1. 5 year old male came to ER with palpitations sweating since past 1 hr. On examination he had HR of 170/min which was regular, BP-120/70mmHg. ECG showed SVT. How will you manage this patient?

   A. Give IV adenosine 24mg stat

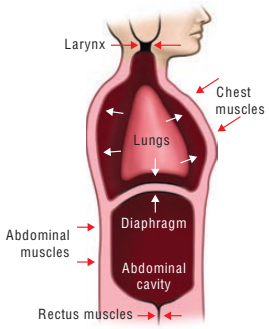
   B. Wait for senior to come to take decision

   C. Valsalva/modified valsalva manouvre

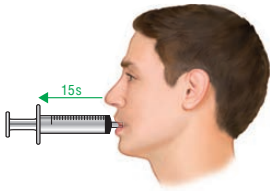
   D. DC cardioversion

Answer: C(Page 1899,1899)

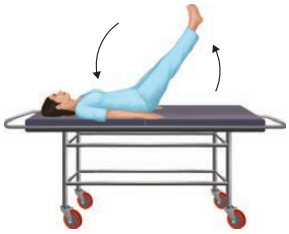
TABLE 246-2 Vagal Maneuvers



Holding breath while bearing down to increase intrathoracic pressure

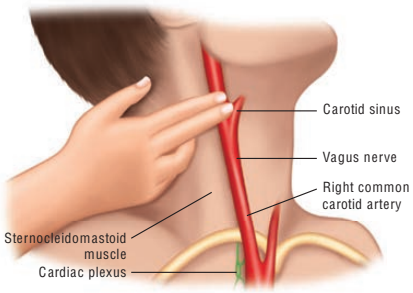


Breathing hard into a syringe against pressure to increase intrathoracic pressure



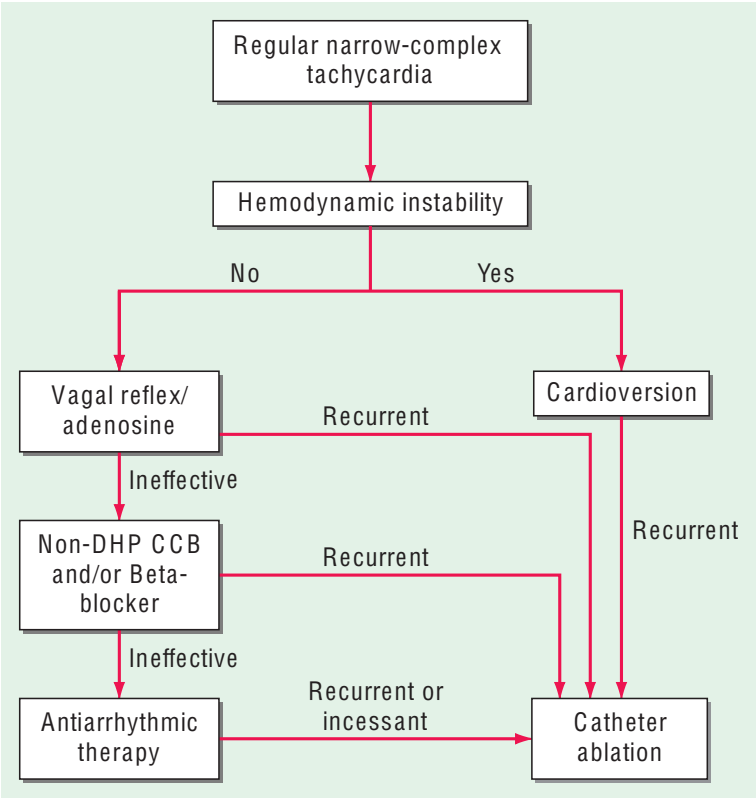
Raise legs abruptly to increase venous return

Submerge face into cold water (diver's reflex)



Carotid sinus massage

Adenosine



2. Which of the following drugs are not typically associated with sinus node dysfunction

   A. Beta blockers

   B. Lithium

   C. Digitalis

   D. Pantoprazole

Answer:D(Page 1875)

Medications Associated with Sinus Node Dysfunction
Antihypertensive Medications
• Beta-adrenergic receptor blockers
• Clonidine
• Methyldopa
• Nondihydropyridine calcium channel blockers
Antiarrhythmic Medications
• Amiodarone
• Dronedarone
• Flecainide
• Procainamide
• Propafenone
• Quinidine
• Sotalol
• Ivabradine
Psychiatric Medications
• Donepezil
• Lithium
• Opioid analgesics
• Phenothiazine antiemetics and antipsychotics
• Phenytoin
• Selective serotonin reuptake inhibitors
• Tricyclic antidepressants
Other
• Anesthetic drugs (propofol)
• Cannabis
• Digoxin
• Muscle relaxants

3. 55 year old known AF with controlled ventricular rate, hypertensive and past history of IC bleed 6 yrs back came with Malena. His creatinine level was 4mg/dL. What would be the ideal next step for this patient?

   A. LAA closure

   B. Pacemaker

   C. Left ventricular assist devices

   D. Angioplasty

Answer: A(Page 1906-07)

Bleeding is the major risk of anticoagulation. Major bleeding requiring transfusion and intracranial bleeding occur in ~1% of patients per year with warfarin. Direct-acting anticoagulants appear to have a lower risk of intracranial bleeding compared with warfarin without sacrificing protective effects against thromboembolism.

Risk factors for bleeding include age >65–75 years, heart failure, renal insufficiency, prior bleeding, and excessive alcohol or non- steroidal anti-inflammatory drug use. In patients who require dual antiplatelet therapy (e.g., aspirin and clopidogrel) after coronary or peripheral arterial stenting, there is a substantially increased bleeding risk when standard oral anticoagulation with warfarin or a direct-acting anticoagulant is added. The optimal combination of agents for patients with AF who also require antiplatelet therapy remains unclear.

Chronic anticoagulation is contraindicated in some patients due to bleeding risks. Because most atrial thrombi likely originate in the left atrial appendage, surgical removal of the appendage, combined with atrial maze surgery, may be considered for patients undergoing surgery, although removal of the appendage has not been unequivocally shown to reduce the risk of thromboembolism. Percutaneously deployed devices that occlude or ligate the left atrial appendage are also available, appear to be noninferior to warfarin in reducing stroke risk, and are considered in patients who have a high risk of thromboembolism but serious bleeding risk from chronic oral anticoagulation

4. Dabigatran is
   A. Factor Xa inhibitor
   B. Vitamin K antagonist
   C. Direct thrombin inhibitor
   D. Factor Xa inhibitor

Answer: A(Page 1906)

The options for anticoagulation are the oral factor Xa inhibitors apixaban, edoxaban, or rivaroxaban; the oral antithrombin inhibitor dabigatran; and the vitamin K antagonist warfarin

The direct-acting anticoagulants dabigatran, rivaroxaban, apixaban, and edoxaban were noninferior to warfarin in individual trials of nonvalvular AF patients, and intent-to-treat analysis of pooled data suggests superiority to warfa- rin by small absolute margins of 0.4–0.7% in reduction of mortality, stroke, major bleeding, and intracranial hemorrhage. Warfarin is required for patients with rheumatic mitral stenosis or mechanical heart valves. The newer, direct-acting anticoagulants have not been tested in rheumatic heart disease, and a direct thrombin inhibitor did not prevent thromboembolism in patients with mechanical heart valves.

5. 90 year old diabetic came with c/o giddiness since one day. Hear rate was 30 per min. Bp was 110/60 mm Hg. Electrolytes, TSH was normal. ECG showed Complete heart

block. Echo showed no RWMA with EF 56%. What would be further course of action for this patient?

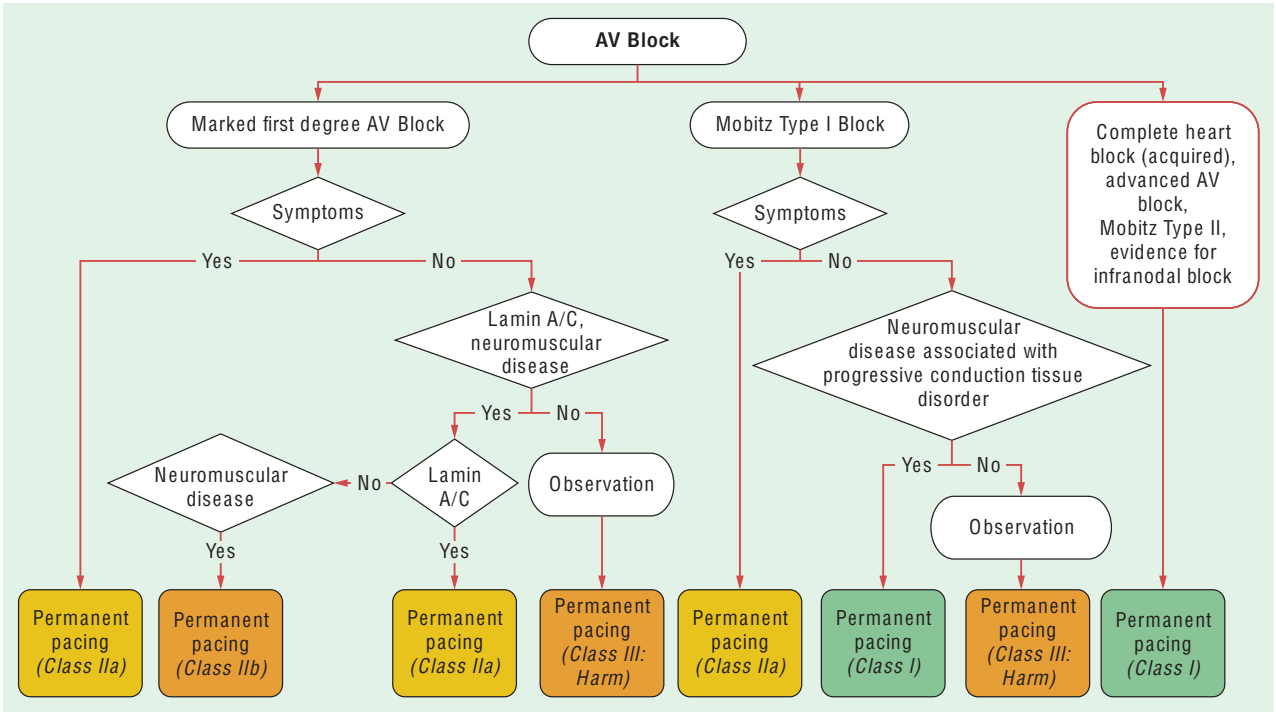
A. Pacemaker

B. Implantable cardioverter defibrillator

C. CRT D

D. Radio frequency ablation

Anwer: A(Page1886)



