persistent Increase

BMC Cardiovasc Dis. 2011;11:6. http://www.biomedcentral.com/1471-2261/11/6.

Clinical Manifestations

J Med Genet. 2001;38:750–60. http://jmg.bmj.com/content/38/11/750.long.

Clinical Predictors

Heart. 2015;101:961–6. http://heart.bmj.com/content/101/12/961?etoc.

Complete AV Block

Heart. 2003;89:e2. doi:10.1136/heart.89.1.e2 http://heart.bmj.com/content/89/1/e2.full#ref-6.

Early ECG Changes

Heart. 2011;97:485–90. http://www.ncbi.nlm.nih.gov/pubmed/21270075.

Enzyme Replacement Therapy

Circulation. 2003;108:1299–301. http://circ.ahajournals.org/content/119/4/524.full?sid=315d335a-edbe-4a2f-8645-f363e9cd07d6.

Familial Globotriaosylceramide-Associated Cardiomyopathy

Heart. 2014;100:1793–8. http://heart.bmj.com/content/100/22/1793.abstract.

Kidney Disease and Outcomes

Heart. 2015;101:287–93. http://heart.bmj.com/content/101/4/287.abstract.

Mimic of Hypertrophic Cardiomyopathy

J Am Coll Cardiol. 2014;63:e43–3. http://content.onlinejacc.org/article.aspx?articleID=1842649.

Mimic of Hypertrophic Cardiomyopathy

Heart. 2003;89:929–30. http://www.ncbi.nlm.nih.gov/pmc/articles/PMC1767790/.

Natural History: Females

J Med Genet. 2006;43:347–52. http://jmg.bmj.com/content/43/4/347.abstract?ijkey=49bcb066248659b8c1049ed467812fa2523097e1&keytype2=tf-ipsecsha.

Neurological Complications

Curr Pharm Des. 2013;19:6014–30 http://www.ncbi.nlm.nih.gov/pubmed/23448452.

Myocardial Fibrosis/Outcomes

Am J Cardiol. 2104;114:895–900. http://www.sciencedirect.com/science/article/pii/S0002914914013666.

Progression Despite Enzyme Replacement Therapy

Circulation. 2013;128:1687–8. http://circ.ahajournals.org/content/128/15/1687.full.

Short PR Interval

Am J Cardiol. 1994;74:203–4. http://www.sciencedirect.com/science/article/pii/0002914994901066#.

Short PR Interval

Pacing Clin Electrophysiol. 2008;31(6):782–3. http://www.ncbi.nlm.nih.gov/pubmed/18507557.

Updates and More

https://clinicalguidecvd.com/fabry

Chapter 50
Fibromuscular Dysplasia

ICD-10 Code

I77.3

Alternate Names/Abbreviation

FMD
FIBROMUSCULAR HYPERPLASIA

Description/Etiology

Nonatheroscertic noninflammatory vascular disease associated with arterial stenosis, occlusion, aneurysm, dissection
Most often involves renal arteries, extracranial carotid and vertebral arteries; usually multivascular
Veins not involved
Cause unknown

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_50,

Comorbid Conditions

HYPERTENSION – SYSTEMIC ARTERIAL
SPONTANEOUS CORONARY ARTERY DISSECTION [12]

Demography

F 9:1 [11]

Pathophysiology

Ischemia/infarction of organs supplied by involved arteries

Signs/Symptoms [4] [11]

ABDOMEN – BRUIT
ABDOMEN – PAIN
ABDOMEN – PAIN, AFTER MEALS (ABDOMINAL ANGINA)
ARTERY, CAROTID – BRUIT
BLOOD PRESSURE, ARTERIAL – INCREASED/ELEVATED
BREATHING – DIFF (DYSPNEA)
CHEST – PAIN
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)
DIZZY/LIGHTHEADED/PRESYNCOPE
EARS – RINGING (TINNITUS)
EARS – RINGING, PULSATILE (PULSATILE TINNITUS)
EARS, HEARING – LOSS (DEAFNESS)
EXTREM, LOWER, BILAT – PAIN, EFFORT (CLAUDICATION)

EYES, VISION, MONOCULAR – DECR/TOTAL LOSS, TRANSIENT (AMAUROSIS FUGAX)
FACE – HORNER SYNDROME
FLANK – PAIN
HEADACHE [1]
NECK – PAIN
SEIZURES
WEIGHT – LOSS

Differentiation

Atherosclerotic CV disease [3]
Other causes of Systemic Arterial Hypertension

Complications

AMI
Arterial Aneurysm
Arterial Dissection
Chronic renal disease/Renal Failure
Mesenteric Thrombosis
Spontaneous Coronary Artery Dissection [12]
Sudden Death
Stroke/TIA [9]

Laboratory

NS
ECG
N/NS ABN

Imaging

ART, CAROTID, INT – TORTUOUS [5]
ART, CAROTID, INT, APPEARANCE – BEADED [5]

ART, CAROTID, INT, FLOW – TURBULENT
ART, RENAL – TORTUOUS [7]
ART, RENAL, FLOW – OBS [6] [7]
ART, RENAL, FLOW – TURBULENT [7]
ART, VERTEB, FLOW – TURBULENT

Other Tests

Arteriography [8]

Treatment: Nonpharmacologic

NS
Atherosclerotic risk factor modification

Treatment: Pharmacologic

NS
Antihypertensives (especially RAAS inhibitors for hypertension)
Antiplatelets for ischemic events
Anticoagulants for ischemic events

Treatment: Surgical/Invasive

Renal arterial revascularization considered for:

- Aneurysm
- Branch renal arterial disease
- Dissection
- Preservation of renal function
- Resistant hypertension
- Short duration hypertension (for cure)

Carotid/vertebral revascularization: indicated only for symptomatic disease

Course

Highly variable

Notes

[1] Affects >50 % and migraine-like in one-half; may occur daily
[2] Described as “swishing” or “swooshing” sound in ears
[3] May occur concomitantly
[4] FMD may be detected as an incidental angiographic finding in patients with no clinical manifestations of FMD
[5] Mid-distal internal carotid artery
[6] Concentric or tubular
[7] Mid-distal
[8] Angiography demonstrating "beaded" appearance" (gold standard for diagnosis)
[9] Cerebral aneurysm, subarachnoid hemorrhage
[10] Systemic Arterial Hypertension and headache most common clinical manifestations, both occurring in >50 % of pts
[11] Sexes differ in clinical features:

Females more often have extracranial cerebrovascular manifestations, including:

Cervical bruit
Neck pain
Pulsatile tinnitus

Males more often have:

Arterial aneurysm
Arterial dissection
Focal disease

[12] Spontaneous Coronary Artery Dissection appears to be primary coronary arterial manifestation of FMD

Guidelines

Management of patients with peripheral artery disease (Compilation of 2005 and 2011 ACCF/AHA Guideline Recommendations)
J Am Coll Cardiol. 2013;61:1555–70. http://content.onlinejacc.org/article.aspx?articleid=1659662.

ESC Guidelines on the diagnosis and treatment of peripheral artery diseases
Eur Heart J. 2011;32:2851–906. http://eurheartj.oxfordjournals.org/content/32/22/2851.full?sid=d4dea8fc-0852-4cde-b488-374f010a51f9.

2013 ESH/ESC Guidelines for the management of arterial hypertension
Eur Heart J. 2013:34;2206. http://www.escardio.org/guidelines-surveys/esc-guidelines/Pages/arterial-hypertension.aspx.

ACR appropriateness criteria renovascular hypertension
http://www.guideline.gov/content.aspx?id=37941&search=fibromuscular+hyperplasia#Section442.

Patient Information

Images

https://www.nlm.nih.gov/medlineplus/spanish/ency/espimagepages/8818.htm.

Medlineplus-Renovascular Hypertension

ENGLISH

https://www.nlm.nih.gov/medlineplus/ency/imagepages/8818.htm.

ESPANOL

https://www.nlm.nih.gov/medlineplus/spanish/ency/article/000204.htm.

AHA

Circulation. 2012;125:e636–9. http://circ.ahajournals.org/content/125/18/e636.full.

NCBI

http://www.ncbi.nlm.nih.gov/sites/ga?disorder=fibromuscular%20dysplasia.

Mayo Clinic

ENGLISH

http://www.mayoclinic.org/diseases-conditions/fibromuscular-dysplasia/basics/definition/con-20034731.

ESPANOL

http://www.mayoclinic.org/espanol.

Professional Information

AHA Scientific Statement

Circulation. 2014;129:1048–78. http://circ.ahajournals.org/content/129/9/1048.full?sid=3838ff96-440b-4dd5-ace8-36f8110404e6.

Recent Developments (2014)

J Am Heart Assoc. 2014;3:e001259. http://jaha.ahajournals.org/content/3/6/e001259?etoc.

Clinical Manifesations: Sex Variation

J Am Coll Cardiol. 2013;62:2026–8. http://content.onlinejacc.org/article.aspx?articleID=1729174.

Coronary Artery Manifestations

J Am Coll Cardiol. 2014;64:1033–46. http://content.onlinejacc.org/article.aspx?articleid=1900746.

Coronary Artery Pathology

Am J Cardiol. 1990;65:12–22. http://www.sciencedirect.com/science/article/pii/000291499090954Y.

Extracoronary Vascular Abnormalities

Am J Cardiol. 2015;115:1672–7. http://www.ajconline.org/article/S0002-9149(15)00966-2/abstract.

Mesenteric Ischemia

J Interv Gastrointerol. 2012;2:19–21. http://www.ncbi.nlm.nih.gov/pmc/articles/PMC3655395/.

Postpartum Coronary Dissection (Case Report)

Eur Heart J. 2008;29:2825. http://eurheartj.oxfordjournals.org/content/29/22/2825.full?sid=62a9da7d-d37b-4249-a375-f7fef0b12ec1.

Renal Artery Imaging

Circulation. 2005;112:e278–9. http://circ.ahajournals.org/content/112/17/e278.full?sid=db49e3cc-4c6b-4029-9e89-79746604fccf.

Renal Artery Morphology in Renovasc HTN

Am J Med. 1967;43:97–112. http://www.sciencedirect.com/science/article/pii/0002934367901519.

Spontaneous Coronary Artery Dissection

Circ Cardiovasc Intervent. 2012;5:134–7. http://circinterventions.ahajournals.org/content/5/1/134.short.

Updates and More

https://clinicalguidecvd.com/fmd

Chapter 51
Giant Cell Myocarditis

ICD-10 Code

I40.1

Alternate Names/Abbreviation

GCM

Description/Etiology [12]

Nonischemic, rapidly progressive necrosis of cardiac myocytes with multinucleated giant cell, lymphocytic, and eosinophilic inflammatory infiltrate

Generally presumed an autoinflammatory disease, sometimes associated with and preceded by other chronic inflammatory disorders (see COMORBID CONDITIONS)

Comorbid Conditions [1] [2]

ALOPECIA TOTALIS VITILIGO
ARRHYTHMOGENIC RIGHT VENTRICULAR DYSPLASIA

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 665
DOI 10.1007/978-1-4471-7293-2_51,

ASYMMETRIC SEPTAL HYPERTROPHY
CHRONIC HEPATITIS
CROHN DIS
CRYOFIBRINOGENEMIA
DIABETES MELLITUS [INSULIN-DEPENDENT]
DRUG HYPERSENSITIVITY [3]
FIBROMYALGIA
HASHIMOTO THYROIDITIS
HEART TRANSPLANT [10]
HYPERTHYROIDISM
HYPOTHYROIDISM
LYMPHOMA
MYASTHENIA GRAVIS
OPTIC NEURITIS
ORBITAL MYOSITIS
PARVOVIRUS B19 INFECTION
PERNICIOUS ANEMIA
RHEUMATOID ARTHRITIS
SJOGREN SYNDROME
SYSTEMIC LUPUS ERYTHEMATOSUS
TAKAYASU ARTERITIS
THYMOMA [4]
ULCERATIVE COLITIS
WEGENER GRANULOMATOSIS

Demography

Age of clinical onset (mean) 42 years [5]
All ethnicities
Gender equal

Pathophysiology

LV myocardial necrosis and fibrosis causing LV dysfunction and atrioventricular heart block

EMB histology:

Eosinophils
Giant cells (early)
Interstitial Fibrosis
Lymphocytes

Signs/Symptoms

BREATHING – DIFF (DYSPNEA)
BREATHING – DIFF, RECLINING FLAT (ORTHOPNEA)
CHEST – PAIN [7] [8]
CHEST – PALPITATIONS
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)
EXTREM, LOWER, BILAT – EDEMA
FATIGUE

Differentiation

AMI [8]
Cardiac Sarcoidosis
Other causes of Myocarditis/Cardiomyopathy/HF
Other causes of heart block

Complications

Cardiac rupture/hemopericardium
Complete heart block [7]
HF [7] [12]
Infections
Renal failure [11]
Sudden death
VT [6] [7]

Laboratory

BLOOD, TROPONIN, CARD – INCR

ECG

AV COND – 1ST DEGREE BLOCK
AV COND – 3RD DEGREE BLOCK
DYSRHYTHMIAS – VENTRICULAR (PVCS/OTHERS) [9]
EPSILON WAVE
Q WAVE – ABN [8]

Imaging

IVS, THICKNESS – INCR (SEPTAL HYPERTROPHY)
LV, CHAMBER, SIZE – INCR
LV, EF – DECR
LV, MYOCARD, WALL THICKNESS – INCR (HYPERTROPHY)
LV, WALL THICKNESS, SEG – INCR
MYOCARD – FIBROSIS
RV, CHAMBER, SIZE – INCR

Other Tests

EMB

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic

Immunosuppression

Treatment: Surgical/Invasive [12]

LVAD
Cardiac transplant

Course

HF with cardiac transplant within 1 year of diagnosis often required
Post-cardiac transplant survival: about 70 % at 5 years
Recurrence common for several years after initial diagnosis

Notes

[1] Most cases not associated with other conditions; some conditions listed based on single case reports
[2] Some conditions, eg, Inflammatory Bowel Disease, may precede onset of GCM by several years
[3] Many drug classes reported, including antiinfectives, antiepileptics, vaccines, antihypertensives
[4] Also reported post-thymoma resection
[5] All ages reported, from infancy to >85 years
[6] Often sustained and refractory
[7] May be initial clinical manifestation
[8] Chest pain syndrome resembling AMI may occur
[9] Especially VT
[10] Many reports in transplants replacing GCM hearts and isolated reports of GCM in transplants for other causes
[11] Due to HF/chronic calcineurin use

[12] Excerpted from ACCF/AHA 2013 Guidelines for Management of Heart Failure (J Am Coll Cardiol 2013;62:e147-e239):

"Sec 5.6.1 Myocarditis

Giant cell myocarditis is a rare form of myocardial inflammation characterized by fulminant HF, often associated with refractory ventricular arrhythmias and a poor prognosis. Histologic findings include diffuse myocardial necrosis with numerous multinucleated giant cells without granuloma formation. Consideration for advanced HF therapies, including immunosuppression, mechanical circulatory support (MCS), and transplantation, is warranted."

Guidelines

2013 ACCF/AHA guideline for the management of heart failure
J Am Coll Cardiol. 2013;62:e147–239. http://content.onlinejacc.org/article.aspx?articleid=1695825.

ESC Guidelines for the diagnosis and treatment of acute and chronic heart failure 2012
Eur Heart J. 2012;33:1787–847. http://eurheartj.oxfordjournals.org/content/ehj/33/14/1787.full.pdf.

Patient Information

Myocarditis Foundation

http://www.myocarditisfoundation.org/about-giant-cell-myocarditis/.

Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/myocarditis/basics/definition/con-20027303.

Medlineplus

ENGLISH
http://www.nlm.nih.gov/medlineplus/ency/article/000149.htm.
ESPANOL
http://www.nlm.nih.gov/medlineplus/spanish/ency/article/000149.htm.

NORD

https://rarediseases.org/rare-diseases/giant-cell-myocarditis/.

Professional Information

Review

Herz. 2012;37:632–6. http://www.ncbi.nlm.nih.gov/pubmed/22930389?dopt=Abstract.

Drug Hypersensitivity

Cardiovasc Path. 2000;8:287–91. http://www.sciencedirect.com/science/article/pii/S1054880700000491.

EMB/Immunosuppression

Circ Heart Failure. 2013;6:15–22. http://circheartfailure.ahajournals.org/content/6/1/15.long.

ECG Epsilon Waves: Case Report

Eur Heart J. 2014;35:9. http://eurheartj.oxfordjournals.org/content/35/1/9.

Imaging: MRI

Circulation. 2014;129:e467–9. http://circ.ahajournals.org/content/129/17/e467.full.

Immunosuppression

Am J Cardiol. 2008;102:1535–9. http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2613862/.

Long-Term Follow-Up

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Natural History/Treatment

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VT as Presenting Manifestation

Heart. 2007;93:119–21. http://heart.bmj.com/content/93/1/119.extract.

Updates and More

https://clinicalguidecvd.com/gcm

Chapter 52
Heart Failure (CHF/ Congestive Heart Failure)

Management Keys

Consider reversible causes, including:

- Acute Myocarditis
- Cardiomyopathy – Peripartum
- Cardiomyopathy – Tachycardia-Induced
- Cardiomyopathy – Takotsubo
- Cardiomyopathy – Toxic (eg, Cobalt) [58]
- Hypocalcemia/Hypomagnesemia [62]

Consider treatment with ICD for primary prevention if LVEF ≤35 % in non-ischemic DCM; or ischemic heart disease at least 40 days post-AMI with EF ≤30 % (NYHA I) or ≤35 % (NYHA II-IV)

Diagnose specific stage/treat specific cause if identified

Diagnose/treat comorbid conditions (both CV and non-CV)

Early diagnosis/treatment of asymptomatic patients with LVEF <40 % [33]

Evaluate for cardiac transplantation/mechanical circulatory support for select patients with stage D HF

Provide multidisciplinary specialist services to enhance QOL and survival while simultaneously decreasing hospital readmissions up to 30–50 %

Provide prompt diagnosis/treatment of decompensated HF including hospitalization

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, DOI 10.1007/978-1-4471-7293-2_52,

Treat with Aldosterone antagonists to decrease morbidity and mortality in NYHA class II-IV pts with LVEF ≤35 % or with LVEF ≤ 40 % post-AMI or Diabetes Mellitus (with acceptable creatinine/potassium blood levels)
Treat with Angiotensin-Converting Enzyme Inhibitors or Angiotensin-II Receptor Blockers in patients with HFrEF to decrease morbidity and mortality
Treat with 1 of 3 specific Beta-blockers (carvedilol, metoprolol, bisoprolol) to decrease morbidity and mortality in patients with HFrEF [67]
Treat with LCZ696 to reduce risk of CV death/HF hospitalization
Treat with CRT if LVEF ≤35 % with sinus rhythm, LBBB and QRS ≥150 ms
Treat with hydralazine combined with isosorbide dinitrate in African-Americans with HFrEF (NYHA Class III-IV) to decrease morbidity and mortality in addition to other guideline-recommended drugs
Treat with caution when using long-term continuous or intermittent IV inotropes, which are potentially harmful and not recommended (other than bridging to definitive treatment/palliative care)
Consider exercise training therapy for patients with HFrEF [25]

ICD-10 Code

I50.22 (Chronic Systolic)
I50.32 (Chronic Diastolic)

Alternate Names/Abbreviation

HF
Congestive heart failure (CHF)
Ischemic Cardiomyopathy (ICM)
Left ventricular failure

Left ventricular diastolic dysfunction
Left ventricular systolic dysfunction (LVSD)
Non-ischemic Cardiomyopathy (NICM)
HFrEF: abnormal systolic function with reduced EF (≤40 %); also termed "systolic heart failure" [7]
HFpEF: abnnormal diastolic function with preserved EF (≥50 %); also termed "diastolic heart failure" [7]

Description/Etiology

Clinical syndrome caused by structural or functional cardiac impairment; recognized by characteristic pattern of hemodynamic, renal, and neurohumoral responses
All forms of heart disease can result in HFrEF in advanced stages
Causal categories:

Ischemic heart disease, including:

AMI [66]
Myocardial "stunning"
Myocardial "hibernation"
Ventricular remodeling post-AMA

Cardiomyopathies, including:

Acquired
Inherited

Myocarditis, including:

Infectious
Infiltrative
Inflammatory
Toxic

Increased ventricular pressure load, including:

Aortic Stenosis – Valvular
Aortic Stenosis – Subvalvular
Aortic Stenosis – Supravalvular
Hypertension – Systemic Arterial

Increased ventricular volume load, including:

- Aortic Regurgitation – Acute
- Aortic Regurgitation – Chronic
- Arteriovenous fistula (including Peripheral Extremity Arteriovenous Fistula)
- Intracardiac shunts/congenital heart disease
- Mitral Regurgitation – Acute
- Mitral Regurgitation – Chronic

Restrictive pericardial disease, including

- Pericardial effusion
- Pericarditis – Constrictive

Restrictive myocardial function, including:

- Decreased myocardial dispensability
- Endocardial fibroelastosis
- Restrictive myocardial disease

Electrical abnormalities, including:

- Tachycardias (including Cardiomyopathy – Tachycardia-Induced)
- Ventricular dyssynchrony

High output states, including:

- Anemia
- Beriberi
- Hyperthyroidism and Graves Disease
- Paget Disease
- Peripheral Arteriovenous Fistula

AHA Stages:

A. High risk for HF without structural heart disease or HF symptoms
B. Structural Heart disease without HF signs/symptoms
C. Structural Heart disease with prior or current HF symptoms
D. Refractory HF requiring specialized interventions

NYHA Functional Classification:

I. No limitation of physical activity; ordinary activity does not cause HF symptoms
II. Slight limitation of physical activity; comfortable at rest, but ordinary activity causes HF symptoms
III. Marked limitation of physical activity; comfortable at rest, but < ordinary activity causes HF symptoms
IV. Unable to carry on any physical activity without HF symptoms, or HF symptoms at rest

Common triggers of acute decompensation/HF progression:

Acute infections (eg, viral illnesses, pneumonia)
Acute Kidney Injury/decreased renal function
Alcohol use (heavy)
Anemia
AF
Brady/tachyarrhythmias
Fluid retention caused by noncardiac medication (eg, NSAID)
Hypertension – Systemic Arterial
Hyperthyrodism and Graves Disease
Hypoxia
Myocardial ischemia

Nonadherence with medications/diet

Comorbid Conditions [42]

MOST COMMON

ACUTE MYOCARDIAL INFARCTION [66]
ALZHEIMER DISEASE/DEMENTIA
ANEMIA [54]
ARTHRITIS
ASTHMA [AGE <65 years]
ATHEROSCLEROSIS IN OTHER CV AREAS

ATRIAL FIBRILLATION [52] [AND OTHER CAUSES OF RAPID VENTRIC RATE]
CARDIOMYOPATHY – DILATED [IDIOPATHIC, INCL FAMILIAL]
CEREBROVASCULAR DISEASE
CHRONIC KIDNEY DISEASE
CHRONIC OBSTRUCTIVE PULMONARY DISEASE (EMPHYSEMA)
CORONARY ARTERY DISEASE [AND OTHER FORMS OF ATHEROSCLEROTIC DIS]
DEPRESSION
DIABETES MELLITUS [49]
DYSLIPIDEMIA
DYSRHYTHMIAS – VENTRICULAR [63]
FUNCTIONAL MITRAL REGURGITATION [32]
HYPERTENSION – SYSTEMIC ARTERIAL
METABOLIC SYNDROME
PERIPHERAL ARTERY DISEASE
VALVULAR HEART DISEASE

OTHERS

ACROMEGALY
ALCOHOL USE/EXCESS
ARRHYTHMOGENIC RIGHT VENTRICULAR DYSPLASIA/CARDIOMYOPATHY
AUTOIMMUNE/CONNECTIVE TISSUE DISEASE
CARDIAC AMYLOIDOSIS
CARDIAC SARCOIDOSIS
CARDIOMYOPATHY – DANON DISEASE
CARDIOMYOPATHY – HIV
CARDIOMYOPATHY – HYPERTROPHIC
CARDIOMYOPATHY – IRON OVERLOAD
CARDIOMYOPATHY – NONCOMPACTION
CARDIOMYOPATHY – PERIPARTUM
CARDIOMYOPATHY – RESTRICTIVE
CARDIOMYOPATHY – TAKOTSUBO
CARDIORENAL SYNDROME
CHAGAS DISEASE

COCAINE
CONGENITAL HEART DISEASE [72] [73]
DRUGS: CANCER THERAPY [CARDIOTOXICITY] [50] [51]
ENDOMYOCARDIAL FIBROSIS
FABRY DISEASE
GROWTH HORMONE DEFICIENCY
HEMOCHROMATOSIS
HIV
HYPERTHYROIDISM
HYPOMAGNESEMIA
HYPOTHYROIDISM
MITOCHONDRIAL DISEASE
MUSCULAR DYSTROPHY
MYOCARDITIS [INCL VIRAL, EOSINOPHILIC, GIANT CELL]
NUTRITIONAL DEFICIENCIES
OBESITY [70]
OBSTRUCTIVE SLEEP APNEA [68]
OSTEOPOROSIS
RELAPSING CATASTROPHIC ANTIPHOSPHOLIPID ANTIBODY SYNDROME
STORAGE DISORDERS
TOXINS

Demography

Varies with etiology
CAD and DM increasingly prevalent etiology; hypertension and valve disease have become less common causes in more economically advanced societies
Increased risk with age: 20 % lifetime risk in persons age >40 year
Highest incidence in USA: non-Hispanic black males
Mortality: 50 % within 5 years of diagnosis (except reversible forms)

Patients with HFpEF more often:

Female
Less likely to have CAD
More likely to have Systemic Arterial Hypertension and AF
Obese
Older age

Pathophysiology

HF is a highly complex syndrome, comprising many physiological abnormalities, including:

1. Abnormal systolic/diastolic cardiac pump function to:
   Meet body's metabolic demands, with associated neuroendocrine compensatory changes (eg, fluid retention)
   Accommodate venous return

2. Ventricular remodeling: spherical LV with decreased contractility comprising numerous myocardial functional/structural functions; begins with hypertrophy in response to wall stress

3. Neurohormone activation:
   Arginine vasopressin
   Endothelin-1
   Natriuretic peptides (ANP, BNP)
   Nitric oxide
   RAAS
   Sympathetic nervous system (earliest response to decreased card output)

4. Myocardial fibrosis

5. Ketone bodies: significant fuel source for oxidative ATP production in hypertrophied and failing heart as cardiac capacity to utilize fatty acids (chief fuel in normal hearts) diminishes

Signs/Symptoms [73]

ABDOMEN – DISTENSION
ABDOMEN – FLUID (ASCITES) [8]
ABDOMEN – FULLNESS
ABDOMEN – PAIN [ESP RUQ] [34]
APPETITE – DECR (ANOREXIA)
APPETITE, SATIETY – EARLY [34]
ARTERIAL PRESSURE, UPRIGHT – DECR (ORTHOSTATIC HYPOTENSION) [11]
ARTERIAL PRESSURE, VALSALVA RESPONSE – ABN
ARTERIAL PULSE PRESSURE – DECR [38]
ARTERIAL PULSE, AMP – ALTERNATING (PULSUS ALTERNANS) [37]
ARTERIAL PULSE, AMP – DECR/ABS
BEHAVIOR – BIZARRE/CHANGED [1]
BLOOD PRESSURE, ARTERIAL – INCREASED/ELEVATED
BODY, APPEARANCE – WASTING (CACHEXIA) [9]
BODY, GEN – EDEMA (ANASARCA)
BOWEL MOVEMENTS – DIARRHEA
BREATH SOUNDS – CRACKLES (RALES) [10]
BREATH SOUNDS – DECR
BREATH SOUNDS – WHEEZES
BREATH SOUNDS, BASILAR – DECR
BREATHING – DIFF (DYSPNEA)
BREATHING – DIFF, NOCTURNAL (DYSPNEA, NOCT)
BREATHING – DIFF, RECLINING FLAT (ORTHOPNEA) [8]
BREATHING – RAPID (TACHYPNEA)
BREATHING – RHYTHMIC CHANGES (CHEYNE-STOKES)
BREATHING, NOCT – PAUSES (SLEEP APNEA)
CAPILLARY REFILL – SLUGGISH
CHEST – PAIN [4]

CHEST – PALPITATIONS
CHEST, ANT – MURMUR, SYS
COGNITION – DEFECT, NS
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)
COUGH
COUGH – NOCT [35]
EXTREM, LOWER, BILAT – EDEMA [5] [8]
EXTREM, LOWER, BILAT – FATIGUE
EXTREM, LOWER, TEMP – DECR [12]
EXTREM, UNILAT – EDEMA
EYES – PROMINENT (EXOPHTHALMOS/ PROPTOSIS) [2]
EYES/SKIN – YELLOW (JAUNDICE)
FATIGUE
GENITALS, SCROTUM – SWOLLEN (EDEMA)
HEADACHE
HEART, LV, APEX, IMP – DISPLACED, INF
HEART, LV, APEX, IMP – DISPLACED, LAT
HEART, P2, INTENSITY – INCR
HEART, RATE – RAPID (TACHYCARDIA)
HEART, RHYTHM – IRREG [3]
HEART, S3 LV
HEART, S3 RV
HEART, S4 LV
HEART, S4 RV
HEART, SOUNDS, INTENSITY – DECR
HYPOTENSION (BLOOD PRESSURE – DECREASED/LOW)
LIVER – ENLARGED (HEPATOMEGALY)
LIVER – TENDER
MENTATION – CONFUSION [1]
MENTATION – WEAKNESS (MALAISE)
MOOD – DEPRESSED
MUSCLES – ATROPHY
MUSCLES – WEAK
NAUSEA [34]
NECK, JVP – ABDOMINOJUGULAR REFLUX [36]

NECK, JVP – ELEV [8] [36]
SKIN – ITCHING (PRURITUS)
SKIN, COLOR – BLUE (CYANOSIS)
SKIN, COLOR – PALE (PALLOR)
SLEEP – DISTURBED (INSOMNIA)
SLEEP – DISTURBED (INSOMNIA)
SPLEEN, SIZE – INCR (SPLENOMEGALY)
SPUTUM – BLOOD (HEMOPTYSIS)
URINATION – NIGHTTIME (NOCTURIA) [35]
VOMITING (EMESIS) [34]
WEIGHT – INCREASED/GAIN [8]

Differentiation

Other causes of abdominal pain
Other causes of dyspnea
Other causes of leg edema
Pulmonary Hypertension

Complications

Acute pulmonary edema
Acute Pulmonary Embolism
Cachexia
Cardiogenic shock
Congestive hepatopathy
Deep Vein Thrombosis – Lower Extremity
Dysrhythmias – atrial (esp AF)
Dysrhythmias – ventricular
Functional MR [32]
Pericardial effusion
Peripheral arterial embolism
Pleural effusion
Pneumonia
Pulmonary Hypertension

Renal failure
Stroke – Ischemic
Sudden death [69]

Laboratory [39]

BLOOD, BILIRUBIN – INCR
BLOOD, BUN – INCR [6]
BLOOD, CHOLESTEROL, TOTAL – DECR
BLOOD, CREATININE – INCREASED
BLOOD, GALECTIN-3 – INCR [56]
BLOOD, GLUCOSE – DECR (HYPOGLYCEMIA)
BLOOD, GLUCOSE – INCR (HYPERGLYCEMIA) [65]
BLOOD, HGB/HCT – DECR (ANEMIA) [54]
BLOOD, LIVER ENZYMES – INCREASED
BLOOD, NT-PROBNP – INCR [13] [14]
BLOOD, SODIUM – DECR (HYPONATREMIA)
BLOOD, ST2 – INCR [57] [SEE APPENDIX A]
BLOOD, TROPONIN – INCR
BLOOD, URIC ACID – INCR [53]
URINE, PROTEIN – INCR (PROTEINURIA)

ECG

DYSRHYTHMIAS – ATRIAL (PACS/OTHERS) [52] [especially AF]
DYSRHYTHMIAS – VENTRICULAR (PVCS/OTHERS)
HEART, RATE, VARIABILITY – DECR [64]
NS/VAR PER COMORBIDITY(S)
QRS – LVH PATTERN

Imaging [16]

CARDIOMEGALY [15]
LA, CHAMBER, SIZE – INCR [41]
LV, CHAMBER, SIZE – INCR [15] [41]

LV, DIAS – DYSF [7]
LV, EF – DECR [7]
LV, FILLING – DECR/RESTRICTED HFpEF
LV, MYOCARD, WALL THICKNESS – INCR (HYPERTROPHY) [ESP WHEN PRECEDED BY LVH] [19]
LV, SYS – DYSF [7]
LV, WALL MOTION – DECR
LV, WALL MOTION, SEG – DECR/AKINETIC [18]
LV, WALL THICKNESS – DECR [ESP WHEN PRECEDED BY DIL CARDIOMYOPATHY]
MV, FLOW – DECR
MV, FLOW – REGURG [20]
PA, MAIN, SIZE – INCR [17]
PERICARD – FLUID
PLEURA – FLUID
PUL, VASCULARITY – INCR [40]
PUL, VEINS – CONGESTED
RA, CHAMBER, SIZE – INCR [41]
RV, CHAMBER, SIZE – INCR [41]
RV, WALL MOTION – DECR
RV, WALL MOTION, SEG – DECR/AKINETIC
TV, FLOW – REGURG

Genomics

NS

Other Tests

Exercise test [69]
Right heart catheterization: to resolve specific clinical or therapeutic question (routine use to guide therapy not effective)
Left heart catheterization and coronary angiography: to determine ischemic versus non-ischemic etiology, and subsequently for possible coronary revascularization (eg, angina/ischemia-related LV dysfunction) [4]

EMB: when specific cause is suspected that would influence therapy (eg, suspect Giant Cell Myocarditis in new onset acute HF)
Pulmonary artery pressure wireless monitoring

Treatment: Nonpharmacologic [21]

Alcohol restriction/avoidance
Cocaine, amphetamine avoidance
CPAP when Obstructive Sleep Apnea present
Dietary sodium restriction [22]
Exercise training [25]
Self-care education (e.g., diet, symptom/weight monitoring, medication adherence, exercise)
Social support

Treatment: Pharmacologic [30]

Adjuvant parenteral therapy in selected decompensated patients
Aldosterone Antagonists, including eplerenone [28] [47]
ACEIs/ARBs [26] [43] [47]
Antithrombotics [48]
BP control (especially patients with HFpEF)
Beta-Blockers [23] [44] [45] [46] [67]
Calcium/magnesium replacement when Hypocalcemia/Hypomagnesemia present [62]
Digoxin if already receiving guideline-directed therapy [29] or control of ventricular rate in AF when beta-blocker insufficient
Diuretics, including furosemide and hydrochlorothiazides [27]
Fluid restriction: select patients
Hospital management of decompensated HF including:

Diuretics and other fluid management strategies
Deep Vein Thrombosis prophylaxis

Hydralazine and isosorbide for self-described African American patients or patients intolerant of RAAS inhibitors
Influenza vaccination [55]
Ivabradine [31]
LCZ696 (angiotensin receptor-neprilysin inhibitor) [61]
Non-dihydropyridine CCBs contraindicated with HFrEF
Omecamtiv mecarbil (investigational)
Omega-3 fatty acids
Other drug avoidance:

- NSAIDS
- Antiarrhythmics except amiodarone and dofetilide
- IV inotropes
- Thiazolidinediones

Prompt diagnosis/treatment of decompensated HF including hospitalization
Serelaxin (acute HF) (investigational)
Statins (Hx of CAD or hyperlipidemia; no indication for HF alone)
Ularitide (acute HF) (investigational)

Treatment: Surgical/Invasive (Consult Current Guidelines for Specific Indications)

Coronary revascularization
CRT
ICD [24]
Mechanical circulatory support, including [59]:

- Intra-aortic balloon pump
- Durable pulsatile or continuous flow VAD
- ECMO
- Temporary percutaneous VAD
- Total artificial heart

MV repair

- Surgical

Transcatheter

Heart transplant
Aquapharesis (investigational)
Bariatric surgery (investigational) [71]
Gene Therapy (investigational)
Stem cell/Regenerative (investigational)
Unidirectional L-R interatrial shunting (investigational)

Prevention

Control atherosclerosis risk factors
Control blood pressure
Control lifestyle risk factors [60]
Early diagnosis and GDMT of Cardiomyopathies, Valvular Heart disease, CAD, Congenital heart disease to prevent progression from stage A or B HF

Course

Variable per cause
Prognosis worse with:

Advanced NYHA functional class
Anemia
Cachexia
Decreased cholesterol
Decreased LVEF
Decreased systolic BP
High dose diuretics
Hyperuricemia
Hyponatremia
Increased relative lymphocyte count
Ischemic etiology

Notes

[1] Especially elderly, patients with low cardiac output
[2] Due to chronically increased venous pressure
[3] From AF or multiple PVCs
[4] Chest pain may occur in absence of demonstrable CAD; coronary angiography may be indicated in select patients
[5] Peripheral edema absent in many patients with volume overload, especially younger persons; may be due to peripheral causes, especially obesity and age-related venous insufficiency
[6] May be due to low cardiac output or venous congestion
[7] Most patients have combination of systolic and diastolic dysfunction; HF with EF 40–50 % considered intermediate group
[8] Along with vital signs, should be assessed at every encounter for volume status, especially JVP
[9] Indicates poorer prognosis
[10] Rales often absent in chronic HF
[11] May indicate volume depletion
[12] May indicate inadequate cardiac output
[13] Useful for:

Diagnostic uncertainty
Gauging HF severity
Prognosis and when HF dx uncertain
(Use for monitoring controversial)

[14] Other causes of increased natriuretic peptides:

Cardiac:

AF
Acute coronary syndrome
Cardiac surgery
Cardioversion
LVH

Myocardial disease
Pericardial disease
Valvular heart disease

Non-cardiac:

Advanced age
Anemia
Bacterial sepsis
Burns (severe)
Critical illnesses
Pulmonary disease
Renal failure
Toxins (cancer chemotherapy, envenomation)

[15] May be normal size on CXR

[16] 2D echo: most useful diagnostic test for HF and should be performed as part of initial evaluation for LV function and associated cardiac conditions and serially with proper clinical indications, such as for LV remodeling

MRI/CT/radionuclide ventriculography: useful in select cases, especially when echo is suboptimal

[17] Pulmonary artery dilation suggests Pulmonary Hypertension, a common consequence of LV dysfunction; primary PAH may also result in secondary RHF

[18] Suggests ischemic cardiomyopathy

[19] Suggests LVH-related HF, especially secondary to Systemic Arterial Hypertension or to infiltrative cardiomyopathy

[20] Functional MV regurgitation common in HF with ventricular dilatation; in absence of significant dilatation or when regurgitation is severe, other causes should be considered (eg, papillary muscle dysfunction, primary valve disease)

[21] Including recommendations for patients in stages A and B with goal of preventing progression to overt HF

[22] Not universal recommendation as evidence for efficacy indefinite and may worsen neurohormonal profile

[23] Caution when starting beta-blockers in decompensated patients, as can cause clinical deterioration; start at very

low dose and up-titrate gradually toward target dose as tolerated by symptoms

[24] For patients with LVEF ≤35 % in non-ischemic dilated or ischemic cardiomyopathy, at least 40 days post-AMI, EF ≤30 % (NYHA class I) or ≤35 % (NYHA class II–III), if already receiving appropriate medical therapy and have reasonable expectation of survival >1 year

[25] Safe with multiple benefits, including improvements in mortality, functional capacity, exercise duration, QOL, decreased hospitalizations

[26] ACEIs first choice but ARBs considered reasonable alternative

[27] All patients with fluid retention, with close monitoring of electrolytes and renal function; loop diuretics (furosemide, torsemide, bumetanide) are first-line, with second line intermittent use of thiazide "boosters" (metolazone, chlorothiazide)

[28] Aldosterone antagonists recommended to decrease morbidity and mortality in NYHA class II–IV with LVEF ≤35 % or with LVEF ≤40 % post-AMI or in DM (providing acceptable blood creatinine and potassium); blood creatinine and K should be closely monitored

[29] Use digoxin with caution including close monitoring of electrolytes and appearance of dysrhythmias, heart block, GI symptoms, neurological changes; reduce hospitalizations but not mortality, which may actually increase with its use

[30] Except for BP control, drug treatment for HFpEF not efficacious, with no proven strategies to decrease mortality

[31] EF ≤35 %, HR ≥70 bpm, persistent symptoms, despite beta-blockers and RAAS-inhibition or when beat-blockers not tolerated

[32] Occurs in >50 % of patients with LVEF <40 % and severity correlates with survival; invasive management remains controversial; also common in patients with HFpEF

[33] Early medical treatment can slow HF progression

[34] Suggests liver congestion
[35] Fluid redistribution while recumbent
[36] Measure of RA pressure that correlates with PCWP; should not be used as a marker of PCWP in AMI-related HF
[37] Highly indicative of decreased cardiac output
[38] Proportional pulse pressure (systolic BP minus diastolic BP/systolic BP) <25 % correlates with cardiac index <2.2 L/min by RH catheterization; decreased pulse pressure associated with poor prognosis in HFrEF but less correlation with prognosis in HFpEF
[39] In addition to this list: cardiac enzyme measurement indicated if ACS suspected; special tests when underlying disease suspected (eg thyroid disease, connective tissue disease, Hemochromatosis); chemistries for monitoring HF treatment (eg, serum K with use of RAAS inhibitors)
[40] Also on CXR: cephalization, interstitial/alveolar edema, Kerley B lines
[41] 4-chamber dilatation suggests possible non-ischemic etiology
[42] About 50 % of HF readmissions due to comorbid conditions; comorbidities may occur more often with HFpEF
[43] Mildly increased blood K, mild decrease in renal function and BP expected; should be carefully monitored but do not require changing therapeutic course; ACEIs and ARBs similar for these effects
[44] Beta-blockers vary greatly in efficacy for HF, and special attention should be paid to GDMT for this class: only 1 of carvedilol, metoprolol, bisoprolol should be prescribed for HF
[45] Decreased BP, bradycardia, lethargy, depression exacerbation, impotence may occur with beta-blockers and occurrence may require down-titration to maximally tolerated dose if possible before discontinuation; when beta-blocker termination needed, should not be abrupt

[46] Beta-blockers in HF patients with:

DM: equal benefit in patients with/without DM

COPD: most patients tolerate well, but use caution with bronchospasm including slow titration and close monitoring

Peripheral vascular disease: may worsen limb ischemia but usually tolerated well

[47] Efficacy/safety of combining ARBs/ACEIs/aldosterone antagonists not supported by evidence and carries added risk of hyperkalemia

[48] No randomized trial data for guidance in absence of AF/history of thromboembolic event/recent AMI/ventricular thrombus

[49] Increased 2 year mortality by up to 70 %

[50] Anthracyclines (most severe; eg, doxorubicin), mitoxantrone, cyclophosphamide, mitomycin, trastuzumab, alemtuzumab, sorafenib, imatinib, paclitaxel, docetaxel

[51] Anthracycline-induced form may occur early during treatment (within 1 year) in acute form or delayed 10–20 years post-initial exposure; delayed form has very poor prognosis

[52] AF: both a trigger for and predictor of HF progression; occurs in >50 % of patients with HF

[53] Especially males and African-Americans hospitalized for worsening HFrEF

[54] Anemia associated with increased mortality/length of hospitalization: greater in patients with HFpEF than HFrEF

[55] May be associated with decreased mortality

[56] Galectin-3: associated with myocardial fibrosis/increased risk of HF/HF mortality

[57] ST2: cardiac biomarker assay; useful for prognosis; may be superior to galectin-3 for risk stratification; also increased in other CV conditions, including:

Acute Myocardial Infarction
Aortic Stenosis –-Valvular
Arterial Hypertension –-Systemic

Cardiomyopathy -–Dilated
Diabetes Mellitus
Kawasaki Disease
Mitral Regurgitation -–Chronic
Peripheral artery disease
Pulmonary Arterial Hypertension
Stable Ischemic heart disease
ST2 also increased in many non-CV conditions

[58] Cobalt toxicity should be considered in patients with prosthetic (metal-on-metal) hip replacement; removal of prosthesis has been associated with improvement/reversal of cardiac dysfunction; other manifestations include:

Hypothyroidism
Neuropathy
Polycythemia

[59] In USA, current CMS criteria should be consulted for LVAD implant

[60] Most common lifestyle risk factors associated with decreased risk of HF:

Modest alcohol intake
Obesity avoidance
Physical activity
Tobacco avoidance

[61] FDA-approved indication for LCZ696: chronic heart failure (NYHA Class II–IV) and reduced EF

[62] Hypocalcemia as a reversible cause of HF should especially be considered in patients with QT prolongation and paresthesias; and in settings known to predispose to hypoparathyroidism, such as post-thyroidectomy; hypomagnesemia must also be taken into account in such patients

[63] Frequent PVCs associated with decreased LVEF, increased incidence of HF, and increased mortality

[64] Increased HR range (as measured by ambulatory ECG) associated with better prognosis

[65] Dysglycemia: common and is associated with increased risk of adverse CV outcomes
[66] AMI: HF frequent complication of first AMI, both during acute phase and shortly after hospital discharge
[67] Beta-blockers: tolerability comparable in HFrEF and HFpEF; short-term efficacy greater in HFrEF
[68] Sleep-Disordered Breathing: moderate-severe form common in patients with HF (prevalence >45 % in SchlaHF Registry); associated with:

Age
BMI
Male sex
Symptom severity/LV function

[69] Exercise test in patients with HFrEF: strong predictors of death including decreased:

Peak Vo2
Exercise duration
% ppVo2

[70] Obesity: compared with subjects with normal BMI, obese patients may have up to 2× risk of HF, especially HFpEF
[71] Bariatric surgery: may be associated with decreased HF exacerbation rate in obese HF patients
[72] Heart Failure due to Congenital Heart Disease (from AHA Scientific Statement: Chronic Heart Failure in Congenital Heart Disease, J Am Coll Cardiol 2008;52:e143–e263):

Major cause of late death (>30 days) in children after pediatric cardiac surgery, contributing to 27 % of deaths and occurring at median age of 5.2 years
Leading cause of death in adults with CHD, described in 26 % of all deaths in a national registry of >8000 adults with CHD, with similar findings in other reports
One study demonstrated that adults with CHD admitted with HF had fivefold increase in mortality compared with those who were not admitted;

also showed 1- and 3- year mortality rates of 24 and 35 % after a first HF admission

[73] Clinical presentation of adult HF patient with congenital heart disease may vary significantly by defect or age; patients can have classic symptoms of fatigue, dyspnea, and exercise intolerance but may manifest more subtle signs of malnutrition, growth failure, or cachexia

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Circulation Heart Fail. 2015;8:565–71. http://circheartfailure.ahajournals.org/content/8/3/565.full.

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Updates and More

https://clinicalguidecvd.com/hf

Chapter 53
Systemic Arterial Hypertension (Essential Hypertension)

Management Keys

Consider secondary causes
Consider white coat syndrome [9]
Diagnose/treat target organ damage
Diagnose/treat co-existing CVD
Hospitalize for hypertension crisis [1]
Prescribe appropriate lifestyle modification [8]
Prescribe BP patient self-monitoring
Treat to target diastolic BP <90 mmHg in persons age <60 years
Treat to target sys BP <150 mmHg persons age >60 years

ICD-10 Code

I10

Alternate Names/Abbreviation

HTN
Essential hypertension

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_53,

Description/Etiology

Diseases/conditions causing decreased intravascular compliance relative to circulating blood volume, leading to non-invasively measured systolic BP >140 mmHg/diastolic BP >90 mmHg

Genetic, environmental, lifestyle (eg, NaCl/K intake) factors, adiposity strongly influence development of primary HTN

Diagnosis: average of at least two readings obtained at three visits: 140/90 mmHg or greater

Secondary HTN occurs in up to 10 % of cases

Blood pressure increased risk for CVD beginning at 115/75 mmHg:

- Linear
- Continuous
- Independent of/additive to other risk factors

Stages (JNC 7) (mm Hg):

Normal:	<120/80
Prehypertensive:	120/80–139/89
Stage 1:	140/90–159/99
Stage 2:	>159/110

Pseudohypertension: false BP elevation due to stiff, incompressible arteries [15]

White coat hypertension: increased office BP with normal home/ambulatory BP monitor [16]

Masked hypertension: normal office BP with increased BP at home/ambulatory BP monitoring [17]

Secondary hypertension causes: see predisposing/comorbid conditions; also suspect in patients with:

- Drug-resistant HTN (>2 drugs at max doses)
- New onset HTN age <25 years/>55 years
- Severe vascular disease, including:
  - Abdominal Aortic Aneurysm
  - Aortic Dissection
  - CAD

Carotid Artery Stenosis
Peripheral Artery Disease
RAS

Resistant HTN: BP that remains above goal in spite of concurrent use of three antihypertensive agents of different classes; ideally, one of the three agents should be a diuretic and all agents should be prescribed at optimal doses [27] [28]

Predisposing/Comorbid Conditions [2] [6]

ACROMEGALY
ALDOSTERONISM
AMPHETAMINES
ATRIAL SEPTAL DEFECT, POST-CLOSURE
BRAIN TUMOR
BROMOCRYPTINE
CADMIUM TOXICITY
CHRONIC KIDNEY DISEASE
COARCTATION OF AORTA
COCAINE
CUSHING DISEASE
CYCLOSPORINE
DIABETES MELLITUS
DIABETIC NEPHROPATHY
DRUGS: CANCER THERAPY [13]
DRUGS: SYSTEMIC CORTICOSTEROIDS
DYSLIPIDEMIA
ECLAMPSIA
FIBROMUSCULAR DYSPLASIA
GLOMERULONEPHRITIS, ACUTE
GOUT
GUILLAIN-BARRE SYNDROME
HEART TRANSPLANT
HELLP SYNDROME
HEMANGIOPERICYTOMA
HYPERCALCEMIA
HYPERPARATHYROIDISM

HYPERTHYROIDISM
HYPERVISCOSITY SYNDROME
HYPOGLYCEMIA
HYPOKALEMIA
HYPOTHYROIDISM
LEAD POISONING
LICORICE
LIDDLE SYNDROME
METABOLIC SYNDROME
MITRAL ANNULAR CALCIFICATION
MULTIPLE ENDOCRINE NEOPLASIA
NEPHROTIC SYNDROME
NEUROBLASTOMA
NEUROFIBROMATOSIS
NONSTEROIDAL ANTIINFLAMMATORY DRUGS
OBESITY
OBSTRUCTIVE SLEEP APNEA
PHEOCHROMOCYTOMA
POLYCYSTIC KIDNEY DISEASE
POLYCYTHEMIA VERA
PREECLAMPSIA
PREGNANCY
QUADRIPLEGIA
RENAL ARTERY STENOSIS
RENAL EMBOLIZATION
RENAL FAILURE (UREMIA)
RENAL TRANSPLANT
SICKLE CELL DISEASE/TRAIT
TAKAYASU ARTERITIS
THALLIUM POISONING
TRANSFUSION
VASCULITIS, INTRARENAL
VON HIPPEL-LINDAU DISEASE

Demography

Age relation 40–70 years: each increment of 20 mmHg systolic/10 mmHg diastolic doubles risk of CVD (in range of 115/75–185/115 mmHg); risk increases

are higher in presence of other CV risk factors (eg, DM, CKD)
Populations: all
Location: global

Pathophysiology

Complex interplay of:

- Aortic stiffness/fibrosis
- Chronic intravascular volume expansion (largely due to excess salt intake)
- Decreased vascular compliance
- Increased vascular resistance

Multiple neurohormonal mechanisms, involved, including:

- Increased RAAS activity
- Increased sympathetic activity

Signs/Symptoms [5]

ARTERIAL PRESSURE – HIGHLY VARIABLE [26]
BLOOD PRESSURE, ARTERIAL – INCREASED/ELEVATED
HEART, LV, APEX, IMP – FORCEFUL/SUSTAINED
HEART, S2, SPLIT – REVERSED (PARADOXICAL) [SEVERE HTN]
HEART, S4 LV

Differentiation [24]

Acromegaly
Acute Glomerulonephritis
Anemia
Anxiety
Aortic Dissection
Aortic Regurgitation – Chronic

- Aortic rigidity (esp aging)
- Atrioventricular Heart Block
- Bath Salts – Recreation & Abuse
- Beriberi
- BZ Poisoning
- Chlorine Poisoning
- Chronic Nephritis
- Coarctation Of Aorta
- Cocaine
- Collagen Vascular Disease
- Congenital Adrenal Hyperplasia
- Cushing Syndrome
- Cyanide Poisoning
- Diabetic Nephropathy
- Drugs
  - Amphetamines
  - Anabolic steroids
  - Antidepressants
  - Appetite suppressants
  - Caffeine
  - Cocaine
  - Cyclosporine
  - Ecstasy
  - Erythropoietin
  - Ethanol
  - Glucocorticoids
  - Herbs
    - Arnica
    - Bitter orange
    - Ephedra
    - Gingko
    - Gingseng
    - Guarana
    - Licorice
    - Senna
    - St Johns wort
  - Ivabradine
  - Mineralocorticoids

- Monamine oxidase inhibitors
- Nasal decongestants
- NSAIDs
- Oral contraceptives
- Phenothiazines
- Phenylephrine
- Phenylcyclidine
- Sympathomimetics
- Tacrolimus
- Tyramine

Dysautonomia
Elemental Mercury Poisoning – Acute
Erdheim-Chester Disease
Ethylene Glycol Poisoning
Extra adrenal chromaffin tumors
Fabry Disease
Fibromuscular Dysplasia
Guillain-Barre Syndrome
Hallucinogens – Recreation
Hydronephrosis
Hyperparathyroidism
Hypothyroidism
Lead Poisoning
Mineralocorticoid overproduction
Neurologic disorders
Nicotine Poisoning
Obstructive Sleep Apnea
Organophosphate Poisoning
Paget Bone Disease
Peripheral Extremity Arteriovenous Fistula
Pheochromocytoma
Polycystic Disease
Postoperative hypertension
Primary Aldosteronism
Quadriplegia
Renal Artery Stenosis
Renal parenchymal disease
Renal vascular disease
Sarin Poisoning

Takayasu Arteritis
Thallium Poisoning
VX Poisoning
Williams Syndrome
White Coat Syndrome

Complications

Atrial Fibrillation
Abdominal Aortic Aneurysm
Aortic Dissection
Cardiac arrhythmias
Cardiac hypertrophy
Carotid Artery Stenosis
Cerebral aneurysm
Cognitive changes [25]
Coronary artery disease
  AMI
  Stable Ischemic Heart Disease
Encephalopathy
HF
Intracerebral hemorrhage
Left ventricular hypertrophy
Nephropathy [4]
Peripheral Artery Disease
Renal failure [4]
Retinopathy
Stroke – Ischemic
Subarachnoid hemorrhage

Laboratory [7]

ASSESS FOR OTHER CAUSATIVE FACTORS, EG, ELECTROLYTES
ASSESS FOR TARGET ORGAN DAMAGE, EG, RENAL FUNCTION [4]
BLOOD, ST2 – INCR [22]

BLOOD, TROPONIN – INCR
BLOOD, URIC ACID – INCR
URINE, PROTEIN – INCR (PROTEINURIA) [NEPHROPATHY]

ECG

AV COND – BIFASCICULAR BLOCK [3]
DYSRHYTHMIAS – ATRIAL (PACS/OTHERS)
QRS – LBBB/LBBB PATTERN [3]
QRS – LVH PATTERN
QRS, AXIS – L

Imaging [10]

LA, CHAMBER, SIZE – INCR
LV, MYOCARD, WALL THICKNESS – INCR (HYPERTROPHY)
MYOCARD, THICKNESS – INCR

Genomics

ACCN3 [NOS3-PREGNANCY-INDUCED OMIM +163729]
APB1 [NOS3-PREGNANCY-INDUCED OMIM +163729]
KCNH2 [NOS3-PREGNANCY-INDUCED OMIM +163729]
NOS3 [NOS3-PREGNANCY-INDUCED OMIM +163729]
NR3C2 [EXACERBATION IN PREGNANCY]

Other Tests

Ambulatory BP monitoring – indications:

- Antihypertensive drug efficacy
- Diagnosis/treatment during pregnancy
- Early AM BP surge [19]
- Episodic normal/autonomic dysfunction

Loss of dipping status [18]

White coat HTN suspected

Masked HTN suspected
Nocturnal HTN
Symptomatic hypotension on therapy (patient may be normotensive)
Unusual BP variability (may be associated with increased risk)

Home/self-measured BP

Treatment: Nonpharmacologic [20]

Appropriate lifestyle modification [8] [29]

Treatment: Pharmacologic [11] [20]

Alpha-blockers [23]

Doxazosin
Prazosin
Terazosin

ACEIs [21]

Enalapril
Fosinopril
Lisinopril
Perindopril
Quinapril
Ramipril

ARBs [21]

Candesartan
Irbesartan
Losartan

Beta-Blockers [23]

- Atenolol
- Carvedilol
- Metoprolol
- Nebivolol
- Propranolol

CCBs

- Amlodipine
- Diltiazem
- Nifedipine
- Verapamil

Central beta-agonists [23]

- Clonidine
- Methyldopa

Direct renin inhibitors

- Aliskiren

Diuretics [21] [23]:

- Chlorothiazide
- Chlorthalidone
- Hydrochlorothiazide
- Indapamide

Guanethidine
Hydralazine

Treatment: Surgical/Invasive [12]

Correction of secondary causes (e.g., aldosterone-secreting tumor resection, RAS angioplasty/stenting)
Renal denervation [14]

Prevention

Adult screening (JNC 7): every 1–2 years depending on BP/other CV risk factors
Diet

Calorie control appropriate for weight management
Decreased Na
Increased K

Notes

[1] Severe hypertension crises requiring hospitalization/emergency care include:

AMI
Aortic Dissection
Eclampsia
HF
Hypertensive encephalopathy
Intracranial hemorrhage
Post-CABG
Post-vascular surgery
Severe catecholamine excess, incl:
Clonidine withdrawal
Pheochromocytoma
Tyramine-MAOI interaction
Intoxication, including:

Cocaine
Phencyclidine
Phenylpropanolamine

Subarachnoid hemorrhage
Thrombotic stroke
Unstable angina

[2] Many in this list cause secondary HTN
[3] Often have coexisting CAD

[4] Renal damage secondary to increased intra-arterial pressures
[5] Does not include signs and symptoms of comorbid conditions/target organ damage
[6] Occult forms may be suspected in treatment-resistant HTN
[7] Basic tests for renal function, DM screen, and dyslipidemia screen indicated in all HTN patients; specific work-up for secondary causes reserved for high clinical suspicion or patients with resistant HTN
[8] Meticulous attention to:

Alcohol consumption (daily for males and 1/2 quantities for females)

2 mixed drinks; or
2 12 oz cans beer; or
2 4-oz glasses wine

CVD risk factors
Dietary sodium intake: no more than 2400 mg/day
Dyslipidemia
Regular aerobic exercise: 30 min most days of week
Tobacco cessation
Weight: maintain normal body weight (BMI 18.5–24.9 kg/m^2)

[9] Diagnostic ambulatory BP monitoring
[10] CXR and echo most often useful
[11] First-line treatment should comprise ACEI/ARB, thiazide, or CCB in non-blacks; avoid ACEI/ARB as first line therapy in black population
[12] Renal artery denervation for resistant HTN currently under investigation
[13] Includes bevacizumab, sorafenib, sunitinib, cisplatin, interferon-alpha
[14] Investigational
[15] Pseudohypertension: diagnose with Osler's maneuver: inflate cuff to 30 mmHg above palpable systolic pressure; try to "roll" brachial/radial artery under fingertips; healthy arteries should not be empty/not palpable; hard tube-like structure suggests pseudohypertension

[16] White coat HTN:

Prevalence: 10–20 %
Increased risk for overt Systemic Arterial Hypertension/CVD
Lifestyle modifications/regular follow-up recommended when diagnosed

[17] Masked HTN: 10–40 % of patients with HTN; associated with increased risk of sustained HTN/CV death

[18] Loss of dipping status: BP decreases <10 % at night compared to daytime BP

[19] Early AM BP surge: systolic BP 55 mmHg difference between sleep and early-hour awakening

[20] Target BP goals:

AHA/ACC: All: <140/90 mmHg
ESC: All: <140 mmHg (sys only)
JNC 8 panel:

Patients <60 year of age: <140/90 mmHg
Patients >60 year of age: <150/90 mmHg

Kidney disease improving global outcomes (KDIGO):

Patients with CKD: 130/80 mmHg
Patients with CKD/excreting >30 mg urine albumin/24 h: <130/80 mmHg

[21] JNC 8 panel members report: start treatment on thiazide diuretic except patients with DM or CKD for whom ACEI or ARB in combination with drug from another class is recommended; however some experts/British National Institute (NICE) recommend that chlorthalidone or indapamide should be used rather than hydrochlorothiazide as first-line drug for HTN

[22] JNC 7: compelling indications for specific drug classes:

CKD:

ACEI
ARB

DM:

ACEI
ARB
B-Blocker
CCB
Diuretic

HF:

ACEI
Aldosterone antagonist
ARB
Beta-blocker
Diuretic

High CAD risk:

ACEI
ARB
Beta-blocker
CCB
Diuretic

Post-AMI:

ACEI
Aldosterone antagonist
B-Blocker

Recurrent stroke prevention

ACEI
Diuretic

[22] Increased ST2 correlates with presence of concentric LVH in patients with Systemic Arterial Hypertension
[23] Anti-sympathetic/adrenergic activity by drug treatment lowers BP but little/no evidence that this improves outcomes
[24] List comprises acute/chronic causes of BP elevation
[25] May be more common in African-Americans
[26] Higher visit-to-visit variability of systolic BP associated with increased risk for CVD and mortality, but unproven if treatment warranted

[27] Patient characteristics of resistant HTN in USA:

- Black race
- CKD
- DM
- Excessive dietary salt ingestion
- Female sex
- High baseline BP
- LVH
- Obesity
- Older age
- Residence in southeastern United States

[28] Medications that interfere with HTN treatment include:

- Alcohol
- Cyclosporine
- Erythropoietin
- Herbal compounds, including:
  - Ephedra
  - Ma huang
- Natural licorice
- Nonnarcotic analgesics
  - Nonsteroidal antiinflammatory agents, including aspirin
  - Selective COX-2 inhibitors
- Oral contraceptives
- Stimulants
  - Amphetamines
  - Dexmethylphenidate
  - Dextroamphetamine
  - Methamphetamines
  - Methylphenidate
  - Modafinil

Sympathomimetic agents

Cocaine
Decongestants
Diet pills

[29] Lifestyle changes: **2013 ESH/ESC Guidelines for the management of arterial hypertension**

"5.1 Lifestyle changes

Appropriate lifestyle changes are the cornerstone for the prevention of hypertension. They are also important for its treatment, although they should never delay the initiation of drug therapy in patients at a high level of risk. Clinical studies show that the BP-lowering effects of targeted lifestyle modifications can be equivalent to drug monotherapy, although the major drawback is the low level of adherence over time—which requires special action to be overcome. Appropriate lifestyle changes may safely and effectively delay or prevent hypertension in non-hypertensive subjects, delay or prevent medical therapy in grade I hypertensive patients and contribute to BP reduction in hypertensive individuals already on medical therapy, allowing reduction of the number and doses of antihypertensive agents. Beside the BP-lowering effect, lifestyle changes contribute to the control of other CV risk factors and clinical conditions.

The recommended lifestyle measures that have been shown to be capable of reducing BP are: (i) salt restriction, (ii) moderation of alcohol consumption, (iii) high consumption of vegetables and fruits and low-fat and other types of diet, (iv) weight reduction and maintenance and (v) regular physical exercise. In addition, insistence on cessation of smoking is mandatory in order to improve CV risk, and because cigarette smoking has an acute pressor effect that may raise daytime ambulatory BP."

Guidelines

AHA/ACC/ASH scientific statement: treatment of hypertension in patients with coronary artery disease
Hypertension. 2015;65:1372–407. http://hyper.ahajournals.org/content/65/6/1372.full.

2013 ESH/ESC guidelines for the management of arterial hypertension
Eur Heart J.2014;34:2159–219. http://www.escardio.org/guidelines-surveys/esc-guidelines/Pages/arterial-hypertension.aspx.

JNC 7: Seventh Report of the Joint National Committee on Prevention, Detection, Evaluation, and Treatment of High Blood Pressure
Hypertension. 2003;42:1206–52. http://hyper.ahajournals.org/content/42/6/1206.long.

NICE: hypertension in adults: diagnosis and management. 2011
http://www.nice.org.uk/guidance/cg127/chapter/1-recommendations#choosing-antihypertensive-drug-treatment-2.

Patient Information

Medlineplus

ENGLISH
http://www.nlm.nih.gov/medlineplus/highbloodpressure.html.
ESPANOL
http://www.nlm.nih.gov/medlineplus/spanish/ency/article/000468.htm.

Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/high-blood-pressure/basics/definition/con-20019580.

Genetics Home Reference

http://ghr.nlm.nih.gov/glossary=hypertension.

CDC

http://www.cdc.gov/bloodpressure/.

CDC: Reducing Sodium in Children's Diet

http://www.cdc.gov/vitalsigns/children-sodium/index.html.

Texas Heart Institute

http://www.texasheart.org/HIC/Topics/Cond/hbp.cfm.

Professional Information

AHA scientific statement: BP goals.
Hypertension. 2014;63:878–85. http://hyper.ahajournals.org/content/63/4/878.
AHA SC8IENTIFIC statement: resistant hypertension.
Hypertension. 2008;51:1403–19. https://c.ymcdn.com/sites/www.nyschp.org/resource/resmgr/Files/Resistant-Hypertension-2008-.pdf.

Review

Ann Intern Med. 2014;161(11):ITC1. http://annals.org/article.aspx?articleid=1984496&resultClick=24.

Review: Dietary Sodium

J Am Coll Cardiol. 2015;65:1042–50. http://content.onlinejacc.org/article.aspx?articleID=2194893.

Review: Drug Therapy

Eur Heart J. 2014;35:501–7. http://eurheartj.oxfordjournals.org/content/35/9/557.

Review: JNC 8/Future Recommendations

J Am Heart Assoc. 2015;4:e002315. http://jaha.ahajournals.org/content/4/12/e002315.full.

Review: Preeclampsia/Pregnancy Related HTN

Hypertension. 2016;67:238–42. http://hyper.ahajournals.org/content/67/2/238.extract?etoc.

Review: Randomized RX Trials

Circulation Res. 2015;116:1058–73. http://circres.ahajournals.org/content/116/6/1058.full.

Review: Resistant HTN/CKD

Lancet. 2015;386:1588–98. http://www.sciencedirect.com/science/article/pii/S0140673615004183.

Review: Resistant HTN

JAMA. 2014;311:2216–24. http://jama.jamanetwork.com/article.aspx?articleid=1877189.

ACE Inhibitor Angioedema

Am J Med. 2015;128:120–5. http://www.sciencedirect.com/science/article/pii/S0002934314005919.

African Americans: Counseling

Circulation. 2014;129:2044–51. http://circ.ahajournals.org/content/129/20/2044.full.

Ambulatory BP Monitoring

Am J Med. 2015;128:14–20. http://www.sciencedirect.com/science/article/pii/S0002934314006597.

Ambulatory BP Monitoring

Circulation Res. 2015; 116:1034–45. http://circres.ahajournals.org/content/116/6/1034.full.

American Style Football

Circulation Cardio Qual Out. 2013;6:716–23. http://circoutcomes.ahajournals.org/content/6/6/716.full.

Aortic Stenosis: Treatment of HTN

Circulation. 2013;128:1349–53. http://circ.ahajournals.org/content/128/12/1349.full.

Aortic Stiffness/Fibrosis

Hypertension. 2016;67:461–8. http://hyper.ahajournals.org/content/67/2/461.abstract?etoc.

ARBS: Neuroprotection

Am J Hypertens. 2015;28:289–99. http://ajh.oxfordjournals.org/content/28/3/289.full?etoc.

Arterial Structural Changes

Circulation Res. 2015;116:1007–21. http://circres.ahajournals.org/content/116/6/1007.full.

Beta-Blockers: Central Systolic Pressure

Hypertension. 2016;67:316–24. http://hyper.ahajournals.org/content/67/2/316.abstract?etoc.

CAD: BP Targets

Heart. 2013;99:601–13. http://heart.bmj.com/content/99/9/601.abstract.

CAD: Treatment-Resistant HTN

Am J Med. 2014;127:71–81. http://www.sciencedirect.com/science/article/pii/S0002934313007651.

Caffeine

Hypertension. 2015;65:691–6. http://hyper.ahajournals.org/content/65/3/691.abstract?etoc.

Cardiorespiratory Fitness

J Am Coll Cardiol. 2014;64:1245–53. http://content.onlinejacc.org/article.aspx?articleID=1905460.

Central Aortic Pressure: Effect of Antihypetensives

Am J Hypertens. 2016;29:448–57. http://ajh.oxfordjournals.org/content/29/4/448.abstract?etoc.

CKD: Resistant HTN

J Am Coll Cardiol. 2013;61:2461–7. http://content.onlinejacc.org/article.aspx?articleID=1696815.

CKD: Resistant HTN/Risk Stratification

J Am Coll Cardiol. 2013;61:2468–70. http://content.onlinejacc.org/article.aspx?articleID=1696808.

Children/Adolescents

N Engl J Med. 2014;370:2316–25. http://www.nejm.org/doi/full/10.1056/NEJMcp1001120.

Cognitive Changes: Racial Disparity

Am J Hypertens. 2016;29:185–93. http://ajh.oxfordjournals.org/content/29/2/185.abstract?etoc.

Cognitive Changes: Women

Am J Hypertens. 2016;29:202–16. http://ajh.oxfordjournals.org/content/29/2/202.abstract?etoc.

Cognitive Impairment: Progression with Excess BP Lowering

JAMA Intern Med. Online 2 Mar 2015. http://archinte.jamanetwork.com/article.aspx?articleID=2173093&utm-source=Silverchair%20Information%20Systems&utm-medium=email&utm-campaign=ArchivesofInternalMedicine%3AOnlineFirst03%2F02%2F2015.

Community-Wide HTN Intervention

Circ Cardiovasc Qual Out. 2014;7:828–34. http://circoutcomes.ahajournals.org/content/7/6/828.full.

CPAP

JAMA. 2013;310:2407–15. http://jama.jamanetwork.com/article.aspx?articleid=1788459.

Diabetes Mellitus: New Onset/Mortality

Eur Heart J. 2016;37:968–74. http://eurheartj.oxfordjournals.org/content/37/12/968.abstract?etoc.

Diabetes Mellitus

JAMA. 2015;313:603–15. http://jama.jamanetwork.com/article.aspx?articleid=2108887.

Diabetes Mellitus

Heart. 2013;99:577–85. http://heart.bmj.com/content/99/8/577.extract.

Dietary Effects on Blood Pressure

Hypertension. 2016;67:733–9. http://hyper.ahajournals.org/content/67/4/733.abstract?etoc.

Dietary Sodium

Circulation Res. 2015;116:1046–57. http://circres.ahajournals.org/content/116/6/1046.full.

Dietary Sodium and Potassium

Mayo Clin Proc. 2013;88:987–95. http://www.sciencedirect.com/science/article/pii/S0025619613004631.

Dietary Sodium: Long-Term Effects on CVD Outcomes

BMJ. 2007;334:885–8. http://www.bmj.com/content/334/7599/885.

Dipping: Nocturnal

Expert Rev Cardiovasc Ther. 2010;8:781–92. http://informahealthcare.com/doi/abs/10.1586/erc.10.29.

Echo: Recommendations of ACE/EACVI

Eur Heart J Cardiovasc Imaging. 2015;16:577–605. http://ehjcimaging.oxfordjournals.org/content/16/6/577.full? .

Exercise: Benefits/Risks

Am J Hypertens. 2015;28:429–39. http://ajh.oxfordjournals.org/content/28/4/429.abstract?etoc.

Garlic

Am J Hypertens. 2015;28:414–23. http://ajh.oxfordjournals.org/content/28/3/414.abstract?etoc.

Genetics

Circulation Res.2015;116:937–59. http://circres.ahajournals.org/content/116/6/937.full.

Home BP Monitoring

J Nurse Pract. 2014;10:607–10. http://www.sciencedirect.com/science/article/pii/S1555415514004127.

Home-Clinic BP Difference: Predictors

Am J Hypertens. 2016;29:614–25. http://ajh.oxfordjournals.org/content/29/5/614.full?etoc.

Hydrochlorothiazide Vs Chlorthalidone

Hypertension. 2004;43:4–9. http://hyper.ahajournals.org/content/43/1/4.full.

Hypertension Guidelines (Editorial)

J Clin Hypertens. 2014;16:251–4. http://onlinelibrary.wiley.com/doi/10.1111/jch.12306/full.

JNC8 Panel Members Report

JAMA. 2014;311:507–20. http://jama.jamanetwork.com/article.aspx?articleid=1791497.

KDIGO: BP Goals

Kidney Int. 2012;[Suppl]:2. http://www.nature.com/kisup/journal/v2/n5/index.html.

Large Scale Program

JAMA. 2013;310:699–705. http://jama.jamanetwork.com/article.aspx?articleid=1730511.

Metabolic Syndrome

J Am Coll Cardiol. 2013;61:12–9. http://content.onlinejacc.org/article.aspx?articleID=1555196.

Misconceptions about Treatment

Am J Med. 2015;128:450–5. http://www.sciencedirect.com/science/article/pii/S0002934314011383.

Morning BP Surge

Circulation. 2003;107:1401–6. http://circ.ahajournals.org/content/107/10/1401.full.

Morning BP Surge: White Population

Hypertension. 2014;64:943–50. http://hyper.ahajournals.org/content/64/5/943.full.

Obesity

Circulation Res. 2015;116:991–1006. http://circres.ahajournals.org/content/116/6/991.full.

Obesity: Role of Aldosterone

Am J Hypertens. 2016;29:415–23. http://ajh.oxfordjournals.org/content/29/4/415.abstract?etoc.

Osler Maneuver

Am J Med. 1986;80:906–10. http://www.sciencedirect.com/science/article/pii/0002934386906364.

Pregnancy

Circulation. 2014;129:1254–61. http://circ.ahajournals.org/content/129/11/1254.full.

Pregnancy

N Engl J Med. 2015;372:407–17. http://www.nejm.org/doi/full/10.1056/NEJMoa1404595?query=TOC.

Pregnancy: Future Risk

Circulation. 2013;127:681–90. http://circ.ahajournals.org/content/127/6/681.full.

Pregnancy/Preeclampsia: Future Risk

J Am Coll Cardiol. 2014;63:1815–22. http://content.onlinejacc.org/article.aspx?articleID=1841463.

Prehypertension: CAD Risk in Asians

J Am Heart Assoc. 2015;4:e001519. http://jaha.ahajournals.org/content/4/2/e001519.full.

RAAS

Circulation Res. 2015;116:960–75. http://circres.ahajournals.org/content/116/6/960.full.

Renal Denervation

N Engl J Med. 2014;370:1393–401. http://www.nejm.org/doi/full/10.1056/NEJMoa1402670.

Renal Denervation

Eur Heart J. 2014;36:219–27. http://dx.doi.org/10.1093/eurheartj/ehu441.

Resistant HTN

Eur Heart J. 2015;36:2686–95. http://eurheartj.oxfordjournals.org/content/36/40/2686.

Resistant HTN

Int J Hypertens. 2011;598694. http://www.ncbi.nlm.nih.gov/pmc/articles/PMC3014709/.

Resistant HTN: Atherothrombosis

Eur Heart J. 2013;34:1204–14. http://eurheartj.oxfordjournals.org/content/34/16/1204.

Resting Heart Rate/Incident HTN

Am J Hypertens. 2016;29:251–7. http://ajh.oxfordjournals.org/content/29/2/251.abstract?etoc.

Screening: USPSTF

Ann Intern Med. 2007;147:783–6. http://annals.org/article.aspx?articleid=737820.

Secondary HTN: Screening

Eur Heart J. 2014;35:1245–54. http://eurheartj.oxfordjournals.org/content/35/19/1245.

Self-monitoring/Self-titration

JAMA. 2014;312:799–808. http://jama.jamanetwork.com/article.aspx?articleid=1899205.

Sex/Ethnicity: Incidence/Outcomes

Heart. 2013;99:715–21. http://heart.bmj.com/content/99/10/715.full.

Sympathetic Nervous System

Circulation Res. 2015;116:976–90. http://circres.ahajournals.org/content/116/6/976.full.

Sympathetic Overactivity

Indian Heart J. 2014;66:686–90. http://www.sciencedirect.com/science/article/pii/S0019483214007202.

ST2

J Clin Hypertension. 2013;15:899–904. http://onlinelibrary.wiley.com/doi/10.1111/jch.12205/full.

Systolic BP and Risk

JAMA Intern Med. 2014;174:1252–61. http://archinte.jamanetwork.com/article.aspx?articleid=1881014.

Treatment: Cost Effectiveness

N Engl J Med. 2015;372:447–55. http://www.nejm.org/doi/full/10.1056/NEJMsa1406751?query=TOC.

Treatment: Outcomes (Value Trial)

Eur Heart J. 2016;37:955–64. http://eurheartj.oxfordjournals.org/content/37/12/955.abstract?etoc.

Treatment: Sodium Reduction/Weight Loss

JAMA. 1998;279:839–46. http://jama.jamanetwork.com/article.aspx?articleid=187347.

Treatment: Systolic BP/Outcomes

Am J Med. 2013;126:501–08. http://www.sciencedirect.com/science/article/pii/S0002934313000363.

Visceral Adiposity

J Am Coll Cardiol. 2014;64:997–1002. http://content.onlinejacc.org/article.aspx?articleID=1900739.

Visit to Visit Variability

Ann Intern Med. 2015;163:329–38. http://annals.org/article.aspx?articleID=2398909.

White Coat/Masked HTN

Hypertension. 2009;54:226–32. http://hyper.ahajournals.org/content/54/2/226.full.

Updates and More

https://clinicalguidecvd.com/htn

Chapter 54
Inappropriate Sinus Tachycardia

ICD-10 Code

R00.0 (TACHYCARDIA UNSPECIFIED)

Alternate Names/Abbreviation

IST

Description/Etiology

Resting daytime HR >100 bpm and 24-h average HR >90 BPM not explainable by physiologic demands or conditions associated with increased HR
Cause unknown

Comorbid Conditions

ANXIETY/ANXIETY DISORDER

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_54,

Demography

Females > males
More common in young adults

Pathophysiology

Form of dysautonomia, or intrinsic sinus node abnormality, or both, with no unified mechanism

Signs/Symptoms

BREATHING – DIFF (DYSPNEA)
CHEST – PALPITATIONS
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)
DIZZY/LIGHTHEADED/PRESYNCOPE
FATIGUE
HEART, RATE – RAPID (TACHYCARDIA) [2]
HEART, RATE, VAR – DECR [1]
MOOD – ANXIOUS

Differentiation

Ablation for Supraventricular Tachycardia
Acute Pulmonary Embolism
AMI
Anemia
Anxiety
Aortic Regurgitation
Dehydration
Drugs/substances including:

- Alcohol
- Anticholinergics
- Beta-blocker withdrawal

Caffeine
Catecholamines
Cocaine
Tobacco

Duchenne Muscular Dystrophy
Exercise-Induced
Fever
Hyperthyroidism
Hypoglycemia
Mitral Regurgitation
Myocardial Ischemia
Pain
Pericarditis
Pneumothorax
Postural Orthostatic Tachycardia

Complications

Tachycardia-induced Cardiomyopathy [3]

Laboratory

NS; useful to R/O other causes of tachycardia, such as:

Hyperthyroidism
Hypoglycemia

ECG

RATE – INCREASED (SINUS TACHYCARDIA) [4]

Imaging

NS
R/O OTHER CAUSES OF TACHYCARDIA

Other Tests

24-h Ambulatory ECG
Tilt-table testing [5]

Treatment: Nonpharmacologic

Eliminate dietary stimulants, e.g., caffeine, alcohol
Psychiatric/psychologic consultation/support

Treatment: Pharmacologic

Benzodiazepines [6]
Beta-blockers [6]
Ivabradine

Treatment: Surgical/Invasive

Radiofrequency Ablation [7]
Surgical ablation [7]

Prevention

Avoid stimulants

Notes

[1] Often not present
[2] Rapid acceleration with exercise and slow return to baseline post-exercise
[3] Probably rare
[4] P wave morphology same during slow and fast rates

[5] For Postural Orthostatic Tachycardia
[6] Beta-blockers alone sometimes ineffective but may be more effective when taken in combination with a benzodiazepine
[7] Limited efficacy

Guidelines

2015 ACC/AHA/HRS Guideline for the Management of Adult Patients with Supraventricular Tachycardia
J Am Coll Cardiol. 2016;67:e27–115. http://content.onlinejacc.org/article.aspx?articleID=2443667.

Patient Information

Cardiac Health

http://www.cardiachealth.org/inappropriate-sinus-tachycardia-ist.

Professional Information

Review

J Am Coll Cardiol. 2013;61:793–801. http://content.onlinejacc.org/article.aspx?articleid=1486711&resultClick=3.

Review

Europace. 2005;7:104–12. http://europace.oxfordjournals.org/content/7/2/104.full.

Autoantibodies

Heart Rhythm. 2011;8:1788–95. http://www.heartrhythmjournal.com/article/S1547-5271(11)00787-9/abstract#SEC8.

RF Ablation

Circulation. 1995;92:2919–28. http://circ.ahajournals.org/content/92/10/2919.full.

Ivabradine

J Am Coll Cardiol. 2012;60:1330–2. http://content.onlinejacc.org/article.aspx?articleid=1358180.

Ivabradine: Efficacy

J Am Coll Cardiol. 2012;60:1323–9. http://content.onlinejacc.org/article.aspx?articleid=1358179.

Sympathetic Tone

J Am Heart Assoc. 2014;3:e000700. http://jaha.ahajournals.org/content/3/2/e000700.full.

Updates and More

https://clinicalguidecvd.com/ist

Chapter 55
Infective Endocarditis (Subacute Bacterial Endocarditis/SBE)

ICD-10 Code

133.0

Alternate Names/Abbreviation

IE
SBE
SUBACUTE BACTERIAL ENDOCARDITIS

Description/Etiology

Bacterial/fungal infection of valve/endocardium/intracardiac foreign material due to (most common):

Streptococcus and Staphylococcus (80 % of cases) [1]
Enterococcus [2]
Hacek organisms [3]
Candida and other fungi [4]
Culture-negative [5]

Associated with increased cancer risk
See Duke Criteria for diagnosis [19]

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 771
DOI 10.1007/978-1-4471-7293-2_55,

Comorbid Conditions

ACUTE MYOCARDIAL INFARCTION [9]
BACTERIAL MENINGITIS [23]
CANCER
CARDIAC SURGERY
CARDIOMYOPATHY – HYPERTROPHIC
COLON POLYP
COLONOSCOPY [ESP WITH POLYP REMOVAL]
COLORECTAL ADENOCARCINOMA
COLOSTOMY
CONGENITAL HEART DISEASE [7] [8]
DENTAL INFECTION
DENTAL PROCEDURE
DIABETES MELLITUS
DIABETIC FOOT ULCER
ESOPHAGEAL VARICES
ESOPHAGUS ULCER
GASTRIC ANTRAL/FUNDUS ATROPHY
GASTROSTOMY
HIV [6]
IMMUNE DEFICIENCY
INSECT BITE
INTRACARDIAC DEVICES
INTRAVENOUS DRUG USE/ABUSE [6]
LOWER GI ANGIODYSPLASIA
MITRAL VALVE PROLAPSE [25]
NONRHEUMATIC VALVE DISEASE
OPERATIVE SITE INFECTION
PENETRATING CARDIAC FOREIGN BODY
PERIODONTITIS
PROSTHETIC VALVE
PRURIGO
RHEUMATIC VALVE DISEASE
SIGMOID DIVERTICULOSIS
SKIN WOUND
SKIN/SOFT TISSUE INFECTION
VENOUS LEG ULCER
VENOUS LINE – CENTRAL
VENOUS LINE – PERIPHERAL

Demography

Per underlying condition
Increased incidence in USA since 2007

Incubation

Varies with infecting agent

Signs/Symptoms

ABDOMEN, LUQ – TENDER [13]
CHEST, ANT, STERNUM – PAIN/TENDERNESS [14] [15]
CHILLS
COUGH [15]
EXTREM, HANDS/FEET – HEMORRHAGES, SUBUNGUAL (SPLINTER)
EXTREM, HANDS/FEET – NODULES, TENDER (OSLER NODES)
EXTREM, LONG BONES – PAIN/TENDERNESS [14] [15]
EYES, CONJUNCTIVAE – PETECHIAE
EYES, RETINA – WHITE SPOT AND HEMORRHAGE (ROTH SPOT)
EYES, VISION – DECR/LOSS
FATIGUE
FEVER [10] [11]
HEART – FRICTION RUB, PERICARD [15]
HEART – MURMUR, REGURG [12]
JOINTS – PAIN (ARTHRALGIA)
SKIN – PETECHIAE/ECCHYMOSES/PURPURA
SKIN, FEET/SOLES – MACULES, RED (JANEWAY LESION)
SPLEEN – TENDER [12]
SPLEEN, SIZE – INCR (SPLENOMEGALY)
URINE – BLOOD (HEMATURIA) [15]
VOICE – HOARSE (LARYNGITIS) [15]

Differentiation

All causes of unexplained fever lasting >72 h

Complications [26]

ACUTE VALVE REGURGITATION
ANNULAR ABSCESS
BLEEDING
CARDIAC FISTULA
CARDIAC PERFORATION
CEREBRAL ABSCESS
CEREBRAL MYCOTIC ANEURYSM
CORONARY ARTERY EMBOLUS
HF
INFECTIVE PERICARDITIS
MYOCARDIAL ABSCESS
PROSTHETIC VALVE SEPARATION FORM ANNULUS
PROSTHETIC VALVE OBSTRUCTION
RENAL INFARCTION
SPLENIC ABSCESS
STROKE
SYSTEMIC ARTERIAL EMBOLUS [27]

Laboratory [19]

BLOOD, CULTURE – POS [5] [19]
BLOOD, ESR – INCR
BLOOD, HGB/HCT – DECR (ANEMIA)
BLOOD,LYMPHOCYTES–INCR(LYMPHOCYTOSIS)
BLOOD, PLATELETS – DECR (THROMBOCYTOPENIA) [15]
BLOOD, RHEUMATOID FACTOR – POS
BLOOD, TROPONIN – INCR [WITH MYOCARD EXTENSION]
BLOOD, WBC – INCR (LEUKOCYTOSIS)
URINE, PROTEIN – INCR (PROTEINURIA)

ECG [16]

N/NS ABN
NS/VAR PER COMORBIDITY(S) [ESP VALVULAR HD/INTRACARD DEVICES]

Imaging [19] [21] [26] [29]

AV, FLOW – REGURG [NEW]
CARD ABSCESS
CARD MASS – OSCILLATING [20]
CARD VALVE, NATIVE – MASS [VEGETATION]
CARDIAC VALVE, PROS – DEHISCENCE
CARDIAC VALVE, PROS – MASS [VEGETATION]
MV, FLOW – REGURG [NEW]

Other Tests

NS

Treatment: Nonpharmacologic

Refer to guidelines for current recommendations

Treatment: Pharmacologic

Antibiotics [17]
Anticoagulation (controversial)
Refer to guidelines for current recommendations

Treatment: Surgical/Invasive [24] [26] [28]

Case-by-case basis for treatment of infection and sequelae of valve leaflet and paravalvular tissue destruction [18] [24]

Prevention

IE prophylaxis [22]

Course

Mortality 40 % at 1 year; 15–20 % in-hospital
Increased long-term risks include:

All-cause mortality
AMI
HF
Stroke
Sudden death
Ventricular arrhythmias

Notes

[1] 80 % of TV infections are Staph Aureus; increased incidence due to Streptococcus since 2007
[2] Usually after GU/GI procedure; patients with malignancy
[3] Hemophilus, Actinobacillus, Cardiobacterium, Eikenella, Kingella species; cause large vegetations, large vessel embolism, HF
[4] Especially prosthetic valve IE
[5] 10 % negative due to prior antibiotic use; Candida; Aspergillus; slow growing organisms, eg, Coxiella Burnetii (Q Fever), Bartonella; Libman-Sacks (Systemic Lupus Erythematosus)
[6] Injection drug use or long-term indwelling central catheter
[7] Atrial Septal Defect uncommon cause except Atrioventricular type
[8] Most often with high pressure gradients
[9] At site of infarction or complicating ventricular aneurysm
[10] May be absent, especially with advanced age, prior antibiotic use, CNS hemorrhage, HF, renal failure
[11] Fever duration >72 h plus any murmur should raise suspicion of IE

[12] Changing or worsening of pre-existing murmur not diagnostic
[13] Splenic origin; acute stage
[14] Especially with severe anemia
[15] Uncommon/rare; when present initially in native left valve endocarditis, an early risk indicator of increased mortality
[16] Monitor for complications of heart block and dysrhythmias associated with septal abscess or Myocarditis
[17] Specific agents guided by causative organisms and susceptibility profiles; refer to current guidelines for detailed recommendations
[18] With heart valve team approach involving cardiologists, surgeons, infectious disease specialists
[19] Duke Criteria (Modified) for Infective Endocarditis (from Circulation 2015;132:1435–1486)

Major criteria

Blood culture positive for IE

Typical microorganisms consistent with IE from two separate blood cultures: Viridans streptococci, *Streptococcus bovis*, HACEK group, *Staphylococcus aureus*; or community-acquired enterococci in the absence of a primary focus, or microorganisms consistent with IE from persistently positive blood cultures defined as follows: at least two positive cultures of blood samples drawn >12 h apart or all 3 or a majority of ≥4 separate cultures of blood (with first and last sample drawn at least 1 h apart)

Single positive blood culture for *Coxiella burnetii* or anti–phase 1 IgG antibody titer ≥1:800

Evidence of endocardial involvement

Echocardiogram positive for IE (TEE recommended for patients with prosthetic valves, rated at least possible IE by clinical criteria, or complicated IE [paravalvular abscess]; TTE as first test in other patients) defined as follows: oscillating intracardiac mass on valve or supporting structures, in the path of regurgitant jets, or on implanted material in the absence of an alternative anatomic explanation; abscess; or new partial dehiscence of prosthetic valve or new valvular regurgitation (worsening or changing or pre-existing murmur not sufficient)

Minor criteria

Predisposition, predisposing heart condition, or IDU

Fever, temperature >38 °C

Vascular phenomena, major arterial emboli, septic pulmonary infarcts, mycotic aneurysm, intracranial hemorrhage, conjunctival hemorrhages, and Janeway lesions

Immunological phenomena: glomerulonephritis, Osler nodes, Roth spots, and rheumatoid factor

Microbiological evidence: positive blood culture but does not meet a major criterion as noted above (excludes single positive cultures for coagulase-negative staphylococci and organisms that do not cause endocarditis) or serological evidence of active infection with organism consistent with IE

[20] On valve, supporting structure, in path of regurgitant jet, implanted material

[21] TTE/TEE highly useful for all listed findings; CT indicated when echo insufficient

[22] Refer to guidelines

[23] Uncommon comorbidity, with mortality >60 %

[24] Surgical adverse outcomes correlate with:

Increasing age
Impaired LVEF (mod-severe)
Prosthetic valve IE
Staph aureus infection

[25] Infection on valve itself or in rare cases due to jet lesion in LA mural endocardium

[26] **Clinical and Echocardiographic Features That Suggest Potential Need for Surgical Intervention:** (from Circulation 2015;132:1435–1486)

Vegetation

Persistent vegetation after systemic embolization
Anterior mitral leaflet vegetation, particularly with size >10 mm ≥1 Embolic events during first 2 weeks of antimicrobial therapy
Increase in vegetation size despite appropriate antimicrobial therapy

Valvular dysfunction

Acute AR or MR with signs of ventricular failure
HF unresponsive to medical therapy

Valve perforation or rupture

Perivalvular extension
Valvular dehiscence, rupture, or fistula
New heart block
Large abscess or extension of abscess despite appropriate antimicrobial therapy

[27] Systemic emboli:

Occur in 22–25 % of cases of IE; rates can be higher if noninvasive imaging (including MRI/CT) routinely done to detect asymptomatic (silent) emboli
Often involve major arterial beds, including brain, lungs, coronary arteries, spleen, bowel, extremities
Up to 65 % of embolic events involve CNS, and >90 % of CNS emboli lodge in distribution of middle cerebral artery
Highest incidence of embolic complications is seen with MV IE (more with anterior rather than posterior mitral leaflet involvement) and with IE caused by *S aureus*, *Candida*, and HACEK organisms.
Emboli can occur before diagnosis, during treatment, or after treatment completion, although most emboli occur within first 2–4 weeks of antimicrobial therapy

[28] Valve surgery should be performed in most cases of fungal IE

[29] **Use of Echocardiography During Diagnosis and Treatment of Endocarditis** (from Circulation 2015;132:1435–1486)

Early

Echocardiography as soon as possible (<12 h after initial evaluation)
TEE preferred; obtain TTE views of any abnormal findings for later comparison

TTE if TEE is not immediately available
TTE may be sufficient in small children

Repeat echocardiography

TEE after positive TTE as soon as possible in patients at high risk for complications
TEE 3–5 days after initial TEE if suspicion exists without diagnosis of IE or with worrisome clinical course during early treatment of IE

Intraoperative/prepump

Identification of vegetations, mechanism of regurgitation, abscesses, fistulas, and pseudoaneurysms

Postpump

Confirmation of successful repair of abnormal findings
Assessment of residual valve dysfunction
Elevated afterload if necessary to avoid underestimating valve insufficiency or presence of residual abnormal flow

Completion of therapy

Establish new baseline for valve function and morphology and ventricular size and function
TTE usually adequate; TEE or review of intraoperative TEE may be needed for complex anatomy to establish new baseline

Guidelines

2014 AHA/ACC guideline for the management of patients with valvular heart disease
J Am Coll Cardiol. 2014;63:e57–185. http://content.onlinejacc.org/article.aspx?articleID=1838843.
2015 ESC guidelines for the management of infective endocarditis
Eur Heart J. 2015;36:3075–128. http://eurheartj.oxfordjournals.org/content/36/44/3075.

Patient Information

IMAGES

http://www.nlm.nih.gov/medlineplus/ency/imagepages/18142.htm.

Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/endocarditis/basics/definition/con-20022403.

AHA

http://www.heart.org/HEARTORG/Conditions/Congenital-HeartDefects/TheImpactofCongenitalHeartDefects/Infective-Endocarditis-UCM-307108-Article.jsp.

Merck

http://www.merckmanuals.com/home/heart-and-blood-vessel-disorders/endocarditis/infective-endocarditis.

Cleveland Clinic

http://my.clevelandclinic.org/services/heart/disorders/heart-valve-disease/endocarditis-protection.

Texas Heart Institute

http://www.texasheart.org/HIC/Topics/Cond/endocard.cfm.

Professional Information

AHA Scientific Statement: Adults

Circulation. 2015;132:1435–86. http://circ.ahajournals.org/content/132/15/1435.full.

AHA Scientific Statement: Children

Circulation. 2015;132:1487–515. http://circ.ahajournals.org/content/132/15/1487.full.

Early Description

Lancet. 1885. http://www.sciencedirect.com/science/article/pii/S0140673602008279.

Review: Critical Questions

J Am Coll Cardiol. 2015;66:1068–76. http://content.onlinejacc.org/article.aspx?articleID=2430614.

Review

N Engl J Med. 2013;368:1425–33. http://www.nejm.org/doi/full/10.1056/NEJMcp1206782.

Review

Arch Intern Med. 2009;169463–73. http://archinte.jamanetwork.com/article.aspx?articleid=414841.

Review: Congenital Heart Disease

Circulation. 2013;128:1412–19. http://circ.ahajournals.org/content/128/13/1412.full.

Review: Transaortic Valve IE

J Infect. 2015;70:565–76. http://www.sciencedirect.com/science/article/pii/S0163445314003843.

Cancer Risk

Am J Med. 2013;126:58–67. http://www.sciencedirect.com/science/article/pii/S0002934312007905.

Duke Criteria for Diagnosis

Clin Infect Dis. 2000;30:633–8. http://cid.oxfordjournals.org/content/30/4/633.full.

Enterococcus Faecalis Endocarditis

Circulation. 2013;127:1810–7. http://circ.ahajournals.org/content/127/17/1810.full.

Entry Portals

J Am Coll Cardiol. 2016;67:151–8. http://content.onlinejacc.org/article.aspx?articleID=2480639.

Epidemiology

J Am Coll Cardiol. 2012;59:1977–8. http://content.onlinejacc.org/article.aspx?articleid=1203852.

Epidemiology

PLoS One. 2013;8:e82665. http://journals.plos.org/plosone/article?id=10.1371/journal.pone.0082665.

IE with Bacterial Meningitis

Circulation. 2013;127:2056–62. http://circ.ahajournals.org/content/127/20/2056.full.

Imaging

Eur Heart J. 2014;35:624–32. http://eurheartj.oxfordjournals.org/content/35/10/624.

Imaging: Echo

Heart. 2004;90:614–7. http://www.ncbi.nlm.nih.gov/pmc/articles/PMC1768290/.

Imaging: CT

J Am Coll Cardiol. 2009;53:436–44. http://content.onlinejacc.org/article.aspx?articleid=1139382.

Imaging: 3D Echo

Circulation Card Imaging. 2014;7:149–54. http://circimaging.ahajournals.org/content/7/1/149.full?sid=aeab4f82-993f-45e1-ab08-efbec6dfe9ee.

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Eur Heart J. 2015;36:2484–90. http://eurheartj.oxfordjournals.org/content/36/37/2484.

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JAMA. 2012;307:1727–35. http://jama.jamanetwork.com/article.aspx?articleid=1148195.

LA Mural Endocarditis Secondary to Jet Lesion

Circulation. 2015;131:1529–30. http://circ.ahajournals.org/content/131/17/1529.full.

Management

Lancet. 2012;379:965–75. http://www.ncbi.nlm.nih.gov/pubmed/22317840?dopt=Abstract.

Neurologic Complications

Circulation. 2013;127:2272–84. http://circ.ahajournals.org/content/127/23/2272.full?sid=87d99194-91b1-4b54-ae6a-80b6c2cdedf9.

Outcomes: Long-Term

Circulation. 2014;130:1684–91. http://circ.ahajournals.org/content/130/19/1684.full.

Persistent Positive Blood Cultures: Prognosis

Eur Heart J. 2013;34:1749–54. http://eurheartj.oxfordjournals.org/content/34/23/1749.

Relapse Versus Reinfection

Clin Infect Dis. 2005;41:406–9. http://cid.oxfordjournals.org/content/41/3/406.abstract?ijkey=d759f594ada1e239672a304801ff9e05f4cd9c55&keytype2=tf-ipsecsha.

Roth Spots

Engl J Med. 2014;370:e38. http://www.nejm.org/doi/full/10.1056/NEJMicm1312093.

Septic Shock

Eur Heart J. 2013;34:1999–2006. http://eurheartj.oxfordjournals.org/content/34/26/1999.

Surgery: Outcomes/Prognostic Factors

Eur J Cardiothorac Surg. 2015;47:826–32. http://ejcts.oxfordjournals.org/content/47/5/826.abstract.

Surgery: Early

N Engl J Med. 2012;366:2466–73. http://www.nejm.org/doi/full/10.1056/NEJMoa1112843#t=articleDiscussion.

Surgery: Indications, Risks, Outcomes

Circulation. 2015;131:131–40. http://circ.ahajournals.org/content/131/2/131.abstract?etoc.

Surgery: Long-Term Outcomes

Eur Heart J. 2014;35:1195–204. http://eurheartj.oxfordjournals.org/content/35/18/1195.

Surgery: Clinical Features/Outcomes

QJM. 2015;108:219–29. http://qjmed.oxfordjournals.org/content/108/3/219.abstract?etoc.

Thrombocytopenia: Risk Marker for INCR Mortality

An J Cardiol. 2015;115:950–5. http://www.ajconline.org/article/S0002-9149(15)00056-9/abstract.

Trends in USA: Hospitalizations/Outcomes

J Am Coll Cardiol. 2013;62:2217–26. http://content.onlinejacc.org/article.aspx?articleID=1732398.

Trends in USA: Incidence, Microbiology, Valve Replacement

J Am Coll Cardiol. 2015;65:2070–6. http://content.onlinejacc.org/article.aspx?articleID=2290815.

Updates and More

https://clinicalguidecvd.com/ie

Chapter 56
Leopard Syndrome

ICD-10 Code

Q87.1

Alternate Names/Abbreviation

Leopard: acronym for Lentigines, ECG-changes, ocular changes (Hypertelorism), Pulmonary Stenosis, anomalies in sex organs, growth retardation, deafness

CARDIOCUTANEOUS SYNDROME
LENTIGINOSIS PROFUSE
MOYNAHAN SYNDROME
MULTIPLE LENTIGINES SYNDROME
PROGRESSIVE CARDIOMYOPATHIC LENTIGINOSIS

Description/Etiology

Autosomal dominant inherited condition affecting many areas of body, especially

Skin: brown spots (Lentigines) resembling freckles and Cafe Au Lait spots

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_56,

Heart:

Conduction system
LVH
PS

Facial structures
Genitalia
Stature: short
Hearing (loss)

High penetrance and variable expression
Closely resembles Noonan Syndrome, especially in childhood [2]

Comorbid Conditions

ATRIAL FIBRILLATION
CARDIOMYOPATHY – HYPERTROPHIC
CELIAC DISEASE
PREEXCITATION SYNDROMES
PULMONARY STENOSIS

Pathophysiology

Cardiac structural: primarily Hypertrophic Cardiomyopathy and Pulmonary Valve Stenosis

Signs/Symptoms [3] [4]

BACK, CURV – LAT (SCOLIOSIS)
BACK, CURV – POST (KYPHOSIS)
BODY, GROWTH – DECR
BODY, HT – DECR
BREATHING – DIFF (DYSPNEA)
CHEST – PAIN, EFFORT (ANGINA PECTORIS)
CHEST – PALPITATIONS

CHEST, SHAPE – BARREL
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)
EARS, HEARING – LOSS (DEAFNESS) [8]
EARS, POSITION – LOW SET
EYES, LIDS – DROOPED (PTOSIS)
EYES, MOTION – JERKY (NYSTAGMUS)
EYES, SEPARATION – WIDE (HYPERTELORISM)
FACE, BROWS – PROTRUSION (FRONTAL BOSSING)
FACE, SHAPE – TRIANGULAR
FATIGUE
FERTILITY – DECR
GENITALS, TESTICLES, DESCENT – DELAYED/ABSENT (CRYPTORCHIDISM)
HEART, LSB, LOWER – THRILL, SYS
HEART, LSB, MID – MURMUR, SYS
HEART, LV, APEX – MURMUR, DIAS
HEART, LV, APEX – MURMUR, SYS
HEART, LV, APEX, IMP – FORCEFUL/SUSTAINED
HEART, LV, APEX, IMP – FORCEFUL/SUSTAINED
HEART, LV, APEX, IMP – DISPLACED, LAT
HEART, LV, APEX, IMP – DOUBLE
HEART, LV, APEX, IMP – TRIPLE
HEART, S2, SPLIT – REVERSED (PARADOXICAL)
HEART, S3 LV
JOINTS, MOVEMENT, RANGE – INCR (HYPERMOBILITY)
LIPS – THICK
MENTATION, LEARNING, DEVELOPMENT – DECR
NECK, POST, SKIN FOLDS – EXCESS
PENIS, OPENING – UNDERSIDE (HYPOSPADIAS)
PUBERTY, ONSET – DELAYED
SEIZURES
SKIN – LENTIGINES [1]
SKIN – SPOTS, HYPERPIGMENTED (CAFE – AU-LAIT) [9]
STERNUM, CURV – ANT (PECTUS CARINATUM)
STERNUM, CURV – POST (PECTUS EXCAVATUM)

Differentiation

Cardiac: other causes of LVOT obstruction

Complications

AF
Arterial dissection
HF
Sudden Death

Laboratory

NS

ECG

AV COND – 3RD DEGREE BLOCK
DYSRHYTHMIAS – ATRIAL (PACS/OTHERS)
DYSRHYTHMIAS – VENTRICULAR (PVCS/ OTHERS)
P WAVE, MORPH – VAR/ABN
Q WAVE – ABN
QRS – LVH PATTERN
QRS – RBBB/RBBB PATTERN
QRS – RVH PATTERN
QRS – S1S2S3 PATTERN
QRS, AXIS – L
QT/QTC INTERVAL – LONG
RATE – DECREASED (SINUS BRADYCARDIA)

Imaging [7]

ART, CORONARY, SIZE – INCR/ANEURYSM
AV, FLOW – REGURG

AV, LEAFLETS, MORPH – ABN
IVS, THICKNESS – INCR (SEPTAL HYPERTROPHY)
LA, CHAMBER, SIZE – INCR
LV, DIAS – DYSF
LV, MYOCARD, WALL THICKNESS – INCR (HYPERTROPHY) [5]
LV, OUTFLOW – OBS
LV, WALL THICKNESS, SEG – INCR [6]
MV, LEAFLETS – PROLAPSE
MV, LEAFLETS, MORPH – ABN
MV, MOTION SYS – ANT
PV, LEAFLETS, MORPH – ABN

Genomics

BRAF
PTPN11 [?]
RAF1

Other Tests

Ambulatory ECG monitoring
Cardiac Catheterization
EEG
Skin biopsy

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic

Variable according to associated abnormalities

Treatment: Surgical/Invasive

Septal ablation for LVOT obstruction
Variable according to associated abnormalities

Course

Highly variable; cardiac prognosis related to presence of LVH

Notes

[1] Brown spots, more common on face, neck, upper body; unaffected by sunlight; cardiac abnormalities may precede their appearance
[2] Differs from Noonan: deafness, Hypertrophic Cardiomyopathy, Lentigines more common in Leopard
[3] Highly variable, even within same family
[4] Cardiac signs/symptoms listed are due to LVOT obstruction caused by HCM; signs/symptoms of PS not listed
[5] LVH may be concentric or nonconcentric
[6] Isolated to cardiac apex reported in a few cases
[7] Great variability among reported cases; asymmetric and concentric LVH most common abnormalities; other rare findings sometimes found and not listed include:

Aortic Stenosis – Subvalvular
AV canal defect
LV dilatation
LV noncompaction
Multiple VSDs
LV apical aneurysm
RV moderator band

[8] Sensorineural; may be present at birth or develop later
[9] May precede Lentigines appearance

Guidelines

2014 ESC guidelines on diagnosis and management of hypertrophic cardiomyopathy
Eur Heart J. 2014;35:2733–79. http://eurheartj.oxfordjournals.org/content/35/39/2733.

Patient Information

Images

http://www.nlm.nih.gov/medlineplus/ency/imagepages/2927.htm.

Medlineplus

ENGLISH
http://www.nlm.nih.gov/medlineplus/ency/article/001473.htm.
ESPANOL
http://www.nlm.nih.gov/medlineplus/spanish/ency/article/001473.htm.

Nord

https://rarediseases.org/rare-diseases/leopard-syndrome/.

Genetics Home Reference

http://ghr.nlm.nih.gov/condition/multiple-lentigines-syndrome.

Professional Information

Review

Rev Esp Cardiol. 2013;66:350–6. http://www.revespcardiol.org/en/leopard-syndrome-a-variant-of/articulo/90198894/.

Arterial Dissection

N Engl J Med. 1995;332:576–9. http://www.nejm.org/doi/full/10.1056/NEJM199503023320905.

Cardiovascular Manifestations

Am J Cardiol. 2007;100:736–41. http://www.sciencedirect.com/science/article/pii/S000291490700968X.

Case Series

J Med Genet. 2004;41:5 e68. http://jmg.bmj.com/content/41/5/e68.full.

Images

Circulation. 2009;119:1328–9. http://circ.ahajournals.org/content/119/9.toc.

Images

Eur Heart J. 2007;28:3066. http://eurheartj.oxfordjournals.org/content/28/24/3066.full?sid=33a157c3-039c-4fd6-9021-f9f559b9c3c5.

Imaging: MRI

Eur Heart J. 2006;27:1407. http://eurheartj.oxfordjournals.org/content/27/12/1407.full?sid=33a157c3-039c-4fd6-9021-f9f559b9c3c5.

Updates and More

https://clinicalguidecvd.com/leopard

Chapter 57
Long QT Syndrome: Acquired (LQTS)

ICD-10 Code

I45.81

Alternate Names/Abbreviation

LQTS

Description/Etiology

Acquired long QT syndrome is manifest by ECG QT interval prolongation and Torsades de Pointes ventricular tachycardia triggered by drugs, electrolyte abnormalities, and other factors (see PREDISPOSING/COMORBID CONDITIONS)

In some persons QTc interval remains prolonged after elimination of triggers, suggesting an underlying genetic substrate as a causative factor

Predisposing/Comorbid Conditions

ANTIINFECTIVE DRUGS [2]
ATRIOVENTRICULAR HEART BLOCK

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 797
DOI 10.1007/978-1-4471-7293-2_57,

AUTONOMIC NEUROPATHY
CANCER CHEMOTHERAPY DRUGS [12]
CARDIOMYOPATHY – TAKOTSUBO
CARDIOVASCULAR DRUGS [1]
CENTRAL NERVOUS SYSTEM DRUGS [3]
CEREBRAL HEMORRHAGE
COCAINE
HIV
HYPOCALCEMIA
HYPOKALEMIA
HYPOMAGNESEMIA
HYPOTHERMIA
HYPOTHYROIDISM
LEFT VENTRICULAR HYPERTROPHY [4]
LONG QT SYNDROME – CONGENITAL
MYOCARDIAL FIBROSIS [4]
MYOCARDIAL ISCHEMIA [4]
NONALCOHOLIC FATTY LIVER DISEASE
ORGANOPHOSPHATES
PHEOCHROMOCYTOMA
POST-ACUTE MYOCARDIAL INFARCTION
PROPRIONIC ACIDEMIA [14]
PROTEIN-SPARING FASTING
SINUS NODE DYSFUNCTION
STROKE
SUBARACHNOID HEMORRHAGE

Demography

Variable per cause

Pathophysiology

Electrophysiological changes include:

Slowed outward repolarizing K currents
Enhanced inward calcium currents
Slowed inactivation of inward sodium currents

Signs/Symptoms

CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)
DIZZY/LIGHTHEADED/PRESYNCOPE

Differentiation

Congenital Long QT Syndrome [15]
Other conditions causing ECG QTc interval prolongation

Complications

Polymorphic VT [5]
SCD

Laboratory

NS [6]

ECG

JT INTERVAL – LONG [13]
QT/QTc INTERVAL – LONG
T WAVE – ALTERNANS [7]
U WAVE – ALTERNANS
U WAVE – PROMINENT

Imaging

NS/VAR WITH COMORBID

Genomics

[15]

Other Tests

Ambulatory ECG monitoring

Treatment: Nonpharmacologic

Correct underlying cause

Treatment: Pharmacologic

Torsades: IV MGSO4 bolus followed by continuous infusion
Torsades: IV K [8]
Torsades: Mexiletine
Isoproterenol [11]
Long-term: beta-blocker

Treatment: Surgical/Invasive

Temporary pacing [9] [10]
Permanent pacing [10]
ICD
Left thoracic sympathectomy

Prevention

Avoid/DC causative agent

Course

Variable according to underlying condition

Notes

[1] Includes but not limited to:

Amiodarone
Azimilide
Bepridil
Disopyramide
Ibutilide
Quinidine
Phenylephrine
Procainamide

[2] Includes but not limited to:

Amantadine
Chloroquine
Erythromycin
Grepafloxacin
Moxifloxacin
Pentamidine
Trimethoprim-sulfamethoxazole

[3] Includes but not limited to:

Arsenic trioxide
Astemizole
Cisapride
Haloperidol
Itraconazole
Ketanserin
Ketoconazole
Papaverine
Phenothiazine
Probucol
Tacrolimus
Tricyclic antidepressants

[4] Promote drug-induced LQTS
[5] Torsades de Pointes; occurs after prolonged QTc interval in preceding sinus beats; usually short-lived and resolves spontaneously but may deteriorate to VF and SCD
[6] Eg, abnormal electrolytes
[7] Beat-to-beat variation in T wave amplitude; associated with marked increased risk of cardiac arrest
[8] Adjunct to IV MGSO4
[9] Prevent short-term recurrence of torsades
[10] Especially effective if precipitated by sinus pause/bradycardia
[11] Contraindicated for Congenital Long QT Syndrome
[12] Includes but not limited to:

Arsenic Trioxide
Dasatinib
Lapatinib
Nilotinib
Vorinostat

[13] JT interval measure may be more accurate than QT in presence of prolonged QRS
[14] Propionic Acidemia: rare metabolic disorder due to propionyl-CoA carboxylase deficiency; affected persons have QT prolongation associated with VT/syncope
[15] Up to one-third of persons with Acquired LQTS may carry Congenital LQTS mutations, most often KCNH2

Guidelines

2013 ESC guidelines on cardiac pacing and cardiac resynchronization therapy

Eur Heart J. 2013:34;2318. http://www.escardio.org/guidelines-surveys/esc-guidelines/Pages/cardiac-pacing-and-cardiac-resynchronisation-therapy.aspx.

Patient Information

NIH

http://www.nhlbi.nih.gov/health/health-topics/topics/qt/.

Genetics Home Reference

http://ghr.nlm.nih.gov/glossary=longqtsyndrome.

ESPANOL
http://es.heart.org/dheart/HEARTORG/Conditions/Answers-by-Heart-Fact-Sheets-Multi-language-Information-UCM-314158-Article.jsp.

Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/long-qt-syndrome/basics/symptoms/con-20025388.

Cleveland Clinic

http://my.clevelandclinic.org/services/heart/disorders/arrhythmia/long-qt-syndrome.

Seattle Childrens

http://www.seattlechildrens.org/medical-conditions/heart-blood-conditions/long-qt-syndrome-symptoms/.

Texas Heart Institute

http://www.texasheart.org/HIC/Topics/Cond/longqts.cfm.

Professional Information

Arizona Health Foundation QT Drug List

http://crediblemeds.org/everyone/composite-list-all-qtdrugs/?rf=All.

Review: Drug-Induced QT Prolongation/ Torsades

J Am Coll Cardiol. 2016;67:1639–50. http://content.onlinejacc.org/article.aspx?articleID=2506371.

Atrial Fibrillation

J Am Coll Cardiol. 2008;51:836–42. http://content.onlinejacc.org/article.aspx?articleid=1138744.

Diagnostic Challenges

J Am Coll Cardiol. 2010;55:1962–4. http://content.onlinejacc.org/article.aspx?articleid=1142778#bib15.

Drug-Induced: Children

J Am Coll Cardiol. 2014;63:2272–9. http://content.onlinejacc.org/article.aspx?articleID=1851430.

Genetics

Eur Heart J. 2016;37:1456–64. http://eurheartj.oxfordjournals.org/content/37/18/1456.abstract?etoc.

JT Interval in Presence of Long QRS

Am J Cardiol. 2015;116:74–8. http://www.ajconline.org/article/S0002-9149(15)01044-9/abstract.

Mexiletine: Torsades

J Am Coll Cardiol EP. 2015;1:315–22. http://electrophysiology.onlinejacc.org/article.aspx?articleid=2411173&resultClick=24.

Nonalcoholic Fatty Liver DIS

J Am Heart Assoc. 2015;4:e001820. http://jaha.ahajournals.org/content/4/7/e001820.full.

Proprionic Acidemia

Heart Rhythm 2016: Feb (on line). http://www.heartrhythmjournal.com/article/S1547-5271(16)00137-5/abstract.

T Wave Alternans

Am J Med. 2015;128:480–3. http://www.sciencedirect.com/science/article/pii/S0002934315000741.

Updates and More

https://clinicalguidecvd.com/acqlqts

Chapter 58
Long QT Syndrome: Congenital

ICD-10 Code

I45.81

Alternate Names/Abbreviation

LQTS
ANDERSEN (ANDERSEN-TAWIL) SYNDROME
JERVELL-LANGE-NIELSEN (JLN) SYNDROME
ROMANO-WARD SYNDROME
TIMOTHY SYNDROME

Description/Etiology

Inherited ion channel disease (except LQT4) mainly manifest by life-threatening arrhythmias triggered by sudden increase in sympathetic activity, mediated by left-sided cardiac sympathetic nerves

Many affected persons also have hearing loss, seizures independent of ventricular dysrhythmias, and other neurodevelopmental anomalies

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_58,

Etiology: genetic mutations affecting transmembrane Na or K ion channel proteins, comprising several Subtypes:

LQT1
LQT2
LQT3
LQT4
LQT5
LQT6
LQT7 [Andersen-Tawil Syndrome] [7]
LQT8 [Timothy Syndrome] [8]
LQT9
LQT10
LQT12
LQT13

Frequency:

LQT1–2: 90 % of cases
LQT3: 5 % of cases
LQT4 +: extremely rare

Predisposing/Comorbid Conditions

ATRIAL FIBRILLATION [12]
FAMILY HX: SUDDEN DEATH
SEIZURES [2]
SENSORINEURAL HEARING LOSS [1]

Demography

About 85 % inherited from 1 parent; remainder sporadic
Family history of SCD
Males at greater risk (especially LQT1) for arrhythmic event/SCD before puberty
Females have greater risk (especially LQT2) during prepuberty, postmenopause, postpartum

Pathophysiology

Abnormal K and Na channel and channel-related proteins cause positive overcharge of myocytes and heterogeneous prolongation of repolarization in myocardial layers and regions

Signs/Symptoms

CHEST – PALPITATIONS
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE) [14]
DIZZY/LIGHTHEADED/PRESYNCOPE
EARS, HEARING – LOSS (DEAFNESS) [1]
HEART, RATE – SLOW (BRADYCARDIA) [13]
SEIZURES

Differentiation

Acquired Long QT Syndrome
ARVD
Brugada Syndrome
Hypertrophc cardiomyopathy
Other causes of seizures
Other causes of sensorineural hearing loss
Other causes of ventricular dysrhythmias

Complications [24]

Polymorphic VT [5] [14]
SCD [14]

Laboratory

NS

ECG [3]

AV COND – 2ND DEGREE BLOCK, MOBITZ I (WENCKEBACH)
DYSRHYTHMIAS – ATRIAL (PACS/OTHERS) [12]
DYSRHYTHMIAS – VENTRICULAR (PVCS/OTHERS) [4]
JT INTERVAL – LONG [23]
QRS, AMP – VAR, RHYTHMIC
QT/QTC INTERVAL – LONG [19]
RATE – DECREASED (SINUS BRADYCARDIA) [12]
T WAVE – ALTERNANS [21]
T WAVE – NOTCHED [17]
T WAVE – POLYPHASIC [12]
T WAVE – TALL/PEAKED [15]
T WAVE – WIDE [18]
T WAVE, AMP – DECR/FLAT [16]
U WAVE – ALTERNANS
U WAVE – PROMINENT

Imaging

NS/VAR WITH COMORBID

Genomics [6] [24]

ACNK2
AKAP9
CACNA1C
CAV3
KCNE1
KCNE2
KCNH2
KCNJ2
KCNJ5

KCNQ1 [?]
SCN4B
SCN5A
SNTA1

Other Tests

Ambulatory 24 h ECG monitoring
Exercise test: QTc prolongation in patients with LQTS
Epinephrine infusion: QTc prolongation in patients with LQTS

Treatment: Nonpharmacologic

Avoid/caution in competitive sports [22]
Stress reduction (avoidance of alarm clocks, noise, etc.)
AED immediately available, e.g., at home

Treatment: Pharmacologic

Torsades: IV MGSO4 bolus followed by cont infusion
Torsades: IV K [9]
LQT1 and LQT2 (long-term): Beta-blockers [11]
LQT3 (long-term): Flecainide, Mexiletine [20]
LQT3 with KPQ mutation (short-term): IV Ranolazine [20]

Treatment: Surgical/Invasive

Temporary pacing [10]
Permament pacing [10]
ICD
Left thoracic sympathectomy

Notes

[1] Due to absent functional KCNQ1-KCNE1 pores in cochlea; occurs only in JLN Syndrome
[2] Independent of dysrhythmias
[3] T wave morphology, ST-T, repolarization pattern differ according to genotype
[4] Especially multifocal PVCs and Torsades de Pointes
[5] Torsades de Pointes; occurs after prolonged QTc in preceding sinus beats; usually short-lived and resolves spontaneously but may deteriorate to VF and SCD
[6] Mutations of KCNQ1, KCNH2, and SCN5A comprise most cases; all LQTS not yet genotyped
[7] Long QTc plus K-sensitive periodic paralysis, ventricular dysrhythmias, dysmorphic features
[8] Long QTc plus bradycardia, AV Block, other congenital cardiac defects and multiple other organ systems; longevity usually short with mean age of death at 2.5 years
[9] Adjunct to IV MGSO4
[10] Especially effective if precipitated by sinus pause/bradycardia
[11] Main efficacy in LQT1 and LQT2; decreased risk for cardiac event by >60 %; syncope occurring while on beta-blocker appears to convey no greater protection than on no drug treatment; efficacy may vary for preventing events in high-risk patients
[12] Especially LQT5, with AF and intense sinus bradycardia common
[13] May worsen with beta-blockers
[14] May be precipitated by:

Awakening from sleep (especially LQT3)
Emotions
Exercise (especially LQT1), particularly while swimming/diving
Stress
Sudden noise (especially LQT2)

[15] Especially LQT3
[16] Especially LQT2

[17] Especially LQT2
[18] Especially LQT1
[19] When >500 msc: highly likely to be LQTS
[20] Limited data but may be efficacious with decreased QTc interval and major reduction of life-threatening arrhythmic events in LQT3 patients
[21] T wave alternans associated with marked increasedly risk of cardiac arrest
[22] Outcome studies lacking; however, limited data show that in treatment-compliant persons, no adverse events occur in association with sports participation
[23] JT interval measure may be more accurate than QT in presence of prolonged QRS
[24] Family members should have genomic testing as asymptomatic carriers have 10× increased risk of cardiac events

Guidelines

HRS/EHRA/APHRS expert consensus statement on the diagnosis and management of patients with inherited primary arrhythmia syndromes

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Patient Information

NIH

http://www.nhlbi.nih.gov/health/health-topics/topics/qt/.

Genetics Home Reference

http://ghr.nlm.nih.gov/glossary=longqtsyndrome.

ESPANOL

http://es.heart.org/dheart/HEARTORG/Conditions/Answers-by-Heart-Fact-Sheets-Multi-language-Information-UCM-314158-Article.jsp.

Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/long-qt-syndrome/basics/symptoms/con-20025388.

Cleveland Clinic

http://my.clevelandclinic.org/services/heart/disorders/arrhythmia/long-qt-syndrome.

Seattle Childrens

http://www.seattlechildrens.org/medical-conditions/heart-blood-conditions/long-qt-syndrome-symptoms/.

Texas Heart Institute

http://www.texasheart.org/HIC/Topics/Cond/longqts.cfm.

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AHA Scientific Statement: Genomics

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Arizona Health Foundation QT Drug List

http://crediblemeds.org/everyone/composite-list-all-qtdrugs/?rf=All.

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Eur Heart J. 2013;34:3109–16. http://eurheartj.oxfordjournals.org/content/34/40/3109.

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Genetics Home Reference: Andersen-Tawil Syndrome

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Genetics Home Reference: Jervell and Lange-Nielsen Syndrome

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Genetics Home Reference: Romano Ward Syndrome

http://ghr.nlm.nih.gov/condition/romano-ward-syndrome.

Genetics Home Reference: Timothy Syndrome

http://ghr.nlm.nih.gov/condition/timothy-syndrome.

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J Am Coll Cardiol. 1999;33:1415–23. http://content.onlinejacc.org/article.aspx?articleid=1125732.

Beta-Blockers: Differing Efficacy

J Am Coll Cardiol. 2014;64:1352–8. http://content.onlinejacc.org/article.aspx?articleID=1909594.

ECG Imaging: Electrophysiologic Substrate

Circulation. 2014;130:1936–43. http://circ.ahajournals.org/content/130/22/1936.full.

EPILEPSY

JACCCEP. 2016. doi:10.1016/j.jacep.2015.10.010. http://electrophysiology.onlinejacc.org/article.aspx?articleid=2482972.

Epinephrine Test

Circulation. 2006;113:1385–92. http://circ.ahajournals.org/content/113/11/1385.full.

Exercise Test

Circulation. 2003;107:838–44. http://circ.ahajournals.org/content/107/6/838.full.

Genotype-Specific ECG Patterns

J Electrocardiol. 2006;39(4 Suppl):S101–6. http://www.ncbi.nlm.nih.gov/pubmed/16963070.

JT Interval in Presence of Long QRS

Am J Cardiol. 2015;116:74–78. http://www.ajconline.org/article/S0002-9149(15)01044-9/abstract.

Longitudinal Family Study

Circulation. 1991;84:1136–44. http://www.ncbi.nlm.nih.gov/pubmed/1884444.

Mask with Many Faces

J Am Coll Cardiol. 2008;51:930–2. http://content.onlinejacc.org/article.aspx?articleid=1138772.

Mexiletine

J Am Coll Cardiol. 2016;67:1053–8. http://content.onlinejacc.org/article.aspx?articleID=2498347.

QT Clock

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(Editorial)

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T Wave Alternans

Am J Med. 2015;128:480–3. http://www.sciencedirect.com/science/article/pii/S0002934315000741.

Ventricular Arrhythmias in Channelopathies: Management

Circulation Arrhythmia Electrophysiol. 2015;8:221–31. http://circep.ahajournals.org/content/8/1/221.extract?etoc.

Updates and More

https://clinicalguidecvd.com/conlqts

Chapter 59
Lower Exremity Artery Disease

ICD-10 Code

I74.3

Alternate Names/Abbreviation

LEAD
ARTERIOSCLEROSIS OBLITERANS
LERICHE SYNDROME [8]
LOWER EXTREMITY ARTERY DISEASE
PERIPHERAL ARTERIAL DISEASE (PAD)

Description/Etiology

A form of peripheral artery disease involving obstruction of arterial blood flow to pelvic organs/structures and lower extremities

Atherosclerosis most common cause but nonatherosclerotic forms exist, including:

Adventitial cysts
Aneurysmal disease
Congenital abnormalities
Entrapment syndromes

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_59,

Fibromuscular Dysplasia
Inflammatory disease
Thromboembolic disease
Trauma

Underdiagnosed due to: [19]

High prevalence of asymptomatic patients with LEAD
Wide range of presenting leg symptoms

Predispoisng/Comorbid Conditions

ADVENTITIAL CYSTS
ATHEROSCLEROSIS IN OTHER CV AREAS [1]
DIABETES MELLITUS
DYSLIPIDEMIA
ENTRAPMENT SYNDROME
GOUT
HYPERCHOLESTEROLEMIA – FAMILIAL
HYPERHOMOCYSTEINEMIA
HYPERTENSION – SYSTEMIC ARTERIAL
INFLAMMATION
THROMBOEMBOLISM
TOBACCO USE
TRAUMA

Demography (Atherosclerotic Forms)

Age usually 70+ years except:

Age <50 years with DM and one or more other atherosclerotic risk factors
Age 50–69 years with history of tobacco use and DM

Pathophysiology

Mechanisms of functional impairment and decline in LEAD poorly understood

Calf muscles in patients with LEAD show:

- Apoptosis
- Atrophy
- Increased connective tissue
- Loss of type II muscle fibers

Signs/Symptoms [10]

ARTERIAL PULSE, DORSALIS PEDIS – DECR/ABSENT [9]
ARTERIAL PULSE, FEMORAL – DECR/ABSENT
ARTERIAL PULSE, ILIAC – DECR/ABSENT
ARTERIAL PULSE, POPLITEAL – DECR/ABSENT
ARTERIAL PULSE, POST TIBILAL – DECR/ABSENT [9]
ARTERY, FEMORAL – BRUIT
ARTERY, ILIAC – BRUIT
ARTERY, POPLITEAL – BRUIT
EARS, BILAT, EARLOBE CREASE, DIAGONAL
EXTREM, BUTTOCK – PAIN, REST [5]
EXTREM, BUTTOCK – PAIN, WALKING (CLAUDICATION) [2] [4] [6]
EXTREM, CALF – PAIN, REST [5]
EXTREM, CALF – PAIN, WALKING (CLAUDICATION) [2] [3] [6]
EXTREM, FOOT, ARCH – PAIN, REST [5]
EXTREM, FOOT, ARCH – PAIN, WALKING (CLAUDICATION) [2] [6]
EXTREM, HIP – PAIN, REST [5]
EXTREM, HIP – PAIN, WALKING (INTERMITTENT CLAUDICATION) [4] [6]
EXTREM, THIGH – PAIN, REST [5] [8]
EXTREM, THIGH – PAIN, WALKING (INTERMITTENT CLAUDICATION) [2] [4] [6] [8]
EXTREM, UNILAT, TEMP – DECR
SEX, FUNCTION – DECR/ABSENT (IMPOTENCE) [8]
VEINS, FILLING TIME – INCR

Differentiation

Arthritis
Baker cyst
Chronic Exertional Compartment Syndrome [7]
DVT
Nerve root compression
Spinal Stenosis
Thromboangiitis Obliterans

Complications

Gangrene
Nonhealing skin ulcers

Laboratory [15]

BLOOD, BUN – INCR [18]
BLOOD, CHOLESTEROL, LDL (LDL-C) – INCR
BLOOD, CHOLESTEROL, TOTAL – INCR
BLOOD, ST2 – INCR [16]
BLOOD, TGS – INCR
BLOOD, TRIGLYCERIDES – INCR
BLOOD, TROPONIN – INCR [11]

ECG

NS/VAR PER COMORBIDITY(S) [ESP CAD]
N/NS ABN [12]

Imaging [13]

NS

Genomics [17]

NS

Other Tests

[13]

Treatment: Nonpharmacologic [14]

Exercise
Atherosclerosis risk factor modification

Treatment: Pharmacologic [14]

Atherosclerosis risk factor modification
Antiplatelet Rx
Phosphodiesterase inhibitor
Pentoxifylline
Vasodilator
Prostaglandins
Statins

Treatment: Surgical/Invasive [14]

Revascularization

Endovascular/stent
Surgical

Prevention

Atherosclerosis risk factor modification
Statins

Course

Highly variable; long-term outcomes usually determined by atherosclerosis in other arterial beds, especially coronary and cerebrovascular

Notes

[1] Careful evaluation for cerebrovascular, coronary, renal artery, and aortic disease important
[2] Relieved by rest within minutes
[3] Typically superficial femoral obstruction
[4] Typically aorto-iliac obstruction
[5] Typically nocturnal, with limb elevated, relieved by placing limb into dependent position
[6] May also be described as cramp, ache, numbness
[7] Exercise-induced muscle pain and swelling, usually in athletes
[8] Leriche Syndrome: aorto-iliac obstruction causing buttock/thigh claudication, impotence (due to decreased flow in pudendal artery), decreased/absent femoral artery pulses; less often: sciatica, paraplegia, renal Infarction
[9] Dorsalis pedis (8 %) and posterior tibial (3 %) arteries congenitally absent in many persons, but <1 % of individuals have congenital absence of both
[10] Significant arterial obstruction can be present in absence of any symptoms; clinical features of other common comorbid conditions, e.g., Carotid Stenosis, CAD, AAA, not listed here, but should be searched for in patients with LEAD
[11] Increased cardiac troponin associated with increased frequency of amputations/mortality
[12] ECG manifestations of CAD often present
[13] See current Guidelines for diagnostic details
[14] See current Guidelines for treatment details
[15] Serum lipids, CRP, other atherosclerosis risk factors often abnormal

[16] ST2 increase: correlates with increased mortality in DM/critical limb ischemia
[17] Genetics of LEAD not established, although associations have been strongly suggested in ongoing investigations
[18] Increased BUN may be independent risk factor for critical limb ischemia
[19] In primary care settings, 30–60 % of patients with LEAD have no exertional leg symptoms and up to 50 % have exertional leg symptoms atypical for classic intermittent claudication

Guidelines

Management of patients with peripheral artery disease (Compilation of 2005 and 2011 ACCF/AHA Guideline Recommendations)
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ESC guidelines on the diagnosis and treatment of peripheral artery diseases
Eur Heart J. 2011;32:2851–906. http://eurheartj.oxfordjournals.org/content/ehj/32/22/2851.full.pdf.

Patient Information

Images

http://www.nlm.nih.gov/medlineplus/ency/imagepages/18077.htm.
http://www.mayoclinic.org/diseases-conditions/peripheral-artery-disease/multimedia/intermittent-claudication/img-20008123.

Medlineplus

ENGLISH
http://www.nlm.nih.gov/medlineplus/ency/article/000170.htm.
ESPANOL
http://www.nlm.nih.gov/medlineplus/spanish/ency/article/000170.htm.

Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/peripheral-artery-disease/basics/definition/con-20028731.

AHA

http://www.heart.org/HEARTORG/Conditions/More/PeripheralArteryDisease/About-Peripheral-Artery-Disease-PAD-UCM-301301-Article.jsp.

Cleveland Clinic

http://my.clevelandclinic.org/services/heart/disorders/pad.

ADA

http://www.diabetes.org/living-with-diabetes/complications/heart-disease/peripheral-arterial-disease.html?referrer=https://www.google.com/" style="text-decoration:none;" target="_blank">https://www.google.com/" style="text-decoration:none;" target="_blank">http://www.diabetes.org/living-with-diabetes/complications/heart-disease/peripheral-arterial-disease.html?referrer=https://www.google.com/.

Merck

http://www.merckmanuals.com/home/heart-and-blood-vessel-disorders/peripheral-arterial-disease/overview-of-peripheral-arterial-disease.

CDC: PAD Fact Sheet

http://www.cdc.gov/dhdsp/data-statistics/fact-sheets/fs-pad.htm.

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Br Med Bull. 2012;104:21–39. http://bmb.oxfordjournals.org/content/104/1/21.

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J Am Heart Assoc. 2014;3:e001270. http://jaha.ahajournals.org/content/3/6/e001270.long.

Angiogenic/Cell RX

Circ Res. 2015;116:1561–78. http://circres.ahajournals.org/content/116/9/1561.full.

Antithrombotic RX

Rev Vasc Med. 2014;2:37–42. http://www.sciencedirect.com/science/article/pii/S2212021113000490.

Bun: Risk Factor for Critical Limb Ischemia

Med. 2015;94:e948. http://journals.lww.com/md-journal/Fulltext/2015/06030/Elevated-Blood-Urea-Nitrogen-is-Associated-With.11.aspx.

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Circ Res. 2015;116:1599–613. http://circres.ahajournals.org/content/116/9/1599.full.

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Prog Cardiovasc Dis. 2008;50:238–63. http://www.sciencedirect.com/science/article/pii/S0033062007000758.

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JAMA. 2014;311:415–7. http://jama.jamanetwork.com/article.aspx?articleid=1817779#jld130034r2.

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Nonatheromatous Lesions

Arteriosclerosis Thromb Vas Biol. 2015;35:439–47. http://atvb.ahajournals.org/content/35/2/439.abstract?etoc.

Pathogenesis of Clinical Manifestations

Circ Res. 2015;116:1527–39. http://circres.ahajournals.org/content/116/9/1527.full.

PAR-1 Inhibition: Vorapaxor

Circulation. 2013;127:1522–9. http://circ.ahajournals.org/content/127/14/1522.full.

PAR-1 Inhibition: Vorapaxor

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Pharmacologic Treatment

Circ Res. 2015;116:1579–98. http://circres.ahajournals.org/content/116/9/1579.full.

Revascularization: Below Knee Balloon Angioplasty

Circulation. 2013;128:615–21. http://circ.ahajournals.org/content/128/6/615.full.

Revascularization: Infrapopliteal

J Am Coll Cardiol. 2014;64:1568–76. http://content.onlinejacc.org/article.aspx?articleID=1911033.

Revascularization: Long-Term Outcomes

Circulation. 2013;127:1241–50. http://circ.ahajournals.org/content/127/11/1241.full.

Revascularization: Post-procedure Management

Circulation. 2013;128:749–57. http://circ.ahajournals.org/content/128/7/749.full.

Revascularization Compared to Noninvasive Therapy: 1 YR Follow-Up

Circulation. 2014;130:939–47. http://circ.ahajournals.org/content/130/12/939.full.

Revascularization: Surgery

Circ Res. 2015;116:1614–28. http://circres.ahajournals.org/content/116/9/1614.full.