6. Patient on dabigatran for atrial fibrillation developed massive UGI bleed. What is the antidote that you would give?

A. Idarasizumab

B. Pcsk9 inhibitor

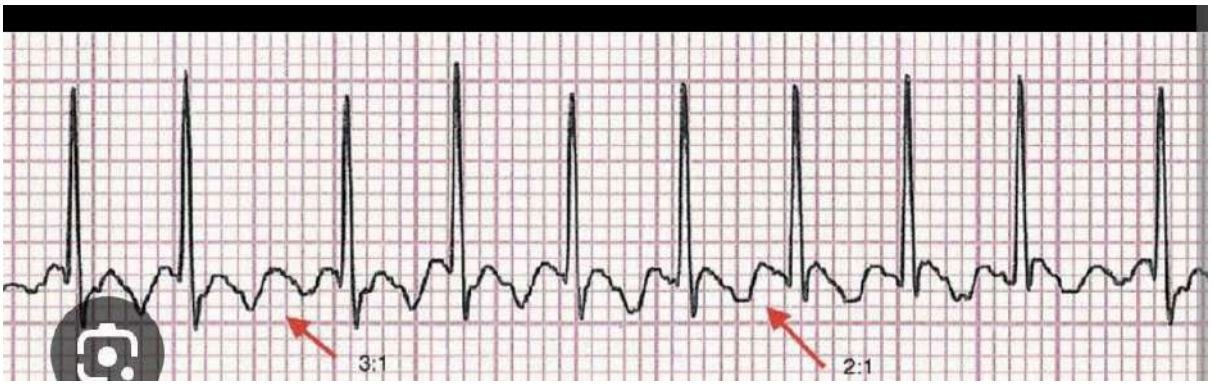
C. Evolocumab

D. Omalizumab

Answer: A(Page 1906)

Warfa- rin anticoagulation can be reversed by administration of fresh frozen plasma, prothrombin complex concentrate, and vitamin K. Reversal agents are available for dabigatran (idarucizumab), and Xa inhibitors are available (andexanet alfa), and both are administered intrave- nously. These agents may be pro-thrombotic and administration must be judicious.

7. Identify the waveform



A. Saw tooth pattern

B. Delta wave

C. Low voltage complex

D. None of the above

Answer:A(Page 1899)

Common or typical right atrial flutter is due to a circuit pathway around the tricuspid valve annulus, bounded anteriorly by the annulus and posteriorly by func- tional conduction block in the crista terminalis. The wavefront passes between the inferior vena cava and the tricuspid valve annulus, known as the sub-Eustachian or cavotricuspid isthmus, where it is susceptible to interruption by catheter ablation. Thus, common atrial flutter is also known as *cavotricuspid isthmus-dependent atrial flutter*. This circuit most commonly revolves in a counterclockwise direction (as viewed looking toward the tricuspid annulus from the ventricular apex), which produces the characteristic negative sawtooth flutter waves in leads II, III, and aVF and positive P waves in lead V_1. When the direction is reversed, clockwise rotation produces the opposite P-wave vector in those leads. The atrial rate is typically 240–300 beats/min but may be slower in the presence of atrial disease or antiarrhythmic drugs. It often conducts to the ventricles with 2:1 AV block, creating a regular tachycardia at 130–150 beats/min, with P waves that may be difficult to discern from the T wave.

Macroreentrant atrial tachycardias (ATs) that are not dependent on conduction through the cavotricuspid isthmus are referred to as *atypical atrial flutters*. They can occur in either atrium and are almost universally associated with areas of atrial scar. Right atrial atypical flut- ter often occurs after cardiac surgery if an atriotomy is performed in the right atrium as part of the surgery. Left atrial flutter and perimitral left atrial flutter are commonly seen after extensive left atrial ablation for atrial fibrillation or atrial surgery

8. Which of the following arrhythmia is not secondary to re-entry phenomena?

A. AVNRT

B. Scar related VT

C. Atrial flutter

D. Digitalis toxicity VT

Answer: D(Page 1869)

Table 243-1 Overview of the Mechanisms of Cardiac Tachyarrhythmias

TACHYARRHYTHMIA CATEGORY	MECHANISM	PROTOTYPICAL ARRHYTHMIAS
Abnormal Automaticity	Enhanced (acceleration of phase 4 repolarization)	Idiopathic VT; AT
	Suppressed (absent or decelerated phase 4 repolarization)	Sinus node dysfunction
Triggered Activity	EADs	TdP in long QT syndrome, PVCs
	DADs	Reperfusion PVCs/VT, AT and VT with digitalis toxicity
Reentry	1) Anatomical or functional confinement of a circuit (i.e., scar, accessory pathway); 2) unidirectional block after a premature impulse; 3) wave of excitation that travels in a single direction returning to its point of origin	AVNRT, AVRT, atrial flutter, scar-related VT

9. Which of the following arrhythmia are not secondary to delayed after depolarisation?

A. Digitalis toxicity

B. Ischemia

C. CPVT

D. TDP in long QT syndrome

Answer:D(Page 1869)

Delayed after depolarisation(Phase 4)	Early after depolarisation(Phase 2-3)
Digitalis toxicity	Bradycardia
Catecholamines	Hypokalemia

Ischemia	Hypomagnesemia
	Electrical remodeeling in CMPs

10. According to Vaughan Williams classification which of this is not a class III anti-arrhythmic drug

    A. Sotalol

    B. Dofetilide

    C. Ibutilide

    D. Flecainide

Answer:D(Page 1871,1872)

The traditional nomenclature of antiarrhythmic drugs (AADs) is known as the Vaughan Williams classification schema. In this schema, there are four classes (I–IV; Table 243-2). Class I AADs primarily target the Na channel, Class II agents target the beta- adrenergic receptor, Class III agents target potassium channels, and Class IV agents target Ca channels. Class I agents are further subdivided into three subclasses based on the kinetics of drug to Na channel interactions. Class IA agents, including procainamide and quinidine, possess intermediate binding kinetics and potency. Class IB agents, including lidocaine and mexiletine, possess rapid binding kinetics and relatively low potency. Class IC agents (fle- cainide, propafenone) possess slow kinetics and high potency. Class II agents consist entirely of beta adrenergic blocking agents. Class III agents (sotalol, dofetilide, ibutilide) specifically target the HERG potassium channel and risk prolongation of the QT interval through effects on the K channel (HERG) that in large part determine phases 2/3 of the AP, and hence ventricular repo- larization. Class IV agents are cardioselective Ca channel blockers including verapamil and diltiazem.

11. Which of the following drugs can promote re-entrant arrhythmia

    A. Sotalol

    B. Ibutilide

    C. Flecainide

    D. Amiodarone

Answer: C(Page 1872)

Table 243-2 Antiarrhythmic Drug Actions

DRUG	CLASS ACTIONS I	II	III	IV	OTHER ACTIONS/COMMON SIDE EFFECTS
Quinidine	++		++		Anticholinergic
Flecainide	+++		+		Can promote reentrant arrhythmias (AFL, VT)
Propafenone	++	+			Mild beta-blocker effect
Amiodarone	++	++	+++	+	Multiorgan toxicity with long-term use
Sotalol		++	+++		Prominent beta-blocker effect
Dofetilide			+++		Prolongation of QT at slower heart rates
Dronedarone	+	+	+	+	Mild effect
Ibutilide			+++		Used only for acute cardioversion
Ranolazine	++		++		Late sodium channel blockade
Lidocaine	++				Used for reperfusion arrhythmias

12. 60 year male with non ischemic dilated cardiomyopathy and EF of 40 percent presents with AV block. Which of the following intervention will be most suitable for him

A. Conventional Dual chamber pacemaker implant

B. Leadless pacemaker implant

C. Coronary angiogram

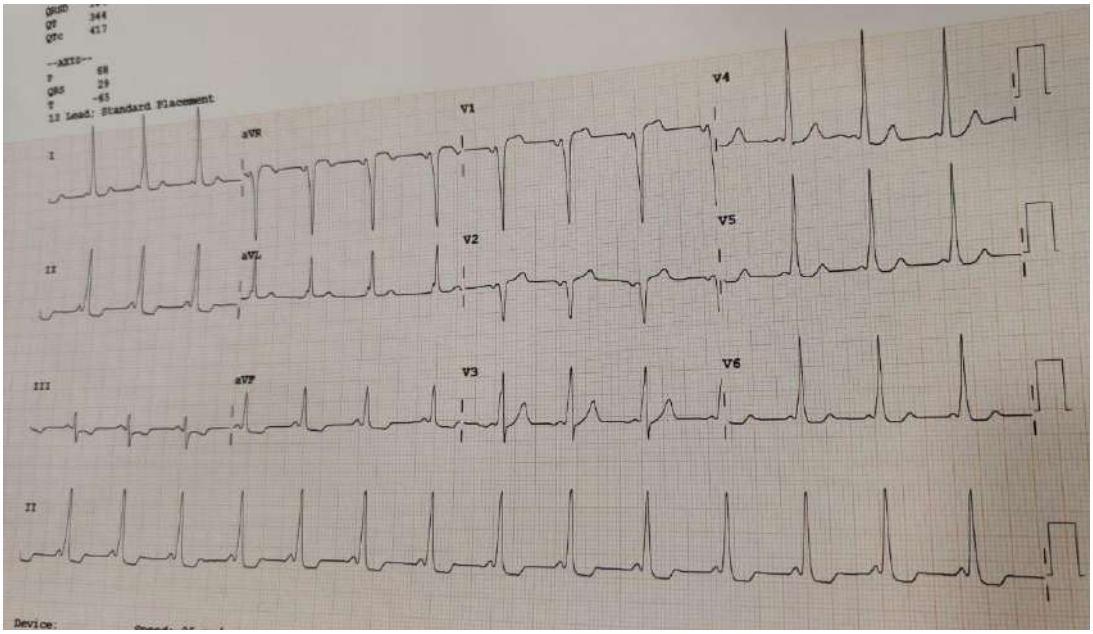
D. His bundle pacing

Answer: D(Page 1886,87)

In patients with left ventricular ejection fraction <50% and AV block who have an indication for permanent pacing and are expected to require ventricular pacing >40% of the time, techniques to provide more physiologic ventric- ular activation are preferred to right ventricular pacing to prevent heart failure. Cardiac resynchronization therapy (CRT) involves placement of an additional pacing lead in a lateral or anterolateral branch of the coronary sinus to allow for simultaneous right ven- tricle and lateral left ventricle pacing leading to a more physiologic left ventricular contraction. CRT pacing has been shown to improve outcomes and mortality in appropriately selected patients. Physiologic ventricular pacing has also been achieved with placement of a ventricular pacing lead in the region of the His bundle. His bundle pacing recruits the specialized conduction system, leading to a more physiologic cardiac contraction. In addition to His bun- dle pacing, left bundle branch area pacing in the proximal inter- ventricular septal region has also been shown to achieve a more physiologic pacing response

13. Below is the ECG of a young patient presenting with palpitations. Localise the accessory pathway.

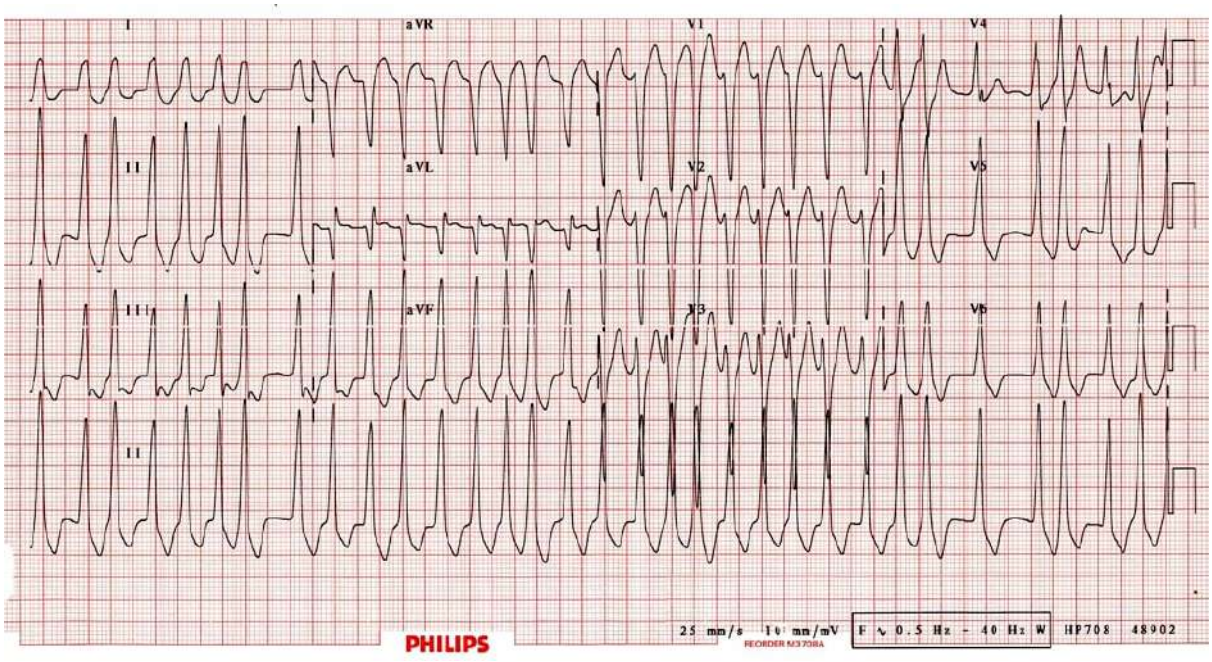
A. Right Anteroseptal
B. Right posteroseptal
C. Right free wall
D. Left anteroseptal



Answer:A (Page 1896)

Right-sided pathways preexcite the right ventricle, producing a left bundle branch block–like configuration in lead V_1, and often create marked preexcitation because of relatively close proximity of the AP to the sinus node (Fig. 249-4). Left-sided pathways preexcite the left ventricle and may produce a right bundle branch–like configuration in lead V_1 and a negative delta wave in aVL, indicating initial depolarization of the lateral portion of the left ventricle that can mimic Q waves of lateral wall infarction (Fig. 249-4). Because of the relatively large distance between the sinus node and left free wall APs, preexcitation may be minimal or absent on 12-lead ECG. Preexcitation due to an AP at the diaphragmatic surface of the heart, typically in the paraseptal region, produces delta waves that are negative in leads III and aVF, mimicking the Q waves of inferior wall infarction

14. Below is the ECG of a patient who presented with sudden onset palpitations and syncope. Which drug should be used?



A. CCBs

B.BBs

C. Adenosine

D. Procainamide

Answer: D(Page 1899)

Very irregular wide-complex tachycardia is most likely preexcited AF or flutter (see above) and should be managed with cardioversion, intravenous procainamide, or ibutilide.

15. A 60 year old male presents with previously untreated but recurrent paroxysmal episodes of atrial fibrillation. What will be the next best choice of treatment?

A. Dabigatran

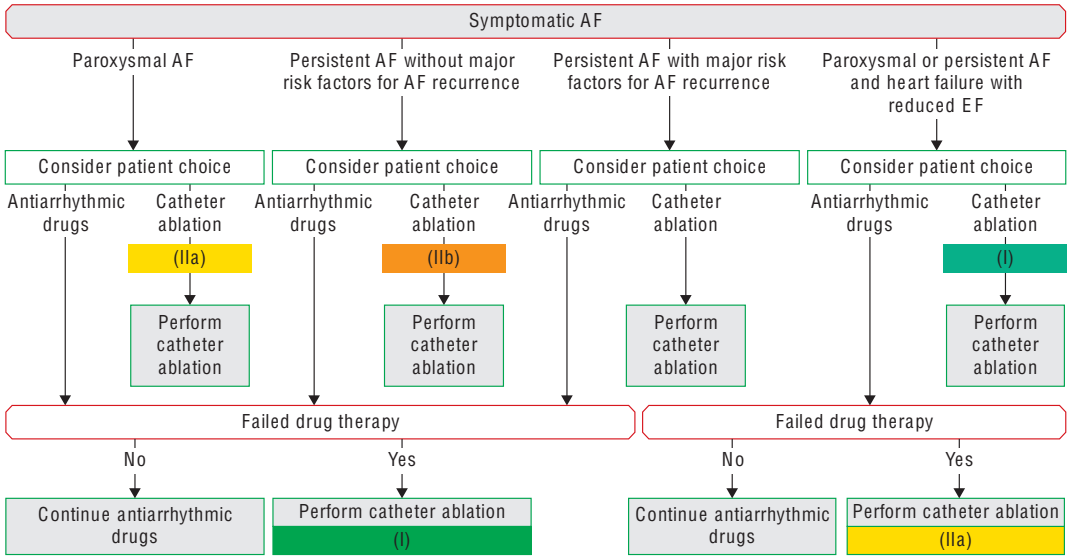
B. Rate control

C. Catheter ablation

D. Surgical ablation

Answer: C(Page 1907-1909)

For patients with previously untreated but recurrent paroxysmal AF, catheter ablation has superior efficacy compared to antiarrhyth- mic drug therapy, and ablation is even more clearly superior to anti- arrhythmic drugs for patients who have recurrent AF despite drug treatment.



16. Which of this is the most common site for idiopathic VT?

A. LV outflow tract

B. RV outflow tract

C. LV fascicular VT

D. Papillary muscle

Answer:B(Page 1919)

1. **Idiopathic ventricular tachycardia (VT) without structural heart disease**
   A. Outflow tract origin
      - Right ventricular (RV) outflow tract: left bundle branch block pattern in V_1 with inferior axis (tall QRS in inferior leads) and late transition in the precordial leads
      - Left ventricular (LV) outflow tract: similar inferiorly directed axis but with early precordial transition with prominent R wave in V_2–V_3

   B. LV fascicular VT: Typical right bundle branch block pattern in V_1 with sharp intrinsicoid deflection and left axis deviation (arising from left posterior fascicle in its most common form)

   C. Papillary muscle VT
      - Posteromedial: atypical right bundle branch block pattern in V_1 with monophasic R wave and left axis deviation
      - Anterolateral: atypical right bundle branch block pattern in V_1 with positive deflection in lead III and negative deflection in lead I

17. A 45 year old male presents with palpitations of 2 months duration. He is being on treatment with 10 mg of bisoprolol however continues to be symptomatic. Holter evaluation showed 15 percent PVC burden. What is the next best line of management

    A. Increase dose of bisoprolol

    B. Pacemaker implantation

    C. ICD implantation

    D. Radio frequency catheter ablation

Answer:D

Treatment is required for symptoms or when frequent or incessant arrhythmias depress ventricular function. Symptoms can be controlled with medications including beta blockers, calcium channel blockers, and sodium channel blockers such as flecainide. Although flecainide is not typically recommended in patients with structural heart disease, it has been used successfully to resolve tachycardia-induced cardiomyopathy in the setting of idiopathic PVCs and VT. Catheter ablation is also indicated for control of symptoms, has an overall success rate of 80%, and is recommended for those with symptomatic VT in whom medications are ineffective or not preferred by the patient. Efficacy and risks of catheter ablation vary with the specific site of origin of the VT, being most favorable for arrhythmias originating in the right ventricular out- flow tract. Failure of ablation is most often due to inability to initiate the arrhythmia for mapping in the electrophysiology laboratory.

18. Which of the following conditions does not typically causes polymorphic VT

    A. In setting of acute ischemia

    B. Scar related VT

C. Congenital long QT syndrome

D.Drug induced VT

Answer: B(Page 1924,25,26)

Acute MI
Congenital Long QT syndrome
Acquired long QT syndrome
Short QT syndrome
Brugada syndrome
Early repolarisation syndrome
CPVT
HCM
Ventricular fibrillation
Genetically determined CMPS

1. Congenital long QT syndromes
   - Long QT syndrome type 1: Reduced repolarizing current I_{Ks} due to mutation in *KCNQ1* gene
   - Long QT syndrome type 2: Reduced repolarizing current I_{Kr} due to mutation in *KCNH2* gene
   - Long QT syndrome type 3: Delayed inactivation of the I_{Na} due to mutations in *SCN5A* gene
   - Others: Several other types of long QT syndromes have been described; long QT syndrome types 1, 2, and 3 account for 80–90% of cases
2. Electrolyte abnormalities: Hypokalemia, hypomagnesemia, hypocalcemia
3. Drug-induced acquired prolongation of QT interval
   - Antiarrhythmic drugs
     - Class IA: Quinidine, disopyramide, procainamide
     - Class III: Sotalol, dronedarone, ranolazine, amiodarone, ibutilide, dofetilide
   - Antibiotics
     - Macrolides: Erythromycin, clarithromycin, azithromycin
     - Fluroquinolones: Levofloxacin, moxifloxacin
     - Trimethoprim-sulfamethoxazole
     - Clindamycin
     - Pentamidine
     - Chloroquine
     - Antifungals: Ketoconazole, itraconazole
     - Antivirals: Amantadine
   - Antipsychotics
     - Haloperidol, phenothiazines, thioridazine, trifluoperazine, sertindole, zimelidine, ziprasidone
     - Tricyclic and tetracyclic antidepressants
   - Antihistamines (histamine 1-receptor antagonists)
     - Astemizole, diphenhydramine, hydroxyzine
   - Other drugs
     - Citrate (massive blood transfusions)
     - Cocaine
     - Methadone
     - Hydroxychloroquine
4. Cardiac conditions
   - Myocardial ischemia and infarction
   - Myocarditis
   - Marked bradycardia
   - Stress cardiomyopathy
5. Endocrine disorders
   - Hypothyroidism
   - Hyperparathyroidism
   - Pheochromocytoma
   - Hyperaldosteronism
6. Intracranial disorders
   - Subarachnoid hemorrhage
   - Thalamic hematoma
   - Cerebrovascular accident
   - Encephalitis
   - Head injury
7. Nutritional disorders
   - Anorexia nervosa
   - Starvation
   - Liquid protein diets
   - Gastroplasty and ileojejunal bypass
   - Celiac disease