ST2 Increase: Predictor of Mortality in DM/Critical Limb Ischemia

Arterioscler Thromb Vasc Biol. 2012;32:e149–60. http://atvb.ahajournals.org/content/32/12/e149.full.

Statins

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Stents: Femoropopliteal

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Tobacco Smoke: Second-Hand

Heart. 2013;99:1342–5. http://heart.bmj.com/content/99/18/1342.abstract.

Tobacco Use

Rev Cardiovasc Med. 2004;5:189–93. http://www.ncbi.nlm.nih.gov/pubmed/15580157.

Tobacco Use

Heart. 2014;100:414–23. http://heart.bmj.com/content/100/5/414.abstract.

Troponin T: Mortality/Amputation Rates

J Am Coll Cardiol. 2014;63:1529–38. http://content.onlinejacc.org/article.aspx?articleID=1700991.

Updates and More

https://clinicalguidecvd.com/lead

Chapter 60
Marfan Syndrome

ICD-10 Code

Q87.40 Unspecified
Q87.41 CV manifestations
Q87.410 Aortic Dilation
Q87.418 Other CV manifestations
Q87.42 Ocular manifestations
Q87.43 Skeletal manifestation

Description/Etiology

Inherited connective disorder (usually dominant inheritance) involving mainly cardiovascular, skeletal, ocular tissues
Usually caused by mutations in FBN1 gene, which encodes matrix protein Fibrillin 1
Ghent diagnostic criteria [35]:

In the absence of family history:

1. Aortic Root Dilatation Z score ≥ 2 and Ectopia Lentis = Marfan syndrome [28].

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_60,

2. Aortic Root Dilatation Z score ≥ 2 and FBN1 = Marfan syndrome [29]
3. Aortic Root Dilatation Z score ≥ 2 and Systemic Score ≥ 7pts = Marfan syndrome [30]
4. Ectopia lentis and FBN1 with known Aortic Root Dilatation = Marfan syndrome [31]

In the presence of family history:

1. Ectopia lentis and Family History of Marfan syndrome (as defined above) = Marfan syndrome [32]
2. A systemic score ≥ 7 points and Family History of Marfan syndrome (as defined above) = Marfan syndrome [33]
3. Aortic Root Dilatation Z score (≥ 2 above 20 years old, ≥ 3 below 20 years old) + Family History of Marfan syndrome (as defined above) = Marfan syndrome [34].

Predisposing/Comorbid Conditions

BICUSPID AORTIC VALVE
CARDIOMYOPATHY – DILATED
COARCTATION OF AORTA
DILATATION: ABDOMINAL AORTA
DILATATION: ASCENDING AORTA
DILATATION: DESCENDING THORACIC AORTA
DILATATION: MAIN PULMONARY ARTERY
ECTOPIA LENTIS [26]
FAMILY HX: SUDDEN DEATH
FH: MARFAN
LUMBOSACRAL DURAL ECTASIA
MITRAL ANNULUS CALCIFICATION
MITRAL VALVE PROLAPSE
RESTRICTIVE LUNG DISEASE
SCOLIOSIS
SPONTANEOUS PNEUMOTHORAX
STRIAE ATROPHICA

Demography

Global
Gender equal
More common in amateur and professional basketball and volleyball athletes

Pathophysiology [13]

FBN1 mutation causes:

- Decreased amount of functional Fibrillin-1 available to form microfibrils
- Decreased tissue elastin content (eg, aortic wall)
- Excess release of growth factors
- Tissue overgrowth/instability

Cardiovascular morphologic changes include:

- Aortic root/descending thoracic aorta dilatation
- Mitral valve prolapse
- Sinuses of Valsalva dilatation (begins in utero)
- Thickened AV valves with regurgitation

Signs/Symptoms

BACK – PAIN [17]
BACK, CURV – LAT (SCOLIOSIS) [5]
BODY, HT – INCR [20]
EXTREM – LONG [2]
EXTREM, ELBOW, EXTENSION – DECR/ABSENT [1]
EXTREM, FEET – FLAT (PES PLANUS)
EXTREM, FEET, ARCH – HIGH (PES CAVUS) **[6]**
EXTREM, FINGERS – CONTRACTURES (CAMPTODACTYLY)
EXTREM, FINGERS – LONG (ARACHNODACTYLY) [3] [4]

EYES – GLAUCOMA
EYES, CORNEA – FLAT
EYES, GLOBE, AXIAL LENGTH – INCR
EYES, IRIS – HYPOPLASTIC
EYES, LENS – DISLOCATION (ECTOPIA LENTIS) [26]
EYES, LENS – OPACITY (CATARACT)
EYES, PALPEBRAL FISS – DOWN-SLANTING
EYES, PUPILS – MIOSIS, INCR [7]
EYES, RETINA – DETACHED
EYES, VISION – MYOPIA
FACE, CHEEKS – FLAT
FACE, CHIN – SMALL (MICROGNATHIA)
FACE, JAW – DISPLACED, POST (RETROGNATHISM)
HEAD, SKULL – LONG/NARROW (DOLICHOCEPHALY)
HEART, LSB, LOWER – CLICK, SYS [8]
HEART, LSB, LOWER – MURMUR, SYS [8]
HEART, LSB, MID – MURMUR, DIAS [10]
HEART, LV, APEX – CLICK(S), SYS [9]
HEART, LV, APEX – MURMUR, SYS [9]
HEART, RSB, UPPER – MURMUR, DIAS [10]
JOINTS, MOVEMENT, RANGE – INCR (HYPERMOBILITY)
MOUTH, PALATE – HIGH/ARCHED
MOUTH, TEETH – CROWDING
SKIN, GEN – STRIAE ATROPHICA [16]
STERNUM, CURV – ANT (PECTUS CARINATUM)
STERNUM, CURV – POST (PECTUS EXCAVATUM)

Differentiation

Aneurysms-Osteoarthritis Syndrome
Ascending Aortic Aneurysm Syndrome
Bicommissural Aortic Valve Syndrome
Cutis Laxa

Ehlers-Danlos Syndrome
Familial Ectopic Lentis
Familial Mitral Valve Prolapse Syndrome
Familial Thoracic Aortic Aneurysm Syndrome
Homocystinuria
Loeys-Dietz Syndrome
MASS phenotype [18]

Complications [13]

AMI [12]
Aortic Aneurysm
Aortic Dissection [21]
Coronary Aneurysm
Dilated Cardiomyopathy [23]
Dysrhythmias – atrial [19]
Dysrhythmias – ventricular
HF
Mitral Regurgitation
Pulmonary Hypertension
Restrictive lung disease [15]
Spontaneous Pneumothorax [14]
Stroke [11]
Sudden Athletic-Related Death
Sudden Infant Death

Laboratory

GENETIC TESTING

ECG

NS/VAR PER COMORBIDITY(S) [ESP CARDIAC ABN]

Imaging [22]

AORTA, ASCEND, SIZE – INCR
AORTA, ROOT, SIZE – INCR [25]
AV, FLOW – REGURG
LV, DIAS – DYSF [23]
LV, SYS – DYSF [23]
MV, ANNULUS – CALCIUM
MV, LEAFLETS – PROLAPSE
TV, LEAFLET, MOTION – PROLAPSE

Genomics

FBN1

Other Tests

Contrast aortography

Treatment: Nonpharmacologic

Avoidance/caution in contact/competitive sports
Avoidance/caution in isometric exercise

Treatment: Pharmacologic

ARBs [27]
Beta-blockers [24]

Treatment: Surgical/Invasive

Elective: aortic root repair with/without AV replacement

Course

Variable according to pathology

Notes

[1] <170°
[2] Arm span to height ratio >1.05
[3] Walker-Murdoch/wrist sign: full overlap of thumb over 5th finger when wrapped over contralateral wrist
[4] Steinberg/thumb sign: distal thumb phalanx fully extends beyond ulnar border of hand when fully extended across palm
[5] >20°
[6] Occurs less often than pes planus
[7] Due to ciliary muscle hypoplasia
[8] TV prolapse/regurgitation
[9] MV prolapse/regurgitation
[10] AR secondary to aortic root dilatation
[11] Involvement of carotid arteries
[12] Involvement of coronary arteries
[13] Pertaining to CV system only
[14] Due to widening of distal lung airspaces
[15] Due to sternum deformities/scoliosis
[16] Occur in areas not subject to stretch, such as anterior shoulder, low back
[17] Due to dural ectasia; usually not symptomatic
[18] MASS: mitral, aortic, skin, skeletal features in persons without true Marfan
[19] Especially AF
[20] Due to long bone overgrowth (dolichostenomelia)
[21] Main risk factors are family history of Aortic Dissection and increased maximal aortic dimension by echo
[22] Echo/Doppler may be used serially for monitoring, especially for aortic root size
[23] LV dysfunction independent of valve regurgitation may occur in some persons

[24] To reduce proximal aortic sheer stress

[25] Echo measures internal aortic diameter; MRI and CT measure external diameter, which is 0.2-0.4 larger than echo measures

[26] Ectopia lentis: affects 60 % of Marfan patients; other causes include:

Familial Ectopia Lentis
Homocystinuria
Weill-Marchesani Syndrome

Brachydactyly
Joint stiffness
Other eye abnormalities
Short stature

[27] Decreases aortic dilatation rates (losartan)

[28] *** Presence of aortic root dilatation (Z-score≥2 when standardized to age and body size) or dissection and ectopia lentis allows unequivocal diagnosis of Marfan syndrome, regardless of presence or absence of systemic features except where these are indicative of Shprintzen Goldberg syndrome, Loeys-Dietz syndrome, or vascular Ehlers Danlos syndrome

[29] *** Presence of aortic root dilatation (Z≥2) or dissection and identification of a bona fide FBN1 mutation are sufficient to establish the diagnosis, even when ectopia lentis is absent.

[30] *** Where aortic root dilatation (Z≥2) or dissection is present, but ectopia lentis is absent and the FBN1 status is either unknown or negative, a Marfan syndrome diagnosis is confirmed by the presence of sufficient systemic findings (≥7 points, according to a scoring system) confirms the diagnosis. However, features suggestive of Shprintzen Goldberg syndrome, Loeys-Dietz syndrome, or vascular Ehlers Danlos syndrome must be excluded and appropriate alternative genetic testing (TGFBR1/2,

SMAD3, TGFB2, TGFB3, collagen biochemistry, COL3A1, and other relevant genetic testing when indicated and available upon the discovery of other genes) should be performed.

[31] *** In the presence of ectopia lentis, but absence of aortic root dilatation/dissection, the identification of an FBN1 mutation previously associated with aortic disease is required before making the diagnosis of Marfan syndrome

[32] *** The presence of ectopia lentis and a family history of Marfan syndrome (as defined in 1–4 above) is sufficient for a diagnosis of Marfan syndrome

[33] *** A systemic score of greater than or equal to 7 points and a family history of Marfan syndrome (as defined in 1–4 above) is sufficient for a diagnosis of Marfan syndrome. However, features suggestive of Shprintzen Goldberg syndrome, Loeys-Dietz syndrome, or vascular Ehlers Danlos syndrome must be excluded and appropriate alternative genetic testing (TGFBR1/2, SMAD3, TGFB2, TGFB3 collagen biochemistry, COL3A1, and other relevant genetic testing when indicated and available upon the discovery of other genes) should be performed

[34] *** The presence of aortic root dilatation ($Z \geq 2$ above 20 year old, ≥ 3 below 20 year old) and a family history of Marfan syndrome (as defined in 1–4 above) is sufficient for a diagnosis of Marfan syndrome. However, features suggestive of Shprintzen Goldberg syndrome, Loeys-Dietz syndrome, or vascular Ehlers Danlos syndrome must be excluded and appropriate alternative genetic testing (TGFBR1/2, SMAD3, TGFB2, TGFB3, collagen biochemistry, COL3A1, and other relevant genetic testing when indicated and available upon the discovery of other genes) should be performed

[35] *** From: The Marfan Foundation https://www.marfan.org/dx/rules

Guidelines

2010 ACCF/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/STS/SVM Guidelines for the Diagnosis and Management of Patients with Thoracic Aortic Disease
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Eur Heart J. doi:10.1093/eurheartj/ehu281. https://www.nvvc.nl/media/richtlijn/187/2014_ESC_Guidelines_on_Diagnosis_and_Treatment_of_Aortic_Disease.pdf.

Patient Information

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Medlineplus

http://www.nlm.nih.gov/medlineplus/marfansyndrome.html.
http://www.nlm.nih.gov/medlineplus/ency/imagepages/9611.htm.

Marfan Foundation

http://www.marfan.org/.

Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/marfan-syndrome/basics/definition/con-20025944.

Espanol

http://www.nlm.nih.gov/medlineplus/spanish/marfansyndrome.html.

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Review

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Review

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Aortic Dilatation/Bicuspid Aortic Valve

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Coronary Aneurysm

Circulation. 2014;129:1791–2. http://circ.ahajournals.org/content/129/17/1791.full.

Cardiovascular Involvement: Natural History/ Followup

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Familial Thoracic Aorta Aneurysms

Curr Opin Cardiol. 2014;29:492–8. http://www.ncbi.nlm.nih.gov/pubmed/25290696.

Ghent Nosology for Marfan Syndrome (Revised)

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Losartan: Decreased Aortic Dilatation Rates

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Mitral Valve Prolapse

Echocardiography. 2009;26:357–64. http://www.ncbi.nlm.nih.gov/pubmed/19054044?dopt=Abstract.

Pregnancy

Int J Cardiol. 2009;136;156–61. http://www.sciencedirect.com/science/article/pii/S0167527308006177.

Pulmonary Artery Aneurysm

Circulation. 2015;131:310–6. http://circ.ahajournals.org/content/131/3/310.extract.

Pulmonary Artery Aneurysm/Dissection

J Am Coll Cardiol. 2013;61:685. http://content.onlinejacc.org/article.aspx?articleID=1567630.

Surgery: Aortic Root Aneurysm

J Thorac Cardiovasc Surg. 2003;125:789–96. http://www.sciencedirect.com/science/article/pii/S0022522302733047.

Surgery: Bentall/Valve-Sparing Aortic Root Procedures

Ann Thor Surg. 2008;85:2003–11. http://www.sciencedirect.com/science/article/pii/S000349750800163X.

Updates and More

https://clinicalguidecvd.com/marfan

Chapter 61
Mitral Regurgitation: Acute

Management Keys

Diagnosis of presence and determination of etiology are urgent in order to initiate appropriate interventional therapy, which may be lifesaving

ICD-10 Code

I34.0

Alternate Names/Abbreviation

ACUTE MR

Description/Etiology

Acute compromise of MV apparatus due to many causes, including:

Chordal rupture [13]
Infective Endocarditis
Papillary muscle rupture [12]
Papillary muscle dysfunction [1]
Prosthetic valve dysfunction

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_61,

Predisposing/Comorbid Conditions

CORONARY ARTERY DISEASE
MYXOMATOUS MV
RHEUMATIC VALVE DISEASE

Demography

Per etiology

Pathophysiology

Sudden increase in LA/LV blood volume without corresponding LA/LV dilatation, causing:

- Decreased LV stoke volume
- Rapid increase in pulmonary venous pressure/acute pulmonary edema
- Systemic Hypotension

Signs/Symptoms

BREATH SOUNDS – CRACKLES (RALES)
BREATHING – DIFF (DYSPNEA)
BREATHING – RAPID (TACHYPNEA)
EXTREM, HANDS/FEET, COLOR – BLUE (ACROCYANOSIS) [5]
FATIGUE
FEVER [6]
HEART, LV, APEX – MURMUR, DIAS [2, 3]
HEART, LV, APEX – MURMUR, SYS [4]
HEART, LV, APEX, IMP – FORCEFUL/SUSTAINED [8]
HEART, RATE – RAPID (TACHYCARDIA)
HEART, S3 LV [2]

HYPOTENSION (BLOOD PRESSURE – DECREASED/LOW)
NECK, JVP, V WAVE – INCR/LARGE
SKIN, TEMP – DECR [5]

Differentiation

HF
Non-cardiac pulmonary edema
Other causes of circulatory collapse
Pneumonia

Complications

Acute pulmonary edema
Dysrhythmias
HF
SHOCK

Laboratory

BLOOD, NT-PROBNP – INCR
BLOOD, WBC – INCR (LEUKOCYTOSIS) [6]

ECG [7]

DYSRHYTHMIAS – ATRIAL (PACS/OTHERS)
DYSRHYTHMIAS – VENTRICULAR (PVCS/OTHERS)
RATE – INCREASED (SINUS TACHYCARDIA)
ST-T WAVE – ABN, NS

Imaging

CARD SIZE MAY BE N [10]
LV, WALL MOTION – INCR/HYPERDYNAMIC
MV, FLOW – REGURG
MV, LEAFLET MORPH – ABN
MV, LEAFLET, MOTION – ABN
PUL – EDEMA [9]

Other Tests

Coronary angiography if ischemic etiology possible

Treatment: Nonpharmacologic

O_2

Treatment: Pharmacologic

Antibiotics [6]
Inotropes [11]
Vasodilators [11]

Treatment: Surgical/Invasive

Intraaortic balloon counterpulsation
MV repair
MV replacement

Prevention

Variable per etiology

Course

Variable per etiology

Notes

[1] Due to acute Myocarditis or acute myocardial ischemia causing papillary muscle displacement
[2] May be only auscultatory findings
[3] Early diastole
[4] May be best heard at cardiac base; may be soft/absent or not holosystolic; may radiate to neck, spine, top of head
[5] Peripheral vasoconstriction
[6] With Infective Endocarditis (bacterial)
[7] Often normal; ischemic changes signify acute coronary etiology of MR
[8] Often absent; if present, hyperdynamic LV in presence of normal cardiac size suggests acute MR
[9] Rarely, edema may be isolated to 1 segment if preferentially directed to a single pulmonary vein
[10] Unless prior chronic MR present
[11] To stabilize prior to surgery only
[12] STEMI, usually inferior AMI
[13] May occur with degenerative MV disease

Guidelines

2014 AHA/ACC Guideline for the Management of Patients with Valvular Heart Disease

J Am Coll Cardiol. 2014;63:e57–e185. http://www.content.onlinejacc.org/article.aspx?articleID=1838843.

Guidelines on the management of valvular heart disease (version 2012)

Eur Heart J. 2012;33:2469–75. http://www.escardio.org/guidelines-surveys/esc-guidelines/Pages/valvular-heart-disease.aspx.

Patient Information

Images

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http://www.nlm.nih.gov/medlineplus/ency/imagepages/1097.htm.

Medlineplus

ENGLISH
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Cleveland Clinic

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Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/mitral-valve-regurgitation/basics/definition/con-20022644.

AHA

http://www.heart.org/HEARTORG/Conditions/More/HeartValveProblemsandDisease/Problem-Mitral-Valve-Regurgitation-UCM-450612-Article.jsp.

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AMI: Shock Trial

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AMI: Outcomes

Ann Intern Med. 1992;117:18–24. http://www.annals.org/article.aspx?articleid=705627&resultClick=3.

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J Heart Valve Dis. 1993;2:512–22. http://www.ncbi.nlm.nih.gov/pubmed/8269160.

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Circulation. 2013;127:e586-8. http://www.circ.ahajournals.org/content/127/18/e586.full.

Post-valvotomy Acute MR

Catheter Cardiovasc Interv. 2013;81:603–8. http://www.onlinelibrary.wiley.com/doi/10.1002/ccd.24417/abstract.

Vasodilator Therapy

Am J Cardiol. 1979;43:773–7. http://www.ajconline.org/article/0002-9149(79)90077-8/abstract.

Updates and More

https://clinicalguidecvd.com/amr

Chapter 62
Mitral Regurgitation: Chronic

ICD-10 Code

I34.1

Alternate Names/Abbreviation

MR
MITRAL INSUFFICIENCY

Description/Etiology

50 % of LV stroke volume diverted to regurgitant flow into LA due to primary or secondary abnormalities of structure/function of mitral apparatus, including:

MV leaflets [1]
MV annulus [2]
Chordae tendineae [3]
Papillary muscles [4]

Stages (secondary form):

A. At risk

Normal valve leaflets/chords/annulus with CAD or cardiomyopathy

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 857
DOI 10.1007/978-1-4471-7293-2_62,

B. Progressive

Abnormal regional wall motion with mild MV leaflet tethering
Annular dilatation with mild loss of central leaflet coaptation

C. Asymptomatic severe

Abnormal regional wall motion/LV dilatation with severe MV leaflet tethering
Annular dilatation with severe loss of central leaflet coaptation

D. Symptomatic severe

Abnormal regional wall motion/LV dilatation with severe MV leaflet tethering
Annular dilatation with severe loss of central leaflet coaptation

Most common cause: degenerative MV disease

Comorbid Conditions

ANTIPHOSPHOLIPID SYNDROME
AORTIC REGURGITATION – CHRONIC
ATRIAL FIBRILLATION
CARCINOID HEART DISEASE
CARDIAC AMYLOIDOSIS
CARDIAC SARCOIDOSIS
CARDIOMYOPATHY – DILATED
CARDIOMYOPATHY – HYPERTROPHIC
CARDIOMYOPATHY – RESTRICTIVE
CARDIOMYOPATHY – TAKOTSUBO
DRUGS: ERGOTAMINES
DRUGS: METHYSERGIDE
DRUGS: PERGOLIDE
ENDOMYOCARDIAL FIBROSIS
GIANT CELL MYOCARDITIS
HEART FAILURE
INFECTIVE ENDOCARDITIS

KAWASAKI DISEASE
MARFAN SYNDROME
MITRAL ANNULUS CALCIFICATION
MITRAL VALVE PROLAPSE [INCL BARLOW]
MYOCARDITIS
MYXOMA – LEFT ATRIUM
RADIATION
RHEUMATIC FEVER [See Appendix A]
RHEUMATOID ARTHRITIS
STABLE ISCHEMIC HEART DISEASE
SYSTEMIC LUPUS ERYTHEMATOSUS
VON WILLEBRAND SYNDROME [ACQUIRED]

Demography

Variable according to etiology
Prevalence: 10 % in persons > age 75 years

Pathophysiology

Abnormal reverse blood flow from LV to LA and resultant LV and LA dilatation and LV compensatory changes to maintain cardiac output
Cardiac function in patients with primary MV abnormalities tends to deteriorate with time as volume overload progresses and effective valve orifice area increases
Both RV systolic dysfunction (due to LV remodeling/abnormal septal function/increased PA pressure) and LV dysfunction contribute to abnormal hemodynamics
Degenerative MV disease consists of myxomatous degeneration of MV leaflets with:

- Elongated/redundant chordal apparatus
- Prolapse of leaflets into LA causing leaflet edge malcoaptation/subsequent regurgitation
- Rupture of chordal structures (sometimes), especially in older males, causing further increase in MR severity due to unsupported leaflet segments

Signs/Symptoms [5]

ARTERIAL PULSE, AMP – DECR/ABS
ARTERIAL PULSE, FALL – RAPID
ARTERIAL PULSE, RISE – RAPID
BREATHING – DIFF (DYSPNEA)
CHEST, ANT, L – BULGE
FATIGUE
HEART, LV, APEX – MURMUR, DIAS
HEART, LV, APEX – MURMUR, SYS [12, 16]
HEART, LV, APEX, IMP – FORCEFUL/SUSTAINED
HEART, LV, APEX, IMP – DIFFUSE
HEART, P2, INTENSITY – INCR
HEART, S2, SPLIT – WIDE [SEVERE; EARLY AV CLOSURE]
HEART, S3 LV [12]
SKIN, TEMP – DECR
SWEATING – INCR (DIAPHORESIS/ HYPERHIDROSIS)

Differentiation (of MR Systolic Murmur)

AS – Valvular
HCM (with LVOT obstruction)
TR
VSD

Complications

AF/Flutter (30 % 10-year incidence)
Bleeding [14]
Other dysrhythmias
HF
IE
Peripheral emboli
Pulmonary edema
SCD [6]

Laboratory

BLOOD, NT-PROBNP – INCR [13]
BLOOD, ST2 – INCR [17]

ECG

P WAVE, DUR – INCR
QRS – LBBB/LBBB PATTERN
QRS – LVH PATTERN

Imaging [7] [15]

CARDIOMEGALY
LUNGS, INTERSTITIUM – EDEMA/INFILTRATES
LA, CHAMBER, SIZE – INCR
LV, CHAMBER, SIZE – INCR
LV, WALL MOTION – INCR/HYPERDYNAMIC
MV, FLOW – REGURG
MV, LEAFLET MORPH – ABN [11]
MV, LEAFLETS, MOTION – ABN [11]

Other Tests

Exercise Doppler [8]
Cardiac catheterization [9]

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic

Asymptomatic: NS
AF: rate control and anticoagulation

Primary versus secondary considerations [19] [20]
Aldosterone antagonists
ACEIs
ARBs
Beta-blockers

Treatment: Surgical/Invasive [10]

MV repair
  Surgical
  Transcatheter [18]
MV replacement with mitral apparatus preservation
MV replacement with mitral apparatus removal

Course

Primary MR asymptomatic 5 year rates:

  Death from any cause: 22 %
  HF: 14 %
  New AF: 14 %

Primary MR predictors of poor outcome include:

  Age
  AF
  Decreased LVEF
  Increased LV end-systolic volume
  LA dilatation
  MR severity
  PAH

Notes

[1] Eg, myxomatous degeneration, rheumatic heart disease, congenital deformity, radiation
[2] Eg, annular calcification, myxomatous degeneration, connective tissue disease

[3] Eg, Rheumatic Carditis, radiation, IE, drugs
[4] Eg, ischemia, DCM, HCM
[5] Patients with mild-moderate MR often are asymptomatic for many years; symptoms of dyspnea/exercise intolerance develop slowly as compensatory mechanisms are overwhelmed by volume overload and irreversible LV dysfunction occurs
[6] May occur in asymptomatic patients
[7] Echo: Principle means of investigation for severity, mechanisms, reparability, consequences, RV function, PA pressure; high degree of operator skill required
[8] To assess patients with severe MR who are asymptomatic for exercise tolerance and effects of exercise on PA pressure and MR severity
[9] When noninvasive testing is unsatisfactory or incongruent with clinical findings or when surgery is contemplated
[10] In patients with primary MR, surgical intervention with repair/replacement indicated with severe MR and symptoms or LV dysfunction (EF <60 % or end systolic diameter >40 mm); surgical repair is preferred treatment for primary MR and is associated with better outcomes than MV replacement; see guidelines for detailed current specific recommendations
[11] According to etiology
[12] Primary MR: increased intensity/long duration of MR murmur and presence of S3 suggest significant MR; in secondary MR, murmur intensity often soft/unrelated to severity
[13] Blood BNP/Pro-BNP level increases/changes may be outcome predictors
[14] Due to acquired von Willebrand syndrome and reversible with MV surgery
[15] CMRI may be superior to echo for quantifying MR
[16] Usually blowing quality; harsh/musical suggests ruptured chordae tendineae/MVP
[17] Increased ST2 correlates with higher level of LVEF and lower LVEDD post-MV repair

[18] Early data: significant MR treated with transcatheter repair results in significant clinical improvements at 12 months
[19] Primary MR: pharmacologic treatment does not alter natural history of severe primary MR; for symptomatic patients with severe primary MR, diuretics and afterload reduction help relieve signs/symptoms of HF, but intervention is ultimate treatment; see guidelines for detailed current specific recommendations
[20] Secondary MR: first-line treatment consists of guideline-directed medical therapy for LV dysfunction, including ACEIs, ARBs, beta-blockers, and aldosterone antagonists; see guidelines for detailed current specific recommendations

Appendix A: World Heart Federation Echo Criteria for Diagnosis of Rheumatic Fever

Individuals Age <20 Years

Definite RHD (either A, B, C, or D):

A. Pathological MR and at least two morphological features of RHD of the MV
B. MS mean gradient = 4 mmHg*
C. Pathological AR and at least two morphological features of RHD of the AV‡
D. Borderline disease of both the AV and MV§

Borderline RHD (either A, B, or C):

A. At least two morphological features of RHD of the MV without pathological MR or MS
B. Pathological MR
C. Pathological AR

Normal echocardiographic findings (all of A, B, C, and D):

A. MR that does not meet all four Doppler echocardiographic criteria (physiological MR)
B. AR that does not meet all four Doppler echocardiographic criteria (physiological AR)
C. An isolated morphological feature of RHD of the MV (for example, valvular thickening) without any associated pathological stenosis or regurgitation
D. Morphological feature of RHD of the AV (for example, valvular thickening) without any associated pathological stenosis or regurgitation

Individuals Age 20+ Years

Definite RHD (either A, B, C, or D):

A. Pathological MR and at least two morphological features of RHD of the MV
B. MS mean gradient =4 mmHg*
C. Pathological AR and at least two morphological features of RHD of the AV, only in individuals aged <35 years‡
D. Pathological AR and at least two morphological features of RHD of the MV

*Congenital MV anomalies must be excluded. Furthermore, inflow obstruction due to nonrheumatic mitral annular calcification must be excluded in adults. ‡Bicuspid AV, dilated aortic root, and hypertension must be excluded. §Combined AR and MR in high prevalence regions and in the absence of congenital heart disease is regarded as rheumatic. Abbreviations: AR, aortic regurgitation; AV, aortic valve; MR, mitral regurgitation; MS, mitral stenosis; MV, mitral valve; RHD, rheumatic heart disease; WHF, World Heart Federation.

Guidelines

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Images

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http://www.nlm.nih.gov/medlineplus/ency/article/000176.htm.

ESPANOL.
http://www.nlm.nih.gov/medlineplus/spanish/ency/article/000176.htm.

Cleveland Clinic

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http://www.mayoclinic.org/diseases-conditions/mitral-valve-regurgitation/basics/definition/con-20022644.

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Post-NSTEMI: Long-Term Follow-Up

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J Am Coll Cardiol. 2014;63:2057–68. http://content.onlinejacc.org/article.aspx?articleID=1838310.

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Nature Reviews Cardiology. 2012;9:297–309. http://www.nature.com/nrcardio/journal/v9/n5/box/nrcardio.2012.7-BX1.html.

Updates and More

https://clinicalguidecvd.com/cmr

Chapter 63
Mitral Stenosis: Acquired

Management Keys

- Perform TTE to establish diagnosis, quantify hemodynamic severity, assess concomitant valvular lesions, and demonstrate valve morphology to determine suitability for mitral commissurotomy
- Anticoagulate for AF (all types), prior systemic embolus, LA thrombus
- Control heart rate for AF with rapid ventricular response and for normal sinus rhythm when symptoms occur with effort
- Perform percutaneous mitral balloon commissurotomy in symptomatic patients with severe MS and favorable valve morphology in absence of LA thrombus or moderate-to-severe MR
- Perform mitral valve surgery (repair, commissurotomy, or valve replacement) in severely symptomatic patients with severe MS who are not high risk for surgery and who are not candidates for or who have failed previous percutaneous mitral balloon commissurotomy

ICD-10 Code

I05.0

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_63,

Alternate Names/Abbreviation

MS

Description/Etiology [8]

Structural obstruction of blood flow through mitral valve, due to:

- Annular calcification
- Rheumatic Carditis [17]

Comorbid Conditions

AORTIC REGURGITATION – CHRONIC [RHEUMATIC HEART DIS]
AORTIC STENOSIS – VALVULAR [RHEUMATIC HEART DIS]
CARCINOID HEART DISEASE [RARE]
HUNTERS SYNDROME
MITRAL ANNULUS CALCIFICATION
MITRAL REGURGITATION – CHRONIC [RHEUMATIC HEART DIS]
RHEUMATIC FEVER [17]
TRICUSPID STENOSIS [RHEUMATIC HEART DIS]

Demography

F > M 2:1
More common in undeveloped countries [2]

Pathophysiology [12]

Morphology:

- MV leaflet thickening
- MV leaflet calcification

MV commissural fusion
MV chordal fusion

Transmitral pressure gradient causes increased LA and pulmonary pressures and LA dilatation
Concomitant MR usually present

Signs/Symptoms [3]

ABDOMEN – PAIN
ARTERIAL PULSE, AMP – DECR/ABS
BREATH SOUNDS – WHEEZES
BREATHING – DIFF (DYSPNEA)
BREATHING – DIFF, NOCTURNAL (DYSPNEA, NOCT)
BREATHING – DIFF, RECLINING FLAT (ORTHOPNEA)
BREATHING – RAPID (TACHYPNEA)
CHEST – PAIN
CHEST – PALPITATIONS
COUGH
EXTREM, LOWER, BILAT – EDEMA
FATIGUE
HEART, BASE – MURMUR, DIAS [10]
HEART, LSB, LOWER – IMP, SYS
HEART, LSB, LOWER – MURMUR, DIAS
HEART, LSB, MID – PERCUSSION, DULLNESS
HEART, LV, APEX – MURMUR, DIAS [11]
HEART, LV, APEX – OPENING SNAP, MITRAL
HEART, LV, APEX – THRILL, DIAS
HEART, P2 – PALPABLE
HEART, P2, INTENSITY – INCR
HEART, RATE – RAPID (TACHYCARDIA)
HEART, RSB – SOUND, SYS EJECTION
HEART, S1, INTENSITY – DECR/ABSENT
HEART, S1, INTENSITY – INCR
HYPOTENSION (BLOOD PRESSURE – DECREASED/LOW)

LIVER – ENLARGED (HEPATOMEGALY)
NECK, JVP – ELEV
NECK, JVP, A WAVE – INCR/LARGE (CANNON WAVE)
SKIN, CHEEKS, COLOR – PINK PURPLE (MALAR FACIES)
SKIN, COLOR – BLUE (CYANOSIS)
SPUTUM – BLOOD (HEMOPTYSIS)
VOICE – HOARSE (LARYNGITIS) [ORTNER-S SYNDROME] [9]

ECG

P WAVE, DUR – INCR, NOTCHED (P MITRALE)
QRS – RVH PATTERN
QRS, AXIS – R

Differentiation

ASD
Cor Triatriatum
Myxoma – Left Atrium
Papillary Fibroelastoma
TV stenosis

Complications

Acute pulmonary edema [5]
Acute Pulmonary Embolism [4]
AF [4]
Pulmonary Hypertension
Stroke – Ischemic
Systemic embolism [4]

Laboratory

NS

Imaging [17]

LA, CHAMBER, SIZE – INCR
LUNGS, INTERSTITIUM – EDEMA, CV ANGLE (KERLEY B LINES) [1]
LUNGS, INTERSTITIUM – EDEMA, HILA (KERLEY A LINES) [1]
LUNGS, INTERSTITIUM – EDEMA/INFILTRATES
MV, ANNULUS – CALCIUM
MV, LEAFLET, MOTION – ABN
MV, LEAFLETS – CALCIUM
MV, LEAFLETS – THICK
MV, LEAFLETS, MORPH – ABN
MV, LEAFLETS, MOTION – DECR
MV, LEAFLETS, MOTION – DOMED
MV, ORIFICE, AREA – DECR
MV, TRANSVALVE PRESS – GRADIENT
PA, MAIN,SIZE – INCR
PA. PRESS – INCR
RA, CHAMBER, SIZE – INCR
RV, CHAMBER, SIZE – INCR

Other Tests

Cardiac catheterization when noninvasive tests are unsatisfactory or discordant with clinical findings

Treatment: Nonpharmacologic

Dietary salt restriction (for pulmonary congestion)

Treatment: Pharmacologic

Anticoagulation for:

- AF (paroxysmal, persistent, permanent)
- LA thrombus
- Prior systemic embolus

Diuretic for pulmonary congestion
Inhaled corticosteroid for bronchial hyperreactivity
Negative chronotropic agent for tachycardia [18]

- Beta-blocker
- CCB

Rheumatic Fever prophylaxis

Treatment: Surgical/Invasive [15]

Percutaneous mitral balloon valvotomy [6]
MV replacement [7]
Surgical commissurotomy

Prevention

Rheumatic fever prevention protocol

Course [12]

Time of Rheumatic Fever to initial symptoms: 20–40 years
Mean age of presentation (developed countries): 5-6th decade
After onset of limiting symptoms, untreated 10-year survival: 0–15 %

Notes

[1] CXR: indicates severe MS

[2] In developed countries, many cases of MS are among immigrants
[3] Symptoms usually absent until valve area decreased to 1.5 cm^2 but other factors (e.g., sinus tachycardia, supraventricular tachycardia, AF, pregnancy, concomitant valve lesions, PAH, anemia) can cause earlier onset of symptoms or symptoms disproportionate to valve area
[4] Sometimes initial MS clinical manifestation
[5] Especially with sudden AF and rapid ventricular response rate
[6] Preferred over surgical commissurotomy, which remains procedure of choice in some developing countries
[7] For patients not candidates for commissurotomy, including:

Significant valve calcification/fibrosis
Subvalvular fusion of MV apparatus

[8] Other uncommon/rare causes of MS include:

Anorectic drugs
Carcinoid Syndrome
Mitral annulus calcification

[9] Due to large LA impingement on recurrent laryngeal nerve
[10] Graham-Steel murmur, due to either pulmonary regurgitation secondary to Pulmonary Hypertension or Rheumatic AR
[11] C-reactive protein may be increased as manifestation of ongoing rheumatic inflammation
[12] Disease progression highly variable between populations and among individuals; usually slow with average decrease in MV area of $0 \cdot 01$ cm^2 per year, but >1/3 of patients have no decline in valve area over several years
[14] Presenting symptom usually dyspnea precipitated by effort, pregnancy, stress, infection, AF with rapid ventricular response
[15] Data inadequate for evidence-based recommendation for mixed valve disease, which predominates with Rheumatic Heart Disease

[16] In selected patients with reduction in transmitral gradient/increased MV area from about 1–2 cm^2, early outcomes include rapid decrease in LA pressure, increased cardiac output, subsequent decrease in PA pressure

[17] **World Heart Federation echo criteria for diagnosis of Rheumatic Fever***

Individuals age <20 years:

Definite RHD (either A, B, C, or D):

A. Pathological MR and at least two morphological features of RHD of MV
B. MS mean gradient: 4 mmHg
C. Pathological AR and at least two morphological features of RHD of AV
D. Borderline disease of both AV and MV§

Borderline RHD (either A, B, or C):

A. At least two morphological features of RHD of MV without pathological MR or MS
B. Pathological MR
C. Pathological AR

Normal echocardiographic findings (all of A, B, C, and D):

A. MR that does not meet all four Doppler echocardiographic criteria (physiological MR)
B. AR that does not meet all four Doppler echocardiographic criteria (physiological AR)
C. An isolated morphological feature of RHD of MV (for example, valvular thickening) without any associated pathological stenosis or regurgitation
D. Morphological feature of RHD of AV (for example, valvular thickening) without any associated pathological stenosis or regurgitation

Individuals age >20 years:

Definite RHD (either A, B, C, or D):

A. Pathological MR and at least two morphological features of RHD of MV

B. MS mean gradient; 4 mmHg
C. Pathological AR and at least two morphological features of RHD of AV, only in individuals aged <35 years
D. Pathological AR and at least two morphological features of RHD of MV

*Congenital MV anomalies must be excluded; inflow obstruction due to nonrheumatic mitral annular calcification must be excluded in adults

[18] For patients with MS in normal sinus rhythm and symptoms associated with exercise

Guidelines

2014 AHA/ACC Guideline for the Management of Patients with Valvular Heart Disease

J Am Coll Cardiol. 2014;63:e57–e185. http://content.onlinejacc.org/article.aspx?articleID=1838843.

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Eur Heart J. 2012;33:2469–75. http://www.escardio.org/guidelines-surveys/esc-guidelines/Pages/valvular-heart-disease.aspx.

Patient Information

Images

http://www.nlm.nih.gov/medlineplus/ency/imagepages/18147.htm.

Medlineplus

ENGLISH
http://www.nlm.nih.gov/medlineplus/ency/article/000175.htm.
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Review: Rheumatic Heart Disease

Lancet. 2016;387:1335–46. http://www.sciencedirect.com/science/article/pii/S014067361600547X.

Annular Calcification

J Am Coll Cardiol. 2015;66:1934–41. http://content.onlinejacc.org/article.aspx?articleid=2461798#tab1.

Annular Calcification (Image)

N Engl J Med. 2015;372:e23. http://www.nejm.org/doi/full/10.1056/NEJMicm1404772.

Balloon Mitral Valvuloplasty

Am J Med. 2014;127:1126.e1–1126.e12. http://www.sciencedirect.com/science/article/pii/S000293431400401X.

Commissurotomy: Reintervention

Eur Heart J. 2013;34:1923–30. http://eurheartj.oxfordjournals.org/content/34/25/1923.

Echo: 2D Versus 3D for MV Area

Heart. 2013;99:253–8. http://heart.bmj.com/content/99/4/253.abstract.

Hunter Syndrome

Circulation. 2013;128:1269–70. http://circ.ahajournals.org/content/128/11/1269.full.

Mitral Commissurotomy for Restenosis After Previous Mitral Commissurotomy

Heart. 2013;99:1336–41. http://heart.bmj.com/content/99/18/1336.

Mitral Valvuloplasty Versus Surgical Treatment in MS with Severe TR

Circulation. 2007;116:I246–I250. http://circ.ahajournals.org/content/116/11_suppl/I-246.full.

Non-rheumatic MS

Eur J Echocardiogr 2009;10:103–105. http://ehjcimaging.oxfordjournals.org/content/10/1/103.

Post-valvuloplasty Tricuspid Regurgitation

Heart. 2013;99:91–7. http://heart.bmj.com/content/99/2/91.abstract.

Rheumatic Heart Disease in Bangladesh

Indian Heart J. 2016;68:88–98. http://www.sciencedirect.com/science/article/pii/S0019483215003181.

Systemic Embolism: Predictors

Ann Intern Med. 1998;128:885–9. http://annals.org/article.aspx?articleid=711449.

Updates and More

https://clinicalguidecvd.com/mvs

Chapter 64
Mitral Valve Prolapse (Barlow/Parachute Mitral Valve Syndrome)

ICD-10 Code

I34.1

Alternate Names/Abbreviation

BARLOW SYNDROME
PARACHUTE MITRAL VALVE SYNDROME

Description/Etiology

The most common cause of chronic MR in developed countries; younger populations with MVP present with Barlow's disease; older populations present with fibroelastic deficiency disease

Barlow's Disease:

Thickened/diffusely redundant myxomatous leaflet tissue with disrupted collagen and elastic layers
Prolapse of most mitral leaflet segments
Severe mitral annular enlargement
Elongated chordae
Patients usually present in young/middle-aged at time of surgery after long history of murmur or MR [15]

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 885
DOI 10.1007/978-1-4471-7293-2_64,

Fibroelastic deficiency:

Decreased connective tissue deficient in collagen, elastin, proteoglycans
Thin, smooth, translucent leaflets without excess tissue
Moderate annulus dilatation
Thin, slightly elongated chordae
Patients often present at older age with chordal rupture and flail leaflet after a short clinical history

Familial [14] and nonfamilial forms

Secondary forms of MVP occurring in other known conditions listed in Predisposing/Comorbid Conditions

Predisposing/Comorbid Conditions

ANEURYSMS-OSTEOARTHRITIS SYNDROME
ATRIAL SEPTAL DEFECT – SECUNDUM
ATRIOVENTRICULAR LEFT-SIDED BYPASS TRACTS
CARDIOMYOPATHY – HYPERTROPHIC
EHLERS-DANLOS SYNDROME
HOMOCYSTINURIA
LOEYS-DIETZ SYNDROME
MARFAN SYNDROME
OPEN ANGLE GLAUCOMA [10]
OSTEOGENESIS IMPERFECTA
PRIMARY HYPOMASTIA
PSEUDOXANTHOMA ELASTICUM
VON WILLEBRAND DISEASE [AND OTHER COAGULOPATHIES]

Demography

2–3 % of general population (global: >176 million persons)
Often familial [14]

Pathophysiology

Morphology of Barlow's:

- Expansion of middle spongiosa layer of MV due to proteoglycan accumulation
- Structural alterations of collagen in all components of leaflets
- Structurally abnormal chordae (composed of increased amounts of glycosaminoglycans) [13]

Systolic billowing of either or both MV leaflets into LA, with or without MR [3]

Signs/Symptoms [1]

CHEST – PAIN [2]
CHEST – PALPITATIONS
CHEST, STERNUM, CURV – POST (PECTUS EXCAVATUM)
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)
DIZZY/LIGHTHEADED/PRESYNCOPE
FATIGUE
HEART, LV, APEX – CLICK(S), SYS [5, 15]
HEART, LV, APEX – MURMUR, DIAS
HEART, LV, APEX – MURMUR, SYS [5, 15]
HEART, LV, APEX – SOUNDS, EARLY DIAS
HYPOTENSION (BLOOD PRESSURE – DECREASED/LOW)
SPINE, THORACIC – STRAIGHT

Differentiation

Other causes of MR

Complications

Fibrin emboli [4]

Infective Endocarditis
Progressive MR
Ruptured chordae [13]
Stroke
Sudden Death

Laboratory

NS

ECG [6]

DYSRHY – PREEXCITATION
DYSRHYTHMIAS – ATRIAL (PACS/OTHERS)
DYSRHYTHMIAS – VENTRICULAR (PVCS/OTHERS)
Q WAVE – ABN
QT/QTC INTERVAL – LONG
ST-T WAVE – ABN, NS
T WAVE – INVER, ABN
U WAVE – PROMINENT

Imaging

MV, FLOW – REGURG
MV, LEAFLETS – THICK [ECHO: ≥5 MM]
MV, LEAFLETS, MOTION – PROLAPSE [ECHO: >2 MM ABOVE ANNULUS IN PARASTERNAL VIEW]

Genomics [14]

DCHS1

Other Tests

Ambulatory ECG monitoring [7]

Treatment: Nonpharmacologic

Reassurance of benign prognosis except in patients with severe forms
DC stimulants

Treatment: Pharmacologic

Endocarditis prophylaxis – patients with:

- Definite MR
- Thick leaflets
- Elongated chordae
- Dilated LA/LV

ASA [8]
Warfarin [8]
Beta-blockers [8]

Treatment: Surgical/Invasive

MV repair

Prevention

Infective Endocarditis prophylaxis for severe forms

Course

Benign in most cases, with no lifestyle restrictions, but may progress to significant MR over subsequent 1–2 decades in up to 1/4 of persons with mild MVP

Notes

[1] A variety of additional symptoms, many neuropsychiatric, such as panic disorder, have been described but are debated as part of a syndrome related to MVP
[2] Seldom resembles angina pectoris
[3] MR severity ranges from mild to severe
[4] May cause ophthalmic abnormalities such as visual field loss
[5] Timing highly variable under different conditions; occurs earlier (closer to S1) with decreased ventricular preload (Valsalva, upright position); occurs later with increased preload (squatting, handgrip)
[6] NS changes; may be normal
[7] Palpitations may be noted in absence of dysrhythmia
[8] No specific guideline recommendations available for medical therapy; drug Rx, if any, should be on case-by-case basis
[9] May be abnormal when secondary to predisposing congenital conditions
[10] MVP and open angle glaucoma may share same pathophysiologic basis involving proteoglycans and glycosaminoglycans
[11] May occur on MV leaflets/apparatus or in rare cases due to jet lesion in LA mural endocardium
[12] Especially in young females; correlates with fibrosis of papillary muscles and inferobasal LV wall, which is structural hallmark and correlates with ventricular arrhythmia origin, possibly due to myocardial stretch by prolapsing leaflet
[13] Chordal rupture: frequent pathological finding; may be secondary to mechanical weakening, combined with abnormal hemodynamic stresses arising from valve leaflet redundancy
[14] Inheritance: autosomal-dominant with variable penetrance influenced by age/sex; marked heterogeneity of clinical presentation including affected members within a family
[15] Classic auscultatory finding in MVP: dynamic, mid-late systolic click often associated with high-pitched, late systolic murmur; careful physical examination is highly sensitive for making MVP diagnosis, but specificity limited with echo as gold standard

Guidelines

2014 AHA/ACC Guideline for the Management of Patients with Valvular Heart Disease
J Am Coll Cardiol. 2014;63:e57–e185. http://content.onlinejacc.org/article.aspx?articleID=1838843.

Guidelines on the management of valvular heart disease (version 2012)
Eur Heart J. 2012;33:2469–75. http://www.escardio.org/guidelines-surveys/esc-guidelines/Pages/valvular-heart-disease.aspx.

Patient Information

Medlineplus

ENGLISH
http://www.nlm.nih.gov/medlineplus/mitralvalveprolapse.html.
ESPANOL
http://www.nlm.nih.gov/medlineplus/spanish/mitralvalveprolapse.html.

Genetics Home Reference

http://ghr.nlm.nih.gov/glossary=mitralvalveprolapse.

Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/mitral-valve-prolapse/basics/definition/con-20024748.

Texas Heart Institute

http://www.texasheartinstitute.org/HIC/Topics/Cond/mvp.cfm.

AHA

http://www.heart.org/HEARTORG/Conditions/More/HeartValveProblemsandDisease/Problem-Mitral-Valve-Prolapse-UCM-450441-Article.jsp.

Cleveland Clinic

http://my.clevelandclinic.org/services/heart/disorders/mitral-valve-prolapse.

Harvard

http://www.health.harvard.edu/heart-health/mitral-valve-prolapse.

Professional Information

DCHS1 Mutation

Nature. 2015. doi:10.1038/nature14670. http://www.nature.com/nature/journal/vaop/ncurrent/full/nature14670.html?WT.ec-id=NATURE-20150813&spMailingID=49306476&spUserID=MTM4MTQzNTcyMTQxS0&spJobID=741956713&spReportId=NzQxOTU2NzEzS0.

Epidemiology/Pathophysiology

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Left Atrial Mural Endocarditis Secondary to MVP JET Lesion

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Mitral Valve Morphology

Circulation. 2013;127:832–41. http://circ.ahajournals.org/content/127/7/832.full.

MR/Flail Leaflet: Effect of Age on Outcomes

Eur Heart J. 2013;34:2600–09. http://eurheartj.oxfordjournals.org/content/34/33/2600.abstract.

Natural History

Circulation. 2002;106:1355–61. http://circ.ahajournals.org/content/106/11/1355.full.pdf+html?sid=1a6c50a2-20e9-4419-a523-b5484f3dcee3.

Open Angle Glaucoma

Heart.2015;101:609–15.http://heart.bmj.com/content/101/8/609?etoc.

Sudden Cardiac Death

Heart Rhythm. 2016;13:498–503. http://www.heartrhythmjournal.com/article/S1547-5271(15)01196-0/abstract.

Sudden Cardiac Death

Circulation. 2015;132:556–66.http://circ.ahajournals.org/content/132/7/556.full.

VF/Sudden Cardiac Death

J Am Coll Cardiol. 2013;62:222–30. http://content.onlinejacc.org/article.aspx?articleID=1709682.

Updates and More

https://clinicalguidecvd.com/mvp

Chapter 65
Myocarditis

Management Keys

Evaluate for underlying/treatable cause
Monitor for possible progression to Dilated Cardiomyopathy and HF
Special vigilance (especially for dysrhythmias) when low voltage or LBBB present on ECG [7]

ICD-10 Code

I40.1

Alternate Names/Abbreviation

NS

Description/Etiology [22]

Cardiac inflammation due to a wide variety of causes (see PREDISPOSING/COMORBID CONDITIONS, below) other than those due to CAD

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_65,

Clinical presentation varies, including: [3]

Acute chest pain
Acute dysrhythmias with

Palpitations
Syncope

Cardiogenic shock
New onset/worsening HF

Predisposing/Comorbid Conditions [1]

AUTOIMMUNE

CHURG-STRAUSS SYNDROME
DIABETES MELLITUS [TYPE 1]
HYPERTHYROIDISM
INFLAMMATORY BOWEL DISEASE
KAWASAKI DIS
MYASTHENIA GRAVIS
POLYMYOSITIS
RHEUMATOID ARTHRITIS
SARCOIDOSIS
SCLERODERMA
SYSTEMIC LUPUS ERYTHEMATOSUS
WEGENER GRANULOMATOSIS

DRUGS – ALLERGIC [23]

AMITRYPTYLINE
CEFACLOR
COLCHICINE
ISONIAZID
LIDOCAINE
LOOP/THIAZIDE DIURETICS
METHYL DOPA
PENICILLIN
PHENYLBUTAZONE
PHENYTOIN

SULFONAMIDES
TETRACYCLINE

DRUGS – TOXIC

AMPHETAMINES
ANTHRACYCLINES
CATECHOLAMINES
CLOZAPINE
COCAINE
CYCLOPHOSPHAMIDE
FLUOURACIL
HEMETINE
INTERLEUKIN 2
LITHIUM
TRASTUZUMAB

HEAVY METAL TOXICITY

COPPER TOXICITY
IRON TOXICITY
LEAD TOXICITY

IMMUNE-MEDIATED

SERUM SICKNESS
TETANUS TOXIOD
VACCINES

INFECTIONS – BACTERIAL

CHAGAS DISEASE
DIPHTHERIA (CORYNEBACTERIUM DIPHTHERIA)
HEMOPHILUS INFLUENZA
LEGIONELLOSIS (LEGIONELLA)
LEPTOSPIROSIS
LYME DISEASE (BORRELIA BURGDORFERI)
MENINGOCOCCEMIA (NEISSERIA MENINGITIDIS)
MYCOBACTERIUM TUBERCULOSIS
MYCOPLASMA PNEUMONIAE

Q FEVER
ROCKY MOUNTAIN SPOTTED FEVER (RICKETTSIA RICKETTSII)
SCRUBTYPHUS(RICKETTSIATSUTSUGAMUSHI)
SHIGELLOSIS (S BOYDII, S DYSENTERIAE, S FLEXNERI, S SONNEI)
TYPHOID FEVER
TYPHUS – SCRUB
YERSINIOSIS (Y ENTEROCOLITICA)

INFECTIONS – VIRAL

ADENOVIRUS [MAINLY CHILDREN]
CHICKENPOX (VARICELLA)
COXSACKIE VIRUS A
COXSACKIE VIRUS B
CYTOMEGALOVIRUS [MAINLY IMMUNOCOMPROMISED PTS]
DENGUE FEVER
EBOLA VIRUS DISEASE
ECHOVIRUS
ENTEROVIRUS
HEMORRHAGIC FEVER WITH RENAL SYNDROME
HEPATITIS C
HERPES SIMPLEX
HIV
HUMAN HERPES VIRUS 6
INFECTIOUS MONONUCLEOSIS
INFLUENZA – SEASONAL
LASSA FEVER
LYMPHOCYTIC CHORIOMENINGITIS
MARBURG FEVER
MUMPS
PARVOVIRUS B19 INFECTION
POLIOMYELITIS- ACUTE

PSITTACOSIS
RABIES
RUBEOLA
SMALLPOX (VARIOLA)
WEST NILE FEVER

OTHERS

ALCOHOL USE/EXCESS
ARSINE POISONING
BEE STING
BERI BERI
CARBON MONOXIDE POISONING
CELIAC DISEASE
ELECTRIC SHOCK
HEART TRANSPLANT [REJECTION]
MALARIA
PHEOCHROMOCYTOMA
PHOSPHOROUS
RADIATION
SCORPION STING
SNAKE BITE
SODIUM AZIDE
SPIDER BITE
WASP STING
CARDIOMYOPATHY – PERIPARTUM
CARDIOMYOPATHY – TAKOTSUBO
GIANT CELL ARTERITIS
INFLUENZA – AVIANl
TRICHINELLOSIS

Demography

Varies with etiology
More common in males
All ages, especially young adults

Pathophysiology

Acute myocyte Injury due to various mechanisms including:

Autoimmune cell invasion of myocardium
Infectious pathogen entry/replication
Intracellular antigen exposure
Immune system activation
Myocyte ischemia/necrosis

Signs/Symptoms [8]

APPETITE – DECR (ANOREXIA) [INCL POOR FEEDING]
BREATHING – DIFF (DYSPNEA) [20]
BREATHING – RAPID (TACHYPNEA)
CHEST – PAIN [6, 11]
CHEST – PALPITATIONS
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)
COUGH
FATIGUE
FEVER [10]
HEART – FRICTION RUB, PERICARD
HEART, LV, APEX – MURMUR, SYS
HEART, RATE – RAPID (TACHYCARDIA) [WITH/WITHOUT FEVER]
HEART, RHYTHM – IRREG
HEART, S1, INTENSITY – DECR/ABSENT
HEART, S3 LV
JOINTS – PAIN (ARTHRALGIA) [10]
LIVER – ENLARGED (HEPATOMEGALY)
MENTATION – WEAKNESS (MALAISE)
MUSCLES – PAIN (MYALGIA) [10]
VOMITING (EMESIS)

Differentiation

AMI
Other causes of HF/LV dysfunction

Complications

Cardiogenic Shock
Dysrhythmias [7]
HF
SCD [7] [9]

Laboratory

BLOOD, TROPONIN – INCR [2]

ECG

T WAVE – INVER, ABN [4]
AV COND – 1ST DEGREE BLOCK
AV COND – 2ND DEGREE BLOCK
AV COND – 3RD DEGREE BLOCK
DYSRHYTHMIAS – ATRIAL (PACS/OTHERS)
DYSRHYTHMIAS – VENTRICULAR (PVCS/OTHERS) [7]
PR SEGMENT – DEPRESSED
Q WAVE – ABN [6] [UNCOMMON]
QRS – LBBB/LBBB PATTERN [18]
QRS – LONG, NS
QRS, AXIS – L [7]
QT/QTC INTERVAL – LONG [7]
RATE – DECREASED (SINUS BRADYCARDIA)

ST SEGMENT – DEPR
ST SEGMENT – ELEV [5] [6]
ST-T WAVE – ABN, NS [4]
VOLTAGE, GEN – DECR [19]

Imaging

CARDIOMEGALY [12]
LUNGS – INFILTRATES
LV, INTRACAVITY – MASS [14]
LV, MYOCARD – LGE [21]
LV, SYS – DYSF [13]
MRI: INTRACELLULAR EDEMA; INTERSTITIAL EDEMA; HYPEREMIA; CAPILLARY LEAKAGE; MYOCARD FIBROSIS/NECROSIS
PERICARD – FLUID
PLEURA – FLUID
PUL, VASCULARITY – INCR

Other Tests

EMB

Treatment: Nonpharmacologic

SCD prevention: withdraw from competitive athletics and vigorous exertion for prescribed period

Treatment: Pharmacologic

Antiarrhythmic [17]
Antivirals [15]
HF protocol
Immunosuppressives [16]

Treatment: Surgical/Invasive

Mechanical circulatory support and card transplant
Pacemaker [17]

Prevention

Varies with etiology

Course

Varies with etiology
Viral Myocarditis: 19 % mortality in <5 years

Notes

[1] Partial list
[2] Cardiac troponins: limited value because:

Do not differentiate from myocardial ischemia
Normal value does not exclude Myocarditis
May be increased in many other conditions

[3] Consider this diagnosis in absence of other explanations of these manifestations, especially CAD
[4] T wave inversion usually occurs after complete normalization of ST-T wave changes in Myocarditis (usually occurs while ST segment elevation still present in AMI)
[5] May resemble AMI
[6] ST segment usually concave upward vs convex upward in myocardial ischemia/AMI
[7] Tachyarrhythmias associated with increased mortality in children
[8] Often asymptomatic; clinical manifestations highly variable
[9] Including young children
[10] Prodromal phase preceding by days-weeks in viral form

[11] Pericardial involvement
[12] Due to chamber enlargement/pericardial effusion
[13] LV dysfunction common; RV dysfunction uncommon
[14] Thrombus; occurs in up to 25 % of patients
[15] Limited effectiveness because viral infection usually precedes Myocarditis by weeks
[16] Eg, corticosteroids and cyclosporine for Giant Cell Myocarditis
[17] When specifically indicated for significant arrhythmias/ high grade HB
[18] LBBB associated with worse outcome (up to 8x in children)
[19] Low voltage is risk factor for dysrhythmias; mechanism unknown
[20] Difficult breathing is most common complaint in children
[21] Presence of LGE may be best independent predictor of all-cause and cardiac mortality in biopsy-proven viral Myocarditis
[22] ACC/AHA Guideline Description***

Sec 5.6.1 Myocarditis

Inflammation of the heart may cause HF in about 10 % of cases of initially unexplained cardiomyopathy. A variety of infectious organisms, as well as toxins and medications, most often postviral in origin, may cause myocarditis. In addition, myocarditis is also seen as part of other systemic diseases such as systemic lupus erythematosus and other myocardial muscle diseases such as HIV cardiomyopathy and possibly peripartum cardiomyopathy. Presentation may be acute, with a distinct onset, severe hemodynamic compromise, and severe LV dysfunction as seen in acute fulminant myocarditis, or it may be subacute, with an indistinct onset and better-tolerated LV dysfunction. Prognosis varies, with spontaneous complete resolution (paradoxically most often seen with acute fulminant myocarditis) to the development of DCM despite immunosuppressive therapy. The role of immunosuppressive therapy is controversial.

Targeting such therapy to specific individuals based on the presence or absence of viral genome in myocardial biopsy samples may improve response to immunosuppressive therapy.

[23] ACC/AHA Guideline description***

Sec 5.7.1 Hypersensitivity Myocarditis
Hypersensitivity to a variety of agents may result in allergic reactions that involve the myocardium, characterized by peripheral eosinophilia and a perivascular infiltration of the myocardium by eosinophils, lymphocytes, and histiocytes. A variety of drugs, most commonly the sulfonamides, penicillins, methyldopa, and other agents such as amphotericin B, streptomycin, phenytoin, isoniazid, tetanus toxoid, hydrochlorothiazide, dobutamine, and chlorthalidone, have been reported to cause allergic hypersensitivity myocarditis. Most patients are not clinically ill but may die suddenly, presumably secondary to an arrhythmia.

***From 2013 ACCF/AHA Guideline for the Management of Heart Failure: A Report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines. J Am Coll Cardiol. 2013;62(16):e147–e239

Guidelines

2013 ACCF/AHA Guideline for the Management of Heart Failure
J Am Coll Cardiol. 2013;62:e147–e239. http://content.onlinejacc.org/article.aspx?articleid=1695825.

ESC Guidelines for the diagnosis and treatment of acute and chronic heart failure 2012
Eur Heart J. 2012;33:1787–1847. http://eurheartj.oxfordjournals.org/content/ehj/33/14/1787.full.pdf.

HFSA: Myocarditis: Current Treatment
J Card Fail. 2010;16:e176–e179. http://www.onlinejcf.com/article/S1071-9164(10)00231-9/fulltext.

Patient Information

Medlineplus

ENGLISH
http://www.nlm.nih.gov/medlineplus/ency/article/000149.htm.
ESPANOL.
http://www.nlm.nih.gov/medlineplus/spanish/ency/article/000149.htm.

Texas Heart Institute

http://www.texasheartinstitute.org/HIC/Topics/Cond/myocard.cfm.

Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/myocarditis/basics/definition/con-20027303.

Myocarditis Foundation

http://www.myocarditisfoundation.org/about-myocarditis/.

Nord

https://rarediseases.org/rare-diseases/giant-cell-myocarditis/.

Cincinnati Childrens: Pediatric Myocarditis

http://www.cincinnatichildrens.org/health/m/myocarditis/.

Johns Hopkins

http://www.hopkinsmedicine.org/heart-vascular-institute/conditions-treatments/conditions/myocarditis.html.

Seattle Childrens

http://www.seattlechildrens.org/medical-conditions/heart-blood-conditions/myocarditis/.

Professional Information

History

Heart Vessels. 1985;1(Supplement):1–3. http://link.springer.com/article/10.1007%2FBF02072348.

ESC Position Statement

Eur Heart J. 2013;34:2636–48. http://eurheartj.oxfordjournals.org/content/34/33/2636.

Review

Lancet. 2012;379:738–47. http://www.sciencedirect.com/science/article/pii/S014067361160648X.

Review

Circulation. 2006;113:876–90. http://www.circ.ahajournals.org/content/113/6/876.full.

Review

Mayo Clin Proc. 2009;84:1001–9. http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2770911/.

Review

Circulation. 2009;119:2615–24. http://circ.ahajournals.org/content/119/19/2615.full.

Review: Clinical Presentation/Diagnosis

Heart. 2015;101:1332–44. http://heart.bmj.com/content/101/16/1332.extract.

Review: Diagnosis/Treatment in Children

Circulation. 2014;129:115–28. http://circ.ahajournals.org/content/129/1/115.extract.

Review: State of Art Update

J Am Coll Cardiol. 2012;59:779–92. http://content.onlinejacc.org/article.aspx?articleid=1201151&resultClick=3.

Autoimmunity

J Clin Invest. 2011;121:1561–73. http://www.jci.org/articles/view/44583.

Children: Clinical Presentation

Am J Emerg Med. 2009;27:942–7. http://www.sciencedirect.com/science/article/pii/S0735675708005573#.

Diagnosis of Myocarditis

J Am Coll Cardiol. 2016;67:1812–4. http://content.onlinejacc.org/article.aspx?articleID=2512991#tab1.

Endomyocardial Biopsy

Eur Heart J. 2007;28:3076–93. http://www.escardio.org/guidelines-surveys/esc-guidelines/Pages/endomyocardial-biopsy.aspx.

Gender/Age Characteristics

Heart. 2013;99:1681–4. http://heart.bmj.com/content/99/22/1681.abstract.

Giant Cell Arteritis (Case Report)

Circ Heart Fail. 2016;9:e002778. http://circheartfailure.ahajournals.org/content/9/2/e002778.full.

Imaging: CMR

J Am Coll Cardiol. 2016;67:1800–11. http://content.onlinejacc.org/article.aspx?articleID=2512985#tab1.

LGE: Long-Term Predictor of Mortality in Viral Myocarditis

J Am Coll Cardiol. 2012;59:1604–15. http://content.onlinejacc.org/article.aspx?articleID=1203136.

Pheochromocytoma

Circulation. 2014;129:1348–9. http://circ.ahajournals.org/content/129/12/1348.full.

Tachyarrhythmias and Increased Mortality in Children

Am J Cardiol. 2014;113:535–40. http://www.sciencedirect.com/science/article/pii/S0002914913021516.

Tuberculous Myocarditis (Case Report)

Circulation. 2013;128:271–7. http://circ.ahajournals.org/content/128/3/271.full.

Viral Myocarditis: Diagnosis

Arch Cardiovasc Dis. 2009;102:559–68. http://www.ncbi.nlm.nih.gov/pubmed/19664576?dopt=Abstract.