19. Which of the following modalities is not used in the setting of acute electrical storm

A. ICD implantation

B. Stellate ganglia block

C. Beta blockers

D. Sedation

Answer: A(Page 1929)

Electrical storm treatments

Speed of Deployment	Stabilize rhythm	Relieve triggers	Reduce sympathetic drive
Rapid	• Defibrillation • Amiodarone • Lidocaine	• Electrolyte management • Volume removal • Coronary revascularization	• Beta blockers • Sedation and intubation • Anxiolytics
	• Quinidine • Ranolazine • Procainamide • Catheter ablation	• Overdrive pacing • Mechanical support (ECMO/IABP)	• Stellate ganglion block (SGB)
Delayed		• Consideration of biopsy/anti-inflammatory therapies	• Cardiac surgical sympathetic denervation

20. Which of the following is the gold standard investigation to measure vessel wall stiffness in the patient's suspected to have hypertension mediated organ damage (HMOD)

a. Ankle brachial index

b. Carotid angiography

c. Pulse wave velocity

d. Femoral artery doppler

Answer: C(page 2075)

An association between arterial stiffness and hypertension is well established. A stiffened vasculature is less able to buffer short-term alterations in flow. Although it has been assumed that arterial stiffness is a manifestation of hypertension, recent evidence suggests that vascu- lar stiffness may also contribute to elevated

arterial pressure. Clinically, noninvasive determination of elevated pulse wave velocity between the carotid and femoral arteries is often interpreted as an indicator of arte- rial stiffness. Due to arterial stiffness, central blood pressures (aortic, carotid) may not correspond to brachial artery pressures. Ejection of blood into the aorta elicits a pressure wave that is propagated at a given velocity. The forward traveling wave generates a reflected wave that travels backward toward the ascending aorta. Although mean arterial pressure is determined by cardiac output and peripheral resistance, pulse pressure is related to the functional properties of large arteries and the amplitude and timing of the incident and reflected waves. Increased arterial stiffness results in increased pulse wave velocity of both incident and reflected waves. The consequence is augmentation of aortic systolic pressure and a reduction of aortic diastolic pressure, i.e., an increase in pulse pressure. The aortic augmentation index, a surrogate index of arterial stiffening, is calculated as the ratio of central arterial pressure to pulse pressure. However, wave reflections are also influenced by left ventricular structure and function. Central blood pressure may be measured directly by placing a sensor in the aorta or noninvasively by radial tonometry. Central blood pressure and the aortic augmentation index are independent predictors of cardiovascu- lar disease and all-cause mortality. Central blood pressure also appears to be more strongly associated with preclinical organ damage than brachial blood pressure.

21. Which of the following patients are having hypertensive urgency

a. 50 year old male with BP- 162/104 with no other complaints

b. 47 female with altered mental status, nausea, headache and BP- 190/120

c. 60 year old male, BP 188/120 with no target organ damage

d. 55 year old female, BP 200/140 with papilledema

Answer: C(page 2086)

Severe asymptomatic hypertension (systolic blood pressure ≥180 mmHg or diastolic blood pressure ≥120 mmHg) is considered a hypertensive “urgency,” but when accompanied by acute target damage, it is considered a hypertensive “emergency.”

22. 55 year old with diabetes, hypertension, dyslipidemia came to emergency with headache, vomiting. On examination he was agitated BP 220/120 and having papilledema. Drug least useful in this situation is

a. Furosemide

b. Nicardipine

c. Labetalol

d. Enalaprilat

Answer: A(page 2087)

TABLE 277-11 Usual Intravenous Doses of Antihypertensive Agents Used in Hypertensive Emergencies[a]

ANTIHYPERTENSIVE AGENT	INTRAVENOUS DOSE
Nitroprusside	Initial 0.3 (g/kg)/min; usual 2–4 (g/kg)/min; maximum 10 (g/kg)/min for 10 min
Nicardipine	Initial 5 mg/h; titrate by 2.5 mg/h at 5–15 min intervals; max 15 mg/h
Labetalol	2 mg/min up to 300 mg *or* 20 mg over 2 min, then 40–80 mg at 10-min intervals up to 300 mg total
Enalaprilat	Usual 0.625–1.25 mg over 5 min every 6–8 h; maximum 5 mg/dose
Esmolol	Initial 80–500 g/kg over 1 min, then 50–300 (g/kg)/min
Phentolamine	5–15 mg bolus
Nitroglycerin	Initial 5 g/min, then titrate by 5 g/min at 3–5-min intervals; if no response is seen at 20 g/min, incremental increases of 10–20 g/min may be used
Hydralazine	10–50 mg at 30-min intervals

23. . A patient with History of frequent exacerabations of COPD undergoing right heart catheterisation for evaluation. Which of the following Mean Pulmonary Arterial Pressure is diagnostic?

a. >20mm/hg

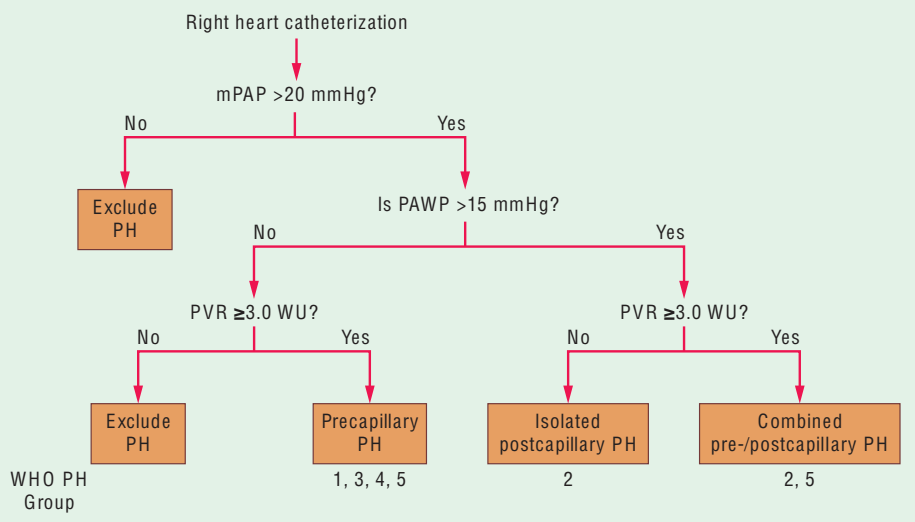
b. >25 mm/hg

c. >32 mm/hg

d. >30 mm/hg

Answer: A(Page 2124)

The RHC remains the gold standard test to both establish the diagnosis of PH and guide selection of appropriate medical therapy. The hemodynamic criteria for diagnosing PH requires, first, an mPAP >20 mmHg. Precapillary and postcapillary PH are then distinguished by virtue of a pulmo- nary artery wedge pressure (PAWP) (or left ventricular end-diastolic pressure [LVEDP]) ≤15 mmHg or >15 mmHg, respectively. Isolated precapillary PH also requires a PVR ≥3.0 Wood units (WU), whereas isolated postcapillary PH is defined by PVR <3.0 WU. Increasingly, combined pre- and postcapillary PH is recognized, defined by elevated mPAP >20 mmHg, PVR ≥3.0 WU, and PAWP >15 mmHg



24. Which of the following groups of patient with PAH has the worst prognosis?

a. HIV associated

b. Scleroderma associated

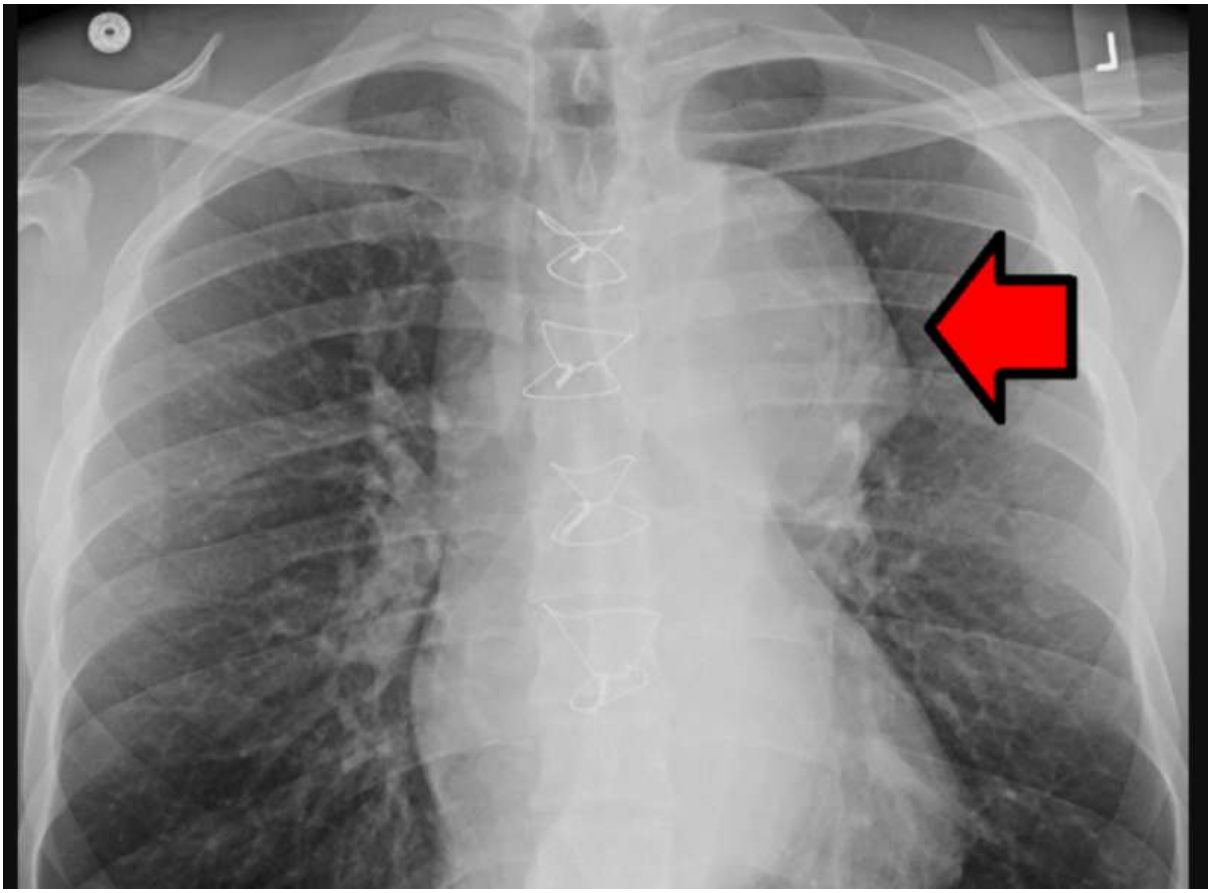
c. Left heart ds associated

d. CKD related

Answer: A(Page A)

The true prevalence of HIV-PAH is not known; however, this PAH subtype is an important cause of mortality in the HIV- infected population, and prognosis in these patients is among the least favorable for all PH subgroups. There is no correlation between the stage of HIV infection and the development of PAH.

25. 40 year old male K/C/O Marfan's Syndrome came to a regular check up. Chest Xray is as shown in the figure below. Following which ECHO was done. Which is the following is the management?



a. Operative repair if diameter is >5 cm

b. Operative repair if diameter is >6 cm

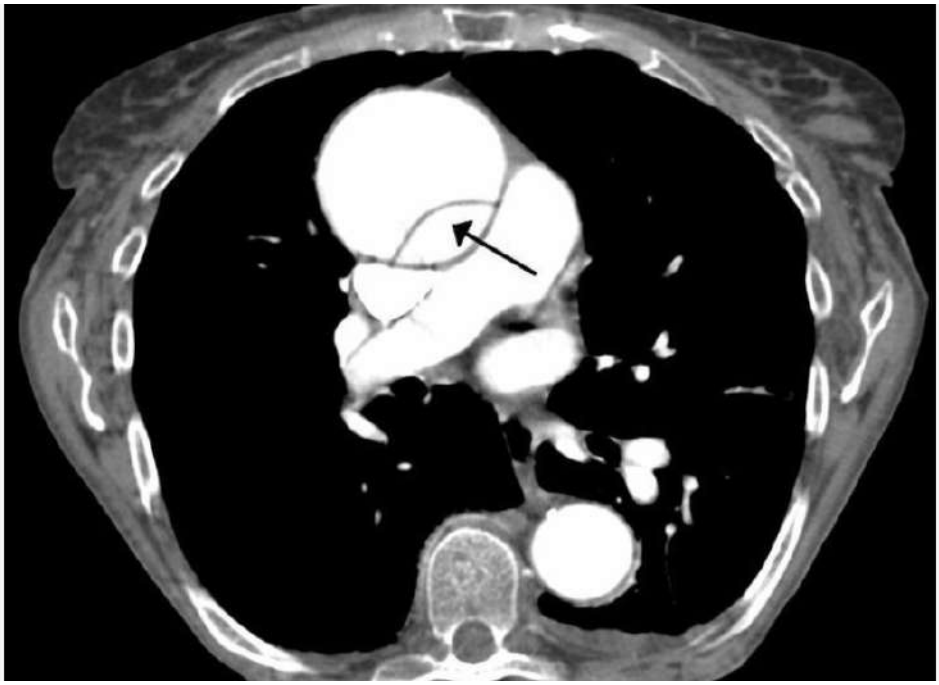
c. Operative repair if diameter is > 5.5 cm

d. Operative repair if diameter is >4.5cm

Answer: D(Page 2103)

Operative repair with placement of a prosthetic graft is indicated in patients with symptomatic ascending thoracic aortic aneurysms, and for most asymptomatic aneurysms, including those associated with bicuspid aortic valves when the aortic root or ascending aor- tic diameter is ≥5.5 cm, or when the growth rate is >0.5 cm per year. Replacement of the ascending aorta >4.5 cm is reasonable in patients with bicuspid aortic valves undergoing aortic valve replace- ment because of severe aortic stenosis or aortic regurgitation. In patients with Marfan's syndrome, ascending thoracic aortic aneu- rysms of 4–5 cm should be considered for surgery. Operative repair is indicated for patients with degenerative descending thoracic aor- tic aneurysms when the diameter is >6 cm, and endovascular repair should be considered if feasible when the diameter is >5.5 cm. Repair is also recommended when the diameter of a descending thoracic aortic aneurysm has increased >1 cm per year.

26. 65 year old male came with acute chest pain radiating to the interscapular region in the back. He is Hypertenisive since 10 years. CT image is as shown in the figure. Drugs contraindicated in this patient is?



a. Hydralazine
b. Esmolol
c. Labetolol
d. Verapamil

Answer: A(Page 2106)

For acute dissection, unless contraindicated, β-adrenergic blockers should be administered parenterally, using intravenous propranolol, metoprolol, or the short-acting esmolol to achieve a heart rate of ~60 beats/min. This should be accompanied by sodium nitroprusside infusion to lower systolic blood pressure to ≤120 mmHg. Labetalol , a drug with both β- and α-adrenergic blocking properties, also may be used as a parenteral agent in acute therapy for dissection.

The calcium channel antagonists verapamil and diltiazem may be used intravenously if nitroprusside or β-adrenergic blockers cannot be employed. The addition of a parenteral angiotensin-converting enzyme (ACE) inhibitor such as enalaprilat to a β-adrenergic blocker also may be considered. Isolated use of a direct

vasodilator such as hydralazine is contraindicated because these agents can increase hydraulic shear and may propagate the dissection.

27. 45 year old female came with complaints of bluish discolouration of finger tips on exposure to cold. Drug to be avoided in this condition is

a. Clonidine

b. Prazosin

c. Diltiazem

d. Isradipine

Answer: A(Page 2113)

TABLE 281-1 Classification of Raynaud's Phenomenon
Primary or idiopathic Raynaud's phenomenon
Secondary Raynaud's phenomenon
Collagen vascular diseases: scleroderma, systemic lupus erythematosus, rheumatoid arthritis, dermatomyositis, polymyositis, mixed connective tissue disease, Sjögren's syndrome
Arterial occlusive diseases: atherosclerosis of the extremities, thromboangiitis obliterans, acute arterial occlusion, thoracic outlet syndrome
Pulmonary hypertension
Neurologic disorders: intervertebral disk disease, syringomyelia, spinal cord tumors, stroke, poliomyelitis, carpal tunnel syndrome, complex regional pain syndrome
Blood dyscrasias: cold agglutinins, cryoglobulinemia, cryofibrinogenemia, myeloproliferative disorders, lymphoplasmacytic lymphoma
Trauma: vibration injury, hammer hand syndrome, electric shock, cold injury, typing, piano playing
Drugs and toxins: ergot derivatives, methysergide, -adrenergic receptor blockers, bleomycin, vinblastine, cisplatin, gemcitabine, vinyl chloride

28. ECHO in a patient with pulmonary embolism shows akinesia of RV free wall while normal motion of RV apex. This sign is?

a. Moose's sign

b. Mc Connel Sign

c. Homan's Sign

d. Lowenback sign

Answer: B(page 2097)

Echocardiography is *not* a reliable diagnostic imaging tool for acute PE because most patients with PE have normal echocardiograms. However, echocardiography is a very useful diag- nostic tool for detecting conditions that may mimic PE, such as acute myocardial infarction, pericardial tamponade, and aortic dissection. Transthoracic echocardiography rarely images thrombus directly. The best-known indirect sign of PE on transthoracic echocardiography is McConnell's sign: hypokinesis of the RV free wall with normal or hyperkinetic motion of the RV apex. One should consider transesoph- ageal echocardiography when CT scanning facilities are not available or when a patient has renal failure or severe contrast allergy that precludes administration of contrast despite premedication with high- dose steroids. This imaging modality can identify saddle, right main, or left main PE.

29. A middle aged man presents to the emergency room with a sudden onset of pain in the right leg. The pain is associated with swelling, redness and tenderness. Which Of the following is false regarding the management of this condition?

a. Recurrence of this condition despite intensive anticoagulation is an indication of IVC filter

b. IVC filters can prevent recurrent PE in patients of right heart failure

c. IVC filters reduce the rate of recurrence of this condition

d. Pulmonary thromboendarterectomy is done in patients with chronic thromboembolic pulmonary hypertension

Answer: B(Page 2098)

Anticoagulation or placement of an inferior vena cava (IVC) filter constitutes *secondary prevention* of VTE. IVC filters are indicated in patients with an absolute contraindication to anticoagulation and for those who have suffered recurrent VTE while receiving therapeutic doses of anticoagulation. Under most circumstances, IVC filters are not indicated for primary prevention of VTE. The IVC filter should be retrieved if the clinician judges that the patient no longer requires it.

30. In a patient with a Well's score of 1.5, which of the following should be initial investigation?

a. D-dimer levels

b. CT-PA

c. V/Q scan

d. Doppler

Answer: A(Page 2095)

TABLE 279-2 Clinical Decision Rules

Low Clinical Likelihood of DVT if Point Score Is Zero or Less; Moderate Likelihood if Score Is 1 to 2; High Likelihood if Score Is 3 or Greater	
CLINICAL VARIABLE	**DVT SCORE**
Active cancer	1
Paralysis, paresis, or recent cast	1
Bedridden for >3 days; major surgery <12 weeks	1
Tenderness along distribution of deep veins	1
Entire leg swelling	1
Unilateral calf swelling >3 cm	1
Pitting edema	1
Collateral superficial nonvaricose veins	1
Alternative diagnosis at least as likely as DVT	–2
High Clinical Likelihood of PE if Point Score Exceeds 4	
CLINICAL VARIABLE	**PE SCORE**
Signs and symptoms of DVT	3.0
Alternative diagnosis less likely than PE	3.0
Heart rate >100/min	1.5
Immobilization >3 days; surgery within 4 weeks	1.5
Prior PE or DVT	1.5
Hemoptysis	1.0
Cancer	1.0

31. 45 year old woman underwent bariatric surgery. On post operative day 4, she developed pulmonary embolism as confirmed by CT-PA. Her BP was 70/44 mm/hd with poor urine output. IV fluids were administered with dobutamine after which the BP increased to 82/54mm/hg.

Which is the most appropriate next step to manage this patient.

a. LMWH

b. IVC filter

c. Mechanical pulmonary thrombectomy

d. Thrombolysis with tPA

Answer: C(Page 2100)

Contraindications to fibrinolysis include intracranial disease, recent surgery, and trauma. The overall major bleeding rate is ~10%, including a 2–3% risk of intracranial hemorrhage. Careful screening of patients for contraindications to fibrinolytic therapy is the best way to minimize bleeding risk.