Updates and More

https://clinicalguidecvd.com/myocard

Chapter 66
Myxoma: Left Atrium

ICD-10 Code

D15.1

Description/Etiology

Most common primary benign cardiac tumor in adults; 75 % of all cardiac myxomas
Etiology either idiopathic or congenital as part of Carney Complex [5]

Predisposing/Comorbid Conditions

BREAST FIBROADENOMA
CARDIAC TUMOR(S)
CARNEY COMPLEX [5]
DYSRHYTHMIAS – ATRIAL [13]

Demography

More common in females
All ages
Cardiac myxomas may be more common and present at younger age in developing countries

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 911
DOI 10.1007/978-1-4471-7293-2_66,

Pathophysiology

Most often attached by stalk to region of fossa ovalis
Composition: gelatinous (myxoid); heterogeneous, often with areas of necrosis, cysts, hemorrhage
Flow obstruction/regurgitation at level of MV due to mobile, pedunculated tumor
Constitutional symptoms (e.g., fever) due to tumor secretion of IL-6
Metastatic embolization [16]

Signs/Symptoms [7] [12]

BEHAVIOR – BIZARRE/CHANGED
BODY, APPEARANCE – WASTING (CACHEXIA)
BREATH SOUNDS – CRACKLES (RALES) [MAY BE LOCAL]
BREATHING – DIFF (DYSPNEA)
BREATHING – DIFF, NOCTURNAL (DYSPNEA, NOCT)
CHEST – PAIN
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)
CONSCIOUSNESS – LOSS, SUDDEN, UPRIGHT (ORTHOSTATIC SYNCOPE)
COUGH
EXTREM – RAYNAUD PHENOMENON
EXTREM, DIGITS – CLUBBED
EXTREM, LOWER, BILAT – EDEMA
EYES, VISION – DECR/LOSS
FATIGUE
FEVER
HEART, LV, APEX – MURMUR, DIAS [2]
HEART, LV, APEX – MURMUR, SYS [2]
HEART, P2, INTENSITY – INCR [2]
HEART, S1, INTENSITY – INCR [2]
HEART, S1, SPLIT – WIDE [2]
HEART, S4 LV [2]

HEART, SOUND – EARLY DIAS [1] [2]
SKIN – LENTIGINES [CARNEY COMPLEX] [5]
SKIN – PETECHIAE/ECCHYMOSES/PURPURA
SKIN, COLOR – PALE (PALLOR)
SPUTUM – BLOOD (HEMOPTYSIS)
WEIGHT – LOSS

Differentiation

Acute Rheumatic Fever
Cerebrovascular Disease
Infective Endocarditis
Mitral Regurgitation
Mitral Stenosis
Myocarditis
Pulmonary disease
Pulmonary Hypertension
Vasculitis

Complications

Acute Mitral Regurgitation [8]
AMI
Acute Pulmonary Edema
Detach/Lodge at Aortic Bifurcation
Endocarditis
Extension Locally
Intracranial Arterial Aneurysm
LV dysfunction
Metastasis [16]
Peripheral emboli [4] [16]
Pulmonary Arterial Hypertension
Stroke
Sudden Death
Visceral infarction

Laboratory [12]

BLOOD, ESR – INCR
BLOOD, HGB/HCT – DECR (ANEMIA) [6]
BLOOD, IGG – INCR
BLOOD, IL-6 – INCR
BLOOD, PLATELETS – DECR (THROMBOCYTOPENIA)

ECG [10] [12]

DYSRHYTHMIAS – ATRIAL (PACS/OTHERS)
N/NS ABN
P WAVE, DUR – INCR
QRS – RBBB/RBBB PATTERN
QRS – RVH PATTERN
QRS, AXIS – R
RATE – INCREASED (SINUS TACHYCARDIA)

Imaging [9] [10] [12]

LA, CHAMBER, SIZE – INCR [MAY BE NORMAL SIZE]
LA, INTRACAVITY – MASS [MAY BE CALCIFIED]
LGE, TUMOR [14]
MV, FLOW – REGURG
MV, LEAFLETS, MOTION – ABN [FLUTTERING]
PA, PRESS – INCR
PUL, VASCULARITY – INCR
RV, CHAMBER, SIZE – INCR [MAY BE NORMAL SIZE]

Genomics

PRKAR1A

Other Tests

Cardiac catheterization/coronary angiography – mainly for CAD
EMB

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic

NS

Treatment: Surgical/Invasive

Surgical resection: usually curative

Course

Recurrence after surgical resection [9]:

About 13 %;
Higher in Carney Complex
Rare after 4 years
Usually at same site

Normal lifespan after complete resection

Notes

[1] "Tumor plop" low-pitch; caused by tumor prolapse across MV; diastolic murmur usually absent

[2] Auscultatory findings may vary with time/patient position
[3] May protrude through MV orifice
[4] Tumor particle or superimposed thrombus; occurs in up to 50 % of patients; most common sites are CNS, retina, coronary artery, extremities; embolic event may be initial clinical manifestation
[5] Carney Complex: autosomal dominant condition, including mammary myxoid fibroadenoma, skin pigmented lesions, endocrine disorders, testicular tumors, psammomatous melanotic schwannoma; 2/3 s have cardiac myxomas
[6] Hemolytic; associated with calcified tumor
[7] Occurrence of symptoms acutely with upright or other certain body positions should raise this diagnostic possibility
[8] Ruptured chordae tendineae due to "wrecking ball effect" of calcified tumor
[9] TEE especially valuable; MRI used when echo inadequate/further characterization needed; semi-annual follow-up echo for 4 years after surgical resection recommended for recurrence detection
[10] Often normal
[11] Adult benign cardiac tumors (frequency):

Myxoma (45 %)
Lipoma (20 %)
Papillary Fibroelastoma (15 %)
Hemangioma (5 %)
Fibroma (3 %)
Rhabdomyoma (1 %)
Teratoma (<1 %)
Others (10 %)

Pediatric benign cardiac tumors:

Rhabdomyoma (45 %)
Fibroma (15 %)
Myxoma (15 %)
Teratoma (15 %)
Hemangioma (5 %)
Others (5 %)

[12] Clinical manifestations highly variable and mainly determined by tumor location/size
[13] Especially Supraventricular Tachycardia, which may be initial manifestation
[14] For tumor vascularity
[16] Embolic tumor cells may remain viable at site of embolization, thereby becoming distant metastases or peripheral tumor masses

Guidelines

NS

Patient Information

Genetics Home Reference: Carney Complex

http://ghr.nlm.nih.gov/condition/carney-complex.

Images

http://www.nlm.nih.gov/medlineplus/ency/imagepages/18078.htm.

Medlineplus

ENGLISH

http://www.nlm.nih.gov/medlineplus/ency/article/007273.htm.

ESPANOL

http://www.nlm.nih.gov/medlineplus/spanish/ency/article/007273.htm.

Patient Journey

BMJ. 2013;347:f4430. http://www.bmj.com/content/347/bmj.f4430.

Pubmed

http://www.ncbi.nlm.nih.gov/pubmedhealth/PMH0004532/.

Merck

http://www.merckmanuals.com/home/heart-and-blood-vessel-disorders/heart-tumors/myxomas.

Professional Information

Carney Complex

J Cardiothorac Surg. 2011;6:25. doi: 10.1186/1749-8090-6-25. http://www.cardiothoracicsurgery.org/content/6/1/25.

Clinical Presentation

Medicine. 2001;80:159–72. http://journals.lww.com/md-journal/Fulltext/2001/05000/Clinical-Presentation-of-Left-Atrial-Cardiac.2.aspx.

Carney Complex

http://ghr.nlm.nih.gov/condition/carney-complex.

Case Report from THIJ: Catastrophic Systemic Embolization

Tex Heart Inst J. 2014;41;64–6. http://www.thij.org/doi/full/10.14503/THIJ-12-2964?=.

Echo

J Am Soc Echocardiogr. 2011;24:618–24. http://www.sciencedirect.com/science/article/pii/S089473171100040X.

Endocarditis

Eur Heart J. 1992;13:1592–3. http://eurheartj.oxfordjournals.org/content/13/11/1592.3.

Familial Multicentric

Eur J Echocardiogr. 2005;6:148–50. http://ehjcimaging.oxfordjournals.org/content/6/2/148.long.

Giant LA Myxoma/Eye Embolism/Endocarditis: Case Report

J Am Coll Cardiol. 2014;63:2049. http://content.onlinejacc.org/article.aspx?articleID=1851444.

Image: Carney's Complex

J Am Coll Cardiol. 2010;55:1395. http://content.onlinejacc.org/article.aspx?articleid=1142663&resultClick=3.

Image: CT

N Engl J Med. 2008;358:728. http://www.nejm.org/doi/full/10.1056/NEJMicm0708551.

Image: Echo

N Engl J Med. 2009;361:e51. http://www.nejm.org/doi/full/10.1056/NEJMicm0807505.

Imaging Cardiac Tumors: MRI/CT

Clin Radiol. 2009;64:1214–30. http://www.ncbi.nlm.nih.gov/pubmed/19913133.

Left Atrial Rhabdomyosarcoma: Case Report

Circulation. 2014;129:e503–e505. http://circ.ahajournals.org/content/129/21/e503.full.

Left Atrial Thrombus Mimicking Myxoma

Circulation. 2006;113:e456–e457. http://circ.ahajournals.org/content/113/11/e456.full?sid=c1d4e4bc-f477-401a-9e82-fa4ac143d123.

Left Ventricular Dysfunction

Angiology. 2006;57:119–22. http://www.ncbi.nlm.nih.gov/pubmed/16444467.

Survival After Resection

Circulation. 2008;118:S7–S15. http://circ.ahajournals.org/content/118/14-suppl-1/S7.full.

Updates and More

https://clinicalguidecvd.com/lamyx

Chapter 67
Myxoma: Left Ventricle

ICD-10 Code

D15.1

Alternate Names/Abbreviation

NS

Description/Etiology

Rare LV intracardiac pedunculated or intramural tumor
Idiopathic or congenital etiology as part of Carney Complex [9]
Clinical features depend on tumor location

Predisposing/Comorbid Conditions

OTHER CARDIAC TUMOR(S)

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_67,

Demography

Females 3:1
Cardiac myxomas may be more common and present at younger age in developing countries

Pathophysiology

Composition: gelatinous (myxoid); heterogeneous, often with areas of necrosis, cysts, hemorrhage
Depending on tumor location/mobility, hemodynamic effects primarily:

LVOT obstruction
MR

Signs/Symptoms [5] [9]

BREATHING – DIFF (DYSPNEA) [3]
CHEST – PAIN [4]
CHEST – PALPITATIONS
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE) [3]
DIZZY/LIGHTHEADED/PRESYNCOPE [3]
EYES, VISION – DECR/LOSS
HEART – MURMURS, CHANGING
HEART, LSB, MID – MURMUR, SYS [6]
SPUTUM – BLOOD (HEMOPTYSIS)

Differentiation

Aortic Stenosis – Subaortic
Aortic Stenosis – Valvular
HF
Ventricular metastatic tumor
Ventricular septal Rhabdomyoma
Ventricular thrombus

Complications [2]

Conduction defects
Dysrhythmias
Stroke
Sudden Death
Systemic embolus [1]

Laboratory

NS

ECG [7]

N/NS ABN
QRS – LVH PATTERN

Imaging [8]

CARDIOMEGALY
LV, INTRACAVITY – MASS

Genomics

PRKAR1A

Other Tests

NS

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic

NS

Treatment: Surgical/Invasive

Tumor resection

Prevention

After resection monitor for tumor recurrence in LV/other cardiac chambers

Notes

[1] 2/3 s of patients; cerebral embolus most common
[2] May be initial presentation
[3] 50 % of patients; may be related to position
[4] May resemble angina pectoris; due to direct tumor obstruction or embolic obstruction of coronary artery
[5] Unlike atrial myxomas, constitutional symptoms uncommon
[6] Resembles AS
[7] May be normal
[8] Incidental finding of LV mass by imaging for another indication may be first manifestation
[9] Carney Complex: autosomal dominant condition, including mammary myxoid fibroadenoma, skin pigmented lesions, endocrine disorders, testicular tumors, psammomatous melanotic schwannoma; 2/3 s have cardiac myxomas

Guidelines

NS

Patient Information

Genetics Home Reference: Carney Complex

http://ghr.nlm.nih.gov/condition/carney-complex.

Images

http://www.nlm.nih.gov/medlineplus/ency/imagepages/18078.htm.

Medlineplus

ENGLISH
http://www.nlm.nih.gov/medlineplus/ency/article/007273.htm.

ESPANOL
http://www.nlm.nih.gov/medlineplus/spanish/ency/article/007273.htm.

Patient Journey

BMJ. 2013;347:f4430. http://www.bmj.com/content/347/bmj.f4430.

Pubmed

http://www.ncbi.nlm.nih.gov/pubmedhealth/PMH0004532/.

Merck

http://www.merckmanuals.com/home/heart-and-blood-vessel-disorders/heart-tumors/myxomas.

Professional Information

Carney Complex

J Cardiothorac Surg. 2011;6:25. doi: 10.1186/1749-8090-6-25. http://www.cardiothoracicsurgery.org/content/6/1/25.

Case Report

Egyptian Heart J. 2014;66:375–7. http://www.sciencedirect.com/science/article/pii/S1110260814000386.

Case Report

Eur J Echocardiog. 2009;10:593–5. http://ehjcimaging.oxfordjournals.org/content/10/4/593.

Missed Metastatic

World J Cardiol. 2013;5:387–90. http://www.ncbi.nlm.nih.gov/pmc/articles/PMC3817281/.

Stroke Presentation (Case Report)

Egyptian Heart J. 2014;66:375–7. http://www.sciencedirect.com/science/article/pii/S1110260814000386.

Ventricular Metastatic Liposarcoma (Case Report)

Circulation. 2013;127:e443–e445. http://circ.ahajournals.org/content/127/6/e443.full.

Updates and More

https://clinicalguidecvd.com/lvmyx

Chapter 68
Myxoma: Right Atrium

ICD-10 Code

D15.1

Description/Etiology

Tumor usually attached to interatrial septum; may be very large, pedunculated, often calcified; may be biatrial with dumbbell shape with common stalk attached to fossa ovalis

Etiology: idiopathic or congenital as part of Carney Complex [10]

Predisposing/Comorbid Conditions

CARDIAC TUMOR(S)

Demography

All ages

Cardiac myxomas may be more common and present at younger age in developing countries

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 927
DOI 10.1007/978-1-4471-7293-2_68,

Pathophysiology

Composition: gelatinous (myxoid); heterogeneous, often with areas of necrosis, cysts, hemorrhage
Functional TV stenosis
R-L shunting in presence of patent foramen ovale may cause paradoxical (systemic) embolism

Signs/Symptoms

ABDOMEN – FLUID (ASCITES)
BODY, APPEARANCE – WASTING (CACHEXIA)
BREATHING – DIFF (DYSPNEA) [9]
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)
CONSCIOUSNESS – LOSS, SUDDEN, UPRIGHT (ORTHOSTATIC SYNCOPE)
EXTREM – RAYNAUD PHENOMENON
EXTREM, DIGITS – CLUBBED
EXTREM, LOWER, BILAT – EDEMA
FATIGUE
FEVER
HEART, LSB, LOWER – MURMUR, DIAS [3] [4] [6] [7]
HEART, LSB, LOWER – MURMUR, SHORT, EARLY, SYS [2] [4] [6] [7]
HEART, LSB, LOWER – MURMUR, SYS [HOLOSYS] [4] [5] [6] [7]
HEART, SOUND – EARLY SYS [1] [6]
JOINTS – PAIN (ARTHRALGIA)
LIVER – ENLARGED (HEPATOMEGALY)
MUSCLES – PAIN (MYALGIA)
NECK, JVP – ELEV
NECK, JVP, V WAVE – INCR/LARGE [5] [6]
SKIN, COLOR – BLUE (CYANOSIS) [8]
SKIN, COLOR – RED (ERYTHEMA)
WEIGHT – LOSS

Differentiation

Carcinoid Syndrome
Constrictive Pericarditis
Ebstein Anomaly
Pulmonary Arterial Hypertension
Pulmonary Embolism
Pulmonary Stenosis
Pulmonary Venous Hypertension
Rheumatic Valve Disease
Tricuspid Stenosis

Complications

Acute Pulmonary Embolism
LV Dysfunction
Paradoxical Systemic Embolism
RHF

Laboratory

BLOOD, ESR – INCR
BLOOD, HGB/HCT – DECR (ANEMIA)
BLOOD, IGG – INCR
BLOOD, IL-6 – INCR
BLOOD, PLATELETS – DECR (THROMBOCYTOPENIA)

ECG

P WAVE – TALL/PEAKED
QRS – RBBB/RBBB PATTERN
QRS, AXIS – R

Imaging

RA, CHAMBER, SIZE – INCR
RA, INTRACAVITY – MASS [MAY BE CALCIFIED]
RV, CHAMBER, SIZE – INCR
TV, FLOW – REGURG

Genomics

PRKAR1A

Other Tests

Cardiac catheterization (caution: has risk of provoking pulmonary embolization)

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic

NS

Treatment: Surgical/Invasive

Tumor resection

Course

Recurrence after surgical resection:

About 13 %; probably lower in sporadic cases, higher in familial forms (eg, Carney Complex)

Usually at same site
Rare after 4 years

High incidence of perioperative arrhythmias but lower long term prevalence
Normal longevity usual after complete resection

Notes

[1] Due to expulsion of tumor from RV; may be associated with palpable tumor shock
[2] Caused by TR due to tumor holding open TV in early systole; precedes early systolic sound
[3] Long or only late diastole
[4] Accentuated by inspiration
[5] Large TR when TV damaged by tumor
[6] May change with position
[7] Murmurs may resemble friction rub
[8] With patent foramen ovale
[9] Most common first symptom
[10] Carney Complex: autosomal dominant condition, includes mammary myxoid fibroadenoma, skin pigmented lesions, endocrine disorders, testicular tumors, psammomatous melanotic schwannoma; 2/3 s have cardiac myxomas

Guidelines

NS

Patient Information

Genetics Home Reference: Carney Complex

http://ghr.nlm.nih.gov/condition/carney-complex.

Images

http://www.nlm.nih.gov/medlineplus/ency/imagepages/18078.htm.

Medlineplus

ENGLISH
http://www.nlm.nih.gov/medlineplus/ency/article/007273.htm.

ESPANOL
http://www.nlm.nih.gov/medlineplus/spanish/ency/article/007273.htm.

Patient Journey

BMJ. 2013;347:f4430. http://www.bmj.com/content/347/bmj.f4430.

Pubmed

http://www.ncbi.nlm.nih.gov/pubmedhealth/PMH0004532/.

Merck

http://www.merckmanuals.com/home/heart-and-blood-vessel-disorders/heart-tumors/myxomas.

Professional Information

Ascites/Extra-Cardiac Manifestations

Angiology. 2005;56:357–60. http://www.ncbi.nlm.nih.gov/pubmed/15889208.

Carney Complex

J Cardiothorac Surg. 2011;6:25. doi: 10.1186/1749-8090-6-25. http://www.cardiothoracicsurgery.org/content/6/1/25.

Images: Right Atrial Hemangioma

J Am Coll Cardiol. 2014;63:e41–e41. http://content.onlinejacc.org/article.aspx?articleID=1841608.

Left Ventricular Dysfunction

Heart Lung Circ. 2013;22:309–11. http://www.ncbi.nlm.nih.gov/pubmed/23098892.

Pulmonary Embolism

J Surg Case Rep. 2014. doi: 10.1093/jscr/rju115. . pii: rju115. http://jscr.oxfordjournals.org/content/2014/10/rju115.long.

Pulmonary Embolism

J Forensic Leg Med. 2008;15:454–6. http://www.ncbi.nlm.nih.gov/pubmed/18761314.

Updates and More

https://clinicalguidecvd.com/ramyx

Chapter 69
Myxoma: Right Ventricle

ICD-10 Code

D15.1

Description/Etiology

Usually attached to free wall or IVS
  Usually pedunculated, sometimes with long stalk
Tend to extend into RVOT tract and sometimes prolapse through PV during ventricular systole [1]
Etiology: idiopathic or congenital as part of Carney Complex [6]

Comorbid Conditions

Other cardiac tumors

Demography

All ages

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_69,

Pathophysiology

Composition: gelatinous (myxoid); heterogeneous, often with areas of necrosis, cysts, hemorrhage
Physiologic effects:

RVOT obstruction
Pulmonary regurgitation

Signs/Symptoms

BREATHING – DIFF (DYSPNEA)
CHEST – PAIN
CHEST – PALPITATIONS
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)
DIZZY/LIGHTHEADED/PRESYNCOPE
HEART, LSB, LOWER – MURMUR, DIAS [5]
HEART, LSB, MID – MURMUR, DIAS [4]
HEART, LSB, MID – MURMUR, SYS [2]
HEART, LSB, UPPER – MURMUR, DIAS [4]
HEART, LSB, UPPER – MURMUR, SYS [2]
HEART, LSB, UPPER – SOUND, DIAS [MID-DIASTOLE] [3]
HEART, LSB, UPPER – SOUND, EJECTION
HEART, LSB, UPPER – THRILL, SYS
NECK, JVP, A WAVE – INCR/LARGE (CANNON WAVE)

Differentiation

Pulmonary Artery Stenosis
Pulmonary Arterial Hypertension
Pulmonary Subvalvar Stenosis
Pulmonary Valve Stenosis

Complications

Pulmonary Embolism
RHF
Sudden death

Laboratory

NS

ECG

QRS – RVH PATTERN
QRS, AXIS – R

Imaging

CARDIOMEGALY [CXR: MILD; MAY BE N SIZE]
RV, CHAMBER, SIZE – INCR
RV, INTRACAVITY – MASS

Other Tests

Cardiac catheterization: determine RVOT gradient

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic

NS

Treatment: Surgical/Invasive

Tumor resection

Prevention

NS

Course

Post-tumor resection longevity approaches normal
Recurrence risk low (2–14 %)

Notes

[1] Consistent with high frequency of RVOT murmurs
[2] Intensity may be highly variable with time and body position but has inconsistent changes with inspiration
[3] "Tumor plop"
[4] Pulmonary regurgitation
[5] Flow across TV
[6] Carney Complex: autosomal dominant condition, including mammary myxoid fibroadenoma, skin pigmented lesions, endocrine disorders, testicular tumors, psammomatous melanotic schwannoma; 2/3 s have cardiac myxomas

Guidelines

NS

Patient Information

Genetics Home Reference: Carney Complex

http://ghr.nlm.nih.gov/condition/carney-complex.

Images

http://www.nlm.nih.gov/medlineplus/ency/imagepages/18078.htm.

Medlineplus

ENGLISH

http://www.nlm.nih.gov/medlineplus/ency/article/007273.htm.

ESPANOL

http://www.nlm.nih.gov/medlineplus/spanish/ency/article/007273.htm.

Patient Journey

BMJ. 2013;347:f4430. http://www.bmj.com/content/347/bmj.f4430.

Pubmed

http://www.ncbi.nlm.nih.gov/pubmedhealth/PMH0004532/.

Merck

http://www.merckmanuals.com/home/heart-and-blood-vessel-disorders/heart-tumors/myxomas.

Professional Information

Carney Complex

J Cardiothorac Surg. 2011;6:25. doi: 10.1186/1749-8090-6-25. http://www.cardiothoracicsurgery.org/content/6/1/25.

Case Report

Circulation. 2000;102:e14–e15. http://circ.ahajournals.org/content/102/2/e14.long.

Case Report

N Engl J Med. 2000;342:295. http://www.nejm.org/doi/full/10.1056/NEJM200001273420418.

Right Ventricular Hemangioma (Case Report)

J Am Coll Cardiol. 2013;61:2388. http://content.onlinejacc.org/article.aspx?articleID=1695217.

Right Ventricular Sarcoma (Case Report)

Eur Heart J. 2014;35:2509. http://eurheartj.oxfordjournals.org/content/35/37/2509.

Updates and More

https://clinicalguidecvd.com/rvmyx

Chapter 70
Nonsustained Ventricular Tachycardia (NSVT)

Management Keys

Post-acute coronary syndrome patients with NSVT should be investigated for evidence of ischemia (e.g., angina, ECG changes) and have evaluation of serum electrolytes, especially those taking diuretics or have had hypokalemia or hypomagnesemia

ICD-10 Code

I47.2

Alternate Names/Abbreviation

NSVT

Description/Etiology [1]

Ventricular tachycardia (≥3 consecutive beats arising below AV node, with wide QRS complex) that:

Does not lead to hemodynamic compromise
<30 s in duration
Rate >125/min

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_70,

May occur secondary to structural heart disease or in idiopathic form in otherwise apparently normal heart [2]

Predisposing/Comorbid Conditions

ACUTE MYOCARDIAL INFARCTION [2]
ARRHYTHMOGENIC RIGHT VENTRICULAR DYSPLASIA/CARDIOMYOPATHY
ATHLETES [8]
BRUGADA SYNDROME
CABG
CARDIAC AMYLOIDOSIS
CARDIOMYOPATHY – DILATED
CARDIOMYOPATHY – HYPERTROPHIC
CARDIOMYOPATHY – RESTRICTIVE
CATECHOLAMINERGIC POLYMORPHIC VENTRICULAR TACHYCARDIA
CHAGAS DISEASE
EARLY REPOLARIZATION [3]
GIANT CELL MYOCARDITIS
HEART FAILURE
HYPERTENSION – SYSTEMIC ARTERIAL
LONG QT SYNDROME – CONGENITAL
OBSTRUCTIVE SLEEP APNEA [9]
VALVULAR HEART DISEASE

Demography

All populations; varies according to etiology

Pathophysiology

Idiopathic: most likely triggered by adrenergic stimulation (eg, exercise)
Usually arises in RV

Signs/Symptoms [4]

CHEST – PALPITATIONS
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)
DIZZY/LIGHTHEADED/PRESYNCOPE
HEART, RATE – RAPID (TACHYCARDIA)
HYPOTENSION (BLOOD PRESSURE – DECREASED/LOW)

Differentiation

ECG: Supraventricular Tachycardia with aberrant conduction
Other causes of syncope

Complications

Tachycardia-Induced Cardiomyopathy
SCD [7] [10]

Laboratory

NS

ECG

DYSRHYTHMIAS – VENTRICULAR (PVCS/OTHERS)
QRS – LBBB/LBBB PATTERN [USUAL] [5]
QRS – LONG, NS [5]

Imaging [6]

NS/VAR WITH COMORBID

Other Tests

Ambulatory ECG monitoring
EP testing

Treatment: Nonpharmacologic

Avoidance of proven triggers

Treatment: Pharmacologic

Beta-blockers
Other drug indications dictated by underlying etiology

Treatment: Surgical/Invasive

ICD
Other interventions dictated by underlying etiology (eg, CABG for CAD)

Prevention

NS

Course

Varies according to etiology
Exercise-induced occurrence in asymptomatic persons: unaffected

Notes

[1] No universal definition
[2] Common post-AMI (40-70 % of pts) in first 24 h
[3] QRS slurring/notching, most often in young male athletes
[4] In addition to findings due to underlying disease
[5] During VT
[6] For risk stratification and decision for ICD
[7] Uncommon in idiopathic form
[8] Considered benign when suppressed by exercise; may be suppressed during deconditioning and resume with conditioning; mechanism unknown
[9] Especially during sleep
[10] High risk features:

Long duration (>7 beats)
Increasing frequency
Polymorphic appearance
ECG changes/biomarker elevation
Recurrent ischemia
Low LVEF
HF (Killip II-IV)
Prior ventricular arrhythmia
Occurrence more than 12–24 h after ACS
BBB

Guidelines

2012 ACCF/AHA/HRS Focused Update Incorporated Into the ACCF/AHA/HRS 2008 Guidelines for Device-Based Therapy of Cardiac Rhythm Abnormalities

J Am Coll Cardiol. 2013;61:e6–e75. http://content.onlinejacc.org/article.aspx?articleid=1486116#tab1.

Patient Information

Medlineplus

ENGLISH

http://www.nlm.nih.gov/medlineplus/ency/article/000187.htm.

ESPANOL

http://www.nlm.nih.gov/medlineplus/spanish/ency/article/000187.htm.

Merck

http://www.merckmanuals.com/home/heart-and-blood-vessel-disorders/abnormal-heart-rhythms/ventricular-tachycardia.

AHA

http://www.heart.org/HEARTORG/Conditions/Arrhythmia/AboutArrhythmia/Tachycardia-Fast-Heart-Rate-UCM-302018-Article.jsp#.

Cleveland Clinic

http://my.clevelandclinic.org/services/heart/disorders/arrhythmia/Ventricular-Tachycardia.

Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/ventricular-tachycardia/basics/definition/con-20036846.

Stanford

https://stanfordhealthcare.org/medical-conditions/blood-heart-circulation/ventricular-tachycardia.html.

Professional Information

Review

J Am Coll Cardiol. 2012;60:1993–2004. http://content.onlinejacc.org/article.aspx?articleid=1378223.

Review

Eur Heart J. 2004;25:1093–9. http://eurheartj.oxfordjournals.org/content/25/13/1093.long.

Acute Myocardial Infarction

Circulation. 1998;98:2030–6. http://circ.ahajournals.org/content/98/19/2030.

ARVD

Circulation, 2010;122:1144–52. http://circ.ahajournals.org/content/122/12/1144.abstract.

Cardiac Amyloidosis

Pacing Clin Electrophysiol. 2001;24:1228–33. http://onlinelibrary.wiley.com/doi/10.1046/j.1460-9592.2001.01228.x/abstract

Chagas Disease

Pacing Clin Electrophysiol. 2011;34:54–62. http://onlinelibrary.wiley.com/doi/10.1111/j.1540-8159.2010.02896.x/abstract.

Dilated Cardiomyopathy

Curr Cardiol Rep. 2005;7:368–75. http://www.ncbi.nlm.nih.gov/pubmed/16105493.

Exercise-Induced: Prognosis/Asymptomatic Persons

J Am Coll Cardiol. 2013;62:595–600. http://content.onlinejacc.org/article.aspx?articleID=1697402.

Giant Cell Myocarditis

N Engl J Med. 1997;336:1860–6. http://www.nejm.org/doi/full/10.1056/NEJM199706263362603.

Post-CABG

J Cardiovasc Electrophysiol. 2002;13:757–63. http://onlinelibrary.wiley.com/doi/10.1046/j.1540-8167.2002.00757.x/abstract.

Post-revascularization

J Cardiovasc Electrophysiol. 2002;13:342–6. http://onlinelibrary.wiley.com/doi/10.1046/j.1540-8167.2002.00342.x/abstract.

Updates and More

https://clinicalguidecvd.com/nsvt

Chapter 71
Obstructive Sleep Apnea

Management Keys

Consider this diagnosis in all pts with CVD/increased risk of CVD [1] [24]

ICD-10 Code

G47.33

Alternate Names/Abbreviation

OSA
SLEEP RELATED BREATHING DISORDER

Description/Etiology

Recurrent collapse of pharyngeal airway causing intermittent hypoxemia and CO_2 retention during sleep, disrupting normal autonomic and hemodynamic sleep responses; excess sympathetic drive may extend to waking hours, causing increased BP and increased HR [1]

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_71,

Apnea: ≥10 s pause in respiration associated with ongoing ventilatory effort [1]
Apnea-hypopnea index (# of apnea and hypopneas/h sleep) >5 and excessive daytime sleepiness [1]

Predisposing/Comorbid Conditions

ASTHMA
ATRIAL FIBRILLATION [22]
CHRONIC OBSTRUCTIVE PULMONARY DISEASE (EMPHYSEMA)
CORONARY ARTERY DISEASE
DEPRESSION
DIABETES MELLITUS
END STAGE RENAL DIS [18]
HEART FAILURE [24]
HYPERTENSION – SYSTEMIC ARTERIAL
HYPOTHYROIDISM [19]
OBESITY
POST-TRAUAMATIC STRESS DISORDER

Demography

M > F
Age <35 years: more common in African Americans

Pathophysiology

Possible mechanisms [likely multiple involved]:

Pharyngeal anatomy
Abnormal muscle tone of upper airway dilator muscles
Unstable ventilatory control
Lower lung volumes
Arousal threshold
Asynchronous activation of upper airway muscles and diaphragm

Other possible factors:

Resting BP/HR variability
Oxidative stress
Insulin resistance
Increased risk of thrombosis
Intrathoracic press changes

Signs/Symptoms

ARTERIAL PRESSURE, VARIABILITY – DECR [15]
BEHAVIOR – BIZARRE/CHANGED [5]
BLOOD PRESSURE, ARTERIAL – INCREASED/ELEVATED [3] [16]
BREATHING, NOCT – GASPING
BREATHING, NOCT – PAUSES (SLEEP APNEA)
BREATHING, NOCT – SNORING [2]
COGNITION – DEFECT, NS [6] [7]
FACE, JAW – DISPLACED, POST (RETROGNATHISM) [9]
FACE, JAW – OVERJET [8] [9]
HEART, RATE, VAR – DECR [15]
HEART, RHYTHM – IRREG [10] [16]
MENTATION – CONCENTRATION IMPAIRED [7]
MENTATION – SLEEPY (SOMNOLENCE) [7]
MOOD – DEPRESSED [7]
MOUTH, PALATE – LOW LYING
MOUTH, PHARYNX – NARROW. LAT
MOUTH, TONSILS, SIZE – INCR [9]
MOUTH, UVULA, SIZE – INCR [9]
NECK, SIZE – INCR [9]
TONGUE, SIZE – INCR (MACROGLOSSIA) [9]

Differentiation

Central Sleep Apnea
Hypothyroidism [19]

Major depression [7]
Other causes of Systemic Arterial Hypertension
Other causes of excessive daytime sleepiness [21]

Complications

AMI [1] [11]
AF [1] [23]
Cor Pulmonale
Growth Impairment
Nocturnal Angina
Nocturnal arrhythmias [10]
Pulmonary Arterial Hypertension [14]
Stroke [1]
Sudden Death [17]

Laboratory

BLOOD, ART, PCO2 – INCR (HYPERCAPNIA) [19]

ECG

AV COND – 2ND DEGREE BLOCK, MOBITZ I (WENCKEBACH)
DYSRHYTHMIAS – ATRIAL (PACS/OTHERS)
DYSRHYTHMIAS – VENTRICULAR (PVCS/OTHERS)
FIBRILLATION WAVES, ATRIAL

Imaging

NS/VAR WITH COMORBID [ESP HTN]

Other Tests

Polysomnography

Treatment: Nonpharmacologic

Continuous positive pressure [22]
Oral appliances
Sleep posture: lateral
Weight loss

Treatment: Pharmacologic

Antihypertensives

Treatment: Surgical/Invasive

Adenotonsillectomy [13]
Uvulopalatopharyngoplasty

Prevention

Maintain normal body weight

Course

Variable depending on comorbidities

Notes

[1] OSA: independent risk factor for Ischemic Stroke for patients with AF; appears to be under-diagnosed
[2] Almost all patients with OSA
[3] Occurs in >50 % of patients with OSA
[4] Occurs in estimated 30 % of patients with Systemic Arterial Hypertension (often undiagnosed in this population)
[5] Especially children
[6] May be lifelong; learning disabilities in children
[7] Mental changes of major depression and OSA overlap
[8] Maxilla extrudes forward in relation to mandible
[9] 1 or more findings of oropharyngeal narrowing, especially tongue and lateral pharyngeal enlargement; occur in most patients with OSA
[10] Most common:

NSVT
2nd degree AV block
PVCS
Sinus arrest

Bradyarrhythmias most common and often occur during sleep with apneic episodes
Significant number of patients with AF have OSA

[11] Especially nocturnal AMI
[12] Numerous congenital maxillofacial, soft tissue, neuromuscular, and inflammatory conditions associated with OSA
[13] Mainstay of treatment in children
[14] Usually mild and may be associated with other pulmonary disorders (eg, COPD); causal effect uncertain
[15] Daytime at rest; associated with poorer outcomes, greater likelihood of Systemic Arterial Hypertension
[16] Often resistant to antihypertensive medications
[17] Most common during sleep
[18] OSA may occur as often as 40–60 % in patients with end-stage renal disease
[19] OSA and Hypothyroidism have similar clinical features and may be causally linked

[20] Transient during apneic episodes; persistent elevation suggests associated COPD

[21] Periodic limb movements of sleep, rotating shift work, narcolepsy, respiratory disease, severe GERD

[22] From ACCF/AHA 2013 Guidelines for Management of Heart Failure (Sec 7.3.1.4. Treatment of Sleep Disorders):

> "The primary treatment for obstructive sleep apnea is nocturnal continuous positive airway pressure. In a major trial, continuous positive airway pressure for obstructive sleep apnea was effective in decreasing the apnea–hypopnea index, improving nocturnal oxygenation, increasing LVEF, lowering norepinephrine levels, and increasing the distance walked in 6 min; these benefits were sustained for up to 2 years. Smaller studies suggest that continuous positive airway pressure can improve cardiac function, sympathetic activity, and HRQOL in patients with HF and obstructive sleep apnea."

[23] OSA predisposition to AF multifactorial, including:

Atrial Stretch
Autonomic imbalance
Inflammation
Oxidative stress

[24] From ACCF/AHA 2013 Guidelines for Management of Heart Failure:

> "Sec 7.3.1.4. Treatment of Sleep Disorders Sleep disorders are common in patients with HF. A study of adults with chronic HF treated with evidence-based therapies found that 61 % had either central or obstructive sleep apnea. Despite having less sleep time and sleep efficiency compared with those without HF, patients with HF, including those with documented sleep disorders, rarely report excessive daytime sleepiness. Thus, a high degree of suspicion for sleep disorders should be maintained for these patients. The decision to refer a patient to a sleep study should be based on clinical judgment."

Guidelines

Diagnosis of Obstructive Sleep Apnea in Adults: A Clinical Practice Guideline From the American College of Physicians
Ann Intern Med. 2014;161:210–20. http://annals.org/article.aspx?articleid=1892620.

2013 ESH/ESC Guidelines for the management of arterial hypertension
Eur Heart J. 34;2199. http://eurheartj.oxfordjournals.org/content/34/28/2159.full.pdf.

Position paper on the management of patients with obstructive sleep apnea and hypertension: Joint recommendations by the European Society of Hypertension, by the European Respiratory Society and by the members of European COST
J Hypertension. 2012;30:633–46. http://journals.lww.com/jhypertension/Abstract/2012/04000/Position-paper-on-the-management-of-patients-with.1.aspx.

Patient Information

AHA

http://circ.ahajournals.org/content/132/6/e114.full.

ASSA

http://www.sleepapnea.org/learn/sleep-apnea/obstructive-sleep-apnea.html.

Boston Childrens: OSA in Children

http://www.childrenshospital.org/conditions-and-treatments/conditions/obstructive-sleep-apnea-osa.

Images

http://www.nlm.nih.gov/medlineplus/ency/imagepages/9701.htm.

Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/obstructive-sleep-apnea/basics/definition/con-20027941.

Medlineplus

ENGLISH

http://www.nlm.nih.gov/medlineplus/ency/article/000811.htm.

ESPANOL

http://www.nlm.nih.gov/medlineplus/spanish/ency/article/000811.htm.

Merck

http://www.merckmanuals.com/home/lung-and-airway-disorders/sleep-apnea/sleep-apnea.

National Sleep Foundation

https://sleepfoundation.org/sleep-disorders-problems/sleep-apnea.

University of Maryland

http://umm.edu/health/medical/reports/articles/obstructive-sleep-apnea.

Professional Information

AHA Scientific Statement

http://circ.ahajournals.org/content/118/10/1080.full.pdf+html.

Review

Eur Heart J. 2013;34:809–15. http://eurheartj.oxfordjournals.org/content/34/11/809.

Acute Myocardial Infarction

Chest. 2009;135:1488–95. http://journal.publications.chestnet.org/article.aspx?articleid=1089845.

Acute Myocardial Infarction: Nocturnal AMI

J Am Coll Cardiol. 2008;52:343–6. http://content.onlinejacc.org/article.aspx?articleid=1139082&resultClick=3.

Atrial Fibrillation/Obesity

J Am Coll Cardiol. 2007;49:565–71. http://content.onlinejacc.org/article.aspx?articleid=1188673&resultClick=3.

Atrial Fibrillation/Stroke

Am J Cardiol. 2015;115:461–5. http://www.sciencedirect.com/science/article/pii/S0002914914021687.

Asthma

JAMA. 2015;313:156–64. http://jama.jamanetwork.com/article.aspx?articleid=2089354.

Children

Anesthesia Cl. 2014;32:237–61. http://www.sciencedirect.com/science/article/pii/S1932227513000827.

CPAP

Sleep Med Clin. 2010;5:383–92. http://www.sciencedirect.com/science/article/pii/S1556407X10000585.

CPAP: HTN/CAD: Long-Term Effects

Am J Hypertens. 2015;28:300–6. http://ajh.oxfordjournals.org/content/28/3/300.full?etoc.

CPAP: Prevention of a FIB Recurrence

JACCCEP. 2015;1:41–51. http://electrophysiology.onlinejacc.org/article.aspx?articleID=2277227.

CPAP: Atrial Fibrillation Recurrence Post-ablation

Heart Rhythm. 2013;10:331–7. http://www.ncbi.nlm.nih.gov/pubmed/23178687.

CPAP: Atrial Fibrillation Recurrence Post-ablation

J Am Coll Cardiol. 2013;62:300–5. http://content.onlinejacc.org/article.aspx?articleID=1685125.

Depression

Sleep Med Rev. 2009;13:437–44. http://www.sciencedirect.com/science/article/pii/S1087079209000392.

Endothelial Dysfunction

Heart. 2013;99:30–4. http://heart.bmj.com/content/99/1/30.abstract.

Heart Failure

J Am Coll Cardiol. 2011;57:119–27. http://www.acc.org/latest-in-cardiology/journal-scans/2011/01/03/15/21/obstructive-sleep-apnea-in-heart-failure?w-nav=S.

Oropharynx Abnormalities

Am J Resp Crit Care Med. 2000;162:740–8. http://www.atsjournals.org/doi/full/10.1164/ajrccm.162.2.9908123#.VMauGWfwuwV.

Post-traumatic Stress Disorder

Chest. 2016;149:483–90. http://journal.publications.chestnet.org/article.aspx?articleID=2430456.

Pulmonary Arterial Hypertension

Am J Cardiol. 2009;104:1300–6. http://www.sciencedirect.com/science/article/pii/S0002914909012806#.

Sudden Cardiac Death

J Am Coll Cardiol. 2013;62:610–6. http://content.onlinejacc.org/article.aspx?articleid=1699335&resultClick=3.

Systemic Arterial Hypertension

Curr Hypertens Rep. 2007;9:529–34. http://www.ncbi.nlm.nih.gov/pubmed/18367017.

Underdiagnosis

Heart. 2015;101:1288–92. http://heart.bmj.com/content/101/16/1288?etoc.

Updates and More

https://clinicalguidecvd.com/osa

Chapter 72
Orthostatic Hypotension

ICD-10 Code

I95.1

Alternate Names/Abbreviation

BRADBURY-EGGLESTON SYNDROME
NEUROGENIC HYPOTENSION
POSTURAL HYPOTENSION
PRIMARY AUTONOMIC INSUFFICIENCY

Description/Etiology

Defined by American Autonomic Society and American Academy of Neurology as a reduction in systolic BP of ≥20 mmHg or diastolic BP of ≥10 mmHg within 3 min of undergoing orthostatic stress.

Can cause transient cerebral hypoperfusion after posture change, with symptoms such as dizziness, weakness, blurred vision, or syncope.

Common condition with prevalence of up to 30 % in older home-dwelling persons

Possible risk factor for CVD [17]

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_72,

Causes (see PREDISPOSING/COMORBID CONDITIONS):

Neurogenic [1]
Non-neurogenic [1]

Predisposing/Comorbid Conditions

ACUTE MYOCARDIAL INFARCTION
ADRENAL INSUFFICIENCY
ALCOHOL USE/EXCESS [10]
AMYLOIDOSIS
ANEMIA
AORTIC STENOSIS [9]
ATRIAL FIBRILLATION
ATRIOVENTRICULAR HEART BLOCK [9]
AUTOIMMUNE/CONNECTIVE TISSUE DISEASE [2]
B12 DEFICIENCY [10]
BLOOD LOSS [7]
BOTULISM
CARCINOID SYNDROME/TUMOR
CARDIOMYOPATHY – RESTRICTIVE [9]
CEREBROVASCULAR DISEASE
CONSTRICTIVE PERICARDITIS [9]
CORONARY ARTERY DISEASE
DEHYDRATION [7]
DIABETES INSIPIDUS
DIABETES MELLITUS [2]
DIARRHEA [7]
DIURETICS [7]
DOPAMINE BETA-HYDROLASE DEFICIENCY
DRUGS [8]
FAMILIAL DYSAUTONOMIA [2]
FRAILTY
GUILLAIN BARRE SYNDROME [5]
HEART FAILURE
HIV [10]
HYPERGLYCEMIA – ACUTE

HYPERTENSION – SYSTEMIC ARTERIAL
HYPOALDOSTERONISM
HYPOKALEMIA
HYPOTHYROIDISM
LEWY BODY DEMENTIA
LUMBAR SYMPATHECTOMY
MULTIPLE MYELOMA
MULTIPLE SCLEROSIS
MULTIPLE SYSTEM ATROPHY (MSA) [19]
NEUROTOXINS [10]
OLIVOPONTOCEREBELLAR ATROPHY
PARANEOPLASTIC SYNDROME [3]
PARKINSON DISEASE [2]
PHEOCHROMOCYTOMA
PHYSICAL DECONDITIONING
PORPHYRIA CUTANEA TARDA [10]
PREGNANCY
PROLONGED BED REST
PULMONARY HYPERTENSION
PURE AUTONOMIC FAILURE
RENAL FAILURE (UREMIA)
SHY-DRAGER SYNDROME
SINUS NODE DYSFUNCTION
SPINAL CORD DIS/TRANSECTION [10]
STROKE
SUBACUTE COMBINED SCLEROSIS [10]
SYMPATHECTOMY
SYRINGOMYELIA [10]
SYSTEMIC MASTOCYTOSIS
TACHYARRHYTHMIAS
TOXIC AUTONOMIC NEUROPATHY [HEAVY METALS, DRUGS]
VALVULAR HEART DISEASE
VENOUS INSUFFICIENCY
VOMITING [7]
WERNICKE KORSAKOFF SYNDROME

Demography

Increased prevalence in:

Advanced age
Males
Persons in institutions

Pathophysiology

When body assumes upright posture, gravity causes downward displacement of 500–1000 mL of blood to lower limbs and abdomen, resulting in decreased venous return to heart and about 20 % decrease in cardiac output. Medulla control centers act to compensate for BP drop by increasing sympathetic and reducing parasympathetic nervous system output, causing reflex tachycardia and increased total peripheral resistance. In heathy persons, orthostatic stabilization is achieved within 1 min of standing

Orthostatic hypotension is due to failure of any of the normal compensatory systems for maintaining upright BP.

Signs/Symptoms [20]

ARTERIAL PRESSURE – HIGHLY VARIABLE
ARTERIAL PRESSURE, POSTPRANDIAL – DECR (POSTPRANDIAL HYPOTENSION) [21]
ARTERIAL PRESSURE, PRONE – INCR [6]
ARTERIAL PRESSURE, UPRIGHT – DECR (ORTHOSTATIC HYPOTENSION) [1]
BOWEL MOVEMENTS – CONSTIPATION [MAY BE SEVERE]
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)
DIZZY/LIGHTHEADED/PRESYNCOPE
EYES, VISION – BLURRED
EYES, VISION, UPRIGHT – ALTERED, NS

FATIGUE
GENITALS, PENIS, ERECTION – DECR (ERECTILE DYSF)
HEART, RATE, RESPONSE – DECR (CHRONOTROPIC INCOMPETENCE)
NAUSEA
SWEATING – DECR (ANHIDROSIS)
SWEATING, FACE – INCR [4]
URINATION – NOCTURNAL (NOCTURIA) [22]
URINATION – RETENTION

Differentiation

Other causes of presyncope/syncope

Complications

DVT – Lower Extremity
Trauma secondary to fall

Laboratory

ASSESS FOR COMORBIDS, EG, PHEOCHROMOCYTOMA, DIABETES

ECG

N/NS ABN
NS/VAR PER COMORBIDITY(S)

Imaging

NS/VAR WITH COMORBID

Other Tests

Autonomic function tests [6]
Supine/standing plasma norepinephrine
Thermoregulatory sweat test
24-h urinary sodium

Treatment: Nonpharmacologic [16]

Do:

- Change positions slowly/carefully
- Dorsiflex feet several times before standing
- Eat small/frequent meals
- Elevate head of bed 5–20°/6-9 in.
- Increase salt/fluid intake (unless contraindicated, eg, HF)
- Maintain hydration
- Perform activities in afternoons, especially regular exercise, such as:
  - Bicycling
  - Rowing
  - Swimming
- Wear abdominal binder/compression waist-high stockings

Avoid:

- Alcohol
- Coughing spells
- Dehydration
- Hot baths/environment
- Hyperventilation
- Large meals
- Rapid ascent to high altitude
- Standing:
  - Motionless
  - Rapidly
  - With legs crossed

Straining with urination or defecation
Working with arms above shoulders

Treatment: Pharmacologic [11] [16]

DC causative/contributory agents when possible (eg, apha-blockers, diuretics)
Adrenergic agonists prn [13]
Droxidopa [18]
Fludrocortisone (to increase intravascular volume) [12]
Splanchnic vasoconstrictor [14]
Treatment combinations [15]

Treatment: Surgical/Invasive

NA

Prevention

See above NONPHARMACOLOGICAL TREATMENT

Course

Usually benign but may be risk factor for CVD [17]
Frequently transient in adolescents

Notes

[1] Neurogenic causes include:

AIDS
Alcoholic polyneuropathy
Guillain-Barré syndrome
Multiple Sclerosis

Multiple system atrophy
Pure autonomic failure
Tabes Dorsalis

Non-neurogenic causes include:

Adrenal insufficiency,
AMI
Diabetes Insipidus
Dysrhythmias
HF
Intravascular volume depletion
Myocarditis
Pericarditis
Postprandial
Prolonged sitting/standing (venous pooling)
Sepsis/other acute infectious process
Venous obstruction

[2] Autonomic Neuropathy, also termed Neurogenic Orthostatic Hypotension
[3] Small Cell Lung Carcinoma, Monoclonal Gammopathies, Light Chain Disease, Amyloidosis
[4] May be compensatory for general anhidrosis
[5] Especially post-infection, eg Influenza, Zika virus
[6] Refer patients to specialized autonomic dysfunction center
[7] Due to volume loss
[8] Including:

Alpha-blockers
Antianginal agents
Antiarrhythmics
Anticholinergics
Antihyperensives
Diuretics
Dopamine agonists
Narcotics
Neuroleptics
Sedatives

Tricyclic antidepressants
Vasodilators
Venodilators

[9] Due to decreased cardiac output
[10] Peripheral neuropathy
[11] Derived from Table 1, ASH position statement (GUIDELINES)
[12] Fludrocortisone 0.1-0.3 mg/D
[13] Midodrine 2.5-10 mg; pyridostigmine 60 mg; pseudoephedrine 30 mg; atomoxetine 18 mg
[14] Octreotide 12.5-25 mg (subcutaneous)
[15] Fludrocortisone 0.1-0.3 mg each AM and midodrine 5–10 mg; midodrine 5–10 mg or pseudoephedrine 30 mg with 16 oz water bolus
[16] Treatment should be undertaken only after search for underlying cause and its correction (eg, hypovolemia, anemia)
[17] Including CAD, Stroke, HF, all-cause death
[18] Droxydopa specific indications include orthostatic symptoms due to:

Dopamine beta-hydroxylase deficiency
Multiple System Atrophy
Non-diabetic Autonomic Neuropathy
Parkinson Disease
Pure Autonomic Failure

[19] Progressive neurodegenerative disease manifest by clinical features related to autonomic nervous system and movement
[20] Many patients are asymptomatic; when symptoms occur, more likely to be in AM after arising and exacerbated (in part due to venous pooling) by:

Alcohol ingestion
Dehydration
Heat
Immobilization
Post-exercise
Urination

[21] Especially with large meals/carbohydrate-rich food, caused by:

Gastric distention
Release of vasodicvlatory peptides
Splanchnic blood pooling

[22] Due to peripheral blood redistribution to central areas during recumbency/forced natriuresis with concomitant supine hypertension; this nocturnal intravascular volume loss further contributes to AM hypotension

Guidelines

ASH Position Paper
J Clin Hypertension. 2013;15:147–53. http://onlinelibrary.wiley.com/doi/10.1111/jch.12062/abstract.
EFNS guidelines on the diagnosis and management of orthostatic hypotension
Eur J Neurol. 2006;13:930–6. http://onlinelibrary.wiley.com/doi/10.1111/j.1468-1331.2006.01512.x/full.

Patient Information

Medlineplus

http://www.nlm.nih.gov/medlineplus/ency/article/007278.htm.

Genetics Home Reference: Familial Dysautonomia

http://ghr.nlm.nih.gov/condition/familial-dysautonomia.

Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/orthostatic-hypotension/basics/definition/con-20031255.

Espanol

http://www.nlm.nih.gov/medlineplus/spanish/ency/article/007278.htm.

Professional Information

Review

J Am Coll Cardiol. 2015;66:848–60. http://content.onlinejacc.org/article.aspx?articleID=2423750.

Review

J Clin Neurol. 2015;11:220–6. http://thejcn.com/DOIx.php?id=10.3988/jcn.2015.11.3.220#B9.

Review

N Engl J Med. 2008;358:615–24. http://www.nejm.org/doi/full/10.1056/NEJMcp074189.

Review

Am Fam Physician. 2011;84:527–36. http://www.aafp.org/afp/2011/0901/p527.html#afp20110901p527-b10.

Review

Clin Auton Res. 2008;18(Suppl 1):8–13. http://www.ncbi.nlm.nih.gov/pubmed/18368301.

Age-Related Changes

Circulation. 2014;130:1780–9. http://circ.ahajournals.org/content/130/20/1780.full.

Deep Vein Thrombosis

Am J Hypertens. 2016;29:634–40. http://ajh.oxfordjournals.org/content/29/5/634.abstract?etoc.

Droxidopa: Parkinsons

Neurology. 2014;83:328–35. http://www.neurology.org/content/83/4/328.long.

Heart Failure Risk Factor

J Gerontol A Biol Sci Med Sci. 2014;69:223–30. http://www.ncbi.nlm.nih.gov/pubmed/23846416.

Mortality Risk

Heart. 2014;100:406–13. http://heart.bmj.com/content/100/5/406.abstract.

Neurogenic Orthostatic Hypotension

N Engl J Med. 2008;358:615–24. http://www.nejm.org/doi/full/10.1056/NEJMcp074189.

Parkinson Disease

Vasc Health Risk Manag. 2014;10:169–76. http://www.ncbi.nlm.nih.gov/pubmed/24729712.

Prognosis in Older Adults

Arch Intern Med. 1999;159:273–80. http://www.ncbi.nlm.nih.gov/pubmed/9989539.

Treatment: Neurogenic Orthostatic Hypotension

Lancet Neurol. 2008;7:451–8. http://www.sciencedirect.com/science/article/pii/S1474442208700887.

Updates and More

https://clinicalguidecvd.com/orthohyp

Chapter 73
Papillary Fibroelastoma

Management Keys

- Papillary adenomas should be surgically resected when possible as they are associated with increased risk of stroke and mortality
- Surgical excision has high likelihood of valve preservation and low recurrence rate when performed at a high-volume tertiary care center
- Surgical excision associated with high frequency of valve preservation when performed in high-volume centers
- Aggressive surgical approach requires significant experience/expertise
- Anticoagulant/antiplatelet therapy should be considered in patients not undergoing surgical resection

ICD-10 Code

D15.1

Alternate Names/Abbreviation

PFE
PAPILLOMA
PAPILLARY FIBROMA

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 977
DOI 10.1007/978-1-4471-7293-2_73,

Description/Etiology

Pedunculated, avascular tumor
Most common benign cardiac neoplasm of adulthood
>90 % arising from cardiac valves (AV > MV > TV > PV) or cardiac wall (LV most often) [7]
Clinical manifestations depend on location; often detected only at autopsy or incidentally during echo, cardiac catheterization, cardiac surgery
Etiology unknown but some cases appear related to prior cardiac surgery or radiation

Predisposing/Comorbid Conditions

ATRIAL FIBRILLATION
CARDIAC SURGERY
RADIATION

Demography

All ages, increasing with age and most often >50 yrs

Pathophysiology

Avascular tumor with fibroelastic tissue surrounded by endocardium
Non-embolic physiologic effects, if any, vary according to location, usually due to MV or AV flow obstruction

Signs/Symptoms [1]

BREATHING – DIFF (DYSPNEA)
CHEST – PAIN, EFFORT (ANGINA PECTORIS)
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)
FEVER [2]
HEART – MURMUR, NS

HEART, LSB, MID – MURMUR, SYS [4]
HEART, LV, APEX – MURMUR, DIAS [5]

Differentiation

Antiphospholipd antibodies
AS
CAD
Infective Endocarditis
MV Stenosis
Systemic Lupus Erythematosus
Other causes of acute CNS ischemia
Other types of cardiac tumors

Complications

Acute Pulmonary Embolism
AMI
Heart block
SCD
Systemic embolus:

- Ischemic Stroke [3]
- Mesenteric ischemia/infarction
- Renal infarction
- Retinal artery [3]
- TIA [3]

Laboratory

NS

ECG

AV COND – 3RD DEGREE BLOCK
DYSRHYTHMIAS – ATRIAL (PACS/OTHERS) [6]

Imaging [9] [10] [11]

AV – MASS
LA, INTRACAVITY – MASS
LV, INTRACAVITY – MASS
MV – MASS
PV – MASS
TV – MASS

Other Tests

Cardiac catheterization

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic

Antiplatelet agents

Treatment: Surgical/Invasive

Surgical resection [8]

Management of smaller, nonmobile masses not well defined

Course

Post-surgical recurrence: 1.6 % for echo-detected recurrence by up to 6 years post-resection

Notes

[1] Often asymptomatic or only with features due to complications, especially TIA/stroke as initial manifestation
[2] Rare
[3] Cerebral embolus; >50 % initially present with acute neurological features
[4] AV location
[5] MV location
[6] Especially AF
[7] Tumor resembles Sea Anemone
[8] Highly effective in large tertiary centers with valve preservation and low recurrence rate
[9] Seen on echo; 90 % on AV or MV; valve dysfunction rare; appears as round/oval/irregular well-demarcated mass
[10] Sonographer familiarity with echo features of Papillary Fibroadenoma very important
[11] As many as 25 % of lesions can be detected by TEE but less often with TTE

Guidelines

NS

Professional Information

Acute Myocardial Infarction: Case Report

Eur Heart J. 2014;35:1970. http://eurheartj.oxfordjournals.org/content/35/29/1970.

Angina Pectoris: Case Report

Circulation. 2014;129:1714. http://circ.ahajournals.org/content/129/16/1714.full.

Angina Pectoris/Syncope in Teen: Case Report

Circulation. 2014;130:520–2. http://circ.ahajournals.org/content/130/6/520.full.

Aortic Valve

Circulation. 2007;115:e3–e6. http://circ.ahajournals.org/content/115/1/e3.full?sid=4463ee99-fe80-48ad-9369-7e0bdae36047.

Clinical and Echo Features

Circulation. 2001;103:2687–93. http://circ.ahajournals.org/content/103/22/2687.full?sid=4463ee99-fe80-48ad-9369-7e0bdae36047.

Images: Ventricular Papilary Fibroelastoma

J Am Coll Cardiol. 2014;63:2170. http://content.onlinejacc.org/article.aspx?articleID=1859525.

Imaging (Case Report)

Am J Med. 2013;126:964–5. http://www.sciencedirect.com/science/article/pii/S0002934313006220.

Left Atrial Free Wall

Circulation. 2001;104:e87–e88. http://circ.ahajournals.org/content/104/17/e87.full?sid=4463ee99-fe80-48ad-9369-7e0bdae36047.

Left Ventricle

Tex Heart Inst J. 2006;33:63–5. http://www.ncbi.nlm.nih.gov/pmc/articles/PMC1413617/.

Mitral Valve

Circulation. 1998;98:1251–2. http://circ.ahajournals.org/content/98/12/1251.full?sid=4463ee99-fe80-48ad-9369-7e0bdae36047.

Multiple Sites

Circulation. 2012;126:242–3. http://circ.ahajournals.org/content/126/2/242.full?sid=49f03f95-f2a4-4e6b-a2d8-03bd1a2c74b9.

Prognosis/Bioepidemiology

J Am Coll Cardiol. 2015;65:2420–9. http://content.onlinejacc.org/article.aspx?articleID=2300733.

Surgical Resection

Tex Heart Inst J. 2016;43:148–51. http://thij.org/doi/full/10.14503/THIJ-14-4889.

Tricuspid Valve

Circulation. 2008;117:e190–e191. http://circ.ahajournals.org/content/117/11/e190.full?sid=4463ee99-fe80-48ad-9369-7e0bdae36047.

Updates and More

https://clinicalguidecvd.com/pfe

Chapter 74
Patent Ductus Arteriosus (PDA)

Management Keys (Adults)

- Routine follow-up recommended for patients with small PDA without evidence of left-sided heart volume overload; recommended every 3–5 years for patients with small PDA without evidence of left heart volume overload
- Consultation with adult congenital heart disease interventional cardiologists recommended before surgical closure selected as method of repair for patients with calcified PDA
- Surgical repair by a surgeon experienced in CHD surgery is recommended when:
  - PDA is too large for device closure
  - Distorted ductal anatomy precludes device closure

ICD-10 Code

Q25.0

Alternate Names/Abbreviation

PDA

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_74,

Description/Etiology

Persistent congenital communication between aorta and PA occurring alone or associated with other congenital lesions, most often ASDs and VSDs

PDA associated with several genetic syndromes, including:

Chromosomal aberrations [12]
Single-gene mutations such as Holt-Oram syndrome
X-linked mutations

Small PDAs: usually asymptomatic and detected in adults during evaluation of heart murmur found during routine cardiac examination or by echo, CT, or chest MRI for unrelated condition

Moderate PDAs: tolerated well in childhood and patients often remain completely asymptomatic in early adulthood but usually develop exercise intolerance/symptoms of LV failure beginning in third decade

No single gene defect specific for PDA identified

Predisposing/Comorbid Conditions

ANEURYSMS-OSTEOARTHRITIS SYNDROME
ANOMALOUS CORONARY ARTERY
ATRIAL SEPTAL DEFECT - SECUNDUM
BICUSPID AORTIC VALVE
CANTU SYNDROME
CHAR SYNDROME [11]
MATERNAL RUBELLA [6]
MATERNAL VALPROIC ACID EXPOSURE [18]
NOONAN SYNDROME
OTHER FORMS OF CONGENITAL HEART DISEASE
TREACHER COLLINS SYNDROME [19]
VENTRICULAR SEPTAL DEFECT

Demography

Females 2–3:1
High altitude persistence: 6x incidence than at sea level [20]
Family history

Pathophysiology

Ductus arteriosus: vascular channel connecting junction of main/left PA with descending aorta immediately distal to left subclavian artery origin during fetal life; normal anatomical closure occurs within 2–3 weeks after birth
Failure of duct closure termed Persistent Ductus Arteriosus after 3 months in term infants and by 1 year in premature infants
L-R shunt varies in magnitude according to size of ductus, PVR, LV function
R-L shunt occurs with onset of PAH

Signs/Symptoms [21]

ARTERIAL PULSE - DOUBLE (BISFERIENS)
ARTERIAL PULSE PRESSURE – INCR [15]
ARTERIAL PULSE, FALL – RAPID
ARTERIAL PULSE, RISE – RAPID
BREATHING – DIFF (DYSPNEA)
CHEST – PAIN
CHEST, ANT, L – BULGE
EXTREM, LOWER, DIGITS – CLUBBING
EXTREM, LOWER, DIGITS, COLOR – BLUE (CYANOSIS)
EXTREM, UPPER, L, DIGITS – CLUBBING
EXTREM, UPPER, L, DIGITS, COLOR – BLUE (CYANOSIS) FATIGUE
HEART, LSB, UPPER – MURMUR, CONT [1]
HEART, LSB, UPPER – THRILL, CONT [13]

HEART, LV, APEX – MURMUR, DIAS [14]
HEART, LV, APEX, IMP – FORCEFUL/SUSTAINED [15]
HEART, LV, APEX, IMP – TRIPLE
HEART, P2, INTENSITY – INCR
HEART, S2, SPLIT – REVERSED (PARADOXICAL)
HEART, S3 LV [14]
STERNUM, CURV – ANT (PECTUS CARINATUM)

Differentiation

Aortopulmonary collateral
Coronary Arteriovenous Fistula
Other causes of PAH
Other causes of wide pulse pressure [2]
Ruptured sinus of Valsalva
VSD with AR

Complications

Ductus aneurysm [9]
Ductus rupture
HF
Infective endarteritis/endocarditis
PAH
Pulmonary artery aneurysm
Recurrent pulmonary infections

Laboratory

NS

ECG [3] [7]

DYSRHYTHMIAS – ATRIAL (PACS/OTHERS)
AV COND – 1ST DEGREE BLOCK

P WAVE – FLAT
P WAVE – TALL/PEAKED [4]
P WAVE, DUR – INCR
QRS – BVH PATTERN
QRS – LVH PATTERN
QRS – RVH PATTERN [4]

Imaging [7] [10]

CARDIOMEGALY [17]
DUCTUS ARTERIOSUS – CALCIUM [5]
LA, CHAMBER, SIZE – INCR [16]
LV, CHAMBER, SIZE – INCR [16]
PA, MAIN, SIZE – INCR
PA, PRESS – INCR
PUL, VASCULARITY – INCR [17]

Genomics

NS

Other Tests

Cardiac catheterization [22]

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic

COX inhibitors (newborns) [23]

Ibuprofen
Indomethacin

Treatment: Surgical/Invasive

Percutaneous closure [8]
Surgical closure [8]

Prevention

Prematernal Rubella immunization

Course

Varies with magnitude of shunt, intervention, comorbidities, presence of PH

Notes

[1] Left infraclavicular area; peak intensity around S2; radiates to LSB/left interscapular area; if PAH present, murmur may be systolic only; murmur may be absent in Eisenmenger because of minimal shunting
[2] Eg, Hyperthyroidism, AR
[3] May be normal; LVH/LA enlargement indicate moderate or large PDA; RVH/RA enlargement indicate advanced stage of Pulmonary Hypertension
[4] In presence of PAH
[5] Calcification associated with increased risk of rupture during surgical repair
[6] Also cataracts, deafness, mental retardation, low birth weight, failure to thrive
[7] May be normal with small shunts
[8] Adult intervention for PDA: ACC/AHA 2008 Guidelines for Adults With CHD (J Am Coll Cardiol 2008;52:e185)

"Surgical closure of PDA in the adult may pose some problems due to the friability and/or calcification of the ductus, atherosclerosis, and aneurysm formation, as well as the presence of other unrelated comorbid conditions, such as coronary atherosclerosis or renal disease, that may adversely affect the perioperative risk. Adults with PDA are better suited for percutaneous closure with either the occlusion device or coils because of its high success and few complications. If the PDA is associated with other conditions that require surgical correction, the ductus may be closed during the same operation, although percutaneous closure of the PDA before other cardiac surgery may decrease the risk of cardiopulmonary bypass."

[9] Usually L-R shunt

[10] Also visualize ductus by echo, which is diagnostic

[11] Char Syndrome: autosomal disorder comprising:

PDA
Facial Dysmorphism
Hand anomalies

[12] Esp Trisomy 21

[13] Thrill indicates moderate/large shunt

[14] Diastolic rumbling murmur/S3 due to increased LA-LV blood flow consistent with moderate/large shunt

[15] Indicates moderate/large shunt

[16] LA/LV dilatation (LA:aorta ratio >1.3) on echo indicates moderate/large shunt

[17] CXR heart size/pulmonary vascularity may be normal with small shunt; if shunt volume is moderate to large, CXR shows increased pulmonary vascular markings with prominent ascending aorta and cardiomegaly with prominent LA and LV; peripheral pruning of vascular markings with large PA shadow and right PA indicates severe PH; calcified ductus may appear in some older adults, especially in lateral CXR view

[18] Fetal Valproate Syndrome:

Valproic acid: anticonvulsant drug
Rare congenital disorder due to fetal exposure to valproic acid (Dalpro, Depakene, Depakote, Depakote sprinkle, Divalproex, Epival, Myproic acid) during first 3 months of pregnancy
Clinical features include, in addition to rare occurrences of PDA:

Spina bifida
Distinctive facial features
Musculoskeletal abnormalities

[19] Treacher Collins Syndrome:

Caused by mutations in TCOF1, POLR1C, POLR1D
Associated with multiple defects, including (in addition to rare PDA):

Cleft palate
Hearing loss/ear deformities
Ocular/visual abnormalities
Normal mental development

[20] Due to exposure to low pO_2
[21] Clinical manifestations primarily determined by volume of extra blood recirculating in PA, capillaries, pulmonary veins, LA, LV, ascending aorta, and defect duration
[22] Cardiac catheterization for diagnosis not indicated for uncomplicated PDA with adequate noninvasive imaging; in adults with PDA cardiac catheterization is performed at time of planned percutaneous closure in patients meeting criteria after initial evaluation by TTE
[23] Cox inhibitor treatment for PDA closure:

Newborns: IV indomethacin and IV ibuprofen lysine are equally effective in closure of PDA, with closure rates of 75–93 %; IV ibuprofen lysine may be associated with decreased incidence of adverse events, especially renal toxicity; IV ibuprofen lysine may have less significant impact on cerebral blood flow and mesenteric blood flow

Adults: adult ductal tissue has no response to COX inhibitors like indomethacin; thus, primary modes of PDA closure for PDA in adults are percutaneous and surgical

Guidelines

ACC/AHA 2008 guidelines for the management of adults with congenital heart disease

J Am Coll Cardiol. 2008;52:e143–263. http://content.onlinejacc.org/article.aspx?articleid=1188032#tab1

ESC guidelines for the management of grown-up congenital heart disease (new version 2010)

Eur Heart J. 2010;31:2915–57. http://www.escardio.org/guidelines-surveys/esc-guidelines/Pages/grown-up-congenital-heart-disease.aspx.

Patient Information

Images

https://www.nlm.nih.gov/medlineplus/ency/imagepages/1056.htm.

https://www.nlm.nih.gov/medlineplus/ency/presentations/100012-1.htm.