For patients with submassive PE who have preserved systolic blood pressure but moderate or severe RV dysfunction, use of fibrinolysis remains controversial. A 2019 American Heart Asso- ciation Scientific Statement suggests considering advanced therapy with thrombolysis or embolectomy in patients with lack of improve- ment, clinical deterioration, severe physical distress with anticoag- ulation alone, clot in transit, severe or persistent RV strain, signs of low cardiac output, low bleeding risk, and good life expectancy.

32. Which one of the following regarding intra vascular imaging and physiology is not true?

A. OCT has a better spatial resolution than IVUS

B. IVUS has a better depth of field

C. A FFR more than 0.80 suggests hemodynamic significant stenosis

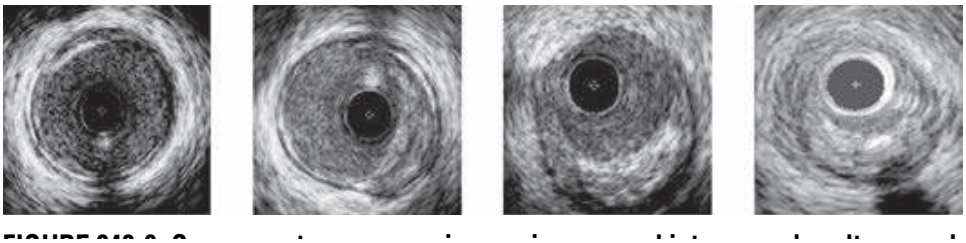
D. A coronary flow reserve less than 2 is considered abnormal

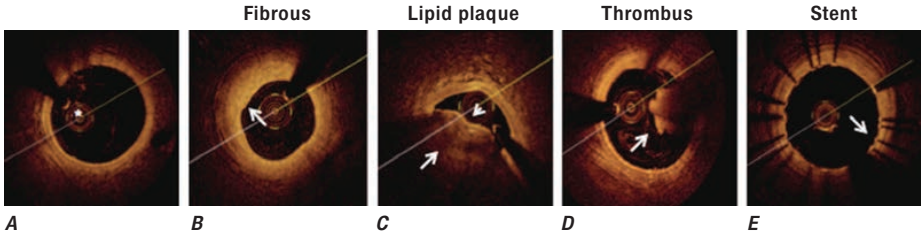
Answer: C(Page 1865)

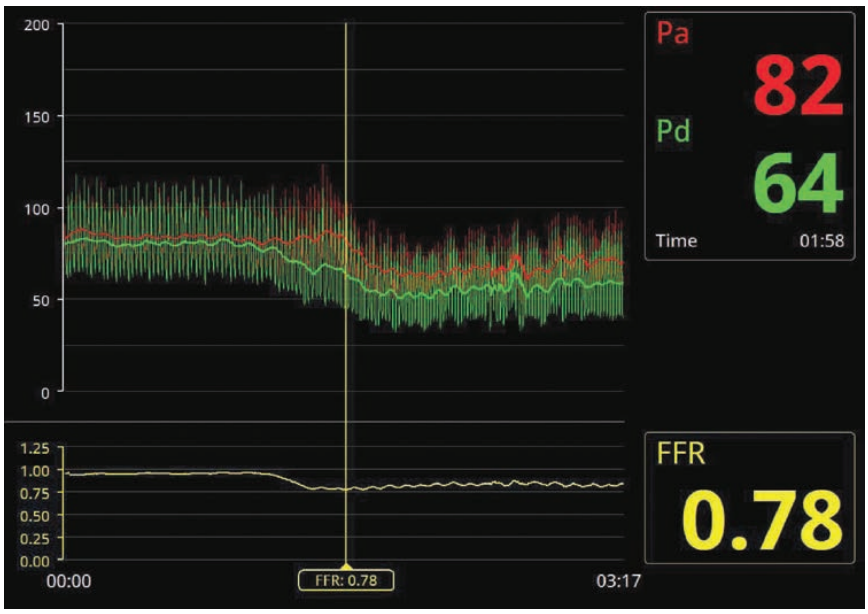
Intravascular ultrasound (IVUS) provides a more accurate anatomic assessment of the coronary artery and the degree of coronary athero- sclerosis . IVUS is performed using a small flexible catheter with a 40-mHz transducer at its tip that is advanced into the coronary artery over a guidewire. Data from IVUS studies may be used to image atherosclerotic plaque precisely, determine luminal cross-sectional area, and measure vessel size; it is also used during or following percutaneous coronary intervention to assess the stenosis and determine the adequacy of stent placement. Optical coherence tomography (OCT) is a catheter- based imaging technique that uses near-infrared light to generate images with better spatial resolution than IVUS (12–18 microns vs 150– 200 microns); however, the depth of field is smaller. The advantage of OCT imaging over IVUS lies in its ability to image characteristics of the atherosclerotic plaque (lipid, fibrous cap) with high definition and to assess coronary stent placement, apposition, and patency

Fractional flow reserve is measured using a coronary pressure–sensor guidewire at rest and at maximal hyperemia following the infusion of adenosine . A fractional flow reserve of <0.80 indicates a hemodynamically significant stenosis that would benefit from intervention.

Microvascular dysfunction can be evaluated by assessing coronary flow reserve, the ratio between coronary blood flow at maximal hyper- emia and rest. Coronary flow reserve is measured using a Doppler wire– or pressure wire–based thermodilution technique in patients with unexplained chest pain or ischemia and no obstructive coronary artery disease. A coronary flow reserve <2.0 is considered abnormal.







33. Which of the following statements regarding restrictive cardiomyopathy is not true?

A. Prominent right atrial y descent

B. Right ventricular systolic pressure more than 60 mm Hg

C. Right and left ventricular systolic pressure relationship concordance

D. RVEDP and LVEDP difference less than 5 mm Hg

Answer: D(Page 1863)

TABLE 242-3 Hemodynamic Findings in Tamponade, Constrictive Pericarditis, and Restrictive Cardiomyopathy

	CARDIAC TAMPONADE	CONSTRICTIVE PERICARDITIS	EFFUSIVE-CONSTRICTIVE PERICARDITIS	RESTRICTIVE CARDIOMYOPATHY
Pericardial pressure				Normal
Right atrium pressure			(Fails to decrease by 50% or to <10 mmHg after pericardiocentesis)	
Right atrium pressure waveform	Prominent "x" descent Diminished or absent "y" descent	Prominent "x" descent Prominent "y" descent	Prominent "x" descent "y" descent less prominent than expected	Prominent "y" descent
Right ventricle systolic pressure	<50 mmHg	<50 mmHg	<50 mmHg	>60 mmHg
Right ventricle end-diastolic pressure	Equals left ventricular end-diastolic pressure within 5 mmHg	>1/3 right ventricular systolic pressure Equals left ventricular end-diastolic pressure within 5 mmHg	>1/3 right ventricular systolic pressure Equals left ventricular end-diastolic pressure within 5 mmHg	<1/3 right ventricular systolic pressure Less than left ventricular end-diastolic pressure by ≥5 mmHg
Right ventricle pressure waveform		Dip and plateau or "square root" sign	Dip and plateau or "square root" sign	Dip and plateau or "square root" sign
Right ventricle–left ventricle systolic pressure relationship with inspiration	Discordant	Discordant	Discordant	Concordant

34. Which of the following normal hemodynamic measurements are not true?

A. Mean RA pressure 0-5 mm Hg

B. Mean PCWP 4-12 mm Hg

C. Cardiac index 2.8-4.2 L-min/m2

D. PVRI 140-200 dyn-s/cm2

Answer: D(Page 1861)

TABLE 242-2 Normal Values for Hemodynamic Measurements

Pressures (mmHg)	
Right atrium	
Mean	0–5
a wave	1–7
v wave	1–7
Right ventricle	
Peak systolic/end diastolic	17–32/1–7
Pulmonary artery	
Peak systolic/end diastolic	17–32/1–7
Mean	9–19
Pulmonary capillary wedge (mean)	4–12
Left atrium	
Mean	4–12
a wave	4–15
v wave	4–15
Left ventricle	
Peak systolic/end diastolic	90–130/5–12
Aorta	
Peak systolic/end diastolic	90–130/60–85
Mean	70–100
Resistances ([dyn-s]/cm^5)	
Systemic vascular resistance	900–1400
Pulmonary vascular resistance	40–120
Oxygen Consumption Index ([L-min]/m^2)	**115–140**
Arteriovenous oxygen difference (vol %)	3.5–4.8
Cardiac index ([L-min]/m^2)	2.8–4.2

35. Which test is used to check the integrity of arcuate system of hand.

A. Harvey’s test.

B. Millers test

C. Allen’s test

D. Harvey’s test

Answer: C(Page 1860)

Cardiac catheterization procedures are performed using a percutaneous technique to enter the femoral or radial artery and femoral, brachial, or internal jugular vein as the access sites for left and right heart catheterization, respectively. A flexible sheath is inserted into the vessel over a guidewire, allowing diagnostic catheters to be introduced into the vessel and advanced toward the heart using fluoroscopic guidance. The radial artery (or rarely the brachial artery) access site is advantageous in patients with peripheral arterial disease that involves the abdominal aorta, iliac, or femoral vessels; severe iliac artery tortuosity; morbid obesity; or preference for early postprocedure ambulation. Use of radial artery access is the preferred access route due to a lower rate of access-site bleeding complications and improved patient comfort. A normal modified Allen’s test or Barbeau test con- firming dual blood supply to the hand from the radial and ulnar arter- ies is recommended prior to access at this site.

36. Which of the following regarding contrast induced AKI is not true?

A. Defined as increase in creatinine more than 25 percent at 48-72 hours

B. Occurs in 2-7 percent population

C. Pretreatment with N- acetyl cysteine has shown to reduce its risk

D. People with DM , old age and heart failure are at increased risk

Answer: C(Page 1860)

Contrast-induced acute kidney injury, defined as an increase in crea- tinine >0.5 mg/dL or 25% above baseline that occurs 48–72 h after con- trast administration, occurs in ~2–7% of patients with rates of 20–30% reported in high-risk patients, including those with diabetes mellitus, congestive heart failure, chronic kidney disease, anemia, older age, or who present with an ST-segment elevation myocardial infarction. Dial- ysis is required in 0.3–0.7% of patients and is associated with a fivefold increase in in-hospital mortality. For all patients, adequate intravascular volume expansion with intravenous 0.9% saline (1.0–1.5 mL/kg per hour) for 3–12 h before and continued 6–24 h after the procedure limits the risk of contrast-induced acute kidney injury by >50%. Pre- treatment with *N*-acetylcysteine (Mucomyst) has not reduced the risk of contrast-induced acute kidney injury consistently and, therefore, is no longer recommended routinely. Diabetic patients treated with met- formin should stop the drug 24 h prior to the procedure and not restart until 48 h after contrast administration to limit the associated risk of lactic acidosis. Other strategies to decrease risk include the adminis- tration of sodium

bicarbonate (3 mL/kg per hour) 1 h before and 6 h after the procedure (similar outcome to saline infusion); use of low- or iso-osmolar contrast agents; and limiting the volume of contrast to <50 mL per procedure.

37. Which of the following regarding Doppler echocardiography is true?

A. Pulse wave Doppler is used to interrogate high velocity flow

B. CW Doppler can interrogate velocity at a specific depth location

C. M mode echo has high temporal resolution and accuracy for making linear measurements

D. Doppler shift is inversely related to pressure gradient between two chambers

Answer:C(Page 1832)

Although M-mode echocardiography has largely been supplanted by two-dimensional (2D) echocardiography, it is still used because of its high temporal resolution and accuracy for making linear measurements.

There are three types of Doppler ultrasound that are typically used in standard echocardiographic examinations: spectral Doppler, which consists of both pulsed wave Doppler and continuous wave Doppler, and color flow Doppler. Both types of spectral Doppler will display a waveform representing the velocity of blood flow, with time on the horizontal axis and velocity on the vertical axis. Pulsed wave Doppler is used to interrogate relatively low velocity flow and has the ability to determine blood flow velocity at a particular location within the heart. Continuous wave Doppler is used to assess high-velocity flow, but it can only identify the highest velocity in a particular direction and can- not interrogate the velocity at a specific depth location.

Echocardiography can be used to interrogate blood flow within the heart and blood vessels by using the Doppler principle to ascertain the velocity of blood flow. When ultra- sound emitted from a transducer reflects off red blood cells that are moving toward the transducer, the reflected ultrasound will return at a slightly higher frequency than emitted; the opposite is true when flow moves away from the transducer. That frequency difference, termed the *Doppler shift*, is directly related to the velocity of the flow of the red blood cells. The velocity of blood flow between two chambers will be directly related to the pressure gradient between those chambers

38. Which of the following is not a feature of moderate or grade II diastolic dysfunction ?

A. A> E

B. Deceleration time > 140 ms

C. E/E' less than 10

D. Pulmonary vein S< D

Answer: C(Page 1839)

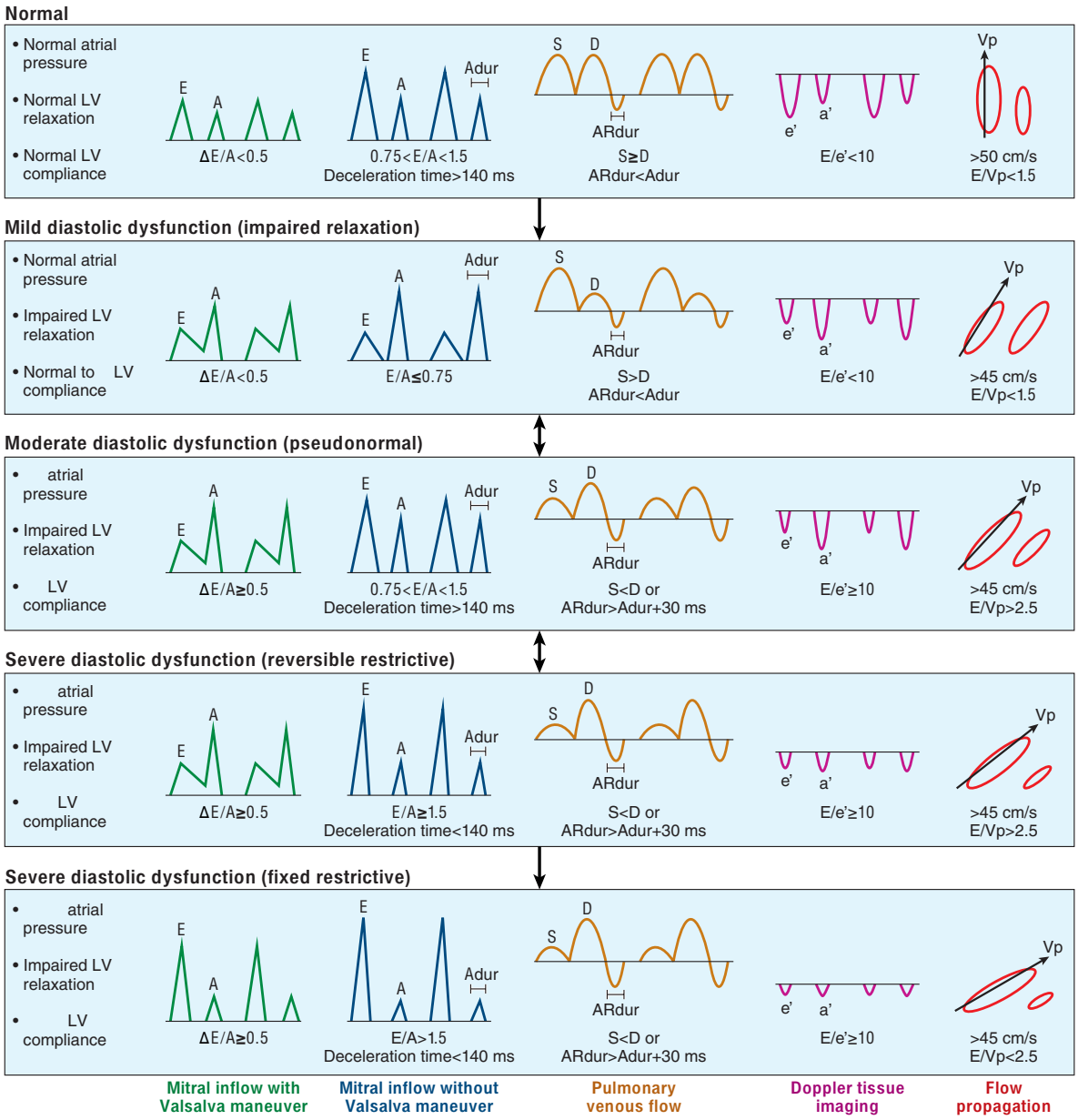


FIGURE 241-8 Stages of diastolic function based on various parameters, including mitral inflow (with and without Valsalva maneuver), Doppler tissue imaging, pulmonary venous flow, and flow propagation. *(Adapted from MM Redfield et al: Burden of systolic and diastolic ventricular dysfunction in the community: appreciating the scope of the heart failure epidemic. JAMA 289:194, 2003.)*

39. Which of the following statement regarding cardiac MRI is false ?

A. T2 weighed sequences can estimate myocardial infarction edema and iron infiltration

B.T1 sequences can assess cardiac function, blood flow and perfusion

C. Nephrogenic systemic fibrosis is common with group II gadolinium based contrast agents

D. CMR has allowed spatial resolution compared to SPECT in diagnosis of ischemia

Answer: C(Page 1837,40,43)

In CMR, T1-weighted pulse sequences are most common, and they assess cardiac structure and function, blood flow, and myocardial perfusion with pharmacologic stress. T2-weighted and $T2^*$-weighted pulse sequences, on the other hand, evaluate myocardial edema and myocardial iron infiltration, respectively

Older generation (group I) linear-structured GBCAs have been associated with a rare but serious condition known as nephrogenic systemic fibrosis (NSF), which is an interstitial inflammatory reaction manifested as fibrosis of tissues or internal organs and even death. Risk factors to developing NSF include high-dose use in presence of severe renal dysfunction (eGFR <30 mL/min per 1.73 m^2), need for hemodialysis, an eGFR <15 mL/min per 1.73 m^2, acute renal deterioration, and concur- rent proinflammatory/systemic illnesses. Newer-generation (group II) macrocyclic-structured GBCAs have a substantially improved safety profile, including in patients with chronic kidney dysfunction and, indeed, have become the agents of choice in most MRI centers. American College of Radiology considers the use of group II agents as safe, including in patients with renal dysfunction or dialysis. With widespread use of group II GBCAs, routine pretest screening, and weight-based dosing, a near-zero incidence of NSF has been reported in the past decade.

The advantages of stress perfusion CMR over SPECT include its higher spatial resolution, which allows detection of subendocardial ischemia or infarction that may be missed by SPECT. As with other imaging modalities, stress CMR studies also provide robust risk stratification. In a recent randomized controlled trial, a stress CMR–guided strategy was shown to improve the guidance toward the use of invasive investigation and coronary revascularization.

40. Which of the following imaging modalities have highest sensitivity in detection of coronary artery disease?

A. Ct coronary angiogram

B. Dobutamine echocardiography

C. Myocardial perfusion PET

D. SPECT MPI

Answer: A(Page 1840)

TABLE 241-3 Comparative Diagnostic Accuracy of Cardiac Imaging Approaches to Coronary Artery Disease

IMAGING MODALITY	PUBLISHED DATA	SENSITIVITY	SPECIFICITY
Exercise echocardiography	15 studies (n = 1849 patients)	84%	82%
Dobutamine echocardiography	28 studies (n = 2246 patients)	80%	84%
SPECT MPI	113 studies (n = 11,212 patients)	88%	76%
Myocardial perfusion PET	9 studies (n = 650 patients)	93%	81%
CMR perfusion	37 studies (n = 2841 patients)	91%	81%
CMR wall motion	14 studies (n = 754 patients)	83%	86%
Coronary CTA	18 studies (n = 1286 patients)	99%	89%

41. Which of the following does not cause mid diastolic murmur at apex

A. Flow murmur of ASD

B. Austin flint murmur

C. Flow murmur of VSD

D. Carey coomb murmur

Answer: A(Page 1822)

MS is the classic cause of a mid- to late diastolic murmur, which is best heard over the apex in the left lateral decubitus position, is low- pitched or rumbling, and is introduced by an OS in the early stages of the rheumatic disease process. Functional" mitral or tricuspid stenosis refers to the generation of mid-diastolic murmurs that are created by increased and accelerated transvalvular diastolic flow, even in the absence of valvular obstruction, in the setting of severe MR, severe TR, or a large ASD with left- to-right shunting. The Austin Flint murmur of chronic severe AR is a low-pitched mid- to late apical diastolic murmur that sometimes can be confused with MS. Unusual causes of a mid-diastolic murmur include atrial myxoma, complete heart block, and acute rheumatic mitral valvulitis.