Medlineplus

ENGLISH

http://www.nlm.nih.gov/medlineplus/ency/article/001560.htm.

ESPANOL

https://www.nlm.nih.gov/medlineplus/spanish/ency/article/001560.htm.

Cleveland Clinic

http://my.clevelandclinic.org/heart/disorders/patent-ductus-arteriousis-adults.aspx.

Genetics Home Reference

http://ghr.nlm.nih.gov/search?query=patent+ductus+arteriosus

Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/patent-ductus-arteriosus/basics/definition/con-20028530.

Texas Heart Institute

http://www.texasheart.org/HIC/Topics/Cond/pda.cfm.

Professional Information

Early Descriptions

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Review

Circulation. 2006;114:1873–82. http://circ.ahajournals.org/content/114/17/1873.long.

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Review

Seminars in Perinatology. 2012;36:146–53. http://www.sciencedirect.com/science/article/pii/S0146000511001716.

Review: Ligation in Premature Infants

Journal of Surgical Research. 2014;190:613–22. http://www.sciencedirect.com/science/article/pii/S0022480414001164..

Review: Transcatheter Closure

Archives of Cardiovascular Diseases. 2014;107:122–32. http://www.sciencedirect.com/science/article/pii/S1875213614000242.

Anomalous Coronary Artery

Res Cardiovasc Med. 2013;2:190–92. http://www.ncbi.nlm.nih.gov/pubmed/25478523.

Char Syndrome

Circulation. 1999;99:3036–42. http://circ.ahajournals.org/content/99/23/3036.full?sid=ab458ca5-62bd-4ff1-be87-5392c85b4fe5.

Eisenmenger (Case Report)

Heart, Lung and Circulation. 2013;22:968–69. http://www.sciencedirect.com/science/article/pii/S1443950613001716.

Indomethacin

J Cardiovasc Thorac Res. 2014;6:257–59. http://www.ncbi.nlm.nih.gov/pubmed/25610559.

Infective Endarteritis

Am J Cardiol. 2004;93:513–15. http://www.sciencedirect.com/science/article/pii/S0002914903016217

Post-ligation Coronary Perfusion/LV Function

J Thor Cardiovasc Surg. 2012;143:1271–78. http://www.sciencedirect.com/science/article/pii/S0022522311011548.

Pulmonary Artery Aneurysm

Curr Cardiol Rev. 2015;11:163–66. http://www.ncbi.nlm.nih.gov/pubmed/25545802.

Transcatheter Occlusion: Followup

Circulation. 1991;84:2313–17. http://circ.ahajournals.org/content/84/6/2313.abstract?sid=ce18dc36-05ee-442a-b607-2428b8c7a019.

Transcatheter Occlusion: Preterm Infants

JACC: Cardiovascular Interventions. 2010;3:550–55. http://www.sciencedirect.com/science/article/pii/S1936879810001743.

Transcatheter Occlusion with Reversible PAH

Heart. 2007;93:514–18. http://heart.bmj.com/content/93/4/514.abstract.

Treacher Collins Syndrome

Int J Pediatr Otorhinolaryngol. 2014; 78:893–98. http://www.ncbi.nlm.nih.gov/pubmed/24690222.

Updates and More

https://clinicalguidecvd.com/pda

Chapter 75
Pericarditis: Acute

Management Keys [26]

- Treat with aspirin or NSAIDs as first-line therapy, with gastro-protection
- Treat with colchicine as first-line therapy as adjunct to aspirin/NSAID therapy
- Use serum CRP as guide to treatment length and for assessing response to therapy
- Corticosteroids are not recommended as first-line therapy for acute pericarditis
- Consider treatment with low-dose corticosteroids:
  - Contraindication/failure of aspirin/NSAIDs and colchicine
  - When infectious cause has been excluded
  - When there is a specific indication such as autoimmune disease
- Consider exercise restriction for non-athletes until resolution of symptoms and normalization of CRP, ECG, echocardiogram
- For athletes, duration of exercise restriction should be considered until resolution of symptoms and normalization of CRP, ECG and echocardiogram for at least 3 months

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_75,

ICD-10 Code

I30.9

Description/Etiology

Inflammatory pericardial syndrome with or without pericardial effusion
Causes:

- Idiopathic [1]
- Adjacent structure dis
- Cancer
  - Metastatic [16]
  - Primary cardiac
- Connective tissue disease/vasculitis
- Drugs
- Infection
- Metabolic disease
- Post-injury syndrome (especially AMI)
- Radiation
- Trauma [21]
  - Non-penetrating
  - Penetrating

Predisposing/Comorbid Conditions

ANOREXIA NERVOSA
INFLAMMATORY BOWEL DISEASE
ACUTE MYOCARDIAL INFARCTION
ACUTE PANCREATITIS
ACUTE PULMONARY EMBOLISM
AMYLOIDOSIS
ANKYLOSING SPONDYLITIS
AORTIC DISSECTION

BEHCET SYNDROME
CANCER [16]
CARDIAC CONTUSION
CHURG-STRAUSS SYNDROME
DERMATOMYOSITIS
DRUGS [4]
EMPYEMA
FAMILIAL MEDITERRANEAN FEVER
GIANT CELL ARTERITIS
GOUT
HORTON DISEASE
HYPEREOSINOPHILIC SYNDROME
HYPOTHYROIDISM
INFECTION [2] [3]
LEUCOCYTOCLASTIC VASCULITIS
LOEFFLER SYNDROME
PNEUMONIA – COMMUNITY-ACQUIRED
POLYARTERITIS
POLYMYOSITIS
POST-MYOCARDIAL INFARCTION SYNDROME
POST-THORACOTOMY/PERICARDIOTOMY SYNDROME [17]
RADIATION [18]
REITER SYNDROME
RENAL DIALYSIS
RENAL FAILURE (UREMIA)
RHEUMATIC FEVER
RHEUMATOID ARTHRITIS
SARCOIDOSIS
SCLERODERMA
SCURVY
SJOGREN SYNDROME
STEVENS-JOHNSON SYNDROME
SYSTEMIC INFLAMMATORY DISEASE
SYSTEMIC LUPUS ERYTHEMATOSUS
TAKAYUSU DISEASE
TEMPORAL ARTERITIS

THROMBOHEMOLYTIC THROMBOCYTOPENIC PURPURA
TRAUMA
TUMOR NECROSIS FACTOR RECEPTOR-ASSOCATED PERIODOC SYNDROME
WEGENER GRANULOMATOSIS
WHIPPLE DISEASE

Demography

More common in males and adults

Pathophysiology

Acute inflammatory changes of pericardium, often extending into superficial myocardium and pleura

Signs/Symptoms

ABDOMEN – PAIN
BREATHING – DIFF (DYSPNEA)
CHEST – PAIN [5]
CHEST, POST – PAIN, PLEURITIC
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)
COUGH
FATIGUE
FEVER [20]
HEART, LSB, LOWER – FRICTION RUB [7]
HEART, LSB, MID – FRICTION RUB [7]
HICCUPS
MENTATION – WEAKNESS (MALAISE) [20]
MUSCLES – PAIN (MYALGIA) [20]
SWALLOWING – DIFFICULT (DYSPHAGIA)

Differentiation

Acute Pulmonary Embolism
AMI
Aortic Dissection
Costochondritis
Gastroesophageal Reflux Disease
Pneumonia

Complications

Cardiac Tamponade [24]
Dysrhythmias (especially AF)
Pericarditis – Constrictive
Recurrent pericarditis

Laboratory

BLOOD, CKMB – INCR
BLOOD, CRP – INCR
BLOOD, ESR – INCR
BLOOD, TROPONIN – INCR
BLOOD, WBC – INCR (LEUKOCYTOSIS)

ECG

DYSRHYTHMIAS – ATRIAL (PACS/OTHERS) [10]
PR SEGMENT – DEPRESSED [9]
ST SEGMENT – ELEV [11]
T WAVE – INVER, ABN [12]
VOLTAGE, GEN – DECR [8]

Imaging

CARDIOMEGALY [LARGE EFFUSION]
PERICARD – FLUID
PLEURA – FLUID

Other Tests

Pericardiocentesis [14]
Pericardial biopsy

Treatment: Nonpharmacologic

Activity restrictions [27]

Treatment: Pharmacologic

Antibiotics: suspected/proven bacterial etiology
Colchicine [25]
NSAIDS
Corticosteroids [28]:

- Severe cases without adequate response to colchicine/NSAIDS
- Tuberculosis-related etiology

Treatment: Surgical/Invasive

Pericardiocentesis [14]

Prevention

NS

Course

Usual resolution in 2–6 weeks [15]
Indicators of poor prognosis:

- Fever (>38° C)
- Large effusion/tamponade
- Pneumonia
- NSAID/ASA/colchicine failure (1 week)
- Sepsis
- Subacute course
- Trauma

Notes

[1] Majority of cases
[2] Including:

- Bacterial
- Fungal
- Leptospiral
- Mycoplasma
- Parasitic
- Rickettsial
- Tuberculosis
- Viral, especially:
  - Adenovirus
  - Cytomegalovirus
  - Enterovirus
  - Herpes simplex
  - HIV
  - Influenza

[3] In immunocompromised patients: Herpes complex and Cytomegalovirus are especially important
[4] Including:

- Diphenylhydantoin

Doxorubicin
Hydralazine
Isoniazid
Penicillin
Procainamide

[5] May radiate to left or right shoulder, arms, elbows, trapezius area (due to common phrenic nerve innervation), jaw, throat, ear, occipital area, interscapular area
[6] Exacerbated by reclining position/relieved by leaning forward
[7] Often absent; presence/character may vary between exams; best heard at end-expiration/patient leaning forward; monophasic/biphasic/triphasic
[8] When pericardial effusion present
[9] Best seen in ECG leads II, AVR, AVF, V4-6
[10] <10 %: AF/A Flutter most common
[11] Most characteristic ECG feature; due to subepicardial inflammation; diffuse; saddle-shaped/concave upward
[12] After ST segments return to baseline
[13] About 25 %; usually on left
[14] Primarily for drainage or when underlying pathology suspected, eg, cancer/infection
[15] For idiopathic/viral cause
[16] Usually secondary, via local invasion/lymphatic/hematogenous spread; >60 % of cardiac metastases involve pericardium
[17] Reported in up to 20 % of patients post-CABG; mean 4 weeks
[18] Especially breast cancer, mediastinal tumors (eg, Hodgkins)
[19] Differentiate from AMI by:

Absence of Q wave/R wave loss in pericarditis
QRS prolongation/QT shortening in leads with ST elevation in AMI but not pericarditis

[20] Prodromal of fever, myalgia, malaise often precedes onset
[21] Prior cardiac surgery most common
[22] Lung adenocarcinoma most common
[23] Criteria for diagnosis of Acute Pericarditis (from 2015 ESC Guidelines)

1. Chest pain (85–90 % of cases) – typically sharp and pleuritic, improved by sitting up and leaning forward
2. Pericardial friction rub (≤33 % of cases) superficial scratchy or squeaking sound best heard with diaphragm of stethoscope over LSB
3. ECG changes (up to 60 % of cases) – new widespread ST elevation or PR depression in acute phase
4. Pericardial effusion (up to 60 % of cases, generally mild)

Additional signs and symptoms may be present according to underlying etiology or systemic disease (i.e., signs and symptoms of systemic infection such as fever and leucocytosis, or systemic inflammatory disease or cancer)

[24] Acute tamponade rare in acute idiopathic pericarditis; more common with specific underlying causes such as cancer, tuberculosis, purulent pericarditis

[25] Colchicine: 15–30 % of patients not treated with colchicine develop recurrent or incessant disease, while colchicine decreases recurrence rate by 50 %

[26] Derived from 2015 ESC Guidelines

[27] Activity restrictions:

Consider exercise restriction for non-athletes until resolution of symptoms and normalization of CRP, ECG, echocardiogram

For athletes, duration of exercise restriction should be considered until resolution of symptoms and normalization of CRP, ECG and echocardiogram for at least 3 months

[28] Use corticosteroids in HIV pts as may increase risk of malignancy

Guidelines

2015 ESC guidelines for the diagnosis and management of pericardial diseases

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American Society of Echocardiography clinical recommendations for multimodality cardiovascular imaging of patients with pericardial disease: endorsed by the Society for Cardiovascular Magnetic Resonance and Society of Cardiovascular Computed Tomography
J Am Soc Echocardiogr. 2013;26:965–1012. http://www.sciencedirect.com/science/article/pii/S0894731713005336.

Patient Information

Images

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http://www.nlm.nih.gov/medlineplus/ency/imagepages/18151.htm.

Medlineplus

ENGLISH

http://www.nlm.nih.gov/medlineplus/ency/article/001103.htm.

ESPANOL

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Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/pericarditis/basics/definition/con-20035562.

Cleveland Clinic

http://my.clevelandclinic.org/services/heart/disorders/pericarditis.

MERCK

http://www.merckmanuals.com/home/heart-and-blood-vessel-disorders/pericardial-disease/acute-pericarditis.
http://www.merckmanuals.com/home/heart-and-blood-vessel-disorders/pericardial-disease/chronic-pericarditis.

AHA

http://www.heart.org/HEARTORG/Conditions/More/Symptoms-and-Diagnosis-of-Pericarditis-UCM-444932-Article.jsp.

Texas Heart Institute

http://www.texasheart.org/HIC/Topics/Cond/pericard.cfm.

Cardiosmart

https://www.cardiosmart.org/Heart-Conditions/Pericarditis

Professional Information

Review

N Engl J Med. 2014;371:2410–16. http://www.nejm.org/doi/full/10.1056/NEJMcp1404070?query=cardiology.

Review

J Nurse Pract. 2015;11:146–48. http://www.sciencedirect.com/science/article/pii/S1555415514007508

Review

Am Fam Physician. 2014;89:553–60. http://www.aafp.org/afp/2014/0401/p553.html.

Review

Lancet. 2004;363:717–27. http://www.sciencedirect.com/science/article/pii/S0140673604156481.

Review: Treatment

Circulation. 2013;127:1723–26. http://circ.ahajournals.org/content/127/16/1723.full.

Atrial Flutter/Fibrillation

Heart. 2015;101:1463–67. http://heart.bmj.com/content/101/18/1463?etoc

Clinical Profiles/Outcomes Hospitalized Patients

Circulation. 2014;130:1601–06. http://circ.ahajournals.org/content/130/18/1601.full?sid=94c2fb71-9fbe-443d-8f15-79263d7ce758.

Colchicine

N Engl J Med. 2013;369:1522–28. http://www.nejm.org/doi/full/10.1056/NEJMoa1208536.

Colchicine: Prevention of Post-Pericardiotomy Syndrome

Eur Heart J. 2010;31:2749–54. http://eurheartj.oxfordjournals.org/content/31/22/2749.

Colchicine: Prevention of Post-Pericardiotomy Syndrome

JAMA. 2014;312:1016–23. http://jama.jamanetwork.com/article.aspx?articleid=1900482.

Corticosteroids

Circulation. 2008;118:667–71. http://circ.ahajournals.org/content/118/6/667.full.

Cystic Tuberculous Pericarditis (Case Report)

J Am Coll Cardiol. 2013;62:1393–93. http://content.onlinejacc.org/article.aspx?articleID=1729179.

ECG (New Criteria): Acute Pericarditis Vs AMI

Am J Med. 2014;27:233–39. http://www.sciencedirect.com/science/article/pii/S0002934313009753.

Etiology

Am J Med. 2015;128:784.e1–784.e8. http://www.sciencedirect.com/science/article/pii/S0002934315001771.

Hydatid Cyst Rupture (Case Report)

Circulation. 2013;128:2073–4. http://circ.ahajournals.org/content/128/18/2073.full.

Metastatic Cancer

Circulation. 2013;128:1790–4. http://circ.ahajournals.org/content/128/16/1790.full

Prognosis

Circulation. 2013;128:42–9. http://circ.ahajournals.org/content/128/1/42.full.

Prognosis: Indicators

Circulation. 2007;115:2739–44. http://circ.ahajournals.org/content/115/21/2739.full.

Triage/Management

Int J Cardiol. 2007;118:286–94. http://www.sciencedirect.com/science/article/pii/S0167527306008862.

Tuberculosis

Heart. 2014;100:135–9. http://heart.bmj.com/content/100/2/135abstract

Updates and More

http://www.cormt.com/acperi
https://clinicalguidecvd.com/aaa

Chapter 76
Pericarditis: Constrictive

Management Keys

Differentiate from Restrictive Cardiomyopathy
Perform pericardiectomy when not responding to medical therapy

ICD-10 Code

I31.1

Description/Etiology

Thick, adherent, fibrotic pericardium causing decreased pericardial compliance and limited diastolic filling
Occurs after many diverse pericardial disease processes

Most common reported causes in developed countries:

Viral and idiopathic: 42–49 %
Post-cardiac surgery: 11–37 %
Post-radiation: 9–31 % [13]
Connective tissue/inflammatory disorder (3–7 %) [8]
Post-infectious (TB or purulent pericarditis): (3–6 %) [15]
Miscellaneous causes: 10 % [14]

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 1013
DOI 10.1007/978-1-4471-7293-2_76,

Risk of progression:

Viral and idiopathic pericarditis: <1 %
Immune-mediated pericarditis and neoplastic pericardial diseases: 2–5 %
Bacterial pericarditis (especially purulent pericarditis): 20–30 %

Rarely follows recurrent pericarditis
Clinical features due to volume overload and decreased cardiac output

Predisposing/Comorbid Conditions

ACUTE PERICARDITIS [ESP INF]
AUTOIMMUNE/CONNECTIVE TISSUE DISEASE [8]
BLUNT CHEST TRAUMA
CANCER
CARCINOID HEART DISEASE
CARDIAC SURGERY
CHRONIC RENAL FAILURE
HEART TRANSPLANT
INFECTIVE ENDOCARDITIS [ESP WITH INCOMPLETE DRAINAGE]
LUNG TRANSPLANT
RADIATION

Demography

Variable according to etiology

Pathophysiology

Anatomical: prominent pericardial thickening and calcifications; however, constriction may be present with normal pericardial thickness in up to 20 % of cases

Physiological:

Restrictive filling and intracardiac-intrathoracic pressure dissociation

Decreased venous filling of RV and ventricular interdependence lead to right heart volume overload and decreased LVSV [10]

Signs/Symptoms

ABDOMEN–DISTENSION
ABDOMEN–FLATULENCE
ABDOMEN–FLUID (ASCITES)
ABDOMEN–FULLNESS [ESP POSTPRANDIAL]
ABDOMEN–PAIN
APPETITE–DECR (ANOREXIA)
ARTERIAL PULSE–PARADOXICAL (PARADOXICAL PULSE)
ARTERIAL PULSE PRESSURE–DECR
BREATHING–DIFF (DYSPNEA)
BREATHING–DIFF, RECLINING FLAT (ORTHOPNEA)
BREATHING–RAPID (TACHYPNEA)
EXTREM, HANDS, PALMS, COLOR–RED (PALMAR ERYTHEMA)
EXTREM, LOWER, BILAT–EDEMA
EYES/SKIN–YELLOW (JAUNDICE)
FATIGUE
GENITALS, SCROTUM–SWOLLEN (EDEMA)
HEART–PERICARD KNOCK
HEART, LV, APEX, IMP–DECR/ABSENT
HEART, LV, APEX, IMP–RETRACTION, SYS
HEART, RATE–RAPID (TACHYCARDIA)
HEART, S2, SPLIT–WIDE
HEART, SOUNDS, INTENSITY–DECR
HYPOTENSION (BLOOD PRESSURE–DECREASED/LOW)
LIVER–ENLARGED (HEPATOMEGALY)

LIVER–PULSATION, PRESYS
LIVER–PULSATION, SYS
MUSCLES–ATROPHY
NECK, JVP–ELEV
NECK, JVP–INSP RISE (KUSSMAUL SIGN)
NECK, JVP, Y DESCENT–RAPID
SKIN–SPIDER ANGIOMAS
SKIN, COLOR, EFFORT–BLUE (CYANOSIS)
SPLEEN, SIZE–INCR (SPLENOMEGALY)
WEIGHT–LOSS

Differentiation

Cardiac Tamponade
Hypertrophic Cardiomyopathy
Hepatic disease
Intraabdominal malignancy
Nephrotic Syndrome
Other causes of HF
Other causes of systemic venous congestion
RA Myxoma
Restrictive Cardiomyopathy
SVC obstruction
TV regurgitation
TV stenosis

Complications

AF [1]
Cardiac Tamponade
Circulatory collapse
Effusive Constrictive Pericarditis
Heart block
HF
Hepatic dysfunction
Protein-losing Enteropathy

Laboratory

BLOOD, NT-PROBNP–INCR

ECG [6]

AV COND–1ST DEGREE BLOCK
AV COND–3RD DEGREE BLOCK
DYSRHYTHMIAS–ATRIAL (PACS/OTHERS)
P WAVE, DUR–INCR [2]
Q WAVE–ABN [3]
QRS–LONG, NS
QRS–RVH PATTERN
QRS–SLURRED
QRS, AMP–DECR
QRS, AXIS–R
RATE–INCREASED (SINUS TACHYCARDIA)
ST-T WAVE–ABN, NS
T WAVE–INVER, ABN
VOLTAGE, GEN–DECR

Imaging [5]

ABDOM–FLUID (ASCITES)
IVS, MOTION, DIAS–ABN [4]
LA, CHAMBER, SIZE–INCR
LIVER, SIZE–INCR (HEPATOMEGALY)
LV, FILLING–DECR/RESTRICTED
PERICARD–CALCIUM
PERICARD–FLUID
PERICARD–THICK
PLEURA–FLUID
RA, CHAMBER, SIZE–INCR
RV, FILLING–RESTRICTED

SPLEEN, SIZE–INCR (SPLENOMEGALY)
VEINS, HEPATIC, SIZE–INCR [WITH RESTRICTED RESP FLUCTUATIONS]
VENA CAVA, INF, SIZE–INCR [WITH RESTRICTED RESP FLUCTUATIONS]

Other Tests

Tests to rule out non-idiopathic causes
Cardiac catheterization [11]

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic

Treatment of congestion/supportive [9]
Treatment of inflammation [7]
Treatment of specific etiologies [16]

Treatment: Surgical/Invasive

Pericardiectomy [12]

Prevention

Treatment of underlying causes before progression to Constrictive Pericarditis [16]

Course

Variable per underlying etiology
Worse prognosis in patients with mixed disease (ie, underlying restrictive Cardiomyopathy, liver disease, prior radiation)

Notes

[1] 1/3 of patients
[2] Also may be notched, resembling P mitrale
[3] "Pseudoinfarction" pattern
[4] Termed septal bounce: "dip-plateau" phenomenon: abnormal early outward/inward IVS diastolic motion; also occurs in Cardiac Tamponade
[5] Transthoracic echocardiography recommended in all patients with suspected Constrictive Pericarditis; CXR (frontal/lateral views) recommended in all patients with suspected Constrictive Pericarditis; CT/CMR indicated as second-level imaging techniques to assess calcifications, pericardial thickness, degree/extension of pericardial involvement
[6] Can be normal
[7] May reverse transient constriction (occurs in 10–20 % of cases) within a few months, as a temporary measure during resolution of pericarditis; elevated CRP and imaging evidence of pericardial inflammation by contrast enhancement on CT/CMR may help identify patients with potentially reversible forms of constriction for whom empiric anti-inflammatory therapy should be considered, preventing need for pericardiectomy
[8] Including:

Amyloidosis
Behcet Disease
Familial Mediterranean Fever
Inflammatory Bowel Disease

Rheumatoid Arthritis
Sarcoidosis
Sjogren Syndrome
Systemic Lupus Erythematosus
Temporal Arteritis
Whipple Disease

[9] Especially diuretics, aimed at controlling symptoms of congestion in advanced cases and when surgery is high risk/contraindicated; medical therapy should never delay surgery
[10] Both ventricles cannot expand/fill at same time due to noncompliant pericardium; expansion of RV causes LV compression
[11] Cardiac catheterization indicated when non-invasive diagnostic methods do not provide a definite diagnosis; used to evaluate for ventricular interdependence, equalization of ventricular end-diastolic pressure, dissociation of intrathoracic/intracardiac pressures
[12] Nontrivial morbidity/mortality, but may be only definitive treatment in subset of patients
[13] Most cases due to Hodgkin's disease or breast cancer
[14] Includes malignancy, trauma, drug-induced, Asbestosis, Sarcoidosis, uremia
[15] Tuberculosis as a cause, however, may be increasing among immigrants from underdeveloped nations and in patients with HIV
[16] Especially tuberculosis, to prevent progression to constriction; antituberculous antibiotics may decrease risk of constriction from >80 % to <10 %

Guidelines

2015 ESC guidelines for the diagnosis and management of pericardial diseases
Eur Heart J. 2015;36:2921–64. http://eurheartj.oxfordjournals.org/content/36/42/2921.

American Society of Echocardiography Clinical Recommendations for multimodality cardiovascular imaging of patients with pericardial disease: endorsed by the Society for Cardiovascular

Magnetic Resonance and Society of Cardiovascular Computed Tomography
J Am Soc Echocardiogr. 2013;26:965–1012. http://www.sciencedirect.com/science/article/pii/S0894731713005336.

Patient Information

Images

http://www.nlm.nih.gov/medlineplus/ency/imagepages/18081.htm.
http://www.nlm.nih.gov/medlineplus/ency/imagepages/18151.htm.

Medlineplus

ENGLISH

http://www.nlm.nih.gov/medlineplus/ency/article/001103.htm.

ESPANOL0

http://www.nlm.nih.gov/medlineplus/spanish/ency/article/001103.htm.

Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/pericarditis/basics/definition/con-20035562.

Cleveland Clinic

http://my.clevelandclinic.org/services/heart/disorders/pericarditis.

MERCK

http://www.merckmanuals.com/home/heart-and-blood-vessel-disorders/pericardial-disease/acute-pericarditis.
http://www.merckmanuals.com/home/heart-and-blood-vessel-disorders/pericardial-disease/chronic-pericarditis.

AHA

http://www.heart.org/HEARTORG/Conditions/More/Symptoms-and-Diagnosis-of-Pericarditis-UCM-444932-Article.jsp.

Texas Heart Institute

http://www.texasheart.org/HIC/Topics/Cond/pericard.cfm.

Cardiosmart

https://www.cardiosmart.org/Heart-Conditions/Pericarditis.

Professional Information

Review

Lancet. 2004;363:717–27. http://www.thelancet.com/journals/lancet/article/PIIS0140-6736(04)15648-1/abstract.

Antiinflammatory Therapy

Circulation. 2011;124:1830–7. http://circ.ahajournals.org/content/124/17/1830.full?sid=3316db6d-3b55-4bc2-bc34-148c5f4022e1.

Imaging: Fungal Effusive Constrictive Pericarditis (Case Report)

Am J Med. 2013;126:25–6. http://www.sciencedirect.com/science/article/pii/S0002934312008017.

Pericardiectomy

Ann Thorac Surg. 2013;96:571–6. http://www.sciencedirect.com/science/article/pii/S0003497513008746.

Septal Bounce

Heart. 2013;99:1376. http://heart.bmj.com/content/99/18/1376.extract.

Syncope (Case Report)

Eur Heart J. 2013;34:1817. http://eurheartj.oxfordjournals.org/content/34/24/1817.

Thoracic Radiation

J Am Coll Cardiol. 2013;61:2319–28. http://content.onlinejacc.org/article.aspx?articleID=1679522.

Updates and More

https://clinicalguidecvd.com/conperi

Chapter 77
Peripheral Extremity Arteriovenous Fistula

ICD-10 Code

I77.0 ACQUIRED

Alternate Names/Abbreviation

A-V MALFORMATION
ARTERIOVENOUS MALFORMATION [2]

Description/Etiology

Abnormal vascular connection between peripheral artery and peripheral vein
Acquired and congenital forms

Predisposing/Comorbid Conditions

ANEURYSM
DEEP VEIN THROMBOSIS
HEMODIALYSIS SHUNT
HEREDITARY TELANGIECTASIA [CONGEN FORM]

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 1025
DOI 10.1007/978-1-4471-7293-2_77,

PERIPHERAL ARTERY DISEASE
SURGERY
TRAUMA [1]

Demography

All populations, with variances according to etiology

Pathophysiology

Local effects due to blood shunting from high press artery (s) to low pressure vein (s) with resulting distal ischemia and impingement on adjacent tissue (bone, nerve, skin)

Signs/Symptoms [3]

ARTERIAL PULSE PRESSURE – INCR
BLOOD PRESSURE, ARTERIAL – INCREASED/ELEVATED [SYS]
BRANHAM SIGN – POS [4]
BREATHING – DIFF (DYSPNEA)
EXTREM, LOCAL – BRUIT
EXTREM, LOCAL – MASS, PULSATILE
EXTREM, LOCAL – THRILL
EXTREM, UNILAT – DEFORMED [6]
EXTREM, UNILAT – EDEMA
EXTREM, UNILAT – ISCHEMIA, DISTAL
EXTREM, UNILAT – PAIN
EXTREM, UNILAT – PARESTHESIA
EXTREM, UNILAT, COLOR – BLUE (CYANOSIS)
FATIGUE
HEART, LV, APEX – MURMUR, SYS [5]

HEART, RATE – RAPID (TACHYCARDIA)
HEART, S3 LV
HEART, S4 LV
SKIN, COLOR – RED (ERYTHEMA)
SKIN, COLOR, LOCAL – HYPERPIGMENTATION
SKIN, LOCAL – ULCER
SKIN, TEMP, LOCAL – INCR
VEINS, SUPERFICIAL – DIL

Differentiation

Arterial aneurysm
Atherosclerotic occlusive disease
DVT

Complications

Bacterial endarteritis
Bone destruction
Cellulitis
HF (high output)
Hemorrhage
Infection/gangrene
Limb deformity
Pulmonary embolism

Laboratory

NS

ECG

QRS – LVH PATTERN

Imaging

CARDIOMEGALY [7]
DUPLEX ULTRASOUND FOR DX
LV, CHAMBER, SIZE – INCR
LV, WALL MOTION – INCR/HYPERDYNAMIC [8]

Other Tests

Angiography

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic

NS

Treatment: Surgical/Invasive

Embolization
Surgical resection

Course

Variable according to site/size of fistula

Notes

[1] Penetrating injuries, eg, gunshot; most often to thigh
[2] Angiographic classification of A-V malformations

Group 1: predominantly arterial or arteriovenous
Group 2: predominantly capillaries and small vessels
Group 3: predominantly venous

[3] Highly variable according to fistula duration, location
[4] Temporary occlusion of proximal artery causes transient reflex slowing of HR
[5] Due to increased cardiac output
[6] Congenital; limb may be elongated
[7] More often present with involvement of liver and lungs
[8] Severe shunts

Guidelines

NS

Patient Information

Medlineplus

ENGLISH

http://www.nlm.nih.gov/medlineplus/arteriovenousmalformations.html.

ESPANOL

http://www.nlm.nih.gov/medlineplus/spanish/arteriovenousmalformations.html.

Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/arteriovenous-fistula/basics/definition/con-20034876.

AANS

http://www.aans.org/Patient%20Information/Conditions%20and%20Treatments/Arteriovenous%20Malformations.aspx.

ASA

http://www.strokeassociation.org/STROKEORG/AboutStroke/TypesofStroke/HemorrhagicBleeds/What-Is-an-Arteriovenous-Malformation-AVM-UCM-310099-Article.jsp.

Johns Hopkins

http://www.hopkinsmedicine.org/neurology-neurosurgery/centers-clinics/cerebrovascular/conditions/arteriovenous-malformations-avm.html.

Stanford

https://stanfordhealthcare.org/medical-conditions/brain-and-nerves/arteriovenous-malformation.html.

NORD

https://rarediseases.org/rare-diseases/arteriovenous-malformation/.

UCSF

http://www.ucsfhealth.org/conditions/arteriovenous-malformation/.

Professional Information

Anterior Tibial Artery (Case Report)

J Vasc Surg. 2007;45:1076–9. http://www.sciencedirect.com/science/article/pii/S0741521406022634.

Imaging

Am J Roentgen. 2009;193:1425–33. http://www.ajronline.org/doi/abs/10.2214/AJR.09.2631.

Mechanisms of High Output HF

Am J Kidney Dis. 2004;43:e17–22. http://www.sciencedirect.com/science/article/pii/S0272638604001519.

Trauma Series

Surgery. 1975;78:817–28. http://www.surgjournal.com/article/0039-6060(75)90209-3/abstract.

Updates and More

https://clinicalguidecvd.com/peavf

Chapter 78
Pheochromocytoma (Chromaffin Tumor/ Paraganglioma)

Management Keys

- Suspect in patients with paroxysmal hypertension, tachycardia, diaphoresis, panic attacks
- Avoid beta-blockade prior to alpha-blockade; may exacerbate alpha effect of catecholamine surge
- Treat acute episodes with alpha-blocker and volume expansion, followed by beta-blocker/related agents after adequate alpha-blockade
- Avoid any form of sympathetic stimulation [7]
- Involve endocrinologist or hypertension expert in management decisions for patients with suspected pheo

ICD-10 Code

C74.10

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 1033
DOI 10.1007/978-1-4471-7293-2_78,

Alternate Names/Abbreviation

Pheo
PCC
PPGL (Pheochromocytoma And Paraganglioma)
Chromaffin Paraganglioma
Chromaffin Tumor
Chromaffinoma
Medullary Paraganglioma

Description/Etiology

Catecholamine secreting tumor that may cause life-threatening hypertension episodes/cardiac arrhythmias
Pheochromocytoma: tumor arising from adrenomedullary chromaffin cells that:

- Commonly produces one or more catecholamines: epinephrine, norepinephrine, and dopamine
- Are rarely biochemically silent

Paraganglioma: tumor derived from extra-adrenal chromaffin cells of thoracic/abdominal/pelvic sympathetic paravertebral ganglia; also arise from parasympathetic ganglia located along glossopharyngeal and vagal nerves in neck and at base of skull, but these do not produce catecholamines
Occur in both sporadic (75 %) and hereditary forms (25 %)

Predisposing/Comorbid Conditions

CARDIOMYOPATHY–DILATED
CARDIOMYOPATHY–HYPERTROPHIC
CARDIOMYOPATHY–TAKOTSUBO
MULTIPLE ENDOCRINE NEOPLASIA TYPE 2 [5]
NEUROFIBROMATOSIS [5]
VON HIPPEL-LINDAU DISEASE [5]
WERMER SYNDROME [9]

Demography

All ages affected, with first manifestations, usually:

Hereditary form: childhood/young adult
Sporadic form: 40–50 years

Pathophysiology

85–90 % located in adrenal medulla; 98 % intraabdominal; less often in sympathetic ganglia (paraganglioma); rarely intracardiac
10 % malignant
Clinical manifestations caused by secretion of excess catecholamines and enhanced sympathetic activity and resultant effects (HTN, tachycardia)
Increased sensitivity to normal sympathetic activity; any sympathetic stimulation to patient with pheo can cause hypertension crisis [**7**]

Signs/Symptoms

ABDOMEN–PAIN
ARTERIAL PRESSURE–HIGHLY VARIABLE [22]
ARTERIAL PRESSURE, UPRIGHT–DECR (ORTHOSTATIC HYPOTENSION)
BLOOD PRESSURE, ARTERIAL–INCREASED/ELEVATED [1]
BOWEL MOVEMENTS–CONSTIPATION [15]
CHEST–PAIN
CHEST–PALPITATIONS [2]
EXTREM, HANDS–TREMOR
EYES, RETINA–RETINOPATHY, HTN
EYES, VISION–BLURRED
FEVER [16]
FLANK–PAIN

HEADACHE [2]
HEART, RATE–RAPID (TACHYCARDIA) [3]
HYPOTENSION (BLOOD PRESSURE–DECREASED/LOW) [11]
MENTATION–FEELING OF DOOM
MOOD–ANXIOUS
MUSCLES–WEAK
NAUSEA
SKIN, COLOR–PALE (PALLOR) [23]
SLEEP–DISTURBED (INSOMNIA)
SWEATING–INCR (DIAPHORESIS/HYPERHIDROSIS) [2] [4]
VOMITING (EMESIS)
WEIGHT–LOSS [6]

Differentiation

Acute Intermittent Porphyria
Anxiety disorders
Alcohol withdrawal
Autonomic seizure disorder
Carcinoid
Chemical abuse [8]
Cluster headaches
CAD
Hyperthyroidism
Illicit drug use (ie, cocaine)
Mastocytosis
Menopausal Syndrome
Migraine
MVP
Panic attacks
Paroxysmal tachyarrhythmias
Poems Syndrome
Polypharmacy
Other causes of Orthostatic Hypotension

Complications [12]

Acute abdomen [10]
Acute Pulmonary Edema
AMI
DCM
Dysrhythmias – Supraventricular
Dysrhytmias – Ventricular
Encephalopathy
HCM
HF
Hypercalcemia
Ischemic Colitis
Lactic Acidosis
Postop Hypotension [21]
Postop Hypoglycemia [21]
Renal Failure
Shock
Stroke
Tumor rupture

Laboratory

BLOOD, ARTERIAL PH–DECREASED (ACIDOSIS)
BLOOD, CALCIUM–INCR [14]
BLOOD, GLUCOSE–INCR (HYPERGLYCEMIA) [13]
BLOOD, HGB/HCT–INCR [24]
BLOOD, LACTATE–INCR
BLOOD, WBC–INCR (LEUKOCYTOSIS) [24]

ECG [17]

DYSRHYTHMIAS–ATRIAL (PACS/OTHERS)
DYSRHYTHMIAS–VENTRICULAR (PVCS/OTHERS)
P WAVE, DUR–INCR
PR INTERVAL–SHORT

QRS–LVH PATTERN
QRS, AXIS–L
QT/QTC INTERVAL–LONG
RATE–INCREASED (SINUS TACHYCARDIA) [3]
ST-T WAVE–ABN, NS [17]

Imaging

IVS, MOTION–PARADOX [18]
LV, MYOCARD, WALL THICKNESS–INCR (HYPERTROPHY)
MV, LEAFLETS, MOTION, SYS–ANT [18]

Genomics

FP/TMEM127
RET (Multiple Endocrine Neoplasia type IIa OMIM 171400)
SDHB
SDHC
SDHA (Paragangliomas 5 OMIM 614165)
SDHAF2 (Paragangliomas 2 OMIM 601650)
SDHD (Paragangliomas 1 OMIM 168000)

Other Tests

Urine (24 h)

- Norepinephrine
- Epinephrine
- Normetanephrine
- Metanephrine [19]
- VMA

Blood

- Noradrenaline
- Adrenaline
- Normetanephrine
- Metanephrine [19]

Treatment: Nonpharmacologic

Avoid sympathetic stimulation [8]

Treatment: Pharmacologic [20]

Alpha - blockers
Beta-blockers
CCBs
Methyl paratyrosine

Treatment: Surgical/Invasive

Tumor excision [21]

Notes

[1] May be constantly (50 %) or part-time (95 %) elevated
[2] Triad of headache, palpitations, diaphoresis on background of hypertension is highly suggestive of pheo; may be precipitated by ingestion of tyramine-containing foods
[3] May be inappropriately increased for level of BP
[4] May be profound
[5] Hereditary forms
[6] Despite normal appetite
[7] Eg, pain, emotional upset, intubation, anesthesia, abdominal trauma
[8] Eg, cocaine, amphetamines, alcohol, MAO inhibitors
[9] Hyperparathyroidism, pituitary adenoma, pancreatic islet cell tumors
[10] Due to numerous causes, eg, tumor necrosis, bowel infarction/obstruction, cholecystitis
[11] Patients may present with hypotension rather than hypertension

[12] This is a partial list as pheos cause many diverse adverse effects on many organ systems
[13] Usually mild and occurs during acute hypertensive state
[14] Uncommon
[15] Due to catecholamine-induced intestinal hypomobility
[16] Due to hypercatabolic state; patients may present with fever of unknown origin
[17] Acute myocardial ischemic changes may also occur during hypertensive crisis
[18] During hypertensive crisis
[19] Metanephrines recommended for screening with >95 % sensitivity
[20] Significantly reduce perioperative mortality
[21] Preoperative and postoperative precautions, eg, pre-op alpha-blockers and post-op monitoring for hypotension and hypoglycemia; example: 2-week course of phenoxybenzamine or doxazosin with progressive dosage escalation until patient is orthostatic; CCBs also effective
[22] Variability is an added independent risk for CV morbidity and mortality
[23] Especially face/upper torso
[24] Due to hemoconcentration

Guidelines

Pheochromocytoma and paraganglioma: an endocrine society clinical practice guideline

J Clin Endocrinol Metab. 2014;99:1915–42. http://press.endocrine.org/doi/10.1210/jc.2014-1498?url_ver=Z39.88-2003&rfr_id=ori%3Arid%3Acrossref.org&rfr_dat=cr_pub%3Dpubmed&.

Patient Information

Mayo

http://www.mayoclinic.org/diseases-conditions/pheochromocytoma/basics/definition/con-20030435.

Medlineplus

https://www.nlm.nih.gov/medlineplus/ency/article/000340.htm.

MERCK

https://www.merckmanuals.com/home/hormonal-and-metabolic-disorders/adrenal-gland-disorders/pheochromocytoma.

UCSF

https://www.ucsfhealth.org/conditions/pheochromocytoma/?gclid=CjwKEAjwyPW5BRCC3JaM7qfW_FwSJACM3jz9WBX_04Tioz1S7CPeAqvm-W8E9UqeWKfdSkYBfKeE6xoC3aHw_wcB.

Professional Information

Review/Update

Circulation. 2014; 130: 1295–8. https://circ.ahajournals.org/content/130/15/1295.full.pdf+html.

Review

Lancet. 2005;366:665–75. http://www.thelancet.com/journals/lancet/article/PIIS0140-6736(05)67139-5/abstract.

Catecholamine Metabolism

Pharmacol Rev. 2004;56:331–49. http://pharmrev.aspetjournals.org/content/56/3/331.

Cardiovascular Manifestations

J Hypertens. 2011;29:2049–60. http://journals.lww.com/jhypertension/pages/articleviewer.aspx?year=2011&issue=11000&article=00001&type=abstract.

Clinical Experience

Ann Surg. 1999;229:755–64. http://journals.lww.com/annalsofsurgery/pages/articleviewer.aspx?year=1999&issue=06000&article=00001&type=abstract.

Genetics

Horm Metab Res. 2012;44:328–33. https://www.thieme-connect.de/DOI/DOI?10.1055/s-0031-1301302.

Metanephrines

J Clin Endocrinol Metab. 1998;83:2175–85. http://press.endocrine.org/doi/10.1210/jcem.83.6.4870.

Metastatic

Horm Metab Res. 2012;44:390–9. https://www.thieme-connect.de/DOI/DOI?10.1055/s-0031-1299707.

MRI

Eur Radiol. 2008;18:2885–92. http://link.springer.com/article/10.1007%2Fs00330-008-1073-z.

Overlooked Diagnosis

Am J Surg. 2000;179:212–5. http://www.americanjournalofsurgery.com/article/S0002-9610(00)00296-8/abstract.

Perioperative Morbidity/Mortality Factors

J Clin Endocrinol Metab. 2001;86:1480–6. http://press.endocrine.org/doi/abs/10.1210/jcem.86.4.7392.

Preoperative Apha Blockade In Normotensive Patients

J Hypertens. 2011;29:2429–32. http://journals.lww.com/jhypertension/pages/articleviewer.aspx?year=2011&issue=12000&article=00020&type=abstract.

Updates and More

https://clinicalguidecvd.com/pheo

Chapter 79
Primary Aldosteronism (Conn Syndrome)

ICD-10 Code

E26.09

Alternate Names/Abbreviation

CONN SYNDROME

Description/Etiology

Constellation of physiological abnormalities due to increased autonomous secretion of aldosterone, most often clinically manifest by Systemic Arterial Hypertension and its effects

Causes:

Adrenal Hyperplasia

Unilateral
Bilateral

Adrenal Carcinoma [2]
Adrenocortical Adenoma (Aldosteronoma) [1]
Ectopic tumors
Familial/hereditary [3]

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_79,

Predisposing/Comorbid Conditions

HYPERTENSION – SYSTEMIC ARTERIAL
HYPOKALEMIA [SYMPTOMATIC]

Demography

Rare in children
Usual initial presentation age 30–50 years

Pathophysiology

Increased levels of serum aldosterone with:

- Decreased renin
- Increased BP
- Increased sodium resorption/K excretion

Direct effects on CV system beyond hypertension-related pathology [12]

Signs/Symptoms

BLOOD PRESSURE, ARTERIAL – INCREASED/ELEVATED [7]
EXTREM – PAIN, SHOOTING (PARESTHESIAS) [5]
MUSCLES – CRAMPS [4]
MUSCLES – SPASM (TETANY) [5]
MUSCLES – WEAK [5]
URINATION – INCR (POLYURIA) [4]
URINATION – NIGHTTIME (NOCTURIA) [4]

Differentiation

Other causes of increased BP, especially (idiopathic) Systemic Arterial Hypertension
Other causes of Hypokalemia

Complications [6]

AF
AMI
LVH
STROKE
SCD

Laboratory

BLOOD, ALDOSTERONE: RENIN RATIO – INCR [13]
BLOOD, GLUCOSE – INCR (HYPERGLYCEMIA) [8]
BLOOD, POTASSIUM (K) – DECR (HYPOKALEMIA) [16]
BLOOD, RENIN – DECR
URINE, POTASSIUM – INCR

ECG [11]

AV COND – 1ST DEGREE BLOCK
AV COND – 2ND DEGREE BLOCK, MOBITZ I (WENCKEBACH)
AV CONDUCTION – AV DISSOCIATION, COMPLETE
DYSRHYTHMIAS – ATRIAL (PACS/OTHERS)
DYSRHYTHMIAS – VENTRICULAR (PVCS/OTHERS) [9]
QRS – LONG, NS
QRS – LVH PATTERN
ST SEGMENT – DEPR
T WAVE, AMP – DECR/FLAT
U WAVE – PROMINENT

Imaging

DEFINE RENAL ANATOMY
EXCLUDE ADRENAL CARCINOMA
LOCALIZE ADRENAL VEINS FOR SAMPLING

Genomics

ATP1A1 [SPORADIC FORM OF ADENOMA]
ATP2B3 [SPORADIC FORM OF ADENOMA]
CACNA1D [SPORADIC FORM OF ADENOMA]
CYP11B1 [FAMILIAL TYPE 1]
CYP11B2 [FAMILIAL TYPE 1]
KCNJ5 [FAMILIAL TYPE 111 OMNI 613677]

Other Tests

Adrenal vein sampling [14]
Confirmatory testing [15]

- Captopril challenge
- Fludrocortisone suppression
- Oral sodium loading
- Saline infusion

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic

Aldosterone antagonists
Eplerenone
Spironolactone

Treatment: Surgical/Invasive

Adrenalectomy

Notes

[1] Small, benign
[2] Rare; often secrete other adrenal hormones; locally invasive/metastatic
[3] Types:

FH 1: Glucocorticoid-Remedial Aldosteronism (autosomal dominant)
FH 2: Non-glucocorticoid Remediable Aldosteronism (5x more common than FH 1)
FH 3: Paradox worsening of HTN with Dexamethasone

[4] Due to hypokalemia
[5] Severe hypokalemia
[6] All significantly more frequent than in patients with essential hypertension
[7] Usually moderate-severe increase; occasionally normal
[8] With glucose load
[9] PVCS, VT
[10] AF, PAT with block
[11] Changes mainly due to hypokalemia/LVH
[12] Fibrosis, inflammation, remodeling, LVH
[13] Preferred screening test for Primary Aldosteronism; positive test: >20 ng/dl per ng/m/h with aldosterone >10 mg/dl
[14] Differentiates between unilateral disease and bilateral disease before adrenalectomy as CT alone may be inadequate
[15] After adrenal vein sampling
[16] May be normal in absence of diet high in sodium or when not taking diuretics
[17] 1/3 of patients with this disease achieve normal BP after adrenalectomy; factors associated with normal BP post-surgery include negative family history of hypertension, <3 antihypertensive agents

Guidelines

Case detection, diagnosis, and treatment of patients with primary aldosteronism: an endocrine society clinical practice guideline
J Clin Endocrinol Metab. 2008;93:9. http://press.endocrine.org/doi/full/10.1210/jc.2008-0104.

2013 ESH/ESC guidelines for the management of arterial hypertension
Eur Heart J. 2014;34:2159–219. http://www.escardio.org/guidelines-surveys/esc-guidelines/Pages/arterial-hypertension.aspx.

The 2012 Canadian Hypertension Education Program recommendations for the management of hypertension: blood pressure measurement, diagnosis, assessment of risk, and therapy
Can J Cardiol. 2012;28:277. http://www.sciencedirect.com/science/article/pii/S0828282X12001365#.

NICE: hypertension in adults: diagnosis and management (2011)
http://www.nice.org.uk/guidance/cg127/chapter/1-recommendations#choosing-antihypertensive-drug-treatment-2.

JNC 7: seventh report of the Joint National Committee on prevention, detection, evaluation, and treatment of high blood pressure
Hypertension. 2003;42:1206–12. http://hyper.ahajournals.org/content/42/6/1206.long.

Patient Information

Images

http://www.nlm.nih.gov/medlineplus/ency/imagepages/1093.htm.
http://www.nlm.nih.gov/medlineplus/ency/imagepages/8719.htm.

Medlineplus

ENGLISH

http://www.nlm.nih.gov/medlineplus/ency/article/000330.htm.

ESPANOL
http://www.nlm.nih.gov/medlineplus/spanish/ency/article/000330.htm.

Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/primary-aldosteronism/basics/definition/con-20030194.

Professional Information

Review

Postgrad Med J. 2001;77:639–44. http://pmj.bmj.com/content/77/912/639.full.

Review

N Engl J Med. 2007;356:601–10. http://www.nejm.org/doi/full/10.1056/NEJMcp065470.

Review

Trends in Endocrinology & Metabolism. 2013;24:421–30. http://www.sciencedirect.com/science/article/pii/S1043276013000829.

Review: Genetic/Autoimmune Triggers

Hypertension. 2015;66:248–53. http://hyper.ahajournals.org/content/66/2/248.full.

Adrenal Hypertension

Arch Intern Med. 1974;133:1001–6. http://archinte.jamanetwork.com/article.aspx?articleid=583081.

Adrenalectomy

Am J Hypertens. 2015;28:312–8. http://ajh.oxfordjournals.org/content/28/3/312.abstract.

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Adrenocortical Function Assessment

N Engl J Med. 1971;285:735–9. http://www.nejm.org/doi/full/10.1056/NEJM197109232851306.

Aldosterone-Renin Ratio

JAMA. 2014;312:184–5. http://jama.jamanetwork.com/article.aspx?articleid=1886170.

Atrial Fibrillation

Eur Heart J. 2011;33:2098–108. http://eurheartj.oxfordjournals.org/content/33/16/2098.

Complications

J Am Coll Cardiol. 2005;45:1243–8. http://content.onlinejacc.org/article.aspx?articleid=1136490.

Cardioavascular Outcomes Post-treatment

Arch Intern Med. 2008;168:80–5. http://archinte.jamanetwork.com/article.aspx?articleid=413688.

Differentiation of Unilateral from Bilateral Adrenal Disease

Ann Intern Med. 2009;151:329–37. http://annals.org/article.aspx?articleid=744703&resultClick=24.

Familial

Endocrinol Metab Clin North Am. 2011;40:343–68. http://www.sciencedirect.com/science/article/pii/S0889852911000089.

Medical Threapy

J Hypertension. 2001;19:353–61. http://www.ncbi.nlm.nih.gov/pubmed/?term=11288803%5Buid%5D.

Post-Surgery BP

Ann Intern Med. 2001;135:258–61. http://annals.org/article.aspx?articleid=714690&resultClick=24.

Updates and More

https://clinicalguidecvd.com/aldo

Chapter 80
Pulmonary Arteriovenous Fistula

ICD-10 Code

Q25.72 CONGENITAL
I28.0 ACQUIRED

Alternate Names/Abbreviation

Pulmonary AV Fistula
Pulmonary Arteriovenous Malformations (PAVMs)

Description/Etiology

- Abnormal direct high flow, low-resistance fistulous connection between pulmonary artery and vein
- Usually congenital with autosomal dominance, most often Hereditary Hemorrhagic Telangiectasia, but may be acquired [1]
- R-L shunt can cause dyspnea but most patients are asymptomatic
- Paradoxical emboli can cause stroke and brain abscess and acute events in other organs

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 1055
DOI 10.1007/978-1-4471-7293-2_80,

PAVMs in hepatopulmonary syndrome: due to failure of hepatic clearance of vasoactive substances (eg, prostaglandins); may be mechanism of PAVM development in children following shunt procedures for congenital cardiac anomalies

Predisposing/Comorbid Conditions

HEPATIC CIRRHOSIS
HEREDITARY HEMORRHAGIC TELANGIECTASIA [CONGEN FORM] [1]
HISTORY: CHEST SURGERY
INFECTION [4]
PENETRATING CHEST TRAUMA
PULMONARY AMYLOIDOSIS
PULMONARY METASTASIS

Demography

F > M [2]
All ages [3]

Pathophysiology

Anatomic types:

Simple: single segmental artery feeding malformation; feeding segmental artery may have multiple subsegmental branches feeding malformation
Complex: multiple segmental feeding arteries

Lesions: >50 % lower lobes and right middle lobe; 70 % unilateral; may be multiple
Depending on shunt size, cardiac/circulatory hemodynamics may be normal (most cases); large shunts may cause significant arterial O_2 desaturation

Signs/Symptoms [1] [5] [9]

BOWEL MOVEMENTS, STOOL–BLOOD (HEMATOCHEZIA) OR BLACK (MELENA) [1]
BREATHING–DIFF (DYSPNEA)
BREATHING–DIFF, UPRIGHT (PLATYPNEA)
CHEST–PAIN
CHEST, LOCAL–MURMUR, SYS [6] [14]
CHEST, LOCAL–THRILL [6] [14]
CHEST. LOCAL–MURMUR, CONT [6] [14]
CONSCIOUSNESS–LOSS, SUDDEN (SYNCOPE) [1] [9]
COUGH
DIZZY/SPINNING (TRUE VERTIGO) [1] [9]
EXTREM, DIGITS–CLUBBED
EXTREM, DIGITS, NAILS–TELANGIECTASIA [1] [12]
EYES, RETINA–TELANGIECTASIA [1] [12]
HEADACHE [1]
HEART, P2, INTENSITY–INCR
LIPS–BLEEDING [1]
LIPS–TELANGIECTASIA [1] [12]
MENTATION–CONFUSION [1] [9]
MOUTH, MUCOUS MEMBRANES–TELANGIECTASIA [1] [12]
NOSE–BLOOD (EPISTAXIS) [1]
NOSE, MUCOSA–TELANGIECTASIA [1] [12]
SKIN–TELANGIECTASES [1] [10] [12]
SKIN, COLOR–BLUE (CYANOSIS)
SPUTUM–BLOOD (HEMOPTYSIS) [7]
SWALLOWING–DIFFICULT (DYSPHAGIA) [1] [9]
TONGUE–TELANGIECTASIA [1] [12]
URINE–BLOOD (HEMATURIA) [1]
VOMITING–BLOOD (HEMATEMESIS) [1]

Differentiation

Atelectasis
CNS Disease
Coagulation disorders
Lung benign and malignant masses
Primary Polycythemia Vera
Pulmonary embolism
Pulmonary Hypertension
Pulmonary infarction

Complications

Cerebral abscess [8] [9]
Hemothorax
Massive hemoptysis [7]
Meningitis [8] [9]
PAH [8]
Seizures [8] [9]
Stroke [8] [9]

Laboratory

BLOOD, ART, PO2–DECR/WORSENS IN UPRIGHT POSITION (ORTHODEOXIA)
BLOOD, ARTERIAL PCO2–DECREASED [11]
BLOOD, ARTERIAL PO2–DECREASED (HYPOXIA) [11] [15]
BLOOD, HGB/HCT–INCR [11]
BLOOD, RBC COUNT–INCR [11]
BLOOD, RETICULOCYTE COUNT–INCR [11]

ECG

N/NS ABN

Imaging

CARDIOMEGALY [LARGE FISTULAE; OTHERWISE N SIZE]
CONTRAST ECHO/CT
LUNG DENSITY(S) [CHEST X-RAY] [13]

Other Tests

Pulmonary arteriography

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic

NS

Treatment: Surgical/Invasive

Embolization [16]
Surgical resection [16]

Course

Highly variable due to associated malformations

Notes

[1] Hereditary Hemorrhagic Telangiectasia (Rendu-Osler-Weber syndrome); >80 % of cases; many clinical features due to CNS involvement, which may be caused by A-V fistulae in brain, paradoxical embolism, or severe polycythemia; other organs (kidneys, GI tract) may also be involved with A-V fistulae causing the listed signs and symptoms
[2] Ratio varies among reports, from about 1:1 up to 2:1 female predominance
[3] Defect present at birth expands with age and usually clinically first manifest age 20–40 years
[4] Including Schistosomiasis, Actinomycosis
[5] Includes features of Hereditary Hemorrhagic Telangiectasia
[6] Over A-V malformation sites, which are usually peripheral in lower lobes and right middle lobe; louder during systole and with deep inspiration
[7] Usually due to fistula rupture; rarely due to endobronchial telangiectasia
[8] May be more common in acquired form
[9] CNS signs and symptoms are presenting manifestations in many patients
[10] Skin lesions are fragile and bleed easily, especially with sunlight exposure
[11] Blood indices correct after definitive treatment of pulmonary fistulae
[12] Small clusters of ruby red lesions
[13] Oval, round, homogeneous lesions; single or multiple; unilateral or bilateral; most often lower lobes and right middle lobe; usually peripheral
[14] May disappear during pregnancy due to diaphragm elevation
[15] Does not correct with pure O_2 inhalation
[16] Historically treated with surgical resection but endovascular embolization now treatment standard, using coils and detachable occlusion balloons; >95 % success rate with embolization but recanalization can occur

Guidelines

NS

Patient Information

Medlineplus

ENGLISH

http://www.nlm.nih.gov/medlineplus/ency/article/001090.htm.

ESPANOL

http://www.nlm.nih.gov/medlineplus/spanish/ency/article/001090.htm.

Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/arteriovenous-fistula/basics/symptoms/con-20034876.

Professional Information

First Description: Churton

Br Med J. 1897;1:1223.

Review

Semin Intervent Radiol. 2011;28:24–31. http://www.ncbi.nlm.nih.gov/pmc/articles/PMC3140246/.

Review

Postgrad Med J. 2002;78:191–97. http://pmj.bmj.com/content/78/918/191.

Review

Am J Resp Crit Care Med. 1998;158:643–61. http://www.atsjournals.org/doi/full/10.1164/ajrccm.158.2.9711041#.Vzs_HvMUV1s.

Review: Hereditary Hemorrhagic Telangiectasia

N Engl J Med. 1995;333:918–24. http://www.nejm.org/doi/full/10.1056/NEJM199510053331407.

Case Report: Hereditary Complex Av Fistula

BMJ Case Reports. 2014. doi:10.1136/bcr-2014-205939. http://casereports.bmj.com/content/2014/bcr-2014-205939.abstract.

Case Series

Chest. 1963;43:449–55. http://journal.publications.chestnet.org/article.aspx?articleid=1055734.

Cavo-Pulmonary Shunt (Children)

Circulation. 1995;92:309–14. http://circ.ahajournals.org/content/92/9/309.full?sid=28b1b275-ef73-45d4-b159-76c7c813e4a8.

Images

N Engl J Med. 2009;360:1769. http://www.nejm.org/doi/full/10.1056/NEJMicm0803889.

Neurological Manifestations

Circulation. 1970;41:123–28. http://circ.ahajournals.org/content/41/1/123.abstract?sid=6831653d-5534-4f24-bd0b-bf8bb3b92815.

Pulmonary Amyloidosis

Am J Respir Crit Care Med. 2014;190:e14–5. http://www.ncbi.nlm.nih.gov/pubmed/25127313.

Pulmonary Amyloidosis

Chest. 1989;96:1435–6. http://journal.publications.chestnet.org/article.aspx?articleid=1062488&resultClick=3.

Surgery: Children

J Pediatr Surg. 2008;43:1365–367. http://www.ncbi.nlm.nih.gov/pubmed/18639698.

Transcatheter Coil Closure

J Interv Cardiol. 2004;17:23–6. http://onlinelibrary.wiley.com/doi/10.1111/j.1540-8183.2004.00287.x/abstract;jsessionid=A0FD3515BFFD722686A689300670D2BF.f01t03.

Transcatheter Occlusion

Heart. 2011;97:A1550. doi:10.1136/heartjnl-2011-300867.451. http://heart.bmj.com/content/97/Suppl_3/A155.1.abstract?sid=12a8325b-4b29-430a-8c29-266d7ac8aa45.

Updates and More

https://clinicalguidecvd.com/pavf

Chapter 81
Pulmonary Hypertension

ICD-10 Code

I27.0

Alternate Names/Abbreviation

CTPH (CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION)
PH (PULMONARY HYPERTENSION)
PAH (PULMONARY ARTERIAL HYPERTENSION)

Description/Etiology

Pulmonary Hypertension: pulmonary arterial pressure ≥ 25 mmHg (mean) at rest measured by right heart catheterization [31]

Pulmonary Arterial Hypertension: PH characterized hemodynamically by presence of pre-capillary PH, defined by a pulmonary artery wedge pressure ≤ 15 mmHg and PVR >3 Wood units (WU) in absence of other causes of pre-capillary PH such as PH due to lung diseases, CTEPH, or other rare diseases

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 1065
DOI 10.1007/978-1-4471-7293-2_81,

Symptoms: non-specific and mainly related to progressive RV dysfunction

2013 Updated Classification [31]

Group 1: Pulmonary Arterial Hypertension

1.1 Idiopathic PAH
1.2 Hereditable PAH

1.2.1 BMPR2
1.2.2 Alk-1, Eng, Smad9, Cav1, Kcnk3
1.2.3 Unknown

1.3 Drug and toxin induced [32]
1.4 Associated with:

1.4.1 Connective tissue disease
1.4.2 HIV infection
1.4.3 Portal Hypertension
1.4.4 Congenital heart disease
1.4.5 Schistosomiasis

1' Pulmonary Veno-Occlusive Disease and/or Pulmonary Capillary Hemangiomatosis
1' Persistent Pulmonary Hypertension of the Newborn (PPHN)

Group 2: Pulmonary Hypertension due to left heart disease

2.1 Left ventricular systolic dysfunction
2.2 Left ventricular diastolic dysfunction
2.3 Valvular disease
2.4 Congenital/acquired left heart inflow/outflow tract obstruction and congenital cardiomyopathies

Group 3: Pulmonary Hypertension due to lung disease and/or hypoxia

3.1 Chronic Obstructive Pulmonary Disease
3.2 Interstitial Lung Disease
3.3 Other pulmonary diseases with mixed restrictive and obstructive pattern
3.4 Sleep-disordered breathing
3.5 Alveolar hypoventilation disorders
3.6 Chronic exposure to high altitude
3.7 Developmental lung disease

Group 4: Chronic Thromboembolic Pulmonary Hypertension (CTEPH)

Group 5: Pulmonary Hypertension with unclear multifactorial mechanisms [29]

5.1 Hematologic disorders:

Chronic hemolytic anemia
Myeloproliferative disorders
Splenectomy

5.2 Systemic disorders:

Sarcoidosis
Pulmonary Histiocytosis
Lymphangiomyomatosis

5.3 Metabolic disorders:

Glycogen Storage Disease
Gaucher Disease
Thyroid Disorders (including Hyperthyroidism, Graves Disease)

5.4 Others:

Tumoral obstruction
Fibrosing Mediastinitis
Chronic renal failure
Segmental PH

Predisposing/Comorbid Conditions

[SEE DISEASES LISTED IN 2013 UPDATED CLASSIFICATION]

Demography

Highly variable according to etiology

Pathophysiology

PH: abnormal cell signaling pathways within alveolar-pulmonary arteriole-right ventricular axis causes increased PVR, which leads to RV dysfunction

PAH: among the many etiologies and varying complex pathogeneses, common mechanisms that trigger abnormal vascular remodeling include:

- Aberrant vascular wall cell proliferation
- Bone morphogenetic protein receptor type 2 gene mutations
- Endothelial dysfunction
- Inflammation

Signs/Symptoms [4]

ABDOMEN – DISTENSION [15]
ABDOMEN – FLUID (ASCITES) [15]
APPETITE, SATIETY – EARLY [15]
ARTERIAL PULSE PRESSURE – DECR
ARTERIAL PULSE, AMP – DECR/ABS
BREATHING – DIFF (DYSPNEA)
BREATHING – DIFF, NOCTURNAL (DYSPNEA, NOCT)
BREATHING – DIFF, RECLINING FLAT (ORTHOPNEA)
CHEST – PAIN

CHEST – PAIN, EFFORT (ANGINA PECTORIS) [16]
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)
EXTREM, LOWER, BILAT – EDEMA
FATIGUE
HEART, LSB, LOWER – IMP, SYS
HEART, LSB, LOWER – MURMUR, SYS [6]
HEART, LSB, UPPER – CLICK, SYS
HEART, LSB, UPPER – MURMUR, DIAS [7]
HEART, P2 – PALPABLE
HEART, P2, INTENSITY – INCR
HEART, RATE – RAPID (TACHYCARDIA)
HEART, RSB, LOWER – HEAVE
HEART, S2, SPLIT – FIXED [3]
HEART, S2, SPLIT – REVERSED (PARADOXICAL) [3]
HEART, S2, SPLIT – WIDE
HEART, S3 RV [UNCOMMON]
HEART, S4 RV
HYPOTENSION (BLOOD PRESSURE – DECREASED/LOW)
LIVER – ENLARGED (HEPATOMEGALY)
LIVER – PULSATION, PRESYS
MENTATION – WEAKNESS (MALAISE)
NECK, JVP, A WAVE – INCR/LARGE (CANNON WAVE)
NECK, JVP, C WAVE – INCR/LARGE
NECK, JVP, V WAVE – INCR/LARGE
NECK, SENSATION – PULSATIONS
SKIN, COLOR – BLUE (CYANOSIS)
SKIN, COLOR, EFFORT – BLUE (CYANOSIS)
SKIN, TEMP, HANDS/FEET – DECR
SPUTUM – BLOOD (HEMOPTYSIS) [17]
SWALLOWING – DIFFICULT (DYSPHAGIA)
VOICE – HOARSE [33]

Differentiation

Chronic Obstructive Pulmonary Disease
Myxoma – Right Ventricle
Other causes of right heart failure

Other causes of cyanosis

Complications [4]

Pneumonia
Pulmonary artery dissection/rupture with Cardiac Tamponade
RV failure
Sudden Death

Laboratory [4] [37]

BLOOD, NT-PROBNP – INCR [18]
BLOOD, PLATELETS – DECR (THROMBO CYTOPENIA) [21]
BLOOD, PLATELETS – INCR (THROMBOCYTOSIS) [21]
BLOOD, ST2 – INCR [PAH]
BLOOD, TROPONIN – INCR [SEVERE]
BLOOD, URIC ACID – INCR [20]

ECG [4] [34] [37]

P WAVE – TALL/PEAKED
QRS – RVH PATTERN [26]
QRS, AXIS – R
T WAVE – INVER, ABN [25]

Imaging [4] [8] [35] [37]

LA, CHAMBER, SIZE – INCR [27]
RA, CHAMBER, SIZE – INCR
PA, BRANCHES, SIZE – DECR
PA, MAIN, PRESS – INCR [12]

PA, MAIN, SIZE – INCR
PERICARD – FLUID [13]
PV, FLOW – REGURG
RV, CHAMBER, SIZE – INCR

Genomics [37]

ALK-1
CAV1
EF2AK4
ENDOGIN
KCNK3
SMAD9
BMPR2 [>80 % OF FAMILIAL CASES of PAH]

Other Tests [8] [37]

Pulse oximetry [19]
Ventilation/perfusion scan for detecting chronic thromboembolic PH
Right heart catheterization/pulmonary angiography: confirm diagnosis; assess hemodynamic severity and vasoreactivity of pulmonary circulation [28]
Coronary angiography: assess for comorbid CAD
6 min walk test [36]

Treatment: Nonpharmacologic [4] [37]

Avoid pregnancy
Exercise rehabilitation
Low sodium diet for volume overload with RV failure
O_2
Smoking cessation

Treatment: Pharmacologic [4] [37]

Antiarrhythmics [10]
Anticoagulants
CCBs
Diuretics
Pulmonary artery vasodilators

Treatment: Surgical/Invasive [4] [37]

Atrial septostomy [11]
Lung transplant
Pulmonary artery denervation (investigational for PAH)

Prevention

Tobacco avoidance/cessation
Vaccination:

- Pneumococcal
- Viral

Course

Variable according to category

Notes

[1] Initial symptoms (mainly dyspnea with effort) usually do not appear until disease is advanced
[2] Higher incidence in males reported in Japan
[3] With severe RV dysfunction
[4] Not including specific manifestations/treatment for associated conditions (eg, Sarcoidosis, Heart Failure, Thyroid

disease, Pulmonary Thromboembolic disease), which must be individualized
[5] Occur mainly in advanced cases
[6] Pansystolic; due to TR
[7] Pulmonary regurgitation; due to dilated main PA, usually due to long-standing increased PA press
[8] Normal CXR does not rule out PH; may assist in differential diagnosis of PH by showing features of lung or left heart disease; may help distinguish between arterial PH (increased artery-vein ratio) and venous PH (decreased artery-vein ratio)
[9] Autoimmune thyroid disease common in group 5
[10] Maintenance of sinus rhythm if possible
[11] Palliative/bridging
[12] Doppler echo estimate of PA systolic pressure correlates well with direct measure
[13] Presence of pericardial fluid correlates with poorer survival
[14] Heart Failure: most common cause of PH; occurs in >80 % of HF patients
[15] Due to abdominal venous congestion
[16] Believed due to decreased right coronary flow to hypertrophied RV causing myocardial ischemia
[17] Hemoptysis uncommon; presence should prompt search for other causes, such as Pulmonary Embolism, Mitral Stenosis
[18] May be sign of impending/overt RV failure, suggesting need for more aggressive treatment
[19] In PAH: normal/low; marked desaturation with exercise suggests possible cardiac shunt
[20] Clubbing uncommon except in Eisenmenger and hypoxic lung disease
[21] Increased platelets: risk factor for PH
[22] Decreased platelets: suggests possible autoimmune disease, hypersplenism, cirrhosis
[23] Increased BNP common in PH: degree correlates with RVH severity/poorer outcomes
[24] Mechanism of increased uric acid uncertain

[25] Anterior leads, may extend to inferior leads
[26] Insensitive but highly specific for RVH secondary to PH
[27] LA dilatation suggests pulmonary venous hypertension rather than PAH
[28] Agents for testing pulmonary vasoreactivity include:

Adenosine
Epoprostenol
Iloprost
Nitric oxide

[29] Also: Hereditary Hemorrhagic Telangiectasia
[30] In PAH and experimental PH, KCNK3 expression/activity strongly reduced in pulmonary artery smooth muscle cells and endothelial cells
[31] Normal mean PA resting pressure: 14 ± 3 mmHg with upper limit of normal about 20 mmHg; clinical significance of PA resting pressure of 21–24 mmHg uncertain but patients with pressures in this range should be followed closely
[31] Clinical classification of Pulmonary Hypertension derived from J Am Coll Cardiol 2004;43 (Suppl 1): S5–S12
[32] Drug/toxin causes of PAH:

Definite

Aminorex
Fenfluramine
Dexfenfluramine
Toxic rapeseed oil
Benfluorex
Selective serotonin reuptake inhibitors

Likely

Amphetamines
Dasatinib
L-tryptophan
Methamphetamines

Possible

Cocaine
Phenylpropanolamine
St John's Wort
Amphetamine-like drugs
Interferon α and β
Some chemotherapeutic agents such as alkylating agents

Mytomycine C
Cyclophosphamide

[33] Due to PA dilatation compressing left recurrent laryngeal nerve
[34] Normal ECG does not rule out PH
[35] ESC/ERS Guideline: Echocardiography should always be performed when PH is suspected and may be used to infer a diagnosis of PH in patients in whom multiple different echocardiographic measurements are consistent with this diagnosis
[36] ESC/ERS Guideline: 6 min walk test is most widely used exercise test in PH centers; test is easy to perform, inexpensive and familiar to patients and centers; as with all PH assessments, test results must always be interpreted in clinical context; distance is influenced by several factors, including sex, age, height, weight, comorbidities, need for O_2, learning curve, motivation
[37] Listed diagnostics and treatments are generic; management of patients with PH/PAH should be conducted by experts in these conditions whenever possible, with careful attention/adherence to most current guidelines

Guidelines

2015 ESC/ERS guidelines for the diagnosis and treatment of pulmonary hypertension
Eur Heart J. 2016;37:67–119. http://eurheartj.oxfordjournals.org/content/37/1/67.

Pharmacologic therapy for pulmonary arterial hypertension in adults: CHEST guideline and expert panel report
Chest 2014;146:449–75. http://journal.publications.chestnet.org/article.aspx?articleid=1881654.
Executive summary: expert consensus statement on the diagnosis and treatment of paediatric pulmonary hypertension. The European Paediatric Pulmonary Vascular Disease Network, endorsed by ISHLT and DGPK
Heart. 2016;102:ii86–100. doi:10.1136/heartjnl-2015-309132. http://heart.bmj.com/content/102/Suppl-2/ii86?etoc.

Patient Information

Genetics Home Reference

http://ghr.nlm.nih.gov/condition/pulmonary-arterial-hypertension.

Medlineplus

ENGLISH
http://www.nlm.nih.gov/medlineplus/pulmonaryhypertension.html

ESPANOL
http://www.nlm.nih.gov/medlineplus/spanish/pulmonaryhypertension.html.

Cleveland Clinic

http://my.clevelandclinic.org/lungs-breathing-allergy/departments-centers/pulmonary-hypertension.aspx.

Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/pulmonary-hypertension/basics/definition/con-20030959.

NHLBI

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AHA

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Updates and More

https://clinicalguidecvd.com/ph

Chapter 82
Pulmonary Stenosis: Supravalvular

ICD-10 Code

Q25.6

Alternate Names/Abbreviation

PULMONARY ARTERY STENOSIS

Description/Etiology

Narrowing (single/combination) of:

- Main pulmonary trunk
- Pulmonary artery bifurcation
- Primary/intrapulmonary artery branches

Variant: hourglass pattern, similar to SupraAS; technically a form of valvular PS as it is due to stenosis at valve commissural ridge

All forms can cause RVOT obstruction and subsequent RV dysfunction/RHF

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*,
DOI 10.1007/978-1-4471-7293-2_82,

Lesions:

- Range from single focal to diffuse hypoplastic to overt occlusion
- May be secondary to previous pulmonary artery band placement
- Arteries distal to patent stenotic lesions often dilated (poststenotic dilation)

Membranous forms of obstruction both above/below pulmonary valve may occur

Comorbid Conditions

AORTIC STENOSIS – SUPRAVALVULAR
ALAGILLE SYNDROME [10]
BEHCET DISEASE
CONGENITAL RUBELLA
EHLERS-DANLOS SYNDROME
KEUTEL SYNDROME [11]
SCARRING AT SITE OF PREVIOUS PA BAND OR AORTICOPULMONARY SHUNT
SILVER SYNDROME [12]
SYSTEMIC VASCULITIS
TAKAYASU ARTERITIS
WILLIAMS SYNDROME

Demography

GENDER EQUAL

Pathophysiology

Lesions: fibrous intimal proliferation with varying degrees of medial hyperplasia and elastic fibers loss
Hemodynamics: hypertension in proximal PA with secondary pulmonary regurgitation leading to:

RV pressure/volume overload
RV dilatation
TR
RV dysfunction/RH failure

Progresses to loss of distal lung parenchyma by adulthood
Peripheral PA stenoses often occur in multiple PA tertiary branches and are progressive

Signs/Symptoms

BREATHING – DIFF (DYSPNEA)
CHEST – PAIN [3]
CHEST, LAT – BRUIT [4]
CHEST, LAT – MURMUR, CONT [4]
CHEST, POST – BRUIT [4]
CHEST, POST – MURMUR, CONT [4]
HEART, RSB, UPPER – MURMUR, DIAS [7]
HEART, S4 RV
NECK, JVP, A WAVE – INCR/LARGE (CANNON WAVE)
NECK, VENOUS PULSE – AWARE [5]
SKIN, COLOR – BLUE (CYANOSIS) [6]
SPUTUM – BLOOD (HEMOPTYSIS) [1]

Differentiation

Myxoma – Right Ventricle
Pulmonary Hypertension [8]
Pulmonary Stenosis – Valvular

Complications

Infective Endocarditis [2]
HF
RV infarction

Laboratory

NS

ECG

QRS – RVH PATTERN
QRS, AXIS – R

Imaging

[MRI/CT SUPERIOR TO ECHO IN IDENTIFYING LESIONS]
PA, MAIN, SIZE – INCR [POSTSTENOTIC DILATATION]
PV, FLOW – REGURG
RV, PRESS, SYS – INCR
RV, WALL THICKNESS – INCR

Genomics

BSCL2
ELASTIN
JAG1
MGP

Other Tests

Cardiac catheterization with contrast angiography: definitive for lesions and severity of PAH [8].

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic

NS

Treatment: Surgical/Invasive [13]

Balloon angioplasty
Stenting
Surgical patch enlargement
Lung transplant [9]

Notes

[1] Due to rupture of thin-walled aneurysm in areas of post-stenotic dilatation
[2] At sites of jet lesions distal to obstruction
[3] Uncommon; may be due to RV myocardial ischemia or PA thrombosis
[4] Increases with inspiration
[5] Correlates with cannon A waves
[6] Increased RA pressure with R-L interatrial shunt
[7] Pulmonary regurgitation
[8] Many adult patients are referred with incorrect diagnosis of Primary Pulmonary Hypertension
[9] Severe peripheral pulmonary stenosis with large loss of lung parenchyma
[10] Dominant inherited multisystem disease involving pulmonary arteries, liver, vertebrae (butterfly), eye, facial dysmorphism
[11] Rare autosomal dominate recessive disease comprising:

Brachytelephalangism
Cartilage calcification
Characteristic physiognomy
Hearing loss
Mental retardation (mild)
Peripheral pulmonary stenosis

[12] Hereditary spastic paraplegia with progressive muscle stiffness

[13] ACC/AHA 2008 Guidelines:

> "Percutaneous interventional therapy is recommended as the treatment of choice in the management of appropriate focal branch and/or peripheral pulmonary artery stenosis with greater than 50 % diameter narrowing, an elevated RV systolic pressure greater than 50 mmHg, and/or symptoms
>
> In patients with the above indications for intervention, surgeons with training and expertise in CHD should perform operations for management of branch pulmonary artery stenosis not anatomically amenable to percutaneous interventional therapy"

Guidelines

ACC/AHA 2008 guidelines for the management of adults with congenital heart disease

J Am Coll Cardiol. 2008;52:e143–263. http://content.onlinejacc.org/article.aspx?articleid=1188032.

ESC guidelines for the management of grown-up congenital heart disease (new version 2010)

Eur Heart J. 2010;31:2915–57. http://www.escardio.org/guidelines-surveys/esc-guidelines/Pages/grown-up-congenital-heart-disease.aspx.

Patient Information

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Medlineplus

ENGLISH

http://www.nlm.nih.gov/medlineplus/ency/imagepages/9380.htm.

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Updates and More

https://clinicalguidecvd.com/pssupra

Chapter 83
Pulmonary Stenosis: Valvular

Management Keys

- Balloon valvotomy recommended for asymptomatic patients with domed pulmonary valve and peak instantaneous Doppler gradient >60 mmHg or mean Doppler gradient >40 mmHg (in association with less than moderate PV regurgitation)
- Balloon valvotomy recommended for symptomatic patients with domed PV and peak instantaneous Doppler gradient >50 mmHg or mean Doppler gradient >30 mmHg (in association with less than moderate pulmonic regurgitation)
- Surgical therapy recommended for patients with severe PS and associated hypoplastic pulmonary annulus, severe pulmonary regurgitation, subvalvular PS, or supravalvular PS; surgery also preferred for most dysplastic pulmonary valves and when there is associated severe TR or need for surgical Maze procedure
- Surgeons with training and expertise in congenital heart disease should perform operations for RVOT and pulmonary valve

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 1093
DOI 10.1007/978-1-4471-7293-2_83,

ICD-10 Code

Q22.1

Alternate Names/Abbreviation

PS

Description/Etiology

- Usually isolated lesion, occurring in approximately 7–12 % of all cases of congenital heart disease
- 80–90 % of all lesions that cause RVOT obstruction
- Inheritance rate low (1.7–3.6 %)
- Approximately 20 % have a dysplastic valve
- If part of Noonan syndrome, patients have autosomal dominant trait with variable penetrance that has been mapped to chromosome 12
- Congenital (usual)
  - Most common:
    - Isolated
    - Tetralogy Of Fallot [4]
  - Others:
    - Complete Atrioventricular Septal Defect
    - Double-Outlet RV
    - Univentricular Atrioventricular Connection
- Acquired
  - Carcinoid
  - Rheumatic Fever (rare)
- Usually presents with asymptomatic systolic murmur; sometimes with exercise intolerance

Rarely progressive when initial gradient is mild, but moderate PS can progress due to progressive valve stenosis or reactive infundibular hypertrophy
Mild forms well-tolerated

Predisposing/Comorbid Conditions

ARTERIOHEPATIC DYSPLASIA
CARCINOID SYNDROME/TUMOR
CONGENITAL RUBELLA
NOONAN SYNDROME
TETRALOGY OF FALLOT
WILLIAMS SYNDROME

Demography

Gender equal
Familial uncommon/rare

Pathophysiology

Morphologic types:

1. Typical dome-shaped:

   Narrow central opening with preserved, mobile valve mechanism
   Three rudimentary raphes are usually present without clear-cut commissures
   Pulmonary trunk dilated, mostly due to inherent medial abnormality
   Jet from stenotic valve tends to favor flow to left PA branch
   Calcification of valve sometimes occurs in older adults

2. Dysplastic pulmonary valve:

   Less common
   Leaflets poorly mobile
   Marked myxomatous thickening with no commissural fusion
   Pulmonary annulus and outflow tract may be narrowed
   Frequent component of Noonan syndrome.

3. Unicuspid or bicuspid PV:

   Usually a feature of Tetralogy of Fallot
   May or may not create significant obstruction

Other morphologic/physiologic abnormalities due to associated conditions
Severity based on peak gradient across valve:

Mild: <30 mmHg
Moderate: 30–50 mmHg
Severe: >50 mmHg.

RV pressure overload/wall stress progresses to RVH/RV failure in advanced stage

Signs/Symptoms [8]

ABDOMEN – FLUID (ASCITES)
ABDOMEN – FULLNESS
ABDOMEN – PULSATION, PRESYS
BREATHING – DIFF (DYSPNEA)
CHEST – PAIN, EFFORT (ANGINA PECTORIS)
CHEST – RV LIFT
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)
EXTREM, LOWER, BILAT – EDEMA
HEART – PULMONARY EJECTION SOUND [6]
HEART, LSB, LOWER – IMP, SYS
HEART, LSB, UPPER – MURMUR, SYS [3]
HEART, LSB, UPPER – THRILL, SYS

HEART, P2, INTENSITY – DECR/ABSENT
HEART, S2 – SINGLE [SEVERE; ONLY A2 AUDIBLE]
HEART, S2, SPLIT – WIDE
HEART, S4 RV
LIVER – ENLARGED (HEPATOMEGALY)
LIVER – PULSATION, PRESYS
NECK, JVP, A WAVE – INCR/LARGE (CANNON WAVE)
NECK, SUPRASTERNAL NOTCH – MURMUR, SYS
NECK, SUPRASTERNAL NOTCH – THRILL, SYS

Differentiation

Myxoma – Right Ventricle
Other causes of RVOT obstruction [7]

Complications

Right heart failure

Laboratory

NS

ECG [9]

P WAVE – TALL/PEAKED
QRS – RVH PATTERN
QRS, AXIS – R

Imaging [10]

CHEST X-RAY: VASC FULLNESS L>R LUNG BASE [1]

PA, MAIN, SIZE – INCR [2]
PV, PRESS – GRADIENT
PV, FLOW – REGURG
PV, LEAFLETS – THICK
PV. LEAFLETS – FUSED
PV/LEAFLETS – DOMED
RA, CHAMBER, SIZE – INCR
RV, CHAMBER, SIZE – INCR
RV, WALL THICKNESS – INCR
TV, FLOW – REGURG

Other Tests

Cardiac catheterization:

Rarely needed for diagnosis
Distinguish valvular from extravalvular stenosis

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic

NS
IE prophylaxis not indicated

Treatment: Surgical/Invasive [12]

Balloon valvotomy [5] [11]
Surgical valvotomy
Valve replacement

Prevention

NA

Course

Variable according to severity/associated defects

Notes

[1] Chen Sign, due to preferential flow to left lung in PS
[2] Dome-type PS
[3] Crescendo/decrescendo occurring later in systole with increased severity; may obscure A2
[4] When associated with Tetralogy of Fallot, most often bicuspid with variable degrees of annular hypoplasia
[5] Except patients with dysplastic valves (Noonan Syndrome)/Subvalvular Stenosis, who should receive surgical valvotomy
[6] Decreased intensity with inspiration; only right heart sound to do so
[7] Non-valvular causes of RVOT obstruction:

- Muscular Infundibular Stenosis, associated with
  - Hypertrophic Cardiomyopathy
  - Pulmonic Valve Stenosis
  - Tetralogy of Fallot (usual)
- Non-Muscular Infundibular Stenosis
  - Fibrous Tags
  - Coronary Sinus
  - Inferior Vena Cava
- Membranous Septum Aneurysm
- Sinus of Valsalva Aneurysm
- Tricuspid Valve Tissue
- Postoperative
  - Conduit Stenosis
  - Peripheral stenosis after prior arterial shunt to pulmonary arteries
  - Valvular
    - Native Valve Restenosis
    - Prosthetic Valve Stenosis

Subinfundibular Obstruction

Double-Chamber RV

Supravalvular Stenosis
Associations – syndromes:

Alagille
Pulmonary Artery Stenosis
Keutel
Rubella
Williams

Hourglass deformity at valve
Peripheral Pulmonary Artery Stenosis
Pulmonary Artery Membrane
Pulmonary Artery Stenosis

[8] Cardiac examination findings depend on stenosis severity, valve pathology, associated cardiac lesions

[9] ECG usually normal when RV systolic pressure <60 mmHg

[10] Cardiac size on CXR is normal unless there is associated cardiac lesion

[11] Balloon valvotomy:

Usually completely eliminates gradient with RVH regression
Mild pulmonary regurgitation post-procedure common/clinically insignificant
Comparable results to surgical valvotomy

[12] 2008 ACC/AHA Guideline recommendations:

- Balloon valvotomy is recommended for asymptomatic patients with a domed pulmonary valve and a peak instantaneous Doppler gradient greater than 60 mmHg or a mean Doppler gradient greater than 40 mmHg (in association with less than moderate pulmonic valve regurgitation). *(Level of Evidence: B)*
- Balloon valvotomy is recommended for symptomatic patients with a domed pulmonary valve and a peak

instantaneous Doppler gradient greater than 50 mmHg or a mean Doppler gradient greater than 30 mmHg (in association with less than moderate pulmonic regurgitation). *(Level of Evidence: C)*

- Surgical therapy is recommended for patients with severe PS and an associated hypoplastic pulmonary annulus, severe pulmonary regurgitation, subvalvular PS, or supravalvular PS. Surgery is also preferred for most dysplastic pulmonary valves and when there is associated severe TR or the need for a surgical Maze procedure. *(Level of Evidence: C)*
- Surgeons with training and expertise in CHD should perform operations for the RVOT and pulmonary valve. *(Level of Evidence: B)*

Class IIb

- Balloon valvotomy may be reasonable in asymptomatic patients with a dysplastic pulmonary valve and a peak instantaneous gradient by Doppler greater than 60 mmHg or a mean Doppler gradient greater than 40 mmHg. *(Level of Evidence: C)*
- Balloon valvotomy may be reasonable in selected symptomatic patients with a dysplastic pulmonary valve and peak instantaneous gradient by Doppler greater than 50 mmHg or a mean Doppler gradient greater than 30 mmHg. *(Level of Evidence: C)*

Class III

- Balloon valvotomy is not recommended for asymptomatic patients with a peak instantaneous gradient by Doppler less than 50 mmHg in the presence of normal cardiac output. *(Level of Evidence: C)*
- Balloon valvotomy is not recommended for symptomatic patients with PS and severe pulmonary regurgitation. *(Level of Evidence: C)*
- Balloon valvotomy is not recommended for symptomatic patients with a peak instantaneous gradient by Doppler less than 30 mmHg. *(Level of Evidence: C)*

Guidelines

ACC/AHA 2008 guidelines for the management of adults with congenital heart disease
J Am Coll Cardiol. 2008;52:e143–263. http://content.onlinejacc.org/article.aspx?articleid=1188032.

ESC guidelines for the management of grown-up congenital heart disease (new version 2010)
Eur Heart J. 2010;31:2915–57. http://www.escardio.org/guidelines-surveys/esc-guidelines/Pages/grown-up-congenital-heart-disease.aspx.

Patient Information

Images

http://www.nlm.nih.gov/medlineplus/ency/imagepages/9380.htm.

Medlineplus

ENGLISH
http://www.nlm.nih.gov/medlineplus/ency/imagepages/9380.htm.

ESPANOL
http://www.nlm.nih.gov/medlineplus/spanish/ency/article/001096.htm.

Mayo Clinic

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AHA

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Stanford Childrens

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MERK

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Professional Information

Review

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Review

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Review

Am J Med. 1949;6:24–40. http://www.sciencedirect.com/science/article/pii/0002934349900042#.

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Long-Term Outcome After Repair

Circulation. 1988;78:1150–6. http://circ.ahajournals.org/content/78/5/1150.full.pdf+html?sid=12160f6c-b471-4e49-be15-b1a85b63ddfd.

Regression of Infundibular Hypertrophy After Valvuloplasty

Am J Cardiol. 1988;62:977–9. http://www.sciencedirect.com/science/article/pii/0002914988909083.

Updates and More

https://clinicalguidecvd.com/psvalv

Chapter 84
Renal Artery Stenosis

ICD-10 Code

I70.1 Atherosclerotic
Q27.1 Congenital

Alternate Names/Abbreviation

RAS

Description/Etiology

Narrowing of renal arteries caused by many conditions (see PREDISPOSING/COMORBID CONDITIONS), most often atherosclerosis (90 % of cases)
May be present in up to 3–6 % of normotensive individuals
>40 % bilateral
In bilateral disease, may induce acute/subacute acceleration of pre-existing essential hypertension including flash pulmonary edema
Renal Artery Stenosis should be suspected in a variety of clinical situations, including:

- Hypertension onset before age 30 years and after 55 years

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 1105
DOI 10.1007/978-1-4471-7293-2_84,

- Hypertension with hypokalemia, especially when receiving thiazide diuretics
- Hypertension and abdominal bruit
- Accelerated hypertension (sudden/persistent worsening of previously controlled hypertension)
- Resistant hypertension (failure of blood-pressure control despite full doses of an appropriate three-drug regimen including a diuretic)
- Malignant hypertension (hypertension with coexistent end-organ damage, i.e., acute renal failure, flash pulmonary edema, hypertensive LV failure, Aortic Dissection, new visual or neurological disturbance, advanced retinopathy)
- New azotemia or worsening renal function after administration of an ACEI or ARB
- Unexplained hypotrophic kidney
- Unexplained renal failure [17]

Predisposing/Comorbid Conditions

ABDOMINAL AORTIC ANEURYSM
ATHEROSCLEROSIS IN OTHER CV AREAS
CAROTID ARTERY STENOSIS
CONGENITAL BANDS
CORONARY ARTERY DISEASE
DIABETES MELLITUS
FIBROMUSCULAR DYSPLASIA [5]
HYPERCOAGULATION STATES
NEUROFIBROMATOSIS
PERIPHERAL ARTERY DISEASE
RADIATION
RENAL ARTERY COMPRESSION
RENAL/AORTIC ANEURYSM
RENAL/AORTIC DISSECTION
TAKAYASU ARTERITIS
THROMBOEMBOLISM
TRAUMA
WILLIAMS SYNDROME [1]

Demography

Follows traditional atherosclerotic CV risk factors
Advanced age
Females more common

Pathophysiology

Varies by etiology but all forms have common natural history of progressive loss of renal mass and function and secondary accelerated, resistant, or malignant hypertension if stenosis not treated
Atherosclerotic form (90 % of cases) involves ostium/proximal 1/3 of renal artery
Hypertension primarily due to activation of RAAS in hypo-perfused kidneys
LVH more common than in Essential Hypertension [18]

Signs/Symptoms

ABDOMEN – BRUIT [4]
ABDOMEN, LUQ – BRUIT [3] [4]
ABDOMEN, RUQ – BRUIT [3] [4]
BLOOD PRESSURE, ARTERIAL – INCREASED/ELEVATED [13]
FLANK – BRUIT [3] [4]

Differentiation

Other causes of abdominal bruit
Other causes of resistant/accelerated Hypertension

Complications [2]

Aortic Dissection
CAD

Flash Pulmonary Edema [14]
LVH/HF
Renal Failure [17]
Resistant/malignant Hypertension
Stroke

Laboratory

BLOOD RENIN/ANGIOTENSIN II LEVELS – INCR [6]
BLOOD, BUN – INCR [7]
BLOOD, CREATININE – INCREASED [7]
URINE, PROTEIN – INCR (PROTEINURIA) [11]

ECG

QRS – LVH PATTERN [2] [18]

Imaging

CT ANGIO: DIAGNOSTIC [9]
ECHO/DOPPLER SCREENING, DEGREE OF STENOSIS, PHYSIOLOGIC PATTERNS [8] [19]
MR ANGIOGRAPHY: DIAGNOSTIC [9]

Other Tests

Ambulatory BP monitoring
Angiography [10]

Treatment: Nonpharmacologic

Hypertension diet
Atherosclerosis risk modification

Treatment: Pharmacologic

Antihypertensives

- ACEIs [12]
- ARBs [12]
- CCBs

Diabetes control
Statins

Treatment: Surgical/Invasive [21]

Percutaneous angioplasty [15]
Stent [16]
Surgical revascularization [20]

Prevention

Atherosclerotic risk factor modification

Course

Data on progression of atherosclerotic RAS are inconsistent
Significant disease progression to high-grade stenosis or occlusion occurs in 1.3–11.1 % of patients
Loss of function after 2 years:

- Unilateral stenosis: 3 %
- Bilateral stenosis: 18 %
- Contralateral occlusion: 55 %

Notes

[1] Includes Supravalvular AS, PA Stenosis, RAS; "elfin" facies, teeth/jaw malformations, mental retardation, hypercalcemia, joint abnormalities, small stature, hypertension, nephrocalcinosis
[2] Incidence of many complications is disproportionate to degree of hypertension, attributed to angiotensin II inflammatory/toxic effects (eg, myocardial fibrosis, arterial medial hypertrophy, plaque rupture, endothelial cell dysfunction, smooth muscle proliferation)
[3] Depending on site of stenosis
[4] High-pitched, long, may extend into part/most of diastole
[5] Second most common cause of RAS; most often in females; involves mid-distal 2/3 of renal artery and branches
[6] Only in acute stages; normalize after a few months; NS
[7] May be normal; may increase as a sign of progressive RAS
[8] First-line screening modality; peak systolic velocity 85 % sensitivity compared to about 98 % for angiography; 92 % specificity compared to about 99 % for angiography
[9] Use contrast agents with caution with decreased renal function; MRI contrast safer but not recommended when serum creatinine clearance <30 ml/min due to risk of Nephrogenic Systemic Fibrosis
[10] Use with caution due to risk of trauma, spasm, thromboembolism
[11] Advanced disease
[12] RAAS inhibitors may transiently decrease GFR; contraindicated in bilateral RAS and RAS in patients with single functioning kidney
[13] Resistant to standard hypertension treatment
[14] Flash pulmonary edema: dramatic form of acute HF; RAS, esp when bilateral, is comman cause
[15] Preferred treatment in cases of Fibromuscular Dysplasia

[16] Angioplasty with/without stenting indications:

>60 % obstruction/symptomatic RAS secondary to atherosclerosis.
Ostial atherosclerotic RAS
RAS with impaired renal function
Unexplained recurrent CHF or sudden pulmonary edema and preserved LV systolic function

[17] Renal failure may occur with severe bilateral RAS or unilateral RAS in a single functional kidney

[18] LVH prevalence in RAS is 79 %, compared to 46 % in patients with essential hypertension, with significant effect on morbidity and mortality

[19] Duplex ultrasonography: first-line screening modality for atherosclerotic RAS; can be applied serially to assess degree of stenosis and physiological patterns, such as flow velocities and vascular resistance

Increased peak systolic velocity in main renal artery associated with post-stenotic turbulence is most frequently used to determine relevant RAS, and corresponds to ≥60 % angiographic RAS with a sensitivity and specificity of 71–98 % and 62–98 %, respectively
Several duplex criteria should be used to identify significant (60 %) stenosis, including:

Imaging of intrarenal interlobar or segmental arteries, including calculation of side difference of intrarenal resistance index
Missing early systolic peak
Retarded acceleration
Increased acceleration time

[20] Surgical revascularization indications:

Patients undergoing aorta surgical repair
Patients with complex renal artery anatomy
Failed endovascular procedure

[21] 2013 ESC Guidelines on the diagnosis and treatment of peripheral artery diseases on renal revascularization:

"The decision regarding the potential revascularization strategy should be based on the patient's individual characteristics, such as life expectancy, co-morbidities, quality of blood pressure control, and renal function. Evidence supporting the benefit of aggressive diagnosis and timing of renal revascularization remains unclear. Among patients receiving medical therapy alone, there is the risk for deterioration of kidney function with worsening morbidity and mortality. Renal artery revascularization can provide immediate improvement in kidney function and blood pressure; however, as with all invasive interventions, it may result in mortality or substantial morbidity in a small percentage of patients. This is particularly the case for renovascular lesions that pose no immediate hazard or risk of progression. There is general consensus that renal revascularization should be performed in patients with anatomically and functionally significant RAS who present with particular clinical scenarios such as sudden onset or 'flash' pulmonary oedema or congestive heart failure with preserved left ventricular function and acute oligo-/anuric renal failure with kidney ischaemia."

Guidelines

ESC guidelines on the diagnosis and treatment of peripheral artery diseases. 2013 ESH/ESC guidelines for the management of arterial hypertension

Eur Heart J. 2011;32:2851–906. http://eurheartj.oxfordjournals.org/content/ehj/32/22/2851.full.pdf.

2013 ESH/ESC guidelines for the management of arterial hypertension

Eur Heart J. 2014;34:2159–219. http://www.escardio.org/guidelines-surveys/esc-guidelines/Pages/arterial-hypertension.aspx.

JNC 7: Seventh report of the Joint National Committee on prevention, detection, evaluation, and treatment of high blood pressure

Hypertension. 2003;42:1206–52. http://hyper.ahajournals.org/content/42/6/1206.long.

NICE: Hypertension in adults: diagnosis and management (2011)
http://www.nice.org.uk/guidance/cg127/chapter/1-recommendations#choosing-antihypertensive-drug-treatment-2.

2011 ACCF/AHA focused update of the guideline for the management of patients with peripheral artery disease (updating the 2005 guideline)
Circulation. 2011;124:2020–45. http://circ.ahajournals.org/content/124/18/2020.long.

Patient Information

Medlineplus

ENGLISH
http://www.nlm.nih.gov/medlineplus/ency/article/000204.htm.

ESPANOL
http://www.nlm.nih.gov/medlineplus/spanish/ency/article/000204.htm.

Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/renal-artery-stenosis/basics/definition/con-20036702.

MERCK

http://www.merckmanuals.com/home/SearchResults?query=Renal+Artery+Stenosis+and+Occlusion&icd9=593.81%3b440.1%3b447.3.

Cleveland Clinic

http://my.clevelandclinic.org/services/heart/disorders/renal-artery-disease.

Professional Information

Review

Mayo Clin Proc. 2011;86:649–57. http://www.mayoclinicproceedings.org/article/S0025-6196(11)60070-0/abstract.

Review

N Engl J Med. 2009;361:1972–8. http://www.nejm.org/doi/full/10.1056/NEJMcp0809200.

Diagnosis: CTA Versus Catheterization

Radiology. 2009;252:299–305. http://pubs.rsna.org/doi/abs/10.1148/radiol.2521081362.

Diagnosis: CTA Versus MRA

Ann Intern Med. 2004;141:674–82. http://annals.org/article.aspx?articleid=717920.

Treatment: Astral Trial (Revasc Versus Medical Therapy)

N Engl J Med. 2009;361:1953–62. http://www.nejm.org/doi/full/10.1056/NEJMoa0905368.

Treatment: Coral Trial (Medical Therapy Versus Stent + MED RX)

J Am Coll Cardiol. 2015;66:2487–94. http://content.onlinejacc.org/article.aspx?articleID=2473755.

Treatment: Coral Trial (Medical Therapy Versus Stent + MED RX)

N Engl J Med. 2014;370:13–22. http://www.nejm.org/doi/full/10.1056/NEJMoa1310753.

Treatment: Star Trial (Medical Therapy Versus Stent + MED RX)

Ann Intern Med. 2009;150:840–8. http://annals.org/article.aspx?articleid=744542.

Updates and More

https://clinicalguidecvd.com/ras

Chapter 85
Short QT Syndrome (SQTS)

ICD-10 Code

NS

Alternate Names/Abbreviation

SQTS

Description/Etiology

Genetically-transmitted cardiac channelopathy in structurally normal hearts associated with dysrhythmias (especially AF and VF) and SCD

Specific triggers of arrhythmic events do not occur

Associated with several mutations affecting cardiac ion channel function responsible for currents that generate cardiac action potential

Classification according to genotype: [11]

SQT1: KCNH2
SQT2: KCNQ1
SQT3: KCNJ2

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 1117
DOI 10.1007/978-1-4471-7293-2_85,

SQT4: CACNA1C
SQT5: CACNB2B

Diagnosis:

Males with QTc <330 ms and females with QTc <340 ms even when asymptomatic
Males with QTc <360 ms and females with <370 ms should be considered for this diagnosis only when symptomatic or have positive family history because these values overlap with healthy population

Predisposing/Comorbid Conditions

DYSRHYTHMIAS – ATRIAL
DYSRHYTHMIAS – VENTRICULAR

Demography

Males 2:1
Mean age of first clinical manifestation about 30 years, but occurs in all ages, from infancy to very aged

Pathophysiology

Shortening of refractory periods and increased dispersion of repolarization predispose to arrhythmias

Signs/Symptoms [1]

CARD ARREST [2]
CHEST – PALPITATIONS [3]
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)

Differentiation [6]

Acidosis
Altered autonomic tone
Digitalis
Hypercalcemia
Hyperkalemia
Hyperthermia

Complications

SCD

Laboratory

NS

ECG

DYSRHYTHMIAS – ATRIAL (PACS/OTHERS) [8]
DYSRHYTHMIAS – VENTRICULAR (PVCS/OTHERS)
J POINT-T PEAK INTERVAL – SHORT [7]
PR SEGMENT – DEPRESSED
QT INTERVAL – SHORT [4]
ST SEGMENT – ELEV [5]
ST SEGMENT – SHORT/ABSENT
T WAVE – TALL/PEAKED

Imaging

NS

Genomics [11]

CACNA1C
CACNB2b
KCNH2
KCNJ2
KCNQ1

Other Tests

Ambulatory ECG [4]
Electrophysiologic testing (controversial)
Exercise test [4]

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic [12]

Quinidine [9]
Disopyramide [9]

Treatment: Surgical/Invasive [12]

ICD [10]

Prevention

Screen family members

Course

Untreated: high mortality

Notes

[1] Often asymptomatic
[2] Often first clinical manifestation
[3] AF/flutter, PVCs
[4] Both QT length and accommodation to HR must be considered:

Persons with SQTS have constant QT values and lack adaptation to HR, with failure to prolong adequately at slower HRs and abnormal shortening during acceleration (pseudonormalization of QT interval at rapid rates)

Serial ECGs, ambulatory ECG monitoring and TST may help prevent unrecognition of SQTS patients with increased HR at baseline and can reduce wrong diagnosis in presence of sinus bradycardia since Bazett formula overcorrects QT interval at slow HRs

[5] Brugada-like, right precordial leads
[6] Causes of short QT interval listed
[7] Degree of shortening correlates with symptoms in affected persons
[8] Both AF and atrial flutter
[9] Both quinidine and disopyramide prolong QT interval but clinical efficacy unproven
[10] ICD: mainstay treatment for SQTS in symptomatic patients; data lacking to definitely support use in asymptomatic persons, especially those with negative family history
[11] Gene mutations not identified in all cases of Short QT Syndrome

[12] HRS/EHRA/APHRS Expert Consensus Statement on the Diagnosis and Management of Patients with Inherited Primary Arrhythmia Syndromes:
"The optimal strategy for primary prevention of cardiac arrest in SQTS is not clear given the lack of independent risk factors, including syncope, for cardiac arrest. Although intuitively it might seem reasonable to suggest that patients with the shortest QTc values are at highest risk, clinical data do not support this hypothesis. However, in a combined symptomatic and asymptomatic group (QTc <360 ms) QTc was the only risk factor for arrhythmic events."

Guidelines

HRS/EHRA/APHRS expert consensus statement on the diagnosis and management of patients with inherited primary arrhythmia syndromes
Heart Rhythm. 2013;10:1932–63.

Patient Information

Genetics Home Reference

http://ghr.nlm.nih.gov/condition/short-qt-syndrome.

Sads Foundation

http://www.sads.org/What-is-SADS/Short-QT-Syndrome#.VYVRRGfJB1s.

Professional Information

Review

Circulation Arrhythmia Electrophysiol. 2010;3:401–8. http://circep.ahajournals.org/content/3/4/401.full.

Review

Ann Noninvasive Electrocardiol. 2005;10:371–7. http://www.ncbi.nlm.nih.gov/pubmed/16029390.

Review

Cardiovasc Res. 2005;67:357–66. http://cardiovascres.oxfordjournals.org/content/67/3/357.long.

Review

Acta Cardiol. 2008;63:553–5. http://www.ncbi.nlm.nih.gov/pubmed/19013996.

Review: Genetics of Sudden Cardiac Death

Circulation Research. 2015;116:1919–36. http://circres.ahajournals.org/content/116/12/1919.full.

Children: Long-Term Follow-Up

J Am Coll Cardiol. 2013;61:1183–91. http://content.onlinejacc.org/article.aspx?articleID=1567306.

Clinical/Medical Genetics

Curr Opin Cardiol. 2008;23:192–8. http://www.ncbi.nlm.nih.gov/pubmed/18382206.

Early Repolarization: Prevalence

Heart Rhythm. 2010;7:647–52. http://www.ncbi.nlm.nih.gov/pubmed/20206319?dopt=Abstract.

Echocardiography

Heart Rhythm. 2016;12:2096–105. http://www.heartrhythmjournal.com/article/S1547-5271(15)00626-8/abstract.

Mutation: KCNJ2 Gene

Proc Natl Acad Sci USA. 2013;110:4291–6. http://www.pnas.org/content/110/11/4291.full.

Mutation: SQTS6 Gene

Eur Heart J. 2011;32:1077–88. http://eurheartj.oxfordjournals.org/content/32/9/1077.

Natural History

J Am Coll Cardiol. 2014;63:1300–13. http://content.onlinejacc.org/article.aspx?articleID=1789345.

Ventricular Arrhythmias in Channelopathies: Management

Circulation Arrhythmia Electrophysiol. 2015;8:221–31. http://circep.ahajournals.org/content/8/1/221.extract?etoc.

Updates and More

https://clinicalguidecvd.com/sqts

Chapter 86
Sinus Node Dysfunction

ICD-10 Code

I49.8

Alternate Names/Abbreviation

SND
BRADYCARDIA-TACHYCARDIA SYNDROME
CHRONOTROPIC INCOMPETENCE
SICK SINUS SYNDROME
SINOATRIAL DISEASE
SINOATRIAL DYSFUNCTION

Description/Etiology

Broad array of abnormalities in sinus node and atrial impulse formation and propagation, including:

- Persistent sinus bradycardia and chronotropic incompetence without identifiable causes
- Paroxysmal or persistent sinus arrest with replacement by subsidiary escape rhythms in atrium, AV junction, or ventricular myocardium

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 1125
DOI 10.1007/978-1-4471-7293-2_86,

In older persons, primary form due to senescence of sinus node and atrial muscle; same degenerative process also affects specialized conduction system, although rate of progression is slow and does not dominate clinical course

Secondary forms may occur at any age due to any condition that affects sinus node cells, such as ischemia or infarction, infiltrative disease, collagen vascular disease, surgical trauma, endocrine abnormalities, autonomic insufficiency

Bradycardia-Tachycardia Syndrome: frequent association of paroxysmal AF and sinus bradycardia or sinus bradyarrhythmias, which may oscillate suddenly from one to the other

Chronotropic incompetence: inadequate HR response to physical activity

Familial forms occur

Predisposing/Comorbid Conditions

ATRIAL FIBRILLATION
ACUTE MYOCARDIAL INFARCTION [1]
AUTOIMMUNE/CONNECTIVE TISSUE DISEASE [4]
CARDIAC AMYLOIDOSIS
CARDIAC INFILTRATIVE DISORDERS [2]
CARDIAC SURGERY [7]
CARDIOMYOPATHY – NONCOMPACTION [18]
CORONARY ARTERY DISEASE
DRUGS [5]
ELECTROLYTE ABNORMALITIES [6]
HYPOTHYROIDISM
INTRACRANIAL HYPERTENSION
MUSCULOSKELETAL DISORDERS [3]
MYOCARDITIS
OBSTRUCTIVE JAUNDICE
PARASYMPATHETIC STIMULATION
PERICARDITIS – ACUTE
STABLE ISCHEMIC HEART DISEASE

Demography

Idiopathic form: advanced age
Less common in African-Americans

Pathophysiology

Abnormal sinus node and atrial impulse formation and propagation, presumed due to senescence of sinus node and atrial muscle

Signs/Symptoms [8]

BREATHING – DIFF (DYSPNEA)
CHEST – PAIN, EFFORT (ANGINA PECTORIS)
CHEST – PALPITATIONS
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE) [14]
DIZZY/LIGHTHEADED/PRESYNCOPE
FATIGUE [19]
HEART, RATE – RAPID (TACHYCARDIA)
HEART, RATE – SLOW (BRADYCARDIA)
HEART, RATE, RESPONSE – DECR (CHRONOTROPIC INCOMPETENCE) [16]
MENTATION – CHANGES, NS
MENTATION, MEMORY – DECR (AMNESIA)
MOOD – LETHARGIC

Differentiation

Other causes of bradycardia
Other causes of syncope
Physiologic bradycardia [21]

Complications

HF
Peripheral embolism
Progressive mental deterioration
Stroke [17] [20]
Death [20]

Laboratory

NS

ECG

AV COND – 1ST DEGREE BLOCK [13]
AV COND – 2ND DEGREE BLOCK
AV COND – 3RD DEGREE BLOCK
DYSRHY – JUNCT [12]
DYSRHYTHMIAS – ATRIAL (PACS/OTHERS) [11]
DYSRHYTHMIAS – VENTRICULAR (PVCS/OTHERS) [15]
P WAVE – ABSENT INTERMITTENT [9]
QRS – LBBB/LBBB PATTERN [13]
QRS – LONG, NS [13]
QRS – RBBB/RBBB PATTERN [13]
RATE – DECREASED (SINUS BRADYCARDIA) [10]
RATE – INCREASED (SINUS TACHYCARDIA) [10]

Imaging

NS/VAR WITH COMORBID

Genomics

HCN4 [18]
MYH6
SCN5A

Other Tests

Ambulatory ECG monitoring
EP
Exercise test

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic [22]

Variable per etiology

Treatment: Surgical/Invasive

Pacing/ablation [23] [24]

Prevention

Variable per etiology

Course

Variable per etiology

Notes

[1] Especially Inferior AMI
[2] Eg, Amyloidosis, Hemochromatosis, tumor
[3] Eg, Duchenne Muscular Dystrophy, Myotonic Dystrophy, Friedreich Ataxia
[4] Eg, SLE, Scleroderma
[5] Eg, antiarrhythmics, sympatholytic antihypertensives, beta-blockers, CCBs, cimetidine, lithium, phenytoin
[6] Especially hyperkalemia
[7] Eg, Mustard procedure, ASD repair
[8] List does not include signs/symptoms due to associated conditions; often asymptommatic
[9] Due to sinus arrest or SA block
[10] Alternating Bradycardia-Tachycardia Syndrome, occurring in >50 %
[11] Especially AF, which fails to convert to sinus rhythm with cardioversion attempt, and may be a component of Bradycardia-Tachycardia Syndrome
[12] Escape rhythm
[13] Persistent or intermittent AV Block may occur
[14] Often due to prolonged asystole following abrupt end of run of PAT
[15] Especially in AMI
[16] Failure to reach appropriate HR for level of exercise
[17] Due to cerebral embolus or reduced cardiac output during Tachycardia/Bradycadia dysrhythmia in presence of atherosclerotic CVD
[18] Isolated reports of familial form with HCN4 mutation and Noncompaction Cardiomyopathy
[19] Easy fatigability due to chronotropic incompetence
[20] Associated with CHADS2/CHA2DS2-VASc score
[21] Especially athletic heart: may have resting HRs of 40–50 BPM while awake, with sleeping HRs as slow as 30 bpm, with sinus pauses or progressive sinus slowing accompanied by AV conduction delay (PR prolongation), some-

times culminating in type I second-degree AV block; distinction depends on correlating bradycardia with symptoms of cerebral hypoperfusion

[22] Treatment generally not indicated in asymptommatic persons; drugs causing brady arrhythmias should be DCd with care when possible

[23] **ACC/AHA/HRS 2008 Guidelines for Device-Based Therapy of Cardiac Rhythm Abnormalities**

Permanent Pacing in Sinus Node Dysfunction

CLASS I

1. Permanent pacemaker implantation is indicated for SND with documented symptomatic bradycardia, including frequent sinus pauses that produce symptoms. *(Level of Evidence: C)*
2. Permanent pacemaker implantation is indicated for symptomatic chronotropic incompetence. *(Level of Evidence: C)*
3. Permanent pacemaker implantation is indicated for symptomatic sinus bradycardia that results from required drug therapy for medical conditions. *(Level of Evidence: C)*

CLASS IIa

1. Permanent pacemaker implantation is reasonable for SND with heart rate less than 40 bpm when a clear association between significant symptoms consistent with bradycardia and the actual presence of bradycardia has not been documented. *(Level of Evidence: C)*
2. Permanent pacemaker implantation is reasonable for syncope of unexplained origin when clinically significant abnormalities of sinus node function are discovered or provoked in electrophysiological studies. *(Level of Evidence: C)*

CLASS IIb

1. Permanent pacemaker implantation may be considered in minimally symptomatic patients with chronic heart rate less than 40 bpm while awake. *(Level of Evidence: C)*

CLASS III

1. Permanent pacemaker implantation is not indicated for SND in asymptomatic patients. *(Level of Evidence: C)*
2. Permanent pacemaker implantation is not indicated for SND in patients for whom the symptoms suggestive of bradycardia have been clearly documented to occur in the absence of bradycardia. *(Level of Evidence: C)*
3. Permanent pacemaker implantation is not indicated for SND with symptomatic bradycardia due to nonessential drug therapy. *(Level of Evidence: C)*

[24] **2013 ESC Guidelines on cardiac pacing and cardiac resynchronization therapy**

Patients with persistent bradycardia

1. Sinus node disease. Pacing is indicated when symptoms can clearly be attributed to bradycardia. Class I
2. Sinus node disease**.** Pacing may be indicated when symptoms are likely to be due to bradycardia, even if the evidence is not conclusive. Class IIb
3. Sinus node disease. Pacing is not indicated in patients with SB which is asymptomatic or due to reversible causes. Class III

Patients with intermittent (documented) bradycardia

1. Sinus node disease (including brady-tachy form). Pacing is indicated in patients affected by sinus node disease

who have the documentation of symptomatic bradycardia due to sinus arrest or sinusatrial block. Class I
2. Intermittent asystolic paroxysmal AV block (including AF with slow ventricular conduction). Pacing is indicated in patients with intermittent/paroxysmal intrinsic third- or second degree AV block. Class I
3. Reflex asystolic syncope. Pacing should be considered in patients ≥40 years with syncopes and documented symptomatic pause/s due to sinus arrest or AV block or the combination of the two. Class IIa
4. Asymptomatic pauses (sinus arrest or AV block). Pacing should be considered in patients with history of syncope and documentation of asymptomatic pauses >6 s due to sinus arrest, sinus-atrial block or AV block. Class IIa
5. Pacing is not indicated in reversible causes of bradycardia. Class III

Guidelines

ACC/AHA/HRS 2008 Guidelines for Device-Based Therapy of Cardiac Rhythm Abnormalities

Circulation. 2008;117:e356. http://circ.ahajournals.org/content/117/21/e350.full.pdf.

2013 ESC Guidelines on cardiac pacing and cardiac resynchronization therapy

Eur Heart J. 2013;34:2285. http://eurheartj.oxfordjournals.org/content/34/29/2281.full.pdf+html?sid=0742d2bc-d672-4b4f-91c9-42e549685d2d.

Patient Information

Genetics Home Reference

http://ghr.nlm.nih.gov/condition/sick-sinus-syndrome.

Medlineplus

ENGLISH
http://www.nlm.nih.gov/medlineplus/ency/article/000161.htm.

ESPANOL
http://www.nlm.nih.gov/medlineplus/spanish/ency/article/000161.htm.

Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/sick-sinus-syndrome/basics/definition/con-20029161.

Cleveland Clinic

http://www.clevelandclinicmeded.com/medicalpubs/diseasemanagement/cardiology/cardiac-arrhythmias/.

Texas Heart Institute

http://www.texasheartinstitute.org/HIC/Topics/Cond/sicksinus.cfm.

Professional Information

Review

Circulation. 2007;115:1921–32. https://circ.ahajournals.org/content/115/14/1921.full.

Atrial Fibrillation

Europace. 2013;15:205–11. http://europace.oxfordjournals.org/content/15/2/205.full.

Atrial Remodeling

Circulation. 2004;109:1514–22. http://circ.ahajournals.org/content/109/12/1514.full.

Autoantibodies

Heart Rhythm. 2011;8:1788–95. http://www.heartrhythmjournal.com/article/S1547-5271(11)00787-9/abstract.

CHADS2/CHA2DS2-VASc Score

Heart. 2013;99:843–8. http://heart.bmj.com/content/99/12/843.abstract.

Congenital Sick Sinus Syndrome

J Clin Invest. 2003;112:1019–28. http://www.jci.org/articles/view/18062.

Heart Failure

Circulation. 2004;110:897–903. http://circ.ahajournals.org/content/110/8/897.full.

Incidence/Risk Factors

J Am Coll Cardiol. 2014;64:531–8. http://content.onlinejacc.org/article.aspx?articleID=1894673.

Mutations: SCN4 Familial/LV Noncompaction

J Am Coll Cardiol. 2014;64:757–67. http://content.onlinejacc.org/article.aspx?articleID=1898539.

Stroke Predictors

J Am Coll Cardiol. 2004;43:1617–22. http://content.onlinejacc.org/article.aspx?articleid=1135557&resultClick=3.

Syncope in Paced Patients

Heart. 2014;100:842–47. http://heart.bmj.com/content/100/11/842.abstract.

Updates and More

https://clinicalguidecvd.com/snd

Chapter 87
Sinus of Valsalva Aneurysm: (Windsock Aneurysm)

ICD-10 Code

Q25.4 [ruptured]

Alternate Names/Abbreviation

SVA
WINDSOCK ANEURYSM

Description/Etiology

Congenital and acquired dilatation of aortic wall between AV annulus and sinotubular ridge, usually protruding into adjacent cardiac chamber; rupture consists of fistula via this connection
Acquired forms due to aortic degenerative disease, infection, trauma [1] [2]

Predisposing/Comorbid Conditions

ABRUPT DECELERATION INJURY [16]
ANEURYSMS-OSTEOARTHRITIS SYNDROME [13]

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 1137
DOI 10.1007/978-1-4471-7293-2_87,

AORTIC STENOSIS – SUBVALVULAR
ATHEROSCLEROSIS IN OTHER CV AREAS [14]
ATRIAL SEPTAL DEFECT – SECUNDUM [13]
BICUSPID AORTIC VALVE [13]
CARDIAC SURGERY [16]
COARCTATION OF AORTA [13]
CORONARY ARTERY ANOMALIES [13]
CYSTIC MEDIAL NECROSIS [14]
DECELERATION TRAUMA
EBSTEIN ANOMALY [13]
EHLERS-DANLOS SYNDROME [13]
INFECTIVE ENDOCARDITIS
INTENSE PHYSICAL ACTIVITY
MARFAN SYNDROME [13]
MITRAL REGURGITATION – CHRONIC [13]
PATENT DUCTUS ARTERIOSUS [13]
SYPHILIS [15]
TETRALOGY OF FALLOT [13]
TRANSPOSITION OF GREAT ARTERIES – CORRECTED [13]
TREACHER COLLINS SYNDROME
TRICUSPID REGURGITATION [13]
TUBERCULOSIS [15]
VENTRICULAR SEPTAL DEFECT [13][17]

Demography

M 4:1
More common in Asians
Initial manifestations (eg, embolism, rupture) occur at all ages [12]

Pathophysiology

AV right coronary cusp most often involved, followed by noncoronary cusp and left coronary cusp

Unruptured (expansion) effects depend on site, including:

RVOT obstruction
LVOT obstruction
Conduction abnormalities
Coronary artery obstruction
Systemic Emboli

Hemodynamic changes with rupture depend on site; most often into:

RV (60 %) (with RV volume overload)
RA (29 %)
LA (6 %)
LV (4 %)
Pericardium (1 %)

Signs/Symptoms [19] [20]

(RUPTURED AND NONRUPTURED) [3]
ABDOMEN – FLUID (ASCITES) [7]
BREATH SOUNDS – CRACKLES (RALES) [7]
BREATHING – DIFF (DYSPNEA)
CHEST – PAIN [RUPTURE]
CHEST – PAIN, EFFORT (ANGINA PECTORIS)
CHEST – PALPITATIONS
COUGH
EXTREM, LOWER, BILAT – EDEMA
FATIGUE
HEART, LSB – MURMUR, CONT [TO-AND-FRO][4] [5]
HEART, LSB – THRILL, DIAS [5]
HEART, LSB, MID – MURMUR, DIAS [8]
HEART, LSB, MID – THRILL, SYS [5]
HEART, LV, APEX – MURMUR, DIAS [8]
HEART, RATE – RAPID (TACHYCARDIA)
HEART, RSB – MURMUR, CONT [TO-AND-FRO][4][6]
HEART, RSB – THRILL, DIAS [6]

HEART, RSB, MID – THRILL, SYS [6]
HEART, RSB, UPPER – MURMUR, DIAS
LIVER – ENLARGED (HEPATOMEGALY) [7]

Differentiation

Other causes of AV regurgitation
Other causes of heart failure
Other causes of right heart volume overload
Patent Ductus Arteriosus

Complications

(INCLUDES BOTH UNRUPTURED AND RUPTURED)
ACUTE MYOCARDIAL INFARCTION [18]
AV BLOCK
CARDIAC TAMPONADE
DYSRHYTHMIAS
HEART FAILURE
INFECTIVE ENDOCARDITIS [19]
MYOCARDIAL ISCHEMIA [18]
PERIPHERAL EMBOLISM
RV OUTFLOW OBSTRUCTION
SUDDEN DEATH
TIA/STROKE

Laboratory

NS

ECG [21]

AV COND – 3RD DEGREE BLOCK [9]
DYSRHYTHMIAS – ATRIAL (PACS/OTHERS)

DYSRHYTHMIAS – VENTRICULAR (PVCS/ OTHERS)
PR INTERVAL – LONG <1ST DEGREE BLOCK
QRS – LBBB/LBBB PATTERN
QRS – LVH PATTERN [10]
QRS – RBBB/RBBB PATTERN
QRS – RVH PATTERN [10]
QRS, AXIS – R
ST-T WAVE – ABN, NS

Imaging [11]

AV, FLOW – REGURG
CARDIOMEGALY
PERICARD – FLUID
SINUS OF VALSALVA, SIZE – INCR

Other Tests

Cardiac catheterization and coronary angiography

Treatment – Nonpharmacologic

NS

Treatment – Pharmacologic

HF protocol when applicable

Treatment – Surgical/Invasive [22]

Surgical repair of aneurysm

Surgical correction of associated lesions (most often VSD, ASD, abnormal AV)
Coronary revascularization
AV repair/replacement

Course

Variable according to severity and complexity
Usual survival without surgery: about 4 years

Notes

[1] Eg, Endocarditis, Tuberculosis, Syphilis
[2] Abrupt deceleration, surgery
[3] Unruptured: usually asymptomatic; clinical onset may be abrupt or gradual, beginning with aneurysmal expansion
[4] Primary clinical sign; loud, harsh, superficial; resembles PDA except located at 3–4th ICS or xiphisternum
[5] Rupture into RV – most cases
[6] Rupture into RA
[7] Due to HF, present in 50 % of ruptured SVAs
[8] Aortic regurgitation
[9] May occur due to impingement on AV conduction
[10] Often biventricular hypertrophy
[11] With adequate quality, echo usually suffices in assessment of aneurysm size, sinus of origin, point of termination; findings highly variable depending on aneurysm severity, location, and presence of HF
[12] Rupture uncommon before puberty
[13] Congenital form
[14] Degenerative form
[15] AV endocarditis is most common cause
[16] Traumatic form
[17] Reported in 12–78 % of patients; usually associated with aneurysm of right coronary sinus

[18] Due to coronary artery compression or emboli
[19] Symptoms of occult infection may be only manifestation
[20] Symptoms due to rupture may subside, then worsen
[21] ECG almost always abnormal with rupture
[22] Definite indications for surgery (in some asymptomatic patients, surgery may be delayed according to circumstances):

Coronary ostial obstruction
Infection
Malignant arrhythmias
Rupture

Guidelines

NS.

Patient Information

Medlineplus

ENGLISH
http://www.nlm.nih.gov/medlineplus/aneurysms.html.

ESPANOL
http://www.nlm.nih.gov/medlineplus/spanish/aneurysms.html.

Professional Information

Review

Am J Cardiol 2007;99:1159–64. http://www.sciencedirect.com/science/article/pii/S0002914907000549.

Review

Semin Thorac Cardiovasc Surg 2006;9:165–76. http://www.sciencedirect.com/science/article/pii/S1092912606000159.

Giant Unruptured Aneurysm: Angina Pectoris

Eur Heart J 2013;34:1608. http://eurheartj.oxfordjournals.org/content/34/21/1608.

Giant Unruptured Aneurysm: Aortic Regurgitation

Heart 2013;99:972. http://heart.bmj.com/content/99/13/972.2.extract.

Long-Term Outcome Post-surgery

Ann Thorac Surg 2002;73:1466–71. http://www.annalsthoracicsurgery.org/article/S0003-4975(02)03493-8/abstract.

Rupture: Case Report

Eur Heart J 2014;35:2123. http://eurheartj.oxfordjournals.org/content/35/31/2123.

Rupture: Percutaneous Vs Surg Closure

Am J Cardiol 2015;115:392–98. http://www.sciencedirect.com/science/article/pii/S0002914914020876.

Rupture to Pericardium

Int Cardiovasc Res J 2014;8:74–7. http://www.ncbi.nlm.nih.gov/pubmed/24936486.

RVOT/Complete Heart Block/LVOT Protrusion (Case Report)

J Am Coll Cardiol 2013;61:e169–e169. http://content.onlinejacc.org/article.aspx?articleID=1667421.

RVOT Obstruction/RHF (Case Report)

Eur Heart J 2014;35:2721. http://eurheartj.oxfordjournals.org/content/35/39/2721.

Updates and More

https://clinicalguidecvd.com/sva

Chapter 88
Spontaneous Coronary Artery Dissection

ICD-10 Code

I25.4

Alternate Names/Abbreviation

SCAD

Description/Etiology [4]

- Nontraumatic nonatherosclerotic coronary artery dissection in younger persons, usually females, presenting as Acute Coronary Syndrome, VF, or SCD
- May have genetic basis in persons with family history of SCAD
- Often associated with Fibromuscular Dysplasia [2]
- Likely under-diagnosed as coronary angiographic appearance may be normal

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 1147
DOI 10.1007/978-1-4471-7293-2_88,

Predisposing/Comorbid Conditions

EHLERS-DANLOS SYNDROME [TYPE 4]
EXTREME PHYSICAL ACTIVITY [1]
FIBROMUSCULAR DYSPLASIA [2]
MARFAN SYNDROME
PERIPARTUM
POSTPARTUM
PSEUDOXANTHOMA ELASTICUM
STRESS [INTENSE]

Demography

More common in females
Age range 30–50 years

Pathophysiology

Dissection of coronary artery intima/media, often with hematoma, luminal stenosis/occlusion
Most often involves LAD coronary artery; multiple arterial dissections occur in 20–25 %
Most common first presentation: AMI (STEMI or NSTEMI, including Unstable Angina)

Signs/Symptoms [3]

ABDOMEN – PAIN
BLOOD PRESSURE, ARTERIAL – INCREASED/ELEVATED
BOWEL MOVEMENTS – DIARRHEA
BREATHING – DIFF (DYSPNEA)
BREATHING – RAPID (TACHYPNEA)
CHEST – FRICTION RUB

CHEST – PAIN
CHEST – PALPITATIONS
COUGH
EXTREM, UPPER – PAIN
FACE, JAW – PAIN
FATIGUE
FEVER
HEART, LV, APEX – MURMUR, SYS
HEART, LV, APEX, IMP – FORCEFUL/SUSTAINED
HEART, LV, APEX, IMP – PRESYS
HEART, RATE – RAPID (TACHYCARDIA)
HEART, RATE – SLOW (BRADYCARDIA)
HEART, RHYTHM – IRREG
HEART, RSB, LOWER – MURMUR, SYS
HEART, S2, SPLIT – REVERSED (PARADOXICAL)
HEART, S3 LV
HEART, S4 LV
HEART, SOUNDS, INTENSITY – DECR
HICCUPS
HYPOTENSION (BLOOD PRESSURE – DECREASED/LOW)
JOINT, SHOULDER – PAIN
JOINT, WRIST – PAIN
MENTATION – CONFUSION
MENTATION – FEELING OF DOOM
MENTATION – WEAKNESS (MALAISE)
MOOD – ANXIOUS
MOOD – DEPRESSED
MOOD – RESTLESS/IRRITABLE/COMBATIVE
NAUSEA
NECK, ANT – PAIN
NECK, JVP – ELEV
SKIN, COLOR – BLUE (CYANOSIS)
SKIN, COLOR – PALE (PALLOR)
SKIN, TEMP – DECR
SWEATING – INCR (DIAPHORESIS/HYPERHIDROSIS)
THROAT – PAIN/TIGHTNESS

VOMITING (EMESIS)

Differentiation

Other causes of Acute Coronary Syndrome

Complications

SCD
VF

Laboratory [3]

BLOOD, CKMB – INCR
BLOOD, ESR – INCR
BLOOD, GLUCOSE – INCR (HYPERGLYCEMIA)
BLOOD, TROPONIN – INCR
BLOOD, WBC – INCR (LEUKOCYTOSIS)

ECG [3]

DYSRHYTHMIAS – ATRIAL (PACS/OTHERS)
DYSRHYTHMIAS – VENTRICULAR (PVCS/OTHERS)
Q WAVE – ABN
QRS, AMP – DECR
RATE – DECREASED (SINUS BRADYCARDIA)
ST SEGMENT – DEPR
ST SEGMENT – ELEV
T WAVE – INVER, ABN

Imaging

[ALSO FINDINGS OF FIBROMUSCULAR DYSPLASIA IN AFFECTED PTS]
LV, EF – DECR

LV, WALL MOTION, SEG – DECR/AKINETIC

Other Tests

Skin biopsy
Coronary angiography: dissection of single/multiple coronary arteries [6]

Treatment: Nonpharmacologic [5]

NS

Treatment: Pharmacologic [5]

NS

Treatment: Surgical/Invasive [5]

NS

Prevention

NS

Course

Long-term follow up excellent in most patients regardless of acute intervention

Notes

[1] Mainly males
[2] Mainly females

[3] Includes features of STEMI/NSTEMI
[4] Tests for Ehlers-Danlos, Pseudoxanthoma Elasticum, Fibrillin Missense Mutation, Fibrillin-1 gene transversion may be positive in some cases
[5] Insufficient data to support any standardized approach, but conservative measures and revascularization (PCI, CABG) all described
[6] Coronary artery tortuosity:

Characteristic of SCAD
Most often seen in left circumflex artery, followed by LAD, RCA
More common in nonculprit arteries
Peripartum-related SCAD may less often have coronary tortuosity
Predictor of recurrent SCAD when severe
May serve as marker or potential mechanism for SCAD

Guidelines

NS

Patient Information

AHA

http://circ.ahajournals.org/content/131/1/e3.extract?etoc.

Mayo Clinic

ENGLISH
http://www.mayoclinic.org/diseases-conditions/spontaneous-coronary-artery-dissection/basics/definition/con-20037794.

ESPANOL
http://www.mayoclinic.org/espanol.

Cleveland Clinic

http://my.clevelandclinic.org/services/heart/disorders/spontaneous-coronary-artery-dissection.

SCAD Research.Org

http://www.scadresearch.org/about/.

Professional Information

Review

Cardiovasc Diagn Ther. 2015;5:37–48. http://www.ncbi.nlm.nih.gov/pmc/articles/PMC4329168/.

Review

Am J Med. 2014;127:1160–3. http://www.sciencedirect.com/science/article/pii/S0002934314006743.

Review

Can J Cardiol. 2013;29:1027–33. http://www.sciencedirect.com/science/article/pii/S0828282X1300007X.

Review

Int J Cardiol. 2014;175:8–20. http://www.sciencedirect.com/science/article/pii/S0167527314008602.

Review

J Cardiol. 2014;63:119–22. http://www.sciencedirect.com/science/article/pii/S0914508713002207.

Case Series

Am J Cardiol. 2011;107:1590–6. http://www.sciencedirect.com/science/article/pii/S0002914911003870#.

Case Series

Circulation. 2012;126:579–88. http://circ.ahajournals.org/content/126/5/579.full?sid=f744ad4b-92e2-4475-b305-9bcd75454b76.

Classification

Catheter Cardiovasc Interv. 2014;84:1115–22. http://www.ncbi.nlm.nih.gov/pubmed/24227590.

Coronary Tortuosity

Circ Cardiovasc Interv. 2014;7:656–62. http://circinterventions.ahajournals.org/content/7/5/656.long.

Editorial

Circulation. 2012;126:667–70. http://circ.ahajournals.org/content/126/6/667.full?sid=f744ad4b-92e2-4475-b305-9bcd75454b76.

Extracoronary Vascular Abnormalities: Fibromuscular Dysplasia

Am J Cardiol. 2015;115:1672–7. http://www.ajconline.org/article/S0002-9149(15)00966-2/abstract.

Extracoronary Vascular Abnormalities

J Am Coll Cardiol. 2014;63: doi:10.1016/S0735-1097(14)62060-X. http://content.onlinejacc.org/article.aspx?articleid=1854882&resultClick=3.

Familial Ocurrences

JAMA Intern Med. Published online March 23, 2015. doi:10.1001/jamainternmed.2014.8307. http://archinte.jamanetwork.com/article.aspx?articleID=2204028&utm-source=Silverchair%20Information%20Systems&utm-medium=email&utm-campaign=ArchivesofInternalMedicine%3AOnlineFirst03%2F23%2F2015.

Imaging

J Am Coll Cardiol. 2013;61:589–9. http://content.onlinejacc.org/article.aspx?articleID=1559948.

Imaging

J Am Coll Cardiol 2013;62:350–350. http://content.onlinejacc.org/article.aspx?articleID=1691040.

Imaging

Circulation. 2006;113:e403–5. http://circ.ahajournals.org/content/113/10/e403.full?sid=f4b4b144-b136-423c-8a71-45fdb84a77b7.

Imaging

Circulation. 1999;99:721. http://circ.ahajournals.org/content/99/5/721.full?sid=f4b4b144-b136-423c-8a71-45fdb84a77b7.

Imaging: Coronary CT Angiography

Heart. 2013;99:672–3. http://heart.bmj.com/content/99/9/672.extract.

Imaging/Ultrasound: Left Main Coronary Artery

Heart. 2004;90:e39. http://heart.bmj.com/content/90/7/e39.full?sid=0432476b-943f-4df4-a992-1f43336f8096.

Long-Term Prognosis/Management

Am J Cardiol. 2015;116:66–73. http://www.ajconline.org/article/S0002-9149(15)01045-0/abstract.

Optical Coherence Tomography

J Am Coll Cardiol. 2012;59:1073–9. http://content.onlinejacc.org/article.aspx?articleID=1201201.

Postpartum with Phospholipid Antibody

Heart. 2004;90:e53. http://heart.bmj.com/content/90/9/e53.full?sid=0432476b-943f-4df4-a992-1f43336f8096.

Pregnancy

Circulation. 2014;130:1915–20. http://circ.ahajournals.org/content/130/21/1915.full.

Pregnancy Risk

J Am Coll Cardiol 2014;63(12-S). doi:10.1016/S0735-1097(14)60005-X. http://content.onlinejacc.org/article.aspx?articleid=1855046&resultClick=3.

Pregnancy: Scad During Delivery

Circulation. 2013;127:1530–5. http://circ.ahajournals.org/content/127/14/1530.full.

Two Vessel Dissection (Case Report)

Heart. 2013;99:970. http://heart.bmj.com/content/99/13/970.2.extract.

Updates and More

https://clinicalguidecvd.com/scad

Chapter 89
Stable Ischemic Heart Disease

Management Keys

- Shared decisions with patients about diagnostic and treatment choices including informing patients about options, risks, benefits, and costs
- Emergency referral for evaluation/treatment of patients with high/intermediate risk unstable angina [29][30]
- Diagnose/treat comorbid causes that may contribute to angina pectoris by increasing myocardial O_2 demand or decreasing myocardial O_2 supply [31][32][33][34]
- Individualized education plan for patients with SIHD to optimize care/promote wellness
- Assess LV function in all patients with SIHD as part of all medical, revascularization, and device-based strategies
- Perform stress test as first line of evaluation for functional capacity in patients capable of exercise and have interpretable ECG
- Perform coronary angiography based on clinical history and noninvasive test results, for both risk stratification and defining coronary artery anatomy for possible revascularization; not all patients especially those in low-moderate risk category, need coronary angiography
- Follow current ACCF/AHA GDMT for medical versus surgical management of symptom relief and prevention of AMI and death

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 1159
DOI 10.1007/978-1-4471-7293-2_89,

ICD-10 Code

I20.0

Alternate Names/Abbreviation

SIHD

Description/Etiology

Stable Ischemic Heart Disease: an established pattern of angina pectoris, a history of myocardial infarction, or presence of plaque documented by imaging (coronary arteriography, IVUS, CTA)

Etiology:

Atherosclerotic CAD (vast majority)
Coronary Artery Entrapment
Calcific Pericarditis
Coronary artery aneurysm
Coronary artery spasm
Congenital anomalies:

Anomalous origin of coronary artery
Coronary A-V fistula
Coronary Ostial Stenosis/Congenital Rubella Syndrome

Coronary (acquired) abnormalities:

Aortic Dissection
Chest radiation
Collagen-Vascular disease
Coronary Embolism
Coronary Extrinsic Compression
Kawasaki Disease
Syphilis

Trauma
Vasculitis
Vasculopathy
HIV
Transplant

Hereditary Disease:

Down Syndrome
Gargoylism
Homocystinuria
Oxaluria – Primary
Progeria
Pseudoxanthoma Elasticum

Angina Pectoris: initial manifestation in >50 % of SIHD patients [24]
Unstable Angina: new onset, or increases in frequency, intensity, duration; or occurring at rest [28]
Risk factors for SIHD:

DM
Family history of premature ischemic heart disease
History of Cerebrovascular disease
History of PAD
Hypercholesterolemia/dyslipidemia
Obesity/Metabolic Syndrome
Physical inactivity
Systemic Arterial Hypertension
Tobacco use

Comorbid Conditions [31] [32] [33] [34]

AMPHETAMINES
ANEMIA
ANXIETY/ANXIETY DISORDER
AORTIC SCLEROSIS
AORTIC STENOSIS – VALVULAR
ARTERIOVENOUS FISTULAE

ARTHRITIS
ASTHMA [7]
ATRIAL FIBRILLATION
CABG
CARDIOMYOPATHY – DILATED
CARDIOMYOPATHY – HYPERTROPHIC
CATARACT
CHRONIC KIDNEY DISEASE
CHRONIC OBSTRUCTIVE PULMONARY DISEASE (EMPHYSEMA)
COCAINE
DIABETES MELLITUS
DYSLIPIDEMIA
DYSRHYTHMIAS – ATRIAL
DYSRHYTHMIAS – VENTRICULAR
ENDOMETRIOSIS
FAMILY HX: ACUTE MYOCARDIAL INFARCTION [PRIOR]
FAMILY HX: ATHEROSCLEROSIS
GOUT
HEART FAILURE
HYPERCHOLESTEROLEMIA
HYPERGAMMAGLOBULINEMIA [8]
HYPERTENSION – SYSTEMIC ARTERIAL
HYPERTHERMIA
HYPERTHYROIDISM
HYPOTHYROIDISM
HYPOXEMIA
INTERSTITIAL PULMONARY FIBROSIS [7]
LEUKEMIA [8]
MICROVASCULAR DISEASE
OBSTRUCTIVE SLEEP APNEA
PERIPHERAL ARTERY DISEASE
PHEOCHROMOCYTOMA
PNEUMONIA – COMMUNITY-ACQUIRED [7]
POLYCYTHEMIA [8]
PORCELAIN AORTA [36]

PULMONARY ARTERIAL HYPERTENSION [7]
PULMONARY EMBOLISM
SICKLE CELL DISEASE/TRAIT
THROMBOCYTOSIS [8]
TOBACCO USE

Demography

Global
More common in males >age 40 years
All ethnicities
Under age 75 years, females with CAD usually present with angina pectoris; males usually present with AMI
Females lag 10 years behind males in terms of first CAD presentation
Pre-menopausal females: low risk of serious CAD manifestations such as AMI
Decreasing mortality and morbidity from CAD since 1975 due to improved preventive measures

Pathophysiology

Myocardial ischemia occurs due to myocardial O_2 demand beyond coronary blood supply, including either or combined:
  Increased myocardial O_2 demand (eg, exercise)
  Insufficient blood supply (eg, CAD)
Most common cause of angina: atherosclerotic plaque obstruction of epicardial coronary arteries
Stable coronary plaques characterized by thick fibrous capsule and calcification; in angina, pathology involves progressive luminal narrowing without plaque rupture (AMI usually caused by rupture of unstable plaques and subsequent acute thrombosis)

Signs/Symptoms

ABDOMEN – PAIN [25]
BLOOD PRESSURE, ARTERIAL – INCREASED/ELEVATED
BREATH SOUNDS – CRACKLES (RALES)
BREATHING – DIFF (DYSPNEA) [25]
CHEST – PAIN [2][24][28] [EFFORT – ANGINA PECTORIS]
CHEST – PALPITATIONS [3]
CHEST, POST – PAIN, NONPLEURITIC EFFORT (ANGINA PECTORIS) [5][25]
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE) [3]
DIZZY/LIGHTHEADED/PRESYNCOPE [3]
EARS, BILAT, EARLOBE CREASE, DIAGONAL
EXTREM, UPPER – PAIN [4][25]
EXTREM, UPPER, SHOULDER – PAIN [25]
FACE, JAW – PAIN [25][26]
HEADACHE [25][26]
HEART, LV, APEX – IMP, DIFFUSE
HEART, LV, APEX – IMP, PARADOX
HEART, LV, APEX – MURMUR, SYS
HEART, S2, SPLIT – REVERSED (PARADOXICAL)
HEART, S3 LV
HEART, S4 LV
HEART, SOUNDS, INTENSITY – DECR
NECK, LAT – PAIN [25]
TEETH – PAIN [25][26]
THROAT – PAIN/TIGHTNESS [25]

Differentiation

AMI
Angina Pectoris – Unstable
Aortic Dissection
Cardiomyopathy
Cholecystitis

Congenital coronary artery anomalies
Costochondritis
Esophageal Reflux
Esophageal Spasm
Esophagitis
Fibromuscular Dysplasia
Fibrositis
Herpes Zoster [before rash]
Intervertebral Disc Disease
Myocarditis
Other causes of abdominal pain
Other causes of chest pain
Pericarditis – Acute
Psychogenic [eg, Anxiety, Hyperventilation, Panic Disorder, Depression]
Pulmonary Hypertension
Rib fracture
Sternoclavicular arthritis
Thoracic aortic aneurysm
Valvular heart disease (especially Aortic Stenosis – Valvular)

Complications

AMI
Dysrhythmias
HF
Major bleeding events
Sudden death

Laboratory

BLOOD, C-REACTIVE PROTEIN (CRP) – INCR [12]
BLOOD, CHOLESTEROL, HDL-C – DECR
BLOOD, CHOLESTEROL, LDL (LDL-C) – INCR
BLOOD, CHOLESTEROL, TOTAL – INCR

BLOOD, NT-PROBNP – INCR [12]
BLOOD, ST2 – INCR [37]
BLOOD, TGS – INCR
BLOOD, TRIGLYCERIDES – INCR
BLOOD, TROPONIN – INCR [39]

ECG

AV COND – 1ST DEGREE BLOCK [28]
AV COND – 2ND DEGREE BLOCK [28]
AV COND – 3RD DEGREE BLOCK [28]
DYSRHYTHMIAS – ATRIAL (PACS/OTHERS) [28][ESP A FIB]
DYSRHYTHMIAS – VENTRICULAR (PVCS/OTHERS)
Q WAVE – ABN [27][28]
QRS – LBBB/LBBB PATTERN [28]
QRS – LVH PATTERN [28]
QRS – RBBB/RBBB PATTERN [28]
QT/QTC INTERVAL – LONG
ST SEGMENT – DEPR
ST-T WAVE – ABN, NS
T WAVE – INVER, ABN
T WAVE – NORMALIZATION
T WAVE – TALL/PEAKED
U WAVE – NEG

Imaging

ART, CAROTID, IMT – INCR
ART, CORONARY – CALCIUM
ART, CORONARY – LESION, OBS [9]
AV, LEAFLETS – CALCIUM
AV, LEAFLETS – THICK
LV, DIAS – DYSF
LV, EF – DECR

LV, SYS – DYSF
LV, WALL MOTION – DECR
LV, WALL MOTION, SEG – DECR/AKINETIC
MYOCARD, PERFUSION – DECR

Other Tests

Exercise ECG test
Exercise test with echo/nuclear MPI [10]
Pharmacological stress test with CMR
Pharmacological stress test with echo/nuclear MPI [10]
Coronary CT angiography
Coronary calcium scoring
CMR angiography
Coronary angiography
Doppler/echo assessment for:

- Abnormalities of heart valves
- Abnormalities of myocardium
- Abnormalities of pericardium
- LV systolic and diastolic function

Treatment – Nonpharmacologic [1] [15]

Patient education
Self-monitoring [13]
Lifestyle modification, including:

- Diet
- Physical activity
- Stress/depression counseling
- Weight loss/control

Cardiac rehabilitation program
Tobacco cessation
Substance abuse counseling (when applicable)

Treatment – Pharmacologic [1] [15] [40]

Prevent AMI/death

Antiplatelet Agents [16]

ASA
Clopidogrel

Beta-Blockers
RAAS Blockers [17]

ACEIs
ARBs

Symptom (angina) relief

Beta-blockers
CCBs
NTG
Ranolazine [22]
Ivabradine
Nicorandil [18]
Trimetazidine [21] [22]

Influenza vaccine

Treatment – Surgical/Invasive [1] [15]

Revascularization to improve survival – left main coronary artery obstruction

CABG
PCI

Revascularization to improve survival – non-left main coronary artery obstruction

CABG
PCI

Revascularization to improve symptoms

CABG
PCI

Refractory angina

Coronary sinus reduction (investigational)
Enhanced external counterpulsation
Spinal cord stimulation
Transmyocardial revascularization (controversial)

Prevention

Atherosclerosis risk factor modification
Statins (primary prevention)
Control of comorbidities

DM
Systemic Arterial Hypertension

Course

CAD progression may occur in absence of changes in angina character/frequency [19]

Notes

[1] Drug and interventional treatment data and outcome measures are highly dynamic, and current guidelines should be consulted for specific therapies
[2] Frequency, duration not increasing
[3] Due to arrhythmia
[4] Ulnar aspect, left more common than right
[5] Interscapular area
[6] Younger ages may occur, especially with strong risk factors

[7] Via hypoxia, usually with underlying coronary atherosclerosis
[8] Via increased blood viscosity, with/without underlying coronary atherosclerosis
[9] Multislice CCTA: high degree of concordance with coronary angiography
[10] According to current ACC/AHA guidelines: recommended for patients with intermediate-high pretest probability of ischemic heart disease with uninterpretable ECG and at least moderate physical functioning or no disabling comorbidity; pharmacologic stress test for patients who cannot exercise and patients with baseline LBBB
[11] Gender and ethnicity NS for risk
[12] Correlates with prognosis, along with other biomarkers
[13] Eg, record home BP, glucose, calorie intake, exercise
[14] Moderate-high dose in absence of contraindications or proven adverse effects
[15] Does not include treatment for comorbid conditions, which should also be addressed for optimal outcomes [31–34]
[16] ASA 75–162 mg/day or clopidogrel 75 mg/day; clinical trials of prasugrel and ticagrelor in patients with stable CAD have not been conducted; due to major bleeding risk, caution when used beyond 1 year after acute coronary event
[17] Especially patients with Systemic Arterial Hypertension, DM, LVEF ≤40 %, CKD
[18] Nitrate derivative that may be added after beta-blockers and CCBS; not FDA-approved
[19] Risk assessment for disease progression:

Socioeconomic: advancing age, low income level
CV risk factors: tobacco use, Systemic Arterial Hypertension, Dyslipidemia, FH premature CAD, obesity, sedentary lifestyle
Coexisting medical conditions: DM, CKD, COPD, Cancer
CV comorbidities: HF, PAD, cerebrovascular disease

Psychosocial: depression, poor social support, poverty, stress
Health status: symptoms, functional capacity, QOL

[20] COURAGE Trial
[21] Anti-ischemic metabolic modulator
[22] Improves glycemic indices in pts with DM
[23] Some genetic diseases cause accelerated atherosclerosis (eg, Down Syndrome) and others may be associated with congenital coronary artery anomalies
[24] Angina pectoris: initial clinical manifestation in at least 50 % of patients with SIHD; incidence rises continuously with age in females; peaks at age 55–65 years in males, then declines; true incidence may be greater than reported; rather than pain, angina often described as squeezing, grip-like, suffocating, heaviness, tightness, pressure
[25] May occur associated with anterior chest pain or in isolation, sometimes termed "atypical angina", more common in females and persons of advanced age
[26] Headache/craniofacial angina associated with inferior wall myocardial ischemia
[27] Abnormal Q wave presence indicates prior AMI, often silent, especially in patients with DM
[28] Indicator of worse long-term outcomes and may warrant more aggressive treatment
[29] Patients with unstable angina and high risk should receive emergency evaluation and treatment; high risk features include one or more of:

Accelerating ischemic symptoms in prior 48 h
Prolonged pain (<20 min rest)
Pulmonary edema
New/worsening MR murmur
New S3 or worsening rales
Hypotension, bradycardia, tachycardia
Age >75 years
Angina at rest with transient ECG ST segment changes >0.5 mm

BBB: new/presumed new
Sustained VT
Elevated cardiac troponin or CKMB

[30] Patients with unstable angina and intermediate risk unstable angina include one or more of the following (no high risk features present):

Prior AMI, Cerebrovasc disease, PAD, CABG
Prior ASA use
Prolonged rest angina (>20 min) with high likelihood of CAD
Rest angina relieved by TNG
Nocturnal angina
New-onset/progressive class III/IV angina in prior 2 weeks without prolonged pain but intermediate/high likelihood of CAD
ECG T wave changes
ECG pathological Q waves or resting ST depression >1 mm in multiple lead groups
Slight elevation of cardiac troponin or CK-MB

[31] Noncardiac comorbid diseases that may contribute to angina pectoris by increasing myocardial O_2 demand include:

Anxiety
Arteriovenous Fistulae
Hyperthermia
Hyperthyroidism
Sympathomimetic Toxicity (eg, cocaine)
Systemic Arterial Hypertension

[32] Cardiac comorbid diseases that may contribute to angina pectoris by increasing myocardial O_2 demand include:

Aortic Stenosis
Dilated Cardiomyopathy
Hypertrophic Cardiomyopathy
Tachycardia (Supraventricular/Ventricular)

[33] Noncardiac comorbid diseases that may contribute to angina pectoris by decreasing myocardial O_2 supply include:

- Anemia
- Asthma
- Chronic Obstructive Pulmonary Disease
- Hypergammaglobulinemia
- Hyperviscosity
  - Leukemia
  - Thrombocytosis
- Hypoxemia
- Interstitial Pulmonary Fibrosis
- Obstructive Sleep Apnea
- Pneumonia
- Polycythemia
- Pulmonary Arterial Hypertension
- Sickle Cell Disease
- Sympathomimetic toxicity (cocaine use, Pheochromocytoma)

[34] Cardiac comorbidities that may contribute to angina pectoris by decreasing myocardial O_2 supply include:

- Aortic Stenosis
- Coronary microvascular disease
- Hypertrophic Cardiomyopathy
- Significant coronary arterial obstruction

[35] Rare but independent predictor of death

[36] Circumferential calcification of ascending aorta due to atherosclerosis; may complicate CABG

[37] Increased ST2 may be long-term predictor of mortality outcome in stable CAD

[38] GWAS studies identifying tag SNPS in loci associated with these genes indicate possible importance in patients with CAD

[39] Baseline troponin elevation associated with increased incidence in subsequent cardiac events and mortality

[40] Optimal medical therapy proven to be as efficacious as revascularization for CAD, but has low rate of utilization

Guidelines

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AHA: TNG/Exercise

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Medlineplus

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http://www.nlm.nih.gov/medlineplus/angina.html.

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http://www.nlm.nih.gov/medlineplus/spanish/angina.html.

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http://ghr.nlm.nih.gov/glossary=angina.

Cleveland Clinic

http://my.clevelandclinic.org/heart/disorders/cad/cadsymptoms.aspx.

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AHA scientific statement: preventing and experiencing ischemic heart disease as a woman: state of the science. Circulation. 2016;133:1302–31. http://circ.ahajournals.org/content/133/13/1302.full.

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Circ Cardiovasc Genet. 2015;8:216–42. http://circgenetics.ahajournals.org/content/8/1/216.

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CABG: Optimizing Outcomes

Eur Heart J. 2013;34:2873–86. http://eurheartj.oxfordjournals.org/content/34/37/2873.

Calcific Pericarditis

Circulation. 2013;128:e30–1. http://circ.ahajournals.org/content/128/3/e30.full.

Cardiac Symptoms Before Death due to CAD

Heart. 2013;99:938–43. http://heart.bmj.com/content/99/13/938.abstract.

Chronic Kidney Disease

Circulation. 2016;133:518–36. http://circ.ahajournals.org/content/133/5/518.full.

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Clin J Am Soc Nephrol. 2009;4:1892–900. http://cjasn.asnjournals.org/content/4/12/1892.long.

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Coronary Arterial External Compression: Giant PA Aneurysm (Case Report)

Circulation. 2013;127:1340–1. http://circ.ahajournals.org/content/127/12/1340.full.

Coronary Ostial Compression: Fibrosing Mediastinitis

J Am Coll Cardiol. 2013;62:163–4. http://content.onlinejacc.org/article.aspx?articleID=1681790.

Coronary Ostial Stenosis/Congenital Rubella Syndrome

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Coronary Sinus Aneurysm (Case Report)

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Coronary Sinus Reduction: Refractory Angina

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J Am Coll Cardiol. 2013;61:702–11. http://content.onlinejacc.org/article.aspx?articleID=1570009.

Diabetes Mellitus: Intensive Glucose Control (ADA/ACCF/AHA Scientific Statement)

Circulation. 2009;119:351–7. http://circ.ahajournals.org/content/119/2/351.full.

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J Am Coll Cardiol. 2013;61:2038–45. http://content.onlinejacc.org/article.aspx?articleid=1666389&resultClick=3.

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Am J Med. 2014;127:905–11. http://www.sciencedirect.com/science/article/pii/S0002934314003933.

Exercise: Cardiac Rehabilitation

Am J Med. 2004;116:682–92. http://www.sciencedirect.com/science/article/pii/S0002934304001238.

Genetic CAD Risk Prediction

Eur Heart J. 2016;37:561–7. http://eurheartj.oxfordjournals.org/content/37/6/561.full?etoc.

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Circulation. 2013;128:1131–8. http://circ.ahajournals.org/content/128/10/1131.full.

Gout: Increased CAD Risk

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Hypertension: Treatment (AHA Scientific Statement)

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Ivabradine

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Ivabradine

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Ranolazine: Terisa Trial

J Am Coll Cardiol. 2013;61:2038–45. http://content.onlinejacc.org/article.aspx?articleID=1666389.

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Statins

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Statins: High-Dose Therapy

Circulation. 2013;127:2485–93. http://circ.ahajournals.org/content/127/25/2485.full.

ST2 Increase: Long-Term Predictor

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Troponin

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Updates and More

https://clinicalguidecvd.com/sihd

Chapter 90
Stroke: Ischemic

Management Keys

PREHOSPITAL
Early recognition by patient/non-professional observers of sudden: [1]

- Arm weakness
- Dizziness
- Facial weakness (facial droop)
- Severe headache
- Slurred speech
- Visual loss

Call 911 when stroke suspected [2]
Prehospital recognition of possible stroke by EMS [See APPENDIX A]
Prehospital interventions/management

- Establish IV line [5]
- Maintain O_2 saturation >94 %
- Hypotension: place head on stretcher flat/isotonic saline
- Hypertension: consult with medical control if systolic BP >220 mmHg
- Glucose <60 mg/dL: IV glucose [5]
- No delay in transport for interventions
- Obtain family/bystander information regarding:

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, DOI 10.1007/978-1-4471-7293-2_90,

- Time of symptom onset (last time patient was normal) [3]
- Seizure activity
- Trauma
- Medications/recent surgery

EMS transport to nearest Primary Stroke/Comprehensive Stroke Center

EMS advance notification of hospital [4]

HOSPITAL

Timely emergency department care with same priority as patients with AMI/serious trauma regardless of neurological deficit severity

Hospitalization for:

- Observation for changes that might prompt added treatment interventions
- Observation/decrease likelihood of bleeding post-rtPA
- Prevention of complications
- Begin long-term treatment to prevent stroke recurrence
- Begin rehabilitation

Admit (consider) to neurocritical care unit for:

- Severe neurological deficit
- Large volume infarcts with potential for significant cerebral edema
- Significant comorbidities
- Blood pressure difficult to control
- Prior IV/intraarterial recanalization interventions

Perform complete CT to rule out hemorrhage before rtPA administration [33]

Early fibrinolytic Rx within 4.5 h of last time patient known to be well [28][31]

Strict adherence to guidelines/protocols for rtPA administration/post-lysis management due to high (6 %) risk of intracranial hemorrhage

Consider intra-arterial thrombectomy for select patient subsets; this should not affect usage of rtPA [30][31]

Monitor for short/long-term complications
Early ambulation
Begin intensive speech/physical/occupational treatment as soon as patient able to participate
Lifestyle changes/medical treatment/appropriate revascularization for secondary prevention of recurrent stroke/other forms of atherosclerotic CVD

ICD-10 Code

I63.9

Alternate Names/Abbreviation

Cerebrovascular accident (CVA)
Wallenberg syndrome (Lateral medullary syndrome) [27]

Description/Etiology

Stroke: acute loss of neurological function due to abnormal brain tissue perfusion; two types:

Ischemic (87 %)
Hemorrhagic (13 %)

Classification:

Large-artery atherosclerosis

Basilar
Internal carotid
Other branches of circle of Willis
Vertebral
Vessel-vessel atheroembolism (eg, carotid to cerebral artery)

Cardiac embolism, including:

- AF
- Cardiac myxomas
- Paradoxical from venous system through congenital shunt
- Severe LV dysfunction
- Valvular fibroelastomas

Small vessel disease, often associated with vascular damage due to: [25]

- DM
- Dyslipidemia
- Systemic Arterial Hypertension
- Tobacco use

Stroke of other determined etiology, including:

- Coagulopathies
- Genetic disease
- Metabolic disease
- Vasculopathies

Stroke of undetermined etiology (diagnosis of exclusion)

Predisposing/Comorbid Conditions

ANKYLOSING SPONDYLITIS
AORTIC DISSECTION
ATRIAL FIBRILLATION [34]
ATRIOVENTRICULAR HEART BLOCK
CARDIOMYOPATHY – DILATED
CARDIOMYOPATHY – HYPERTROPHIC
CAROTID ARTERY STENOSIS
COCAINE
CORONARY ARTERY DISEASE
DIABETES MELLITUS
DYSLIPIDEMIA
FABRY DISEASE [MAY OCCUR AT YOUNG AGE]

FH: STROKE
GOUT
HEART FAILURE
HERPES VIRUS INFECTION [36]
HYPERCOAGULATION STATES
HYPERTENSION – SYSTEMIC ARTERIAL
INFECTIVE ENDOCARDITIS
MYXOMA – LEFT ATRIUM
MYXOMA – LEFT VENTRICLE
PERIPHERAL ARTERY DISEASE
PSORIASIS
RHEUMATIC DISEASES [37]
RHEUMATOID ARTHRITIS
SICKLE CELL DISEASE/TRAIT
SYSTEMIC LUPUS ERYTHEMATOSUS
TAKAYASU ARTERITIS
TOBACCO USE

Demography

Increased in persons:

Age >55 years
Blacks
Males

Pathophysiology

Ischemic stroke: obstruction/occlusion of:

Anterior cerebral artery

Frontal pole
Mesial frontal pole

Anterior cerebellar artery

Lateral pontine

Middle cerebral artery

Parietal lobe
Posterior frontal lobe
Temporal lobe

Posterior cerebral artery

Occipital lobe

Posterior inferior cerebellar artery [27]

Lateral medulla

Vertebral artery [27]

Lateral medulla
Medial medulla

Signs/Symptoms [Appendix A]

BLOOD PRESSURE, ARTERIAL – INCREASED/ELEVATED [15]
COGNITION – DEFECT, NS
COGNITION, COMMAND RESPONSE – DECR
CONSCIOUSNESS – ALTERED [DROWSY/OBTUNDED]
CONSCIOUSNESS – LOSS, PROLONGED (COMA)
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE) [38]
DIZZY/LIGHTHEADED/PRESYNCOPE [38]
EARS, BILAT, EARLOBE CREASE, DIAGONAL
EXTREM – NUMB, FOCAL
EXTREM, UNILAT, SENSORY – DECR/ABSENT
EXTREM, UPPER – ARM DRIFT [29]
EYES, GAZE – DECR/ABS (GAZE PALSY)
EYES, MOTION – DECR/PARALYZED (OPHTHALMOPLEGIA)
EYES, MOTION – JERKY (NYSTAGMUS)
EYES, VISION – DECR/LOSS
FACE – HORNER SYNDROME
FACE – PAIN (UNILATERAL)

FACE – SENSORY (PAIN/TEMP) – DECR/ABSENT
FACE, MUSCLES, UNILAT – WEAK/PARALYZED (FACIAL DROOPING)
FALL [38]
FEVER [14]
GAIT – UNSTEADY (ATAXIA)
HEADACHE
HICCUPS
HYPOTENSION (BLOOD PRESSURE – DECREASED/LOW) [20] RARE
MENTATION – ATTENTION – IMPAIRED/ABSENT
MENTATION – CONFUSION [26]
MENTATION – DISORIENTED [26] [38]
MENTATION, CALCULATING – ABSENT (ACALCULIA)
MENTATION, DECISION-MAKING – DECR/ABSENT (ABULIA) [ESP FRONTAL LOBE INVOLV]
MENTATION, READING – ABSENT (ALEXIA)
MENTATION, WRITING – ABSENT (AGRAPHIA)
MOOD – LETHARGIC [38]
MUSCLES – WEAK, FOCAL
MUSCLES – QUADRIPLEGIA [PONS INFARCT; PROGRESSIVE]
MUSCLES, MOVEMENT – UNCOORDINATED (ATAXIA)
MUSCLES, MOVEMENT – UNCOORDINATED (ATAXIA)
MUSCLES, MOVEMENT – UNCOORDINATED (DYSMETRIA) [38]
MUSCLES, UNILAT – WEAK/PARALYSIS (HEMIPARESIS/HEMIPLEGIA)
NAUSEA
SEIZURES [38] [ESP AT ONSET]
SPEECH – DISTURBED/ABSENT (DYSPHASIA/APHASIA)
SPEECH – INARTICULATE (DYSARTHRIA)
SWALLOWING – DIFFICULT (DYSPHAGIA)

TONGUE – WEAKNESS [ESP VERTEBRAL/ MEDULLA]
VOMITING (EMESIS)

Differentiation

CNS abscess [6]
CNS tumor [7]
Conversion disorder
Drug toxicity, including:

- Carbamazine
- Lithium
- Phenytoin

Electrolyte disturbance
Hypertensive encephalopathy [8]
Hypoglycemia [9]
Hyperglycemia
Meningitis/encephalitis
Migraine/auras
Multiple sclerosis exacerbation
Other causes of acute focal neurological deficits
Other causes of coma
Other causes of seizures
Peripheral Vertigo
Psychogenic [11]
Seizure
Sepsis
Subdural hemorrhage
Wernickes Encephalopathy [10]

Complications

Acute Pulmonary Embolism
AMI
Bleeding post-rtPA
Cerebral edema/brain herniation
Cognitive decline

Decubitus ulcer
DVT
GI ulcers/bleeding
Hemorrhagic transformation of infarct
Pneumonia/aspiration pneumonitis
Seizures
Spasticity
Stroke progression/expansion/recurrence

Laboratory

BLOOD, GLUCOSE – DECR (HYPOGLYCEMIA) [16] [RARE]
BLOOD, GLUCOSE – INCR (HYPERGLYCEMIA) [17][COMMON]

Imaging [12][13][33]

ART, BASILAR, FLOW – OBS [32]
ART, BASILAR, LESION – OBS [32]
ART, CAROTID, FLOW – OBS
ART, CEREBRAL, FLOW – OBS [32]
ART, CEREBRAL, LESION – OBS [32]
ART, VERTEBRAL, FLOW – OBS
ART, VERTEBRAL, LESION – OBS
BRAIN, CEREBRUM – ISCHEMIA/INFARCT
BRAIN, CEREBRUM, REGIONAL PERFUSION – DECR/ABSENT

Other Tests

Brain angiography: superior to noninvasive imaging for:

Arterial narrowing at specific sites
Identifying nonatherosclerotic disease, including:

- Dissection
- Fibromuscular dysplasia
- Moyomoya disease
- Vasculitis

Planning surgical/endovascular procedures

Cardiac monitoring: continuous for at least 24 h

Treatment: Nonpharmacologic [24]

Endotracheal intubation:

When airway threatened
With mechanical ventilatory assist in management of elevated intracranial pressure/malignant brain edema

IV fluids: [21]

Avoid hypervolemia
Glucose [16]
Maintenance fluids only unless hypovolemic, as excess fluids may cause/exacerbate:

- Brain ischemia
- Renal insufficiency
- Thrombosis

Patient position [19]

Non-hypoxic: supine
Hypoxic: upright
Risk for airway obstruction/aspiration, possible elevated intracranial pressure: head of bed elevated 15°–30° (with frequent monitoring of airway, O_2, neurological status)

Supplemental O_2: maintain pO_2 >94 % [18]

Treatment: Pharmacologic [24]

Anticoagulants: [23]

- Unfractionated heparin
- Low molecular weight heparins/danaparoid

Antiplatelet agents/anticoagulants – oral (contraindicated during first 24 h after IV rtPA)

- ASA
- Clopidogrel

Antiplatelet agents – IV: contraindicated during first 24 h after IV rtPA

Fibrinolysis – IV rtPA [22] [28][31]

Fibrinolysis – intra-arterial (eg, alteplase): consideration for patients ineligible for IV rtPA [22] [28] [31]

Thrombin inhibitors: alternative to anticoagulants

- Dabigatran

Blood pressure control [15]

Hyperthermia: ASA/acetaminophen

Treatment: Surgical/Invasive [24]

Acute angioplasty/stenting: usefulness not established

- Extracranial
- Intracranial

Acute carotid endarterectomy: usefulness not established

Intra-arterial thrombectomy/mechanical clot disruption/extraction: [30] [31] [35]

- Merci Retrivel System
- Penumbra System
- Solitaire
- Trevo

Prevention

Primary

- Anticoagulation for patients with AF
- ASA (high risk pts)
- Carotid Artery Stenosis (asymp)
  - Aggressive treatment of all atherosclerotic risk factors
  - ASA unless contraindicated
  - Cartotid endarterectomy/angioplasty/stent considered in select patients
- Hypertension control (with stricter control in patients with DM)
- Lipid control (including statins for high risk patients, eg, those with CAD)
- Physical activity
- Tobacco cessation

Secondary

- Risk factor reduction
  - Lifestyle
  - Medications
- Anticoagulation for AF
- Carotid revascularization for symptomatic Carotid Stenosis

Course

Most functional recovery occurs within 3 months
1-year mortality after first stroke:

- Age 40–69 years: 14–24 %
- Age >69 years: 22–27 %

Notes

[1] Effective/repetitive public education about these features is essential and proven effective
[2] EMS involvement results in shorter pre-hospital delays/earlier diagnostic testing (CT/MRI)
[3] Needed information for possible fibrinolytic therapy
[4] Shortens time to be seen by ER physician/brain imaging/increased use of IV rtPA alteplase
[5] Caution: avoid excess IV fluids
[6] Suspect in patients with:

History of drug abuse
Endocarditis
Medical device implant with fever

[7] Suspect in patients with:

Gradual symptom progression
Other primary malignancy
Seizure at onset

[8] Suspect in patients with:

Cerebral edema
Cortical blindness
Delirium
Headache
Seizure
Severe hypertension

[9] Suspect in patients with:

Decreased level of consciousness
DM
Low serum glucose

[10] Suspect in patients with:

Ataxia
Confusion
History of alcohol abuse
Ophthalmoplegia

[11] Suspect in patients with:

Inconsistent physical findings
Lack of objective cranial nerve findings
Neurological findings in nonvascular distribution

[12] Brain intracranial imaging studies performed to detect:

Bleeding
Cerebral hemodynamic status
Degree of possible reversibility
Fibrinolysis contraindications
Infarct location
Infarct size
Infarct vascular distribution
Large vessel occlusion
Stroke severity

[13] Brain extracranial imaging studies performed to detect obstruction amenable to revascularization

[14] Hyperthermia present in about 1/3 and associated with poor neurological outcome; primary/secondary causes of fever should be sought, including:

Infective Endocarditis
Pneumonia
Sepsis
Urinary tract infection

[15] Increased arterial pressure common in acute stroke (>75 %), especially patients with history of hypertension; typically spontaneously decreases within 90 min of onset; extreme hypertension detrimental, causing encephalopathy, cardiac dysfunction, renal insufficiency

Optimal BP during Acute Ischemic Stroke has not been established; ideal BP range likely depends on stroke subtype and individual patient comorbidities

[16] Hypoglycemia: rare in acute stroke and should be treated (when <60 mg/dL) as soon as detected as may cause autonomic/neurological symptoms/seizures/permanent brain damage

[17] Hyperglycemia: occurs in >40 % of patients (especially those with DM); due to nonfasting state/stress; associated with worse outcomes; no evidence that targeting specific glucose levels for treatment improves outcomes and aggressive treatment carries risk of hypoglycemia, which should be avoided

[18] Hypoxia: occurs in in >60 % of pts within 48 h of onset (100 % of pts with history of cardiac/pulmonary disease); due to:

- Aspiration
- Atelectasis
- Hypoventilation
- Partial airway obstruction
- Pneumonia

[19] Patient position affects:

- Cerebral perfusion pressure
- Intracranial pressure
- O_2 saturation
- MCA mean flow velocity

[20] Hypotension rare (<1 %)/suggests another cause, incl:

- Aortic Dissection
- Cardiac dysrhythmias
- Myocardial ischemia
- Shock

[21] Isotonic solutions preferable to minimize ischemic brain edema; relation between hydration/outcomes indefinite in acute stroke

[22] Data supporting relative efficacy of rtPA versus intra-arterial treatment lacking

[23] Efficacy uncertain in this setting; potential benefits for emergency use:

- Halt neurological worsening/improve neurological outcomes
- Prevent early recurrent embolization

[24] Many agents/procedures that have been tried and have uncertain efficacy/not recommended/investigational include:

- Albumin
- Hyperbaric O_2
- Hypervolemia/hemodilution
- Hypothermia
- Induced HTN
- Mechanical flow augmentation, including:
  - Willisian/leptomeningeal collaterals
  - Extracorpreal counterpulsation
- Near-infrared therapy
- Neuroprotective agents
  - Antiinflamatory agents
  - CCBs
  - Citicoline
  - Clomethiazole
  - Free-radical trapping agents
  - Hematopoietic growth factors
  - N-methyl-aspartate agents
  - Statins
- Surgical decompression
- Vasodilatation

[25] Especially lacunar infarcts (small size: <15 mm^2) typically located in deep structures, including:

- Basal ganglia
- Internal capsule
- Pons
- Thalamus

[26] Confusion/disorientation may be real or perceived due to expressive/receptive aphasia/visuospatial neglect abnormality

[27] Wallenberg Syndrome: due to obstruction of:

Vertebral artery – distal branches
Vertebral artery – superior lateral medullary artery
Posterior inferior cerebellar artery (less common than vertebral)

Signs/symptoms include:

Ataxia
Dysphagia
Facial pain/temporary sensory loss
Hemisensory pain/temporary loss (contralateral; all others ipsilateral)
Hiccups
Hoarseness
Horner syndrome
Nausea/vomiting
Nystagmus
Vertigo

[28] Dose: 0.9 mg/kg (max dose 90 mg) over 1 h; give first 10 % as bolus over 1 min

[29] Arm drift: patient closes eyes and extends both arms straight out for 10 s

Normal: both arms move the same, or both arms do not move at all
Abnormal: one arm either does not move, or one arm drifts down compared to the other

[30] Intra-arterial thrombectomy considerations (consult latest Guidelines for most current recommendations):

Documented occlusion in distal internal carotid/proximal cerebral artery
Relatively normal noncontrast head CT
Severe neurological deficit
Can be performed within 6 h of when patient last seen to be normal
Clearly benefits patients receiving rtPA before intra-arterial thrombectomy
rtPA should not be withheld if patient meets criteria

Benefit in patients who do not receive rtPA or have rtPA exclusions requires further study
Favorable results occur when

Performed in endovascular stroke center by coordinated multidisciplinary team that extends from prehospital stage to endovascular suite

Minimizes time to recanalization
Uses stent-retriever devices
Avoids general anesthesia

[31] rtPA highly effective in recanalizing smaller distal thrombi but dissolves proximal large thrombi in only 15–25 %: reason for considering added intra-arterial embolectomy in appropriate patients
[32] However, 19–39 % of acute ischemic strokes have no identifiable intracranial occlusion
[33] Avoid imaging that may delay stroke workflow
[34] Strokes related to AF have poorer outcomes than non-AF related stroke, including worse functional impairment, recurrence, death
[35] Mechanical thrombectomy meta-analysis (2015) conclusion: In acute ischemic stroke due to large artery occlusion, mechanical thrombectomy after usual care was associated with improved functional outcomes compared with usual care alone, and was found to be relatively safe, with no excess in intracranial hemorrhage. There was a trend for reduction in all-cause mortality with mechanical thrombectomy.
[36] Herpes viruses may trigger childhood ischemic stroke, even if infection is subclinical
[37] Including Rheumatoid Arthritis, Systemic Lupus Erythematosus, Ankylosing Spondylitis, Gout, Psoriasis
[38] Symptom associated with missed diagnosis of stroke

Appendix A: National Institutes of Health Stroke Scale

IA Level of consciousness

0 – Alert
1 – Drowsy
2 – Obtunded
3 – Coma/unresponsive

IB Orientation questions

0 – Answers both correctly
1 – Answers 1 correctly
2 – Answers neither correctly

IC Response to commands

0 – Performs both tasks correctly
1 – Performs 1 task correctly
2 – Performs neither

2 Gaze

0 – Normal horizontal movements
1 – Partial gaze palsy
2 – Complete gaze palsy

3 Visual fields

0 – No visual field defect
1 – Partial hemianopia
2 – Complete hemianopia
3 – Bilateral hemianopia

4 Facial movement

0 – Normal
1 – Minor facial weakness
2 – Partial facial weakness
3 – Complete unilateral palsy

5 Motor function (arm)

a. Left
b. Right
0 – No drift
1 – Drift before 5 s
2 – Falls before 10 s
3 – No effort against gravity
4 – No movement

6 Motor function (leg)

a. Left
b. Right
0 – No drift
1 – Drift before 5 s
2 – Falls before 5 s
3 – No effort against gravity
4 – No movement

7 Limb ataxia

0 – No ataxia
1 – Ataxia in 1 limb
2 – Ataxia in 2 limbs

8 Sensory

0 – No sensory loss
1 – Mild sensory loss
2 – Severe sensory loss

9 Language

0 – Normal
1 – Mild aphasia
2 – Severe aphasia
3 – Mute or global aphasia

10 Articulation

0 – Normal
1 – Mild dysarthria
2 – Severe dysarthria

11 Extinction/inattention

0 – Absent
1 – Mild (loss 1 sensory modality lost)
2 – Severe (loss 2 modalities lost)

Guidelines

AHA/ASA: guidelines for the early management of patients with acute ischemic stroke
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Patient Information

Images

http://www.mayoclinic.org/ischemic-stroke/img-20009031.

Medlineplus

ENGLISH

https://www.nlm.nih.gov/medlineplus/ischemicstroke.html.

ESPANOL

https://www.nlm.nih.gov/medlineplus/spanish/ischemicstroke.html.

CDC: Types of Stroke

http://www.cdc.gov/stroke/types-of-stroke.htm.

Stroke Awareness Foundation

http://strokeinfo.org/.

National Stroke Association

http://www.stroke.org/understand-stroke/what-stroke/ischemic-stroke.

Merck

http://www.merckmanuals.com/home/brain-spinal-cord-and-nerve--disorders/stroke-cva/ischemic-stroke.

Cedars-Sinai

http://www.cedars-sinai.edu/Patients/Programs-and-Services/Stroke-Program/Stroke-Resources/Acute-Ischemic-Stroke.aspx.

ASA

http://www.strokeassociation.org/STROKEORG/AboutStroke/TypesofStroke/IschemicClots/Ischemic-Strokes-Clots-UCM-310939-Article.jsp.

Mayo Clinic

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Tex Heart Inst J. 2015;42:131–5. http://thij.org/doi/full/10.14503/THIJ-14-4262.

Cancer: Young Adults

Stroke.2015;46:1601–6.http://stroke.ahajournals.org/content/46/6/1601.full.

Cincinnati Prehospital Stroke Assessment

Ann Emerg Med. 1999;33:373–8. http://www.annemergmed.com/article/S0196-0644(99)70299-4/abstract.

Cocaine Use/Risk

Stroke. 2016;115.011417. http://stroke.ahajournals.org/content/early/2016/03/10/STROKEAHA.115.011417.abstract.

Cognitive Decline Post-stroke

JAMA. 2015;314:41–51. http://jama.jamanetwork.com/article.aspx?articleid=2382979.

Depression: Nursing Assessment

Stroke. 2016;47:e1–3. http://stroke.ahajournals.org/content/47/1/e1.extract?etoc.

Diabetes Mellitus: Stroke Risk/Glycemic Control

J Am Coll Cardiol. 2016;67:239–47. http://content.onlinejacc.org/article.aspx?articleID=2481278.

Earlobe Crease

Am J Cardiol. 2015;116:286–93. http://www.ajconline.org/article/S0002-9149(15)01120-0/abstract.

Herpes Virus Infection in Childhood Ischemic Stroke

Circulation. 2016;133:732–41. http://circ.ahajournals.org/content/133/8/732.full.

Imaging for Endovascular Interventions

Stroke. 2015;46:1453–61. http://stroke.ahajournals.org/content/46/6/1453.full.

Intracranial Hemorrhage After Ischemic Stroke

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Updates and More

https://clinicalguidecvd.com/stroke

Chapter 91
Supraventricular Tachycardia: (SVT/Paroxysmal Supraventicular Tachycardia/ PSVT)

ICD-10 Code

I47.1

Alternate Names/Abbreviation

SVT (Supraventricular Tachycardia): three forms:

- AVNRT: Atrioventricular Node Reentry Tachycardia (most common form)
- AVRT: Atrioventricular Reentry Tachycardia [2]
- AT: Atrial Tachycardia [20]

PSVT (Paroxysmal Supraventicular Tachycardia):

- AVNRT
- AVRT

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 1213
DOI 10.1007/978-1-4471-7293-2_91,

Description/Etiology [21]

Supraventricular Tachycardia: nonspecific term describing atrial and/or ventricular rates >100 bpm at rest; mechanism involves cardiac tissue from His bundle or above; abrupt onset and cessation; duration lasts seconds-hours [1]
May occur in isolated form or associated with structural heart disease

Predisposing/Comorbid Conditions

HYPERMETABOLIC CONDITIONS
PREEXCITATION SYNDROMES
STRUCTURAL HEART DISEASE

Demography

All ages/populations
Onset of AVRT and AVNRT most often age <20 year
AVRT: males more often
AVNRT: females 2:1
AT: frequency increases with age

Pathophysiology

Two mechanisms: triggered and reentry
Triggered activity: impulse generation dependent on a preceding action potential

Early after-depolarization, which is promoted by:

Slow heart rate
Decreased outward currents
Increased outward currents

Delayed after-depolarizations

States of intracellular Ca^{++} overload

Reentry: due to abnormal conduction, and requires:

Slow conduction
Unidirectional block

Signs/Symptoms

ABDOMEN – BELCHING
ABDOMEN – FULLNESS
ABDOMEN – PULSATION, SENSE OF
CHEST – PAIN, EFFORT (ANGINA PECTORIS) [3]
CHEST – PALPITATIONS [15]
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE) [4]
DIZZY/LIGHTHEADED/PRESYNCOPE
EXTREM, TEMP – DECR [6]
HEAD – PULSATIONS, SENSE OF
HEAD, SENSATION – FULLNESS
HEART, RATE – RAPID (TACHYCARDIA) [7]
HEART, S1, INTENSITY – INCR [DURING TACHY: AVNRT/AVRT]
HEART, S1, INTENSITY – VAR [DURING TACHY: AT]
HYPOTENSION (BLOOD PRESSURE – DECREASED/LOW) [6]
MENTATION – WEAKNESS (MALAISE)
MOOD – ANXIOUS
MOOD, COMBATIVE
MOUTH, SALIVATION – INCR (PTYALISM)
NAUSEA
NECK, JVP, A WAVE – INCR/LARGE (CANNON WAVE) [[16] DURING TACHY: AVNRT/AVRT]
NECK, SENSATION – FULLNESS
NECK, SENSATION – PULSATIONS
SWEATING – INCR (DIAPHORESIS/HYPERHIDROSIS)
URINATION – INCR (POLYURIA)
VOMITING (EMESIS) [5]

Differentiation

Wide QRS Complex: VT
Narrow QRS Complex: Fascicular VT [18]

Complications

HF [8]
Tachycardia-Induced Cardiomyopathy

Laboratory

NS

ECG [13]

P WAVE – ABSENT
P WAVE – FOLLOWS QRS [17]
P WAVE – IMBEDDED WITHIN QRS [17]
P WAVE, MORPH – VAR/ABN
QRS – LONG, NS [9]
QRS – LVH PATTERN [9]
QRS – NORMAL [9]
QRS – RBBB/RBBB PATTERN [9]
RATE, ATRIAL – RAPID [11]
ST SEGMENT – DEPR [10] [12]
T WAVE – INVER, ABN [10] [12]

Imaging

NS/VAR WITH COMORBID

Other Tests

Ambulatory ECG monitoring
EP testing

Treatment: Nonpharmacologic: AVNRT [19]

Vagal stimulation [14]
Electrical cardioversion: always warranted in presence of hemodynamic instability [22]

Treatment: Pharmacologic: AVNRT [19]

Adenosine [23]
Beta-blockers (oral) [25]
Amiodrone [26]
Diltiazem [24]
Verapamil [24]

Treatment: Surgical/Invasive [19]

RF ablation

Notes

[1] Usually does not include AF/Flutter or Multifocal Atrial Tachycardia
[2] WPW
[3] May resemble classic angina pectoris, or may be true angina due to precipitation of myocardial ischemia in patients with co-existing CAD
[4] Onset; uncommon unless associated with Sinus Node Dysfunction; occurs more often in older persons
[5] May terminate the event

[6] With prolonged episodes at very fast rates, especially with co-existing CVD
[7] 150–300 BPM; absolutely regular, unchanging with position, effort, breathing
[8] Especially when associated with structural heart disease
[9] QRS typically narrow in PSVT except in setting of BBB or antegrade preexcitation, requiring differentiation from VT; VT can have narrow complex QRS (fascicular VT) also requiring differentiation from PSVT
[10] May persist for days after cardioversion; not necessarily due to CAD
[11] Unaffected by vagal stimulation (eg, carotid massage)
[12] ST-T wave changes may occur both in presence or absence of CAD
[13] Many different ECG patterns may occur, dictated by location of originating site and conduction path
[14] **Vagal maneuvers** ***

For acute conversion of AVNRT, vagal maneuvers, including Valsalva and carotid sinus massage, can be performed quickly and should be the first-line intervention to terminate SVT. These maneuvers should be performed with the patient in the supine position. There is no "gold standard" for proper Valsalva maneuver technique, but in general, the patient raises intrathoracic pressure by bearing down against a closed glottis for 10–30 s, equivalent to at least 30–40 mmHg. Carotid massage is performed after absence of bruit has been confirmed by auscultation, by applying steady pressure over the right or left carotid sinus for 5–10 s. Another vagal maneuver based on the classic diving reflex consists of applying an ice-cold, wet towel to the face (85); in a laboratory setting, facial immersion in water at 10 °C (50 °F) has proved effective in terminating tachycardia, as well. One study involving 148 patients with SVT demonstrated that Valsalva was more successful than carotid sinus massage, and switching from one technique to the other resulted in an overall success rate of 27.7 %. The practice of applying pressure to the eyeball is potentially dangerous and has been abandoned.

[15] Palpitations with abrupt onset/termination and without clear trigger more often due to AVNT/AVRT; converse true for AT
[16] Cannon waves regular in AVNRT/AVRT; irregular in AT
[17] Short R-P tachycardia usually AVNRT/AVRT; long R-P tachycardia usually AT
[18] Conduction via specialized/rapid pathways
[19] Treatment should be determined by electrophysiology expert when possible, especially with drug therapy
[20] AT more often associated with underlying cardiac disease and has gradual onset
[21] **Definitions of supraventricular tachycardias** ***

Supraventricular tachycardia (SVT): An umbrella term used to describe tachycardias (atrial and/or ventricular rates in excess of 100 bpm at rest), the mechanism of which involves tissue from the His bundle or above. These SVTs include inappropriate sinus tachycardia, AT (including focal and multifocal AT), macroreentrant AT (including typical atrial flutter), junctional tachycardia, AVNRT, and various forms of accessory pathway-mediated reentrant tachycardias.

Paroxysmal supraventricular tachycardia (PSVT): A clinical syndrome characterized by the presence of a regular and rapid tachycardia of abrupt onset and termination. These features are characteristic of AVNRT or AVRT, and, less frequently, AT. PSVT represents a subset of SVT.

Atrial fibrillation (AF): A supraventricular arrhythmia with uncoordinated atrial activation and, consequently, ineffective atrial contraction. ECG characteristics include: (1) irregular atrial activity, (2) absence of distinct P waves, and (3) irregular R-R intervals (when atrioventricular conduction is present).

Sinus tachycardia: Rhythm arising from the sinus node in which the rate of impulses exceeds 100 bpm.

- **Physiologic sinus tachycardia**: Appropriate increased sinus rate in response to exercise and other situations that increase sympathetic tone.

- **Inappropriate sinus tachycardia:** Sinus heart rate >100 bpm at rest, with a mean 24-h heart rate >90 bpm not due to appropriate physiological responses or primary causes such as hyperthyroidism or anemia

Atrial tachycardia (AT)

- **Focal AT:** An SVT arising from a localized atrial site, characterized by regular, organized atrial activity with discrete P waves and typically an isoelectric segment between P waves. At times, irregularity is seen, especially at onset ("warm-up") and termination ("warm-down"). Atrial mapping reveals a focal point of origin.
- **Sinus node reentry tachycardia:** A specific type of focal AT that is due to microreentry arising from the sinus node complex, characterized by abrupt onset and termination, resulting in a P-wave morphology that is indistinguishable from sinus rhythm.
- **Multifocal atrial tachycardia (MAT):** An irregular SVT characterized by ≥3 distinct P-wave morphologies and/or patterns of atrial activation at different rates. The rhythm is always irregular.

Atrial flutter

- **Cavotricuspid isthmus–dependent atrial flutter**: typical: Macroreentrant AT propagating around the tricuspid annulus, proceeding superiorly along the atrial septum, inferiorly along the right atrial wall, and through the cavotricuspid isthmus between the tricuspid valve annulus and the Eustachian valve and ridge. This activation sequence produces predominantly negative "sawtooth" flutter waves on the ECG in leads 2, 3, and aVF and a late positive deflection in V1. The atrial rate can be slower than the typical 300 bpm (cycle length 200 ms) in the presence of antiarrhythmic drugs or scarring. It is also known as "typical atrial flutter" or "cavotricuspid isthmus–dependent atrial flutter" or "counterclockwise atrial flutter."

- **Cavotricuspid isthmus–dependent atrial flutter**: reverse typical: Macroreentrant AT that propagates around in the direction reverse that of typical atrial flutter. Flutter waves typically appear positive in the inferior leads and negative in V1. This type of atrial flutter is also referred to as "reverse typical" atrial flutter or "clockwise typical atrial flutter."

- **Atypical or non–cavotricuspid isthmus–dependent atrial flutter**: Macroreentrant ATs that do not involve the cavotricuspid isthmus. A variety of reentrant circuits may include reentry around the mitral valve annulus or scar tissue within the left or right atrium. A variety of terms have been applied to these arrhythmias according to the re-entry circuit location, including particular forms, such as "LA flutter" and "LA macroreentrant tachycardia" or incisional atrial re-entrant tachycardia due to re-entry around surgical scars.

Junctional tachycardia: A nonreentrant SVT that arises from the AV junction (including the His bundle).

Atrioventricular nodal reentrant tachycardia (AVNRT): A reentrant tachycardia involving two functionally distinct pathways, generally referred to as "fast" and "slow" pathways. Most commonly, the fast pathway is located near the apex of Koch's triangle, and the slow pathway inferoposterior to the compact AV node tissue. Variant pathways have been described, allowing for "slow-slow" AVNRT.

- **Typical AVNRT**: AVNRT in which a slow pathway serves as the anterograde limb of the circuit and the fast pathway serves as the retrograde limb (also called "slow-fast AVNRT").

- **Atypical AVNRT**: AVNRT in which the fast pathway serves as the anterograde limb of the circuit and a slow pathway serves as the retrograde limb (also called "fast-slow AV node reentry") or a slow pathway serves as the anterograde limb and a second slow pathway serves as the retrograde limb (also called "slow-slow AVNRT").

Accessory pathway: an accessory pathway is defined as an extranodal AV pathway that connects the myocardium of the atrium to the ventricle across the AV groove. Accessory pathways can be classified by their location, type of conduction (decremental or nondecremental), and whether they are capable of conducting anterogradely, retrogradely, or in both directions. Of note, accessory pathways of other types (such as atriofascicular, nodo-fascicular, nodo-ventricular, and fasciculoventricular pathways) are uncommon.

- **Manifest accessory pathway**: A pathway that conducts anterogradely to cause ventricular pre-excitation pattern on the ECG.
- **Concealed accessory pathway**: A pathway that conducts only retrogradely and does not affect the ECG pattern during sinus rhythm.
- **Pre-excitation pattern**: An ECG pattern reflecting the presence of a manifest accessory pathway connecting the atrium to the ventricle. Pre-excited ventricular activation over the accessory pathway competes with the anterograde conduction over the AV node and spreads from the accessory pathway insertion point in the ventricular myocardium. Depending on the relative contribution from ventricular activation by the normal AV nodal/His Purkinje system versus the manifest accessory pathway, a variable degree of pre-excitation, with its characteristic pattern of a short P-R interval with slurring of the initial upstroke of the QRS complex (delta wave), is observed. Pre-excitation can be intermittent or not easily appreciated for some pathways capable of anterograde conduction; this is usually associated with a low-risk pathway, but exceptions occur.
- **Asymptomatic pre-excitation (isolated pre-excitation)**: The abnormal pre-excitation ECG pattern in

the absence of documented SVT or symptoms consistent with SVT.

- **Wolff-Parkinson-White syndrome**: Syndrome characterized by documented SVT or symptoms consistent with SVT in a patient with ventricular pre-excitation during sinus rhythm

Atrioventricular reentrant tachycardia (AVRT): A reentrant tachycardia, the electrical pathway of which requires an accessory pathway, the atrium, atrioventricular node (or second accessory pathway), and ventricle.

- **Orthodromic AVRT:** An AVRT in which the reentrant impulse uses the accessory pathway in the retrograde direction from the ventricle to the atrium, and the AV node in the anterograde direction. The QRS complex is generally narrow or may be wide because of pre-existing bundle-branch block or aberrant conduction

- **Antidromic AVRT**: An AVRT in which the reentrant impulse uses the accessory pathway in the anterograde direction from the atrium to the ventricle, and the AV node for the retrograde direction. Occasionally, instead of the AV node, another accessory pathway can be used in the retrograde direction, which is referred to as pre-excited AVRT. The QRS complex is wide (maximally pre-excited).

Permanent form of junctional reciprocating tachycardia (PJRT): A rare form of nearly incessant orthodromic AVRT involving a slowly conducting, concealed, usually posteroseptal accessory pathway.

Pre-excited AF: AF with ventricular pre-excitation caused by conduction over ≥1 accessory pathway(s).

[22] **Synchronized cardioversion** ***

Should be performed for acute treatment in hemodynamically unstable patients with AVNRT when adenosine and vagal maneuvers do not terminate the tachycardia or are not feasible. Synchronized cardioversion is highly effective in terminating SVT (including AVRT and

AVNRT). Most stable patients with SVT respond to pharmacological therapy, with success rates of 80–98 % for agents such as verapamil, diltiazem, or adenosine. In some resistant cases, a second drug bolus or higher dose of initial drug agent is often effective. Nevertheless, in rare instances, drugs may fail to successfully restore sinus rhythm, necessitating synchronized cardioversion.

[23] **Adenosine** ***

Can be considered as both a therapeutic and diagnostic agent in narrow-complex tachyarrhythmias. It will acutely terminate AVNRT in approximately 95 % of patients and will unmask atrial activity in arrhythmias, such as atrial flutter or AT.

[24] **Diltiazem and verapamil** ***

Intravenous diltiazem and verapamil are particularly effective in converting AVNRT to sinus rhythm. These drugs should be used only in hemodynamically stable patients. It is important to ensure the absence of VT or pre-excited AF, because patients with these rhythms may become hemodynamically unstable and develop ventricular fibrillation if administered diltiazem or verapamil. Diltiazem or verapamil should also be avoided in patients with suspected systolic heart failure. Evidence for the effectiveness of beta blockers to terminate AVNRT is limited. In a trial that compared esmolol with diltiazem, diltiazem was more effective in terminating SVT (237). Nonetheless, beta blockers have an excellent safety profile, so it is reasonable to use them to attempt to terminate SVT in hemodynamically stable patients.

[25] **Oral beta-blockers** ***

Overall, there are no data specifically studying the effect of oral beta-blocker monotherapy for the acute termination of AVNRT. However, two studies have demonstrated success with the combination of oral diltiazem and propranolol to terminate AVNRT or AVRT. Oral beta blockers have an excellent safety profile, and administration (particularly in patients without intravenous access) can be performed in conjunction with vagal maneuvers.

[26] **Amiodarone** ***

Intravenous amiodarone may be considered for acute treatment in hemodynamically stable patients with AVNRT when other therapies are ineffective or contraindicated.

In a small cohort study, intravenous amiodarone was effective in terminating AVNRT. Long-term toxicity is not seen with intravenous amiodarone if given for a short period of time.

*** **Extracted verbatim from ACC/AHA/HRS 2015 Guideline for the Management of Adult Patients With Supraventricular Tachycardia: A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Rhythm Society.** J Am Coll Cardiol 2016;67:e27–e115

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J Am Coll Cardiol. 2016;67:e27–115. http://content.onlinejacc.org/article.aspx?articleID=2443667.

Patient Information

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Cleveland Clinic

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Mayo Clinic

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Johns Hopkins

http://www.hopkinsmedicine.org/heart-vascular-institute/conditions-treatments/conditions/supraventricular-tachycardia.html.

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http://www.heart.org/HEARTORG/Conditions/Arrhythmia/AboutArrhythmia/Tachycardia-Fast-Heart-Rate-UCM-302018-Article.jsp#.

Stanford

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Seattle Childrens Hospital

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Updates and More

https://clinicalguidecvd.com/svt

Chapter 92
Takayasu Arteritis

Management Keys

Prompt recognition, evaluation, and treatment [14]

ICD-10 Code

M31.4

Alternate Names/Abbreviation

Pulseless Disease

Description/Etiology

Chronic large vessel inflammatory vasculitis with protean manifestations secondary to multiorgan tissue hypoperfusion, mainly involving aorta and large branch arteries, including:

Brachiocephalic
Carotid
Coronary
Pulmonary artery and branches

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 1229
DOI 10.1007/978-1-4471-7293-2_92,

Renal
Subclavian
Vertebral

Aortic arch most common area involved
Cause unknown; cell-mediated autoimmunity likely

Predisposing/Comorbid Conditions

ACUTE FEBRILE ILLNESS [1]
MYOCARDITIS

Demography

Females 8:1 (varies among reporting countries)
Age of onset 10–40 years
Most commonly reported in Japan, SE Asia, India, Mexico [10]

Pathophysiology

Vascular injury mainly involving arterial media/adventia, leading to aneurysm formation/secondary vascular stiffening and atherosclerosis, composed of:

Myointimal proliferation
Wall thickening
Luminal Stenosis
Destruction of wall tissue leading to aneurysm formation and secondary vascular stiffening and atherosclerosis

Vascular distribution varies among populations within different reporting countries

Signs/Symptoms [13]

ABDOMEN – PAIN, AFTER MEALS (ABDOMINAL ANGINA)
APPETITE – DECR (ANOREXIA) [1]

ARTERIAL PRESSURE – LE > RE
ARTERIAL PULSE, BRACHIAL, L – DECR/ABSENT
ARTERIAL PULSE, BRACHIAL, R – DECR/ABSENT
ARTERIAL PULSE, CAROTID – DECR/ABSENT
ARTERIAL PULSE, UE – ASYMMETRIC
ARTERIES – PAIN
ARTERY, CAROTID – PAIN
ARTERY, CAROTID – TENDER
ARTERY, MULTIPLE LOCATIONS – BRUITS [12]
BLOOD PRESSURE, ARTERIAL – INCREASED/ELEVATED
BOWEL MOVEMENTS – DIARRHEA
CHEST – PAIN [1]
CHEST – PAIN, EFFORT (ANGINA PECTORIS)
CHEST, LAT – PAIN, PLEURITIC [1]
CHEST, POST – PAIN, PLEURITIC [1]
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)
DIZZY/LIGHTHEADED/PRESYNCOPE [11]
EARS, HEARING – LOSS (DEAFNESS)
EXTREM, LOWER, BILAT – PAIN, EFFORT (CLAUDICATION)
EXTREM, UPPER, USE – PAIN
EYES, LENS – OPACITY (CATARACT) [2]
EYES, RETINA – CENTRAL VENOUS RETINOPATHY
EYES, RETINA – DETACHED
EYES, VISION – ABN, NS
EYES, VISION – DECR/LOSS
FACE, JAW – PAIN, CHEWING
FACE, JAW – WEAKNESS, CHEWING
FATIGUE
FEVER [1]
HEADACHE
HEART, LSB, MID – MURMUR, DIAS
JOINTS – PAIN (ARTHRALGIA) [1]
MOUTH, PALATE – ULCERATION
MUSCLES – PAIN (MYALGIA) [1]
MUSCLES, UNILAT – WEAK (HEMIPARESIS) [3]
MUSCLES, UNILAT – WEAK/PARALYSIS (HEMIPARESIS/HEMIPLEGIA) [3]
SEIZURES
SKIN, FACE – ATROPHY

SKIN, FACE – PIGMENTATION
SKIN, NOSE – ULCER
SPEECH – ABSENT (APHONIA)/LOSS (APHASIA)
SWEATING, NOCT – INCR (DIAPHORESIS, NOCT) [1]
WEIGHT – LOSS [1]

Differentiation

Bechets Disease
Infectious aneurysms [6]
Infective Endocarditis
Other congenital and acquired causes of aortic dilatation/aortic regurgitation [4]
Other causes of arteritis involving aorta [5]

Complications [7]

AMI [8]
Aortic Dissection
Aortic Regurgitation
Giant Cell Myocarditis
HF
Premature Atherosclerosis
Renal Failure
Renovascular Hypertension
Retinopathy/Blindness
Stroke – Ischemic
Sudden Death

Laboratory

BLOOD, C-REACTIVE PROTEIN (CRP) – INCR
BLOOD, ESR – INCR
BLOOD, HGB/HCT – DECR (ANEMIA)
BLOOD, IGG – INCR
BLOOD, WBC – INCR (LEUKOCYTOSIS)

ECG

N/NS ABN

Imaging [9]

AV, LEAFLETS – THICK
IVS, THICKNESS – INCR (SEPTAL HYPERTROPHY)
LV, WALL MOTION, SEG – DECR/AKINETIC
MV, LEAFLETS – THICK
MYOCARD – SCAR(S)

Other Tests

NS

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic [14]

Antihypertensives
Biologics (investigational)

- Tocilizumab
- Tumor necrosis factor-α antagonists

Corticosteroids

Treatment: Surgical/Invasive

Discrete stenotic angioplasty and/or artery bypass [15]

Prevention

NS

Course

Prognosis improving with earlier diagnosis and treatment interventions

Main cause of death: renovascular hypertension complications

Notes

[1] Acute episode
[2] Develop rapidly
[3] Transient
[4] Eg, Fibromuscular Dysplasia, Marfan Syndrome, Ehlers-Danlos Syndrome
[5] Eg, Giant Cell Arteritis, Ankylosing Spondylitis, Reiter Syndrome, Bechet Syndrome, Psoriatic Arthritis, Relapsing Polychondritis, Systemic Lupus Erythematosus
[6] Tuberculosis, Syphilis, Staph Aureus
[7] Aneurysm rupture rare
[8] Coronary involvement in 10–13 %, including obstruction of coronary ostia/main epicardial arteries and coronary artery aneurysms
[9] Imaging: main modality for diagnosis; list does not include findings due to vascular dilatation/stenosis; imaging abnormalities of myocardium may occur in absence of ischemic symptoms
[10] Reported as more common in rainy season
[11] Especially when looking upward/looking back/raising arms
[12] Including back, neck, chest, abdomen
[13] Clinical manifestations due to tissue ischemia secondary to arterial stenoses/thrombus formation

[14] 2010 Thoracic Aortic Disease Guidelines – Sec. 7.1: Recommendations for Takayasu Arteritis and Giant Cell Arteritis

Class I

1. Initial therapy for active Takayasu arteritis and active giant cell arteritis should be corticosteroids at a high dose (prednisone 40–60 mg daily at initiation or its equivalent) to reduce the active inflammatory state. (Level of Evidence: B)
2. The success of treatment of patients with Takayasu arteritis and giant cell arteritis should be periodically evaluated to determine disease activity by repeated physical examination and either an erythrocyte sedimentation rate or C-reactive protein level. (Level of Evidence: B)
3. Elective revascularization of patients with Takayasu arteritis and giant cell arteritis should be delayed until the acute inflammatory state is treated and quiescent. (Level of Evidence: B)
4. The initial evaluation of Takayasu arteritis or giant cell arteritis should include thoracic aorta and branch vessel computed tomographic imaging or magnetic resonance imaging to investigate the possibility of aneurysm or occlusive disease in these vessels. (Level of Evidence: C)

Class IIa

1. It is reasonable to treat patients with Takayasu arteritis receiving corticosteroids with an additional anti-inflammatory agent if there is evidence of progression of vascular disease, recurrence of constitutional symptoms, or re-elevation of inflammatory marker. (Level of Evidence: C)

[15] 2010 ACCF/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/STS/SVM Guidelines for the Diagnosis and Management of Patients with Thoracic Aortic Disease

Class I

3. Elective revascularization of patients with Takayasu arteritis and giant cell arteritis should be delayed until the acute inflammatory state is treated and quiescent. (Level of Evidence: B)

Guidelines

2014 ESC guidelines on the diagnosis and treatment of aortic diseases

Eur Heart J. 2014;35:2873–926. http://eurheartj.oxfordjournals.org/content/35/41/2873.

2010 ACCF/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/STS/SVM guidelines for the diagnosis and management of patients with thoracic aortic disease.

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Patient Information

Images

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Medlineplus

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Mayo Clinic

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ACR

http://www.rheumatology.org/I-Am-A/Patient-Caregiver/Diseases-Conditions/Takayasus-Arteritis.

Johns Hopkins

http://www.hopkinsvasculitis.org/types-vasculitis/takayasus-arteritis/.

Vasculitis Foundation

http://www.vasculitisfoundation.org/education/forms/takayasus-arteritis/.

Merck

http://www.merckmanuals.com/home/bone-joint-and-muscle-disorders/vasculitic-disorders/takayasu-arteritis.

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https://my.clevelandclinic.org/services/heart/disorders/hic-Takayasus-Arteritis.

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Biologics Threapy

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Images: Inferior AMI/Dilated Cardiomyopathy

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Ischemic Stroke (Case Report)

QJM. 2016:109:45–6. http://qjmed.oxfordjournals.org/content/109/1/45.

Multimodality Imaging

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Myocarditis (Case Report)

Eur Heart J. 2015;36:2564. http://eurheartj.oxfordjournals.org/content/36/38/2564.extract?etoc.

Pathophysiology

Eur J Int Med. 2006;17:241–6. http://www.sciencedirect.com/science/article/pii/S0953620506000434#.

Updates and More

https://clinicalguidecvd.com/takayusu

Chapter 93
Tetralogy of Fallot

ICD-10 Code

Q21.3

Alternate Names/Abbreviation

TOF

Description/Etiology

Four components:

1. Subpulmonary infundibular stenosis
2. VSD
3. Aorta overriding VSD by 50 % of its diameter
4. RVH

Varying degrees of PV stenosis, supravalvular stenosis, and pulmonary artery branch stenosis may coexist

Secundum ASD present in about 10 % of patients

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 1241
DOI 10.1007/978-1-4471-7293-2_93,

Clinical presentation – unrepaired adults:

Rare in societies with access to surgical correction except among immigrants
Mild defects (pink tetralogy)
Loud precordial murmur

Clinical presentation – repaired adults

Usually asymptomatic
Hemodynamic abnormalities suggested by exertional limitations or dysrhythmias

Comorbid Conditions

ATRIAL SEPTAL DEFECT – SECUNDUM
ATRIOVENTRICULAR SEPTAL DEFECT
CORONARY ARTERY ANOMALIES
DIGEORGE SYNDROME
DOWN SYNDROME
NOONAN SYNDROME
PULMONARY ARTERY ANOMALIES
RIGHT AORTIC ARCH
VACTERL SYNDROME

Demography

Gender equal

Pathophysiology

Reduced pulmonary blood flow and R-L intracardiac shunting secondary to RVOT obstruction with:

Widely varying degrees of RVOT obstruction
Intracardiac R-L shunting causing cyanosis
Severity of cyanosis depending upon severity of outflow obstruction as well as hemoglobin

Severity of outflow tract obstruction function of fixed valve/subvalvular/supravalvular and dynamic (heart rate) components

All factors combine to determine clinical severity

Signs/Symptoms [1]

BODY, POSTURE – SQUATTING
BREATHING – DIFF (DYSPNEA)
BREATHING – RAPID (TACHYPNEA)
CHEST, ANT – MURMUR, CONT
CHEST, LAT – MURMUR, CONT
CHEST, POST – MURMUR, CONT
CONSCIOUSNESS – LOSS, SUDDEN (SYNCOPE)
HEART, LSB, MID – MURMUR, SYS
HEART, LSB, UPPER – MURMUR, DIAS
HEART, LSB, UPPER – MURMUR, SYS
HEART, P2, INTENSITY – DECR/ABSENT
HEART, S1, SPLIT – NARROW/SINGLE
HEART, S2 – PALPABLE
NECK, STERNOCLAV JOINT, R – PULSATION
SKIN, COLOR – BLUE (CYANOSIS)

Differentiation

Pulmonary Valve Stenosis
VSD

Complications

AF/Flutter
AV Block
Branch PA Stenosis or Hypoplasia
HF
Progressive AR
Residual outflow obstruction

Residual Pulmonary Regurgitation
RV dysfunction
Sudden Death
Sustained VT

Laboratory

NS

ECG

P WAVE – TALL/PEAKED
QRS – RBBB/RBBB PATTERN [2]
QRS – RVH PATTERN

Imaging

AORTA, ARCH – R
AORTA, DESCEND – R
AV, FLOW – REGURG
IVS – DEFECT
PA, MAIN – CONCAVE
PUL, VASCULARITY – DECR
PV, FLOW – REGURG
RV, OUTFLOW – OBS
TV, FLOW – REGURG

Genomics

CSX
NKX2.5
TBX5

Other Tests

Exercise test for functional capacity and exertional arrhythmias
Cardiac catheterization

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic

NS

Treatment: Surgical/Invasive

Complete repair [4]
VSD closure
Relief of RVOT obstruction [3]

Course

Variable; most TOF repair occurs in infancy/childhood but lifetime care/follow-up required
Pulmonary valve replacement ultimately indicated in most post-repair adults
Post-repair: most common problem in adults is pulmonary regurgitation, often missed on clinical examination because murmur is short/quiet and frequently missed on echo as well

Notes

[1] Post-repair patients are usually asymptomatic in absence of RV enlargement/failure and arrhythmia, with these findings:

Soft RVOT ejection systolic murmur
Pulmonary regurgitation murmur: low-pitched, late diastolic
Absent P2
Diastolic AR murmur
Pansystolic murmur from VSD patch leak
RBBB if correction made before mid-1990s (transventricular repair)

[2] Especially post-transventricular repair
[3] Including infundibular muscle resection; patch augmentation; transannular patch; extracardiac conduit; pulmonary valvotomy; pulmonary valve resection
[4] In adults, may require pulmonary valve replacement

Guidelines

ACC/AHA 2008 guidelines for the management of adults with congenital heart disease
J Am Coll Cardiol. 2008;52:e143–263. http://content.onlinejacc.org/article.aspx?articleid=1188032#tab1.

ESC guidelines for the management of grown-up congenital heart disease (new version 2010)
Eur Heart J. 2010; 31:2915–57. http://eurheartj.oxfordjournals.org/content/ehj/31/23/2915.full.pdf.

Patient Information

AHA

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Images

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http://ghr.nlm.nih.gov/glossary=tetralogyoffallot.

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http://my.clevelandclinic.org/childrens-hospital/health-info/diseases-conditions/heart/tetralogy-of-fallot-childrens-overview.aspx.

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Coronary Artery Disease

Am J Cardiol. 2009;103:1445–50. http://www.ajconline.org/article/S0002-9149(09)00473-1/abstract.

ICD Follow-Up

Circulation. 2008;117:363–70. http://circ.ahajournals.org/content/117/3/363.full?sid=d5f5835f-da5c-4dbf-951f-4b1e7beacd78.

Long-Term 40 Year Follow-Up

Circulation. 2014;130:1944–53. http://circ.ahajournals.org/content/130/22/1944.full.

Physiologic/Phenotypic Characteristics of Late Survivors

Circulation. 2013;128:1861–8. http://circ.ahajournals.org/content/128/17/1861.full?sid=d5f5835f-da5c-4dbf-951f-4b1e7beacd78.

Predictors of Death/VT – Indicator Registry

Heart. 2014;100:247–53. http://heart.bmj.com/content/100/3/247.full.

Pregnancy

Am Heart J. 2011;161:307–13. http://www.ncbi.nlm.nih.gov/pubmed/21315213.

Pulmonary Valve Replacement

Circulation. 2014;130:795–8. http://circ.ahajournals.org/content/130/9/795.full.

Pulmonary Valve Replacement

Circulation. 2013;128:1855–7. http://circ.ahajournals.org/content/128/17/1855.full.

Transcatheter Pulmonary Valve Replacement: FDA Approval of Melody Valve

http://www.fda.gov/MedicalDevices/ProductsandMedicalProcedures/DeviceApprovalsandClearances/Recently-ApprovedDevices/ucm431866.htm.

Unilateral Rib Notching (Case Report)

J Am Coll Cardiol. 2013;61:e171–e171. http://content.onlinejacc.org/article.aspx?articleID=1673113.

Updates and More

https://clinicalguidecvd.com/tof

Chapter 94
Thromboangiitis Obliterans (Buerger Disease)

Management Keys

Discontinue tobacco use is definitive treatment; even a few cigarettes a day may cause disease progression

ICD-10 Code

I73.1

Alternate Names/Abbreviation

BUERGER DISEASE

Description/Etiology

Nonatherosclerotic inflammation of small/medium arteries, veins, and nerves, most often in extremities but may also involve cerebral, coronary, renal, mesenteric, and pulmonary arteries

Typical presentation due to ischemic symptoms caused by stenosis/occlusion of distal small arteries and veins

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 1251
DOI 10.1007/978-1-4471-7293-2_94,

Involvement of both upper and lower extremities and size/location of affected vessels help distinguish from atherosclerosis

Symptoms typically begin in peripheral area of a single limb and often progress proximally and involve multiple extremities, often as intermittent claudication of feet, legs, hands, or arms

Critical limb ischemia causing rest pain, ulcerations, and digital gangrene, occur in patients with more advanced disease.

Raynaud's phenomenon is present in >40 % of patients and may be asymmetrical

Comorbid Conditions

PERIODONTAL DISEASE
SUPERFICIAL THROMBOPHLEBITIS [2]
TOBACCO USE [1]

Demography

Males more often affected
Onset age <45 years
Most prevalent in Near East and Far East countries

Pathophysiology

Three Phases:

1. Acute thrombus occlusion of lumen but not vessel wall
2. Progressive thrombus organization
3. Organized thrombus/vascular fibrosis without inflammation

Signs/Symptoms [5]

EXTREM – PAIN, SHOOTING (PARESTHESIAS)
EXTREM – RAYNAUD PHENOMENON
EXTREM, ARMS – PAIN, EFFORT/REST [4]
EXTREM, FEET – PAIN, EFFORT/REST [4]
EXTREM, HANDS – PAIN, EFFORT/REST [4]
EXTREM, LEGS – PAIN, EFFORT/REST [4]
JOINTS – PAIN (ARTHRALGIA) [3]
RAYNAUD PHENOMENON
SKIN, EXTREM – NECROSIS
SKIN, EXTREM – ULCERS
VEINS, SUPERFICIAL – HARD, CORDLIKE [2]
VEINS, SUPERFICIAL – NODULES [2]

Differentiation

ATHEROSCLEROTIC PERIPHERAL VASCULAR DISEASE
AUTOIMMUNE DISEASE
CARDIAC EMBOLI
DIABETES MELLITUS
THROMBOPHILIA

Complications

ABDOMINAL ARTERY OBSTRUCTION [6]
CEREBRAL ARTERY OBSTRUCTION [7]
CORONARY ARTERY OBSTRUCTION [8]
PREMATURE ATHEROSCLEROSIS
RENAL ARTERY OBSTRUCTION

Laboratory

NS

ECG

N/NS ABN

Imaging

NS/VAR WITH COMORBID

Other Tests

Angiography [10]
Angiography also may be indicated for excluding large artery occlusion

Treatment: Nonpharmacologic

Tobacco cessation [9]
Exercise

Treatment: Pharmacologic

Prostanoids

Treatment: Surgical/Invasive

Revascularization [12]
Amputation [11]

Prevention

Total tobacco cessation/avoidance

Course

Variable with success of smoking cessation
Risk for tissue loss/extremity amputation may approach 40 % if tobacco use continued

Notes

[1] Invariable
[2] Migratory, tender; may precede Ischemia
[3] Monoarthritis; may precede onset of disease by years
[4] Intermittent claudication and rest pain
[5] LE more often involved, usually below popliteal artery
[6] Ischemia/infarction small intestine, colon, spleen, pancreas
[7] Stroke
[8] Myocardial Ischemia/Infarction
[9] Most important and definitive; must be 100 % cessation
[10] Occlusion of distal extremity arteries with corkscrew-shaped collateral arteries (Martinelli Sign)
[11] Often UE
[12] Surgical revascularization usually not feasible due to distal/diffuse nature of disease; however, select patents with severe ischemia and suitable distal target vessels may be candidates for bypass surgery

Guidelines

Management of patients with peripheral artery disease (compilation of 2005 and 2011 ACCF/AHA guideline recommendations)
J Am Coll Cardiol. 2013;61:1555–70. http://content.onlinejacc.org/article.aspx?articleid=1659662.

ESC guidelines on the diagnosis and treatment of peripheral artery diseases
Eur Heart J. 2011; 32:2851–906. http://eurheartj.oxfordjournals.org/content/ehj/32/22/2851.full.pdf.

Patient Information

Images

http://www.nlm.nih.gov/medlineplus/ency/imagepages/18089.htm.
http://www.nlm.nih.gov/medlineplus/ency/imagepages/8747.htm.

Medlineplus

ENGLISH

http://www.nlm.nih.gov/medlineplus/ency/article/000172.htm.

ESPANOL

http://www.nlm.nih.gov/medlineplus/spanish/ency/article/000172.htm.

Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/buergers-disease/basics/definition/con-20029501.

Johns Hopkins

http://www.hopkinsvasculitis.org/types-vasculitis/buergers-disease/.

CDC

http://www.cdc.gov/tobacco/campaign/tips/diseases/buergers-disease.html.

NORD

https://rarediseases.org/rare-diseases/buergers-disease/.

UC Davis

https://www.ucdmc.ucdavis.edu/vascular/diseases/buergers-disease.html.

Vasculitis Foundation

http://www.vasculitisfoundation.org/education/forms/buergers-disease/.

Professional Information

Review/Update

Circulation. 2010;121:1858–61. http://circ.ahajournals.org/content/121/16/1858.full.

Review

N Engl J Med. 2000;343:864–9. http://www.nejm.org/doi/full/10.1056/NEJM200009213431207.

Corkscrew DSA Image

Circulation. 2007;116:e539–40. http://circ.ahajournals.org/content/116/21/e539.full?sid=8bf135ab-ce19-4511-af6f-078fbe47d1ba.

Lower Extremity Nonatherosclerotic PAD

Circulation. 2012;126:213–22. http://circ.ahajournals.org/content/126/2/213.long.

Updates and More

https://clinicalguidecvd.com/throb

Chapter 95
Tricuspid Regurgitation

ICD-10 Code

I36.1 (Nonrheumatic)
I07.1 (Rheumatic)

Alternate Names/Abbreviation

TR
TRICUSPID VALVE INSUFFICIENCY

Description/Etiology

About 80 % of cases of significant TR are functional and related to tricuspid annular dilation and leaflet tethering in setting of RV remodeling due to pressure/volume overload.

Mild degrees of TR are common in persons with normal anatomical TV and are of no physiologic significance

Most cases of TR are clinically silent and noted incidentally on physical exam or part of echo study

Primary TV disorders as etiology:

Carcinoid
Catheter-related trauma (esp Endomyocardial biopsy)

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 1259
DOI 10.1007/978-1-4471-7293-2_95,

Congenital
Infective Endocarditis
Radiation
Rheumatic
TV Prolapse

Iatrogenic causes:

Blunt chest wall trauma
RV pacemaker/ICD leads

Functional TR (80 % of cases of significant TR): due to TV annulus dilatation and leaflet tethering associated with RV remodeling/volume overload

Stages:

A At risk of TR [9]
B Progressive TR [10]
C Asymptomatic, severe TR [11]
D Symptomatic severe TR [12]

Predisposing/Comorbid Conditions

[SEE ETIOLOGY]
AORTIC STENOSIS – VALVULAR [FUNCTIONAL TR]
ATRIAL FIBRILLATION [FUNCTIONAL TR]
MITRAL STENOSIS – ACQUIRED [FUNCTIONAL TR]
PULMONARY HYPERTENSION [FUNCTIONAL TR]

Demography

Variable according to associated conditions

Pathophysiology

Most TR cases functional due to tricuspid annular dilation and leaflet tethering in setting of RV remodeling due to pressure/volume overload
TV annulus:

- Saddle-shaped ellipsoid that becomes planar and circular as it dilates in an anterior-posterior direction
- Often does not return to its normal size and configuration after relief of RV overload.

Severe TR causes:

- Increased RA size/pressure and systemic venous pressure
- RV enlargement
- Progressive TR from annular dilatation

Signs/Symptoms [1]

ABDOMEN – DISTENSION
ABDOMEN – FLUID (ASCITES)
APPETITE – DECR (ANOREXIA)
BREATHING – DIFF (DYSPNEA)
BREATHING – RAPID (TACHYPNEA)
CHEST – PALPITATIONS [3]
EXTREM, LOWER, BILAT – EDEMA
FATIGUE
HEART, LSB, LOWER – IMP, SYS
HEART, LSB, LOWER – MURMUR, DIAS
HEART, LSB, LOWER – MURMUR, SYS [2] [4]
HEART, LSB, LOWER – THRILL, SYS
HEART, S2, SPLIT – REVERSED (PARADOXICAL) [SEVERE; EARLY PV CLOSURE]
LIVER – ENLARGED (HEPATOMEGALY)
LIVER – PULSATION, SYS
NAUSEA

NECK, JVP, V WAVE – INCR/LARGE [2]
NECK, JVP, Y DESCENT – RAPID [2]
NECK, SENSATION – PULSATIONS
SPLEEN, SIZE – INCR (SPLENOMEGALY)

Differentiation

Other causes of RV volume overload/failure

Complications

Progression of TR
Chronic/significant increased systemic vascular resistance may predispose to:

- Hepatic dysfunction
- Hepatocellular carcinoma
- Protein-losing enteropathy
- Systemic venous varices

Laboratory [1]

BLOOD, BILIRUBIN – INCR
BLOOD, LIVER ENZYMES – INCREASED

ECG [1]

DYSRHYTHMIAS – ATRIAL (PACS/OTHERS)
P WAVE – TALL/PEAKED
PR INTERVAL – LONG <1ST DEGREE BLOCK
QRS, AXIS – R

Imaging [1] [6]

RA, CHAMBER, SIZE – INCR
RV, CHAMBER, SIZE – INCR
TV, ANNULUS, DIAM – INCR [5]
TV, FLOW – REGURG

Other Tests [1]

Stress test [7]
Cardiac catheterization [8]

Treatment: Nonpharmacologic [1]

NS

Treatment: Pharmacologic [1]

Diuretics

Treatment: Surgical/Invasive

Surgical correction [13]

Course

Variable per type/etiology

Notes

[1] Does not include findings or recommendations for functional TR associated abnormalities, eg, LV failure, PH

[2] Murmur may be absent even in severe TR, and jugular vein findings may be only detectable feature on physical exam
[3] Especially if AF present
[4] Increases with inspiration
[5] Correlates with TV regurgitant volume
[6] TTE (TEE when suboptimal) useful for:

- Distinguishing primary from functional TR
- Detecting left heart disease
- Estimating PA pressure

[7] Usually limited by left heart disease, but may be useful for detecting symptoms not recognized by patient in guiding earlier intervention
[8] May be indicated when TTE/TEE are inadequate; also to determine cause of PH, severity of TR, RV function
[9] Stage A:

- Primary
  - Mild rheumatic change
  - Mild prolapse
  - Other (eg, IE with vegetation, early carcinoid deposition, radiation)
  - Intra-annular RV pacemaker or ICD lead
  - Postcardiac transplant (biopsy related)
- Functional
  - Normal
  - Early annular dilation

[10] Stage B

- Primary
  - Progressive leaflet deterioration/destruction
  - Moderate-severe prolapse
  - Limited chordal rupture
- Functional
  - Early annular dilation
  - Moderate leaflet tethering

[11] Stage C

Primary

Flail or grossly distorted leaflets

Functional

Severe annular dilation (>40 mm or 21 mm/m^2)
Marked leaflet tethering

[12] Stage D

Primary

Flail or grossly distorted leaflets

Functional

Severe annular dilation (>40 mm or >21 mm/m^2)
Marked leaflet tethering

[13] Surgical correction of TR most often considered at time of MV or AV surgery; severe TR (primary or functional) may not improve after treatment of left heart valve lesions and reduction of RV afterload; thus, severe TR often should be corrected as part of index procedure

Guidelines

2014 AHA/ACC guideline for the management of patients with valvular heart disease

J Am Coll Cardiol. 2014;63:e57–185. http://content.onlinejacc.org/article.aspx?articleID=1838843.

Guidelines on the management of valvular heart disease (version 2012)

Eur Heart J. 2012;33:2478–80. http://www.escardio.org/guidelines-surveys/esc-guidelines/Pages/valvular-heart-disease.aspx.

Patient Information

Images

http://www.nlm.nih.gov/medlineplus/ency/imagepages/18091.htm.
http://www.nlm.nih.gov/medlineplus/ency/imagepages/18090.htm.

Medlineplus

ENGLISH

http://www.nlm.nih.gov/medlineplus/ency/article/000169.htm.

ESPANOL

http://www.nlm.nih.gov/medlineplus/spanish/ency/article/000169.htm.

Cleveland Clinic

http://my.clevelandclinic.org/heart/disorders/valve/tricuspid.aspx.

Mayo Clinic

http://www.mayoclinic.org/diseases-conditions/tricuspid-valve-disease/basics/definition/con-20036723.

Texas Heart Institute

http://www.texasheartinstitute.org/HIC/Topics/Cond/vtricus.cfm.

Merck

http://www.merckmanuals.com/home/SearchResults?query=Tricuspid+Regurgitation&icd9=424.2.

Professional Information

Review

Circulation. 2009;119:2718–25. http://circ.ahajournals.org/content/119/20/2718.full.

Review

J Am Coll Cardiol. 2015;65:2331–6. http://content.onlinejacc.org/article.aspx?articleID=2297643.

Assessment: Functional TR

Eur Heart J. 2013;34:1875–85. http://eurheartj.oxfordjournals.org/content/34/25/1875.

Association with MV Disease

J Am Coll Cardiol. 2009;53:401–8. http://www.sciencedirect.com/science/article/pii/S0735109708036619#.

C-V Waves

N Engl J Med. 2013;369:e27. http://www.nejm.org/doi/full/10.1056/NEJMicm1103312.

C-V Waves

N Engl J Med. 2012;366:e5. http://www.nejm.org/doi/full/10.1056/NEJMicm1012843.

Heart Failure: Impact of TR on Survival

Eur Heart J. 2013;34:844–52. http://eurheartj.oxfordjournals.org/content/34/11/844.

Mammoth Right Atrium

J Am Coll Cardiol. 2014;63:e21–e21. http://content.onlinejacc.org/article.aspx?articleID=1827543.

Pacemaker/ICD Leads

Heart. 2014;100:960–8. http://heart.bmj.com/content/100/12/960.full.

Pacemaker/ICD Leads

J Am Coll Cardiol. 2005;45:1672–5. http://content.onlinejacc.org/article.aspx?articleid=1136588&resultClick=3.

Pacemaker Implant: Case Study

Circulation. 2014;130:e23–5. http://circ.ahajournals.org/content/130/4/e23.full.

Post-mitral Valvuloplasty

Heart. 2013;99:91–7. http://heart.bmj.com/content/99/2/91.abstract.

Progression After MV Repair

Circulation. 2005;112:I453-7. http://circ.ahajournals.org/content/112/9-suppl/I-453.full.pdf+html?sid=ec03751f-c1be-4d34-b47c-4b5cdf314436.

Secondary TR

J Am Coll Cardiol. 2012;59:703–10. http://content.onlinejacc.org/article.aspx?articleid=1201134&resultClick=3.

Surgical Outcomes (Severe TR)

Heart. 2013;99:181–7. http://heart.bmj.com/content/99/3/181.abstract.

Transcatheter Mitraclip Repair

Eur Heart J. 2016 37: 849–53. http://eurheartj.oxfordjournals.org/content/37/10/849.abstract?etoc.

Trauma: TR/AMI

Circulation. 2014;129:e496–8. http://circ.ahajournals.org/content/129/20/e496.full.

Valve Replacement: Transfemoral

J Am Coll Cardiol. 2013;61:1929–31. http://content.onlinejacc.org/article.aspx?articleID=1662642.

Updates and More

https://clinicalguidecvd.com/tr

Chapter 96
Tricuspid Valve Stenosis

Management Keys

- Suspect this diagnosis in patients with MS who do not have symptoms of pulmonary congestion
- Suspect non-rheumatic etiology in absence of other valvular disease
- Treat clinically significant TS surgically because medical treatment relatively ineffective

ICD-10 Code

- Rheumatic: I0.70
- Congenital: Q22.4 (Tricuspid Atresia)

Description/Etiology

- Most common cause: Rheumatic Heart Disease [1]
- Characterized by diastolic pressure gradient between RA and RV; mean diastolic gradient >2 mmHg is diagnostic
- Clinical manifestations far overshadowed by associated left-sided (especially MV) valve disease [10]
- Usually accompanied by TR
- TTE essential for diagnosis in most cases

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 1271
DOI 10.1007/978-1-4471-7293-2_96,

Other causes:

- Anorectic Drugs [3]
- Carcinoid
- Congenital
- Infective Endocarditis
- Fabry Disease
- Systemic Lupus Erythematosus
- Whipple Disease

Predisposing/Comorbid Conditions

LEFT HEART VALVE DISEASE [2]
RHEUMATIC FEVER
TRICUSPID REGURGITATION

Demography

Rheumatic: Underdeveloped Countries
Females > Males

Pathophysiology

Obstruction to flow from RA to RV at level of TV orifice with resulting:

- Decreased RA emptying
- RA/IVC enlargement
- Increased RA pressure
- Decreased RV filling
- Decreased cardiac output
- Hepatic congestion
- Peripheral edema

Severe TS: thickened, distorted, calcified leaflets

Signs/Symptoms [2] [10]

ABDOMEN – DISTENSION
ABDOMEN – FLUID (ASCITES)
ABDOMEN – FULLNESS
BODY, GROWTH – DECR
EXTREM, LOWER, BILAT – EDEMA
FATIGUE
HEART, LSB, LOWER – MURMUR, DIAS
HEART, LSB, LOWER – OPENING SNAP
HEART, LSB, LOWER – THRILL, DIAS
HEART, RSB, LOWER – IMP, PRESYS
LIVER – ENLARGED (HEPATOMEGALY)
LIVER – PULSATION, PRESYS
NECK, JVP – ELEV
NECK, JVP, A WAVE – INCR/LARGE (CANNON WAVE)
NECK, JVP, Y DESCENT – SLOW
NECK, SENSATION – FLUTTERING
SKIN – SWELLING, EDEMA (ANASARCA)

Differentiation

Ebstein Anomaly
Pacemaker lead obstruction [4]
Pulmonary Valve Stenosis
RA Myxoma [4]
RA Thrombus [4]
RV Myxoma [4]
Tricuspid Atresia

Complications

Infective Endocarditis
Pulmonary embolism

Laboratory

NS

ECG

DYSRHYTHMIAS – ATRIAL (PACS/OTHERS)
P WAVE – TALL/PEAKED [5]

Imaging [8] [11]

CARDIOMEGALY
RA, CHAMBER, SIZE – INCR
TV, FLOW – OBS
TV, FLOW – REGURG
TV, LEAFLET, SEPARATION, DIAS – DECR
TV, LEAFLETS – THICK
TV, MOTION, DIAS – DOMING
VEIN, AZYGOS, SIZE – INCR
VENA CAVA, SUP, SIZE – INCR

Other Tests

Cardiac catheterization [6]

Treatment: Nonpharmacological

NS

Treatment: Pharmacological

NS

Treatment: Surgical/Invasive [7] [9]

TV valvuloplasty/repair
TV replacement

Course

Highly variable according to etiology and severity of associated lesions, especially left heart involvement in rheumatic form

Notes

[1] Most common cause globally, but rare in developed countries
[2] In rheumatic disease, involvement of other valves, especially MS, often dominates clinical picture; described changes are only those of pure TS, which is rare
[3] Eg, ergotamine, methysergide, anorexiants, pergolide
[4] In addition to directly obstructing flow, trauma from these sources may damage TV and cause valve stenosis
[5] Biphasic if MS present
[6] Hemodynamic assessment of TS:

Rarely performed for acquired disease but may be performed in select cases at time of invasive study for another indication, such as MS with PH
Direct assessment of absolute RA/RV diastolic pressure may be useful in determining TS contribution to clinical findings
Trans-tricuspid diastolic gradient is highly variable and is affected by heart rate, forward flow, and phases of the respiratory cycle; severe TS usually has mean pressure gradients >5–10 mmHg at HR of 70 bpm

[7] Surgery for severe TS:

- Most often performed at time of operation for left-sided valve disease, mainly rheumatic MS/MR
- If repair not adequate or feasible due to valve destruction or multiple levels of pathological involvement, replacement may be necessary
- Choice of prosthesis should be individualized
- Perioperative mortality rates are higher for mitral plus tricuspid versus either isolated mitral or tricuspid surgery alone.

[8] Other TTE findings indicative of severe TS:

- Mean pressure gradient >5 mmHg
- Pressure half-time = 190 ms
- Valve area = 1.0 cm^2 (continuity equation)
- RA/IVC enlargement.
- Assessment of TS severity with TTE is often technically limited; these values less well validated than those for MS

[9] 2014 AHA/ACC Guideline recommendations:
Tricuspid valve surgery is recommended for patients with severe TS at the time of operation for left-sided valve disease. *(Level of Evidence: C)*
Tricuspid valve surgery is recommended for patients with isolated, symptomatic severe TS *(Level of Evidence: C)*
Percutaneous balloon tricuspid commissurotomy might be considered in patients with isolated, symptomatic severe TS without accompanying TR. *(Level of Evidence: C)*

[10] Absence of symptoms of pulmonary congestion (eg, dyspnea) in presence of MS suggests TS

[11] CXR: dilated RA, normal size PA, clear lungs

Guidelines

2014 AHA/ACC guideline for the management of patients with valvular heart disease

J Am Coll Cardiol. 2014;63:e57–185. http://content.onlinejacc.org/article.aspx?articleid=1838843.

Guidelines on the management of valvular heart disease (version 2012)

Eur Heart J. 2012;33:2451–96. http://www.escardio.org/guidelines-surveys/esc-guidelines/Pages/valvular-heart-disease.aspx.

Patient Information

AHA

http://www.heart.org/HEARTORG/Conditions/More/HeartValveProblemsandDisease/Problem-Tricuspid-Valve-Stenosis_UCM_450390_Article.jsp#.Vzx4QfMUV1s.

Mayo

http://www.mayoclinic.org/diseases-conditions/tricuspid-valve-disease/home/ovc-20168105.

Merck

http://www.merckmanuals.com/home/heart-and-blood-vessel-disorders/heart-valve-disorders/tricuspid-stenosis.

Texas Heart Institute

http://www.mayoclinic.org/diseases-conditions/tricuspid-valve-disease/home/ovc-20168105.

Professional Information

Anatomic Appearance in Rheumatic Heart Disease

Br Heart J. 1957;19:211–6. http://europepmc.org/backend/ptpmcrender.fcgi?accid=PMC479617&blobtype=pdf.

Auscultatory Findings/RA Pressure Pulse

Am J Med. 1985;78:375–84. http://www.sciencedirect.com/science/article/pii/0002934385903274.

Cardiac Catheterization

Am Heart J. 1985;110:60–4. http://www.sciencedirect.com/science/article/pii/0002870385905150.

Diagnosis/Treatment

Br Heart J. 1964;26:354–79. http://europepmc.org/backend/ptpmcrender.fcgi?accid=PMC1018152&blobtype=pdf.

Diagnosis in Presence of Atrial Fibrillation

Circulation. 1966;33:26–33. http://circ.ahajournals.org/content/33/1/26.full.pdf.

Echo

Eur J Echocardiogr. 2009;10:1–25. http://ehjcimaging.oxfordjournals.org/content/10/1/1.

Hemodynamics

Br Heart J. 1971;33: 16–31. http://www.ncbi.nlm.nih.gov/pmc/articles/PMC487135/.

Isolated TS

Circulation. 1971;44:729–32. http://circ.ahajournals.org/content/44/4/729.full.pdf+html.

Percutaneous Balloon Valvuloplasty

Am J Cardiol. 1993;71:353–4. http://www.sciencedirect.com/science/article/pii/000291499390808P.

Mechanical Valve Replacement

J Thorac Cardiovasc Surg. 2014;148:603–8. http://www.sciencedirect.com/science/article/pii/S0022522313011409.

Updates and More

https://clinicalguidecvd.com/ts

Chapter 97
Ventricular Septal Defect: Congenital

Management Keys

- Suspect VSD in asymptomatic patients or patients with dyspnea with pansystolic LSB or apical murmur
- Use echo as primary diagnostic tool in patients with VSD, including determining morphologic type, shunt magnitude, associated defects
- Perform device or surgical VSD closure by experienced operators in patients according to current Guideline recommendations
- Consider vasodilator theapy for adults with VSDs with progressive/severe pulmonary vascular disease
- Counsel for activity and pregnancy in patients with VSD

ICD-10 Code

Q21.0

Alternate Names/Abbreviation

VSD

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 1281
DOI 10.1007/978-1-4471-7293-2_97,

Description/Etiology

Most common congenital heart defect at birth (3–3.5/1000 live births); high rate of spontaneous closure of small VSDs; incidence much less in adults

Anatomical types:

- Type 1: lie in RVOT; spontaneous closure uncommon; 6 % of defects in non-Asian populations; up to 33 % occur in Asian patients
- Type 2: perimembranous; almost 80 % of VSDs; defect is in membranous septum and adjacent to TV septal leaflet, which can become adherent to the VSD, forming a pouch or "aneurysm" of IVS; pouch limits L-R shunting and can cause partial or complete closure of defect; on LV side of septum, defect is adjacent to AV
- Type 3: inlet VSD; located in lower part of RV and adjacent to TV; typically occurs in Down syndrome
- Type 4: muscular VSD; located centrally (midmuscular), apically, or at margin of IVS and RV free wall; can be multiple; spontaneous closure common, low incidence in adults

Usually occurs as isolated lesion, but also associated with many other defects (see Predisposing/Comorbid Condtions)

Clinical presentation: Isolated VSD depends largely on defect size and PVR

- Small defects ≤25 % size of aortic annulus diameter have small L-R shunts, no left ventricle volume overload, no PAH, and present as systolic murmurs
- Moderate defects: >25 % but <75 % of aortic diameter, have small-moderate L-R shunts, mild-moderate LV volume overload, no/mild PAH; patients may remain asymptomatic or develop symptoms of mild HF; symptoms usually abate with medical treatment and time as VSD size decreases
- Large defects: >75 % of aortic diameter usually have moderate-large L-R shunts, LV volume overload,

PAH; most adult patients with large VSDs have history of HF in infancy; rarely, do not develop large L-R shunts and do not have normal postnatal fall in PVR; can present with R-L shunting and Eisenmenger syndrome later in childhood or as young adults.

Predisposing/Comorbid Conditions

AORTIC REGURGITATION - CHRONIC
AORTIC STENOSIS - SUBVALVULAR
ATRIOVENTRICULAR SEPTAL DEFECT
BICUSPID AORTIC VALVE
COARCTATION OF AORTA
DIGEORGE SYNDROME
DOWN SYNDROME
NOONAN SYNDROME
PULMONARY STENOSIS
TETRALOGY OF FALLOT
TRANSPOSITION OF GREAT ARTERIES - CORRECTED
TREACHER COLLINS SYNDROME

Demography

No gender predilection

Pathophysiology

Varies according to anatomical type; shunt direction and magnitude determined by

PVR
Defect size
LV/RV systolic and diastolic function
Presence of RVOT obstruction

Usual shunt direction is L-R; when significant, can cause:

Increased pulmonary blood flow and pulmonary venous return
LA/LV volume overload and enlargement

Significant L-R shunt:

Ratio of pulmonary to systemic blood flow >1.5/1.0
and/or
LH chamber dilatatation

Progressive increase in PVR leads to PAH

Signs/Symptoms

BODY, GROWTH - DECR
BREATHING - DIFF (DYSPNEA)
BREATHING - DIFF, RECLINING FLAT (ORTHOPNEA)
CHEST, ANT, L - BULGE
CHEST, RIBS, LOWER - DEFORMED (HARRISON GROOVES)
EXTREM, LOWER, BILAT - EDEMA
FATIGUE
HEART, LSB, MID - MURMUR, SYS [1]
HEART, LSB, UPPER - IMP, SYS
HEART, LV, APEX - MURMUR, DIAS
HEART, LV, APEX - MURMUR, SYS [1]
HEART, LV, APEX - THRILL, SYS
HEART, LV, APEX, IMP - FORCEFUL/SUSTAINED
HEART, P2 – INCR [3]
JOINT, SHOULDER - PAIN
SKIN - FLUSHING
SKIN, COLOR - BLUE (CYANOSIS) [2]
SKIN, COLOR - PALE (PALLOR)
SPUTUM - BLOOD (HEMOPTYSIS)
SWEATING - INCR (DIAPHORESIS/HYPERHIDROSIS)

Differentiation

Atrioventricular septal defect
Double-outlet RV with normally related great arteries
Infundibular Pulmonary Stenosis
PDA
Subaortic stenosis

Complications

Dysrhythmias (especially post-repair)
Heart block (especially post-repair)
HF
Infective Endocarditis
Pneumonia
PAH
SCD

Laboratory

NS

ECG [4]

AV COND - 1ST DEGREE BLOCK
Q WAVE - ABN
QRS - BVH PATTERN
QRS - LVH PATTERN
QRS, AXIS - L
QRS, R WAVE - TALL

Imaging [7] [8]

ASSESS FOR ASSD ABNORMALITIES AND VSD LOCALIZATION/NUMBER OF DEFECTS
CARDIOMEGALY
IVS - DEFECT
IVS, FLOW ACROSS - BIDIRECTIONAL
IVS, FLOW ACROSS - LEFT TO RIGHT
LA, CHAMBER, SIZE - INCR [LARGE SHUNTS] [5]
LV, CHAMBER, SIZE - INCR [LARGE SHUNTS] [5]
PA, BRANCHES, SIZE - INCR
PA, MAIN, SIZE - INCR

Genomics

CSX
DTNA
NKX2.5
TBX5

Other Tests

Cardiac catheterization [9]

Treatment: Nonpharmacologic

Activity counseling [13]
Pregnancy counseling [14]

Treatment: Pharmacologic

Vasodilators [10]

Treatment: Surgical/Invasive

Device closure [12]
Surgical closure [11]

Course

Adults with no PAH have normal/close to normal life expectancy post-VSD closure

Notes

[1] Murmur features:

Determined by blood flow velocity across the defect
Loud/pansystolic with high pressure difference between LV and RV
Small defects loud; intensity can decrease in late systole as muscular contraction reduces defect size
As RV pressure increases, murmur shortens and becomes lower pitched

[2] With R-L shunt, indicates PAH
[3] Increased P2 indicates elevated PVR/PA pressure
[4] ECG may be normal with small shunts/no PAH
[5] L-R shunt magnitude across VSD reflected by LA/LA volume overload
[6] CMR: useful when inlet or apical VSD cannot be well seen on echo; also used to quantify AR severity and for LV size/function
[7] CXR: Small VSD – normal

Significant L-R shunt – LA and LV enlargement with increased pulmonary vascular markings
Significant PAH – no LV enlargement with prominent PA segment and decreased pulmonary vascular markings at lung periphery

[8] Echo mainstay of diagnosis, for:

Number/location of defects
Chamber sizes
Ventricular function
Presence or absence of aortic valve prolapse and/or regurgitation
Presence or absence of RV or LV outflow obstruction
Presence or absence of TR
Estimation of RV systolic pressure from TR jet, VSD jet, and/or septal configuration should be a part of the study
In adults with poor echo windows, TEE may be necessary

[9] ACC/AHA 2008 Guidelines: Recommendations for cardiac catheterization:

Class I

- Cardiac catheterization to assess the operability of adults with VSD and PAH should be performed in an ACHD regional center in collaboration with experts *(Level of Evidence: C)*

Class IIa

- Cardiac catheterization can be useful for adults with VSD in whom noninvasive data are inconclusive and further information is needed for management. Data to be obtained include the following:
  - Quantification of shunting. *(Level of Evidence: B)*
  - Assessment of pulmonary pressure and resistance in patients with suspected PAH. Reversibility of PAH should be tested with various vasodilators. *(Level of Evidence: B)*
  - Evaluation of other lesions such as AR and double-chambered right ventricle. *(Level of Evidence: C)*

- Determination of whether multiple VSDs are present before surgery. *(Level of Evidence: C)*
- Performance of coronary arteriography is indicated in patients at risk for coronary artery disease. *(Level of Evidence: C)*
- VSD anatomy, especially if device closure is contemplated. *(Level of Evidence: C)*

[10] ACC/AHA 2008 Guidelines: Recommendation for Medical Therapy

Class IIb

Pulmonary vasodilator therapy may be considered for adults with VSDs with progressive/severe pulmonary vascular disease (*Level of Evidence: B)*

[11] ACC/AHA 2008 Guidelines: Recommendations for Surgical Ventricular Septal Defect Closure

Class I

- Surgeons with training and expertise in CHD should perform VSD closure operations. *(Level of Evidence: C)*
- Closure of a VSD is indicated when there is a Qp/Qs (pulmonary–to–systemic blood flow ratio) of 2.0 or more and clinical evidence of LV volume overload. *(Level of Evidence: B)*
- Closure of a VSD is indicated when the patient has a history of IE. *(Level of Evidence: C)*

Class IIa

- Closure of a VSD is reasonable when net left-to-right shunting is present at a Qp/Qs greater than 1.5 with pulmonary artery pressure less than two thirds of systemic pressure and PVR less than two thirds of systemic vascular resistance. *(Level of Evidence: B)*
- Closure of a VSD is reasonable when net left-to-right shunting is present at a Qp/Qs greater than 1.5 in the presence of LV systolic or diastolic failure. *(Level of Evidence: B)*

Class III

- VSD closure is not recommended in patients with severe irreversible PAH. *(Level of Evidence: B)*

[12] ACC/AHA 2008 Guidelines: Recommendations for device closure

Class IIb

- Device closure of a muscular VSD may be considered, especially if the VSD is remote from the tricuspid valve and the aorta, if the VSD is associated with severe left-sided heart chamber enlargement, or if there is PAH. *(Level of Evidence: C)*

[13] ACC/AHA 2008 Guidelines: Recommendations for activity:

No activity restrictions are indicated for patients with small VSDs, no associated lesions, and normal ventricular function. If pulmonary vascular disease is present, activity is usually self-restricted, but patients should be advised against strenuous exercise or travel to altitudes above 5000 feet. Long-distance air travel should be approached with caution to avoid dehydration, with specific recommendation by an ACHD specialist concerning the need for supplemental oxygen

[14] ACC/AHA 2008 Guidelines Recommendations for pregnancy:

Class III

- Pregnancy in patients with VSD and severe PAH (Eisenmenger syndrome) is not recommended owing to excessive maternal and fetal mortality and should be strongly discouraged. *(Level of Evidence: A)*

Guidelines

ACC/AHA 2008 guidelines for the management of adults with congenital heart disease

J Am Coll Cardiol. 2008;52:e143–e263. http://content.onlinejacc.org/article.aspx?articleid=1188032#tab1.

ESC guidelines for the management of grown-up congenital heart disease (new version 2010)

Eur Heart. J 2010;31:2915–57. http://eurheartj.oxfordjournals.org/content/ehj/31/23/2915.full.pdf.

Patient Information

AHA

http://www.heart.org/HEARTORG/Conditions/CongenitalHeartDefects/AboutCongenitalHeartDefects/Ventricular-Septal-Defect-VSD_UCM_307041_Article.jsp#.Vz73T_MUV1s.

CDC

http://www.cdc.gov/ncbddd/heartdefects/ventricularseptaldefect.html.

MAYO

http://www.mayoclinic.org/diseases-conditions/ventricular-septal-defect/basics/definition/con-20024118.

Medlineplus

https://www.nlm.nih.gov/medlineplus/ency/article/001099.htm.

Stanford

http://www.stanfordchildrens.org/en/topic/default?id=ventricular-septal-defect-vsd-90-P01829.

Professional Information

Anatomical Types

Ann Thorac Surg. 2000;69:S25–S35. http://www.sciencedirect.com/science/article/pii/S0003497599012709.

Device Closure

J Am Coll Cardiol. 2004;43:1257–63. http://www.sciencedirect.com/science/article/pii/S0735109704000816.

Natural Course in Adolescents

Pediatr Cardiol. 1998;19:230–4. http://link.springer.com/article/10.1007%2Fs002469900291.

Pathophysiology

Circulation. 2008;117:1090–9. http://circ.ahajournals.org/content/117/8/1090.full.

Small VSDs in Adults

Eur Heart J. 1998;19:1573–82. http://eurheartj.oxfordjournals.org/content/19/10/1573.

Spontaneous VSD Closure after School Age

Pediatr Int. 2008;50:632–5. http://www.ncbi.nlm.nih.gov/pubmed/19261109.

Unrepaired Perimembranous VSDS in Adults

Am J Cardiol. 2010;105:404–7. http://www.sciencedirect.com/science/article/pii/S0002914909024187.

VSD Closure: Long-Term Followup

J Am Coll Cardiol. 2015;65:1941–51. http://www.sciencedirect.com/science/article/pii/S0735109715009043.

Updates and More

https://clinicalguidecvd.com/vsd

Chapter 98
Williams Syndrome

Management Keys

- Careful routine monitoring, including four-extremity BPs, ECG (especially for QT interval), and close examination of peripheral vasculature
- Take particular caution during invasive procedures
- Avoid treating with RAAS inhibitors for Systemic Arterial Hypertension until Renal Artery Stenosis excluded

ICD-10 Code

Q78.8

Alternate Names/Abbreviation

WS
WILLIAMS-BEUREN SYNDROME
ELFIN FACIES SYNDROME
BEUREN SYNDROME
HYPERCALCEMIA-AORTIC STENOSIS

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 1295
DOI 10.1007/978-1-4471-7293-2_98,

Description/Etiology

Multisystem autosomal dominant developmental disorder caused by deletions of portions of Chromosome 7, involving one or more combinations of 25 genes

Phenotypic expressions include distinctive facial features, mental retardation, developmental delays, hypercalcemia, Supravalvular Aortic Stenosis, Systemic Aterial Hypertension, cerebellar abnormalities

CV defects:

- Most common cause of death in patients with WS
- Structural CV abnormalities occur in about 80 % of all WS patients
- Present in up to 93 % of WS patients at age 1 year
- Most consist of some form of arterial stenosis
- Supravalvular AS most common CV abnormality (45–75 %), occurring in two forms:
  - Discrete, hourglass narrowing at sinotubular junction
  - Diffuse, long-segment stenosis of ascending aorta (associated with brachiocephalic arterial stenosis)

Familial cases rare

Predisposing/Comorbid Conditions

ANXIETY/ANXIETY DISORDER
AORTIC STENOSIS - SUPRAVALVULAR [1]
ATTENTION DEFICIT HYPERACTIVITY DISORDER
CARDIAC VALVE ABNORMALITIES [23]
CAROTID ARTERY STENOSIS
CELIAC DISEASE
CORONARY ARTERY DISEASE [22]
DIABETES MELLITUS [13]

DIVERTICULOSIS
HYPERCALCEMIA [15]
HYPERTENSION - SYSTEMIC ARTERIAL
HYPOTHYROIDISM [16]
INTRACARDIAC DEFECTS [24]
MENTAL RETARDATION
NEPHROCALCINOSIS
OSTEOPENIA
OSTEOPOROSIS
PHOBIC DISORDER
PULMONARY ARTERY STENOSIS [PERIPHERAL] [19]
PULMONARY STENOSIS - SUPRAVALVULAR
RENAL ARTERY STENOSIS
THORACIC AORTA STENOSIS
TYPE I CHIARI FORMATION

Pathophysiology

ELN gene mutation affects elastin, which comprises about 50 % of dry weight of normal aorta

Elastin: characterized by high degree of reversible distensibility, including ability to deform significantly with small forces; in arterial system, this characteristic allows energy storage in form of arterial distension during systole and subsequent release of stored energy via vascular recoil during diastole (Windkessel effect), greatly improving CV system efficiency

In arteries, smooth muscle cells produce most elastin; some also produced by endothelial cells and adventitial fibroblasts

Signs/Symptoms [18]

ABDOMEN - BRUIT [27]
ABDOMEN - PAIN [2]

ABDOMEN, LLQ - HERNIA, INGUINAL
ANORECTUM - PROLAPSE
ARTERIAL PRESSURE, UE, SYS - R>L [6]
ARTERIAL PULSE PRESSURE - DECR [6]
ARTERIAL PULSE, CAROTID - R>L [6]
ARTERIAL PULSE, DOWNSLOPE - GRADUAL [6]
ARTERIAL PULSE, PEAK - SUSTAINED [6]
ARTERIAL PULSE, RISE - SLOW [6]
ARTERIAL PULSE, UE - ASYMMETRIC [6] [7]
ARTERY, CAROTID - BRUIT [29]
ARTERY, CAROTID - THRILL [6]
BACK, CURV - ANT (LORDOSIS)
BACK, CURV - LAT (SCOLIOSIS)
BEHAVIOR - HYPERACTIVE [5]
BEHAVIOR - OBSESSIVE-COMPULSIVE
BLOOD PRESSURE, ARTERIAL - INCREASED/ELEVATED
BODY, EQUILIBRIUM - DECR
BODY, HT - DECR
BOWEL MOVEMENTS - CONSTIPATION
BREATHING - DIFF (DYSPNEA) [6]
COGNITION - DEFECT, NS [11]
CONSCIOUSNESS - LOSS, SUDDEN, EFFORT (EFFORT SYNCOPE) [6]
DIZZY/LIGHTHEADED, EFFORT [6]
EARS, HEARING - FEAR, LOUD SOUNDS (PHONOPHOBIA)
EARS, HEARING - LOSS (DEAFNESS) [3]
EARS, HEARING - SENSATION, INCREASED (HYPERACUSIS)
EYES, IRIS - STELLATE
EYES, LACRIMAL DUCT - NARROW
EYES, MOTION - WANDERING (STRABISMUS)
EYES, PERIORBITAL - FULLNESS
EYES, VISION - DECR/LOSS [SUBTOTAL]
EYES, VISION, 3D - DECR (STEREOPSIS)
FACE, CHEEKS - PROMINENT [4]
FACE, CHIN - POINTED

FACE, FOREHEAD - BROAD/PROMINENT
FATIGUE [6]
FLANK - BRUIT [27]
HAIR, COLOR - GRAY, PREMATURE
HEADACHE [28]
HEART, LV, APEX, IMP - FORCEFUL/SUSTAINED [6]
HEART, LV, APEX, IMP - FORCEFUL/SUSTAINED [6]
HEART, RSB, UPPER - MURMUR, SYS [6][8]
HEART, S2, SPLIT - REVERSED (PARADOXICAL) [6]
HEART, S4 LV [6]
JOINTS - CONTRACTURES [9]
JOINTS, MOVEMENT, RANGE - INCR (HYPERMOBILITY)
LIPS - THICK
MENTATION - ATTENTION DEFICIT/ HYPERACTIVITY DIS
MENTATION, LEARNING, DEVELOPMENT - DECR [10]
MOOD - ANXIOUS
MOOD - DEPRESSED [12]
MOUTH, SMILE - WIDE
MUSCLES, TONE - DECR (HYPOTONIA)
NECK, JVP, A WAVE - INCR/LARGE (CANNON WAVE) [6]
NECK, SUPRASTERNAL NOTCH - THRILL, SYS [6]
NOSE - SHORT
NOSE - UPTURNED
PERSONALITY - EBULLIENT
PUBERTY,DEVELOPMENT - EARLY (PRECOCIOUS PUBERTY)
SKIN, TEXTURE - SOFT
SLEEP - DISTURBED (INSOMNIA)
SWEATING - INCR (DIAPHORESIS/ HYPERHIDROSIS) [6]

SYNDROME - RESTLESS LEGS
TEETH - MALOCCLUSION
TEETH - SMALL
TENDON, REFLEXES - INCR (HYPERACTIVE REFLEXES)
URINATION - BEDWETTING (ENURESIS)
URINATION - INCR (POLYURIA)
URINATION - URGENCY
WEIGHT - INCREASED/GAIN

Differentiation

Other syndromes that include physical/behavioral features and congenital heart disease, including:

Digeorge Syndrome
Noonan Synrome
Turner Syndrome
Fragile X Syndrome
Fetal Alcohol Syndrome

Complications [21]

AMI [14]
Dental Caries
Recurrent Otitis Media
Recurrent Urinary Tract Infection
Stroke
Sudden Death [25]

Laboratory [18]

BLOOD, CALCIUM - INCR [15]
BLOOD, GLUCOSE - INCR (HYPERGLYCEMIA) [13]
URINE, CALCIUM - INCR

ECG [18]

DYSRHYTHMIAS - VENTRICULAR (PVCS/OTHERS)
JT INTERVAL - LONG [33]
QRS - LVH PATTERN [6][26]
QRS - RBBB/RBBB PATTERN [26]
QT/QTC INTERVAL - LONG [32]
ST SEGMENT - DEPR [6]
T WAVE - INVER, ABN [6]

Imaging [CV ONLY] [18]

AORTA, ASCEND, SIZE - DECR [6]
AORTA, ASCEND, SYS - GRADIENT [6]
ART, CORONARY, SIZE - INCR/ANEURYSM
LV, MYOCARD, WALL THICKNESS - INCR (HYPERTROPHY) [6]

Genomics

BAZ1B
CLIP2
ELN
FZD9
GTF2I
GTF2IRD1
LIMK1
NCF1
STX1A

Other Tests

Ambulatory ECG monitoring (especially when QT prolonged)

Treatment: Nonpharmacologic [18]

Dental hygiene
Dietary calcium restriction

Treatment: Pharmacologic [18]

Antihypertensives (RAAS inhibitors contraindicated until RAS excluded)
Beta-blocker for QT prolongation
Oral hypoglycemics

Treatment: Surgical/Invasive [18][19][31]

Complex aorta patching for Supravalvular AS [30]
Transcatheter intervention for PS (when necessary) [19]
Reconstruction of coronary ostia
CABG

Prevention

Pregnancy counseling of affected persons

Course

Variable according to abnormalities

Notes

[1] >50 % of patients with Supravalvular AS have WS
[2] May occur without apparent cause
[3] Sensorineural; detected in adolescence and adults

[4] Young children: face described as “pixie-like”, “cute”, “elfin”
[5] Declines after childhood
[6] Features of Supravalvular AS
[7] With Supravalvular AS, due to Coanda effect: preferential blood flow into right brachiocephalic artery
[8] Crescendo-decrescendo; ejection click absent; radiates to right carotid artery
[9] Especially lower extremities
[10] Mean IQ approximately 55 (sometimes normal)
[11] Williams-Beuren syndrome cognitive profile: includes relative strengths in memory/language and weaknesses in spatial/motor skills; strength in facial recognition
[12] Usually not major; more often dysthymia
[13] >50 % have DM/abnormal glucose metabolism
[14] May occur due to coronary ostial stenosis, which can occur in absence of Supravalvular AS
[15] Usually mild, but may be moderate-severe elevation in infants/young children; usually resolves in childhood
[16] Usually subclinical
[17] Relates to hypertension
[18] Pathology highly variable and treatment individualized according to associated conditions; life-long supportive care with team approach often necessary
[19] Peripheral pulmonary stenosis often resolves spontaneously
[20] May be secondary to RAS or occur in its absence, related to NCF1 mutation
[21] Cardiovascular disease most common cause of death
[22] Due to coronary artery diffuse stenosis, aneurysm, ostial stenosis, AV inflow obstruction, sinotubular ridge, or combinations
[23] Valvular AS, MVP, MR; TV involvement rare; Ebstein Anomaly reported
[24] Especially VSD, usually muscular
[25] 25–100x risk in general population; cause uncertain in many cases
[26] 60 % RVH, 40 % LVH

[27] Common, often due to RAS
[28] May indicate intracranial stenosis
[29] May indicate brachiocephalic stenosis
[30] Angioplasty ineffective
[31] Periprocedural CV collapse reported in WS patients: particular caution indicated at these times
[32] Due to uncertain mechanism in WS, but may also be prolonged by use of CNS drugs for ADHD, anxiety, etc., including but not limited to: phenothiazines, haloperidol, tricyclic antidepressants, astemizole, ketoconazole, itraconazole, probucol, ketanserin, cisapride, papaverine, tacrolimus, arsenic trioxide
[33] JT interval rather than QT interval measure may be preferable in presence of prolonged QRS

Guidelines

ACC/AHA 2008 guidelines for the management of adults with congenital heart disease
J Am Coll Cardiol. 2008;52:e143–e263. http://content.onlinejacc.org/article.aspx?articleid=1188032#tab1.

ESC guidelines for the management of grown-up congenital heart disease (new version 2010)
Eur Heart J. 2010;31:2915–57. http://eurheartj.oxfordjournals.org/content/ehj/31/23/2915.full.pdf.

Patient Information

Images

https://www.nlm.nih.gov/medlineplus/ency/imagepages/17243.htm.

Medlineplus

ENGLISH

https://www.nlm.nih.gov/medlineplus/ency/article/001116.htm.

ESPANOL

https://www.nlm.nih.gov/medlineplus/spanish/ency/article/001116.htm.

Genetics Home REF

http://ghr.nlm.nih.gov/condition/williams-syndrome.

Williams Syndrome ASSN

http://williams-syndrome.org/ws.

Cleveland Clinic

http://my.clevelandclinic.org/disorders/genetic-disorders/hic-williams-syndrome.aspx.

NORD

https://rarediseases.org/rare-diseases/williams-syndrome/.

Professional Information

Review

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Review

Circulation. 2013;127:2125–34. http://circ.ahajournals.org/content/127/21/2125.full.

Acquired Coarctation

Heart. 1998;80:205–6. http://heart.bmj.com/content/80/2/205.full#ref-1.

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Eur Heart J. 2012. doi: 10.1093/eurheartj/ehs206. http://eurheartj.oxfordjournals.org/content/early/2012/07/19/eurheartj.ehs206.full.

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JT Interval

Am J Cardiol. 2010;106:1029–33. http://www.ncbi.nlm.nih.gov/pubmed/20854969.

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J Child Psychol Psychiatry. 2008;49:576–608. http://onlinelibrary.wiley.com/doi/10.1111/j.1469-7610.2008.01887.x/full.

Updates and More

https://clinicalguidecvd.com/williams

Chapter 99
Wolff-Parkinson-White Syndrome

ICD-10 Code

I45.6

Alternate Names/Abbreviation

ACCELERATED AV CONDUCTION
PRE-EXCITATION AV CONDUCTION
PREEXCITATION SYNDROME
WPW

Description/Etiology

- Premature excitation of ventricular myocardium by impulse bypass of normal conduction pathway via accessory pathways [6], causing tachyarrhythmias in many affected persons [5]
- Patients with WPW typically present with palpitations or presyncope caused by an atrioventricular reciprocating tachycardia or, less commonly, a primary atrial tachycardia
- Persons with ECG preexcitation pattern may be asymptomatic [20]

V.E. Friedewald, *Clinical Guide to Cardiovascular Disease*, 1309
DOI 10.1007/978-1-4471-7293-2_99,

Rapid conduction of AF over an accessory pathway resulting in VF rare but may be first manifestation
Accessory pathways thought to be an embryologic remnant
Also occurs in patients with myopathic and structural congenital heart disease, particularly Ebstein anomaly
Uncommonly, may coexist with cardiac rhabdomyoma, usually discovered in newborns, associated with tumors located at AV groove or septum, and believed caused by disruption of AV annulus electrical integrity rather than being true accessory pathways.
HCM may be associated with WPW, often associated with specific gene mutations

Comorbid Conditions

ATRIAL SEPTAL DEFECT - SECUNDUM
CARDIOMYOPATHY - DANON DISEASE
CARDIOMYOPATHY - DILATED
CARDIOMYOPATHY - HYPERTROPHIC
COARCTATION OF AORTA
EBSTEIN ANOMALY
GRAVES DISEASE
HYPERTHYROIDISM
LEBER HEREDITARY OPTIC NEUROPATHY [7]
MITRAL VALVE PROLAPSE
POMPE DISEASE
PRKAG2 SYNDROME
TETRALOGY OF FALLOT
TOTAL ANOMALOUS VENOUS RETURN
TRANSPOSITION OF GREAT ARTERIES - CORRECTED
TRICUSPID ATRESIA
TUBEROUS SCLEROSIS
VENTRICULAR SEPTAL DEFECT

Demography

All populations
Sometimes familial

Pathophysiology

Antegrade conduction of impulse via accessory pathway, initiating ventricular depolarization (manifest as delta wave on ECG) before arrival of normally conducted impulse
Accessory pathways make possible retrograde ventricular-atrial impulse conduction and resultant tachyarrhythmias
Ventricular septal dyskinesis reported in some persons (reversible with ablation)
Majority of WPW cases have normal cardiac anatomy

Signs/Symptoms

CHEST - PALPITATIONS
CONSCIOUSNESS - LOSS, SUDDEN (SYNCOPE)
DIZZY/LIGHTHEADED/PRESYNCOPE
HEART, RATE - RAPID (TACHYCARDIA)
HEART, RHYTHM - IRREG
HEART, S1, INTENSITY - INCR
HEART, S2, SPLIT - REVERSED (PARADOXICAL)

Differentiation

AMI
Fabry Disease [10]
LBBB or RBBB
PVT
RVH

Complications

Cardiac Arrest
Rapid AF
SCD [12]
VF

Laboratory

NS

ECG [11]

DELTA WAVE [2]
DYSRHYTHMIAS - ATRIAL (PACS/OTHERS) [5]
DYSRHYTHMIAS - VENTRICULAR (PVCS/OTHERS)
PR INTERVAL - SHORT [3]
QRS - LONG, NS [4]
ST-T WAVE - ABN, NS

Imaging

NS/VAR WITH COMORBID

Genomics

PRKAG2

Other Tests

Ambulatory ECG monitoring

EP testing [9]
Stress test

Treatment: Nonpharmacologic

NS

Treatment: Pharmacologic

NS

Treatment: Surgical/Invasive [9]

Pathway ablation
Antitachycardia atrial pacemaker

Course

Highly variable

Notes

[1] During tachyarrhythmia
[2] Type A: initial positive QRS deflection in V1-2
Type B: initial negative QRS deflection in V1-2
Type C: initial negative QRS deflection in lateral leads
[3] <0.12 SEC
[4] >0.10 SEC; configuration dependent on location of bypass tract
[5] PSVT; sudden AF may be serious
[6] Accessory pathway: an extranodal AV pathway that connects atrial myocardium to ventricle across the AV groove; accessory pathways include AV tracks, nodofas-

cicular tracts, and many other variations; most common insertion is into left lateral LV free wall

[7] Inherited disorder of mitochondria causing subacute blindness, mainly in young men; WPW may also affect maternal carriers without blindness

[8] For comorbid cardiac conditions

[9] Consult current Guidelines for detailed recommendations of EP testing and treatment for asymptomatic and symptomatic persons with preexcitation findings

[10] Short PR in absence of preexcitation characteristic of Fabry Disease; uncertain mechanism

[11] Preexcitation ECG pattern:

Reflects presence of a manifest accessory pathway connecting the atrium to the ventricle

Pre-excited ventricular activation over accessory pathway competes with anterograde conduction over AV node and spreads from accessory pathway insertion point in ventricular myocardium

Variable degree of pre-excitation, with characteristic pattern of short P-R interval with slurring of the initial upstroke of the QRS complex (delta wave) may occur, depending on relative contribution from ventricular activation by normal AV nodal/His Purkinje system versus the manifest accessory pathway

Can be intermittent or not easily appreciated with pathways capable of anterograde conduction; usually associated with low-risk pathways, with exceptions

[12] Increased risk of SCD in WPW associated with:

History of symptomatic tachycardia

Multiple accessory pathways

Shortest pre-excited R-R interval of <250 ms during AF

First 2 decades of life

Guidelines

2015 ACC/AHA/HRS Guideline for the management of adult patients with supraventricular tachycardia: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Rhythm Society

J Am Coll Cardiol. 2016;67:e27–e115. http://www.sciencedirect.com/science/article/pii/S0735109715058404.

2012 PACES/HRS expert consensus statement on the management of the asymptomatic young patient with a Wolff-Parkinson-White (WPW, Ventricular Preexcitation) electrocardiographic pattern

Heart Rhythm. 2012;9:1006–1024. http://www.hrsonline.org/Practice-Guidance/Clinical-Guidelines-Documents/2012%20Management%20of%20the%20Asymptomatic%20Young%20Patient%20with%20a%20Wolff-Parkinson-White.

Patient Information

Images

http://www.nlm.nih.gov/medlineplus/ency/imagepages/8810.htm.

Genetics Home Reference

http://ghr.nlm.nih.gov/condition/wolff-parkinson-white-syndrome.

Cleveland Clinic

http://my.clevelandclinic.org/heart/disorders/electric/wpw.aspx.

Medline Plus

ENGLISH

http://www.nlm.nih.gov/medlineplus/ency/article/000151.htm.

ESPANOL

http://www.nlm.nih.gov/medlineplus/spanish/ency/article/000151.htm.

MAYO Clinic

http://www.mayoclinic.org/diseases-conditions/wolff-parkinson-white/basics/causes/con-20043508.

SADS Foundation

http://www.sads.org/What-is-SADS/Wolff-Parkinson-White-Syndrome#.VYVRh2fJB1s.

Texas Heart Institute

http://texasheart.org/HIC/Topics/Cond/arrhycat.cfm.

Professional Information

Early Description

Am Heart J. 1930;5:685–704. http://www.sciencedirect.com/science/article/pii/S0002870330900865.

Atrial Fibrillation

Am J Cardiol. 1992;70:A38–A43. http://www.sciencedirect.com/science/article/pii/000291499291076G.

Ablation: Asymptomatic Patients

Card Electrophysiol Clin. 2012;4:281–5. http://www.sciencedirect.com/science/article/pii/S1877918212000755.

Catheter Ablation/EP

Circulation. 2014;130:811–9. http://circ.ahajournals.org/content/130/10/811.long.

Diagnosis in Emergency Department

Am Journal Emerg Med. 2009;27:878–88. http://www.sciencedirect.com/science/article/pii/S0735675708004701.

Malignant Arrhythmias

Circulation. 2012;125:661–8. http://circ.ahajournals.org/content/125/5/661.long.

Natural History

Am J Cardiol. 2013;112:961–5. http://www.sciencedirect.com/science/article/pii/S0002914913012332.

Population Study: SCD

Circulation. 1993;87:866–73. http://circ.ahajournals.org/content/87/3/866.abstract?ijkey=480a8af85ba8cfb030f29fe8c3f2ce878730cdb5&keytype2=tf-ipsecsha.

Registry: Prognosis/Ablation

Circulation. 2014;130:811–9. http://circ.ahajournals.org/content/130/10/811.full.

Septal Dyskinesis

Circulation. 2014;130:e196–8. http://circ.ahajournals.org/content/130/23/e196.full.

Tuberous Sclerosis

Arch Dis Child. 1998;78:159–62. http://adc.bmj.com/content/78/2/159.full.

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https://clinicalguidecvd.com/wpw