42. Reverse splitting of S2 is seen in all except:

A. LBBB

B. RV Pacing

C. LV pacing

D. LV dysfunction

Answer: B(Page 1820)

Causes

LBBB

RV Pacing

Acute Myocardial Ischemia

Severe AS

LV dysfunction

43. continuous murmur is found in all except:

A. mitral stenosis with mitral regurgitation

B. patent ductus arteriosus

C. Rupture of sinus of Valsalva

D. systemic arteriovenous fistula

Answer: A(Page 1822)

A continuous murmur is predicated on a pressure gradient that persists between two cardiac chambers or blood vessels across systole and diastole. The murmurs typically begin in sys- tole, envelop the second heart sound (S_2), and continue through some portion of diastole. They can often be difficult to distinguish from individual systolic and diastolic murmurs in patients with mixed valvular heart disease. The classic example of a continuous murmur is that associated with a PDA, which usually is heard in the second or third interspace at a slight distance from the sternal border. Other causes of a continuous murmur include a ruptured sinus of Valsalva aneurysm with creation of an aortic–right atrial or right ventricular fistula, a cor- onary or great vessel arteriovenous fistula, and an arteriovenous fistula constructed to provide dialysis access. There are two types of benign continuous murmurs. The cervical venous hum is heard in children or adolescents in the supraclavicular

fossa. It can be obliterated with firm pressure applied to the diaphragm of the stethoscope, especially when the subject turns his or her head toward the examiner. The mammary soufflé of pregnancy relates to enhanced arterial blood flow through engorged breasts. The diastolic component of the murmur can be oblit- erated with firm pressure over the stethoscope.

44. Which of the following is not true?

A. pulmonary ejection click increased on inspiration

B. with squatting the murmur of MVP decreased

C. the murmur of MS increases on expiration

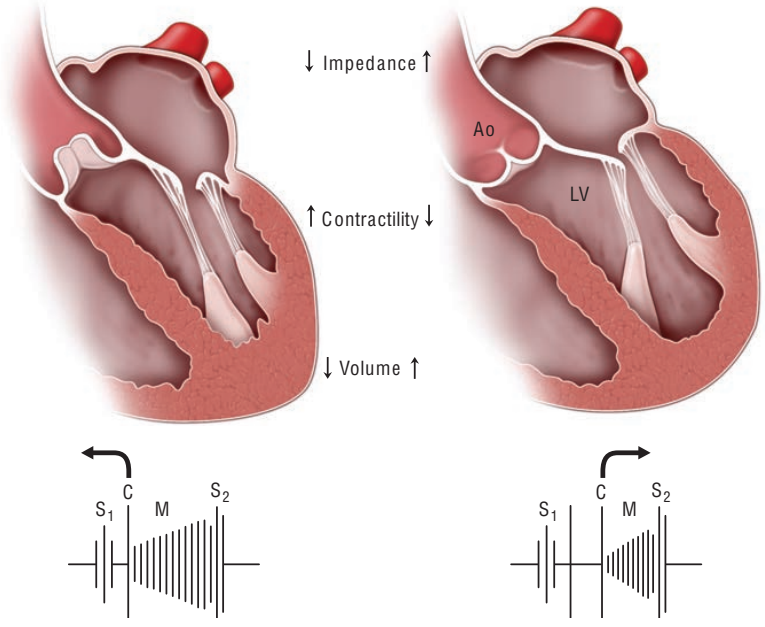
D. the murmur of MR increases on hand gripping

Answer: B(Page 1822,23)

Except for the pulmonic ejection sound, right-sided events increase in intensity with inspiration and decrease with expiration; left-sided events behave oppositely (100% sensitivity, 88% specificity).

The murmur of MR is best heard over the cardiac apex. The intensity of the murmur increases with maneuvers that increase LV afterload, such as sustained hand grip. The murmur of a VSD (without significant pulmonary hypertension) is holosystolic and loudest at the mid-left sternal border, where a thrill is usually present. The murmur of TR is loudest at the lower left sternal border, increases in intensity with inspiration (Carvallo's sign), and is accompanied by visible *cv* waves in the jugular venous wave form and, on occasion, by pulsatile hepatomegaly.

TABLE 239-1 Effects of Physiologic Interventions on the Intensity of Heart Murmurs and Sounds
Respiration
Right-sided murmurs and sounds generally increase with inspiration, except for the PES. Left-sided murmurs and sounds are usually louder during expiration.
Valsalva Maneuver
Most murmurs decrease in length and intensity. Two exceptions are the systolic murmur of HOCM, which usually becomes much louder, and that of MVP, which becomes longer and often louder. After release of the Valsalva maneuver, right-sided murmurs tend to return to control intensity earlier than do left-sided murmurs.
After VPB or AF
Murmurs originating at normal or stenotic semilunar valves increase in the cardiac cycle after a VPB or in the cycle after a long cycle length in AF. By contrast, systolic murmurs due to AV valve regurgitation do not change or become shorter (MVP).
Positional Changes
With *standing*, most murmurs diminish, with two exceptions being the murmur of HOCM, which becomes louder, and that of MVP, which lengthens and often is intensified. With *squatting*, most murmurs become louder, but those of HOCM and MVP usually soften and may disappear. Passive leg raising usually produces the same results.
Exercise
Murmurs due to blood flow across normal or obstructed valves (e.g., PS, MS) become louder with both isotonic and submaximal isometric (hand grip) exercise. Murmurs of MR, VSD, and AR also increase with hand grip exercise. However, the murmur of HOCM often decreases with nearly maximum hand grip exercise. Left-sided S_4 and S_3 sounds are often accentuated by exercise, particularly when due to ischemic heart disease.



45. Which of the following is a not a cause of cannon waves in JVP

A. Junctional rhythm

B. Complete heart block

C. Ventricular tachycardia

D. Avnrt

Answer: D

A prominent *a* wave is seen in patients with reduced right ventricular compliance; a cannon *a* wave occurs with atrioventricular (AV) dissociation and right atrial contraction against a closed tricuspid valve. In a patient with a wide complex tachycardia, the appreciation of cannon *a* waves in the jugular venous waveform identifies the rhythm as ventricular in origin.

46.Which of the following doesn't cause Kussamauls sign in JVP

A. Constrictive pericarditis

B. Advanced LV systolic failure

C. Cardiac tamponade

D. RCM

Answer:C(Page 1817)

Normally, the venous pressure should fall by at least 3 mmHg with inspiration. Kussmaul's sign is defined by either a rise or a lack of fall of the JVP with inspiration and is classically associated with constrictive pericarditis, although it has been reported in patients with restrictive cardiomyopathy, massive pulmonary embolism, right ventricular infarction, and advanced left ventricular (LV) systolic heart failure. It is also a common, isolated finding in patients after cardiac surgery without other hemodynamic abnormalities.

47. Which of the following does not cause discrepancy in both upper limb blood pressures?

A. Subclavian artery ds

B. Supravalvular AS

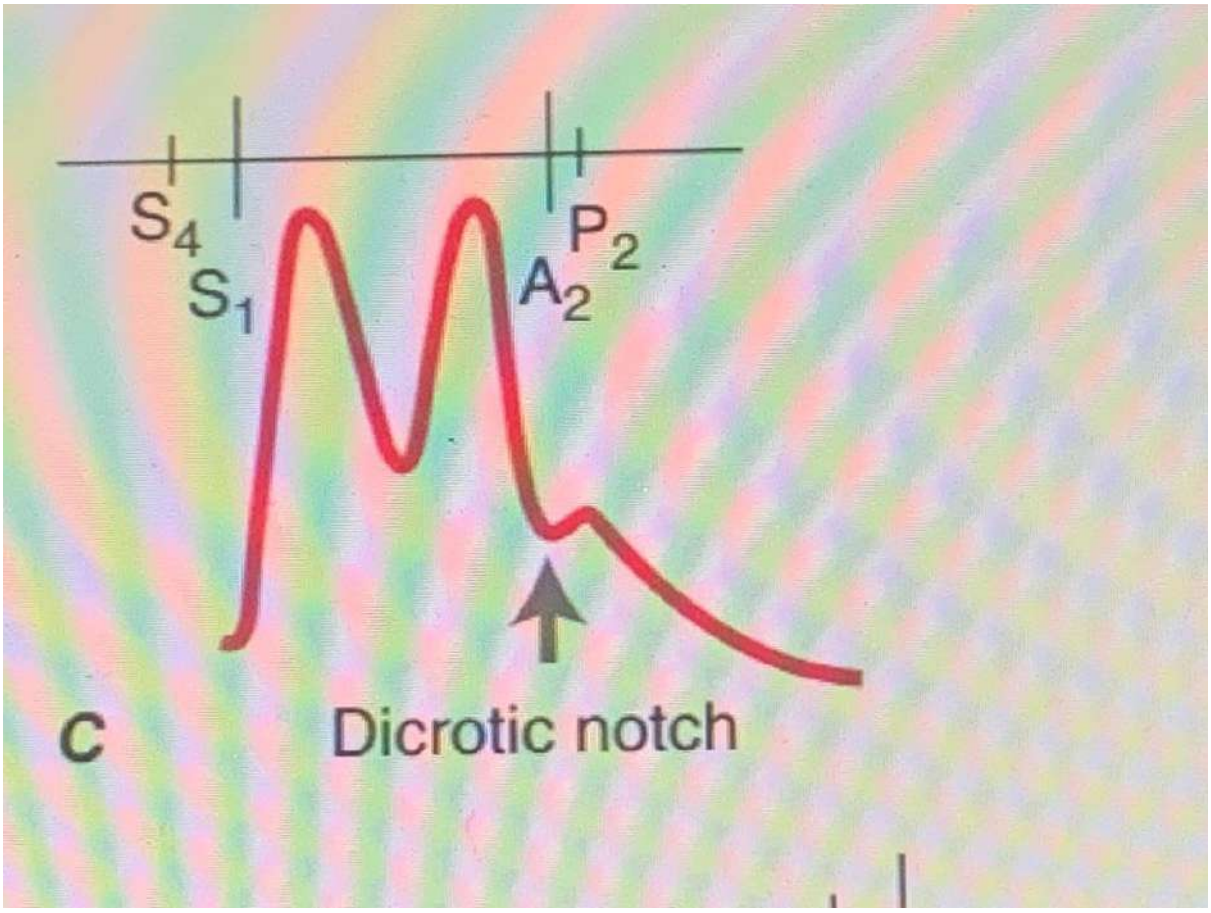
C. Aortic dissection

D. Aortic coarctation with post ductal coarctation

Answer: D(Page 1818)

Blood pressure should be mea- sured in both arms, and the difference should be <10 mmHg. A blood pressure differential that exceeds this threshold may be associated with atherosclerotic or inflammatory subclavian artery disease, supravalvu- lar aortic stenosis, aortic coarctation, or aortic dissection. Systolic leg pressures are usually as much as 20 mmHg higher than systolic arm pressures. Greater leg–arm pressure differences are seen in patients with chronic severe AR as well as patients with extensive and calcified lower extremity peripheral arterial disease. The ankle-brachial index (systolic pressure in the dorsalis pedis and/or posterior tibial artery divided by the higher of the two brachial artery pressures) is a powerful predictor of long-term cardiovascular mortality.

48. Following figure 1. shows configurational change in carotid pulse. Which condition is associated with the given tracing



A. HOCM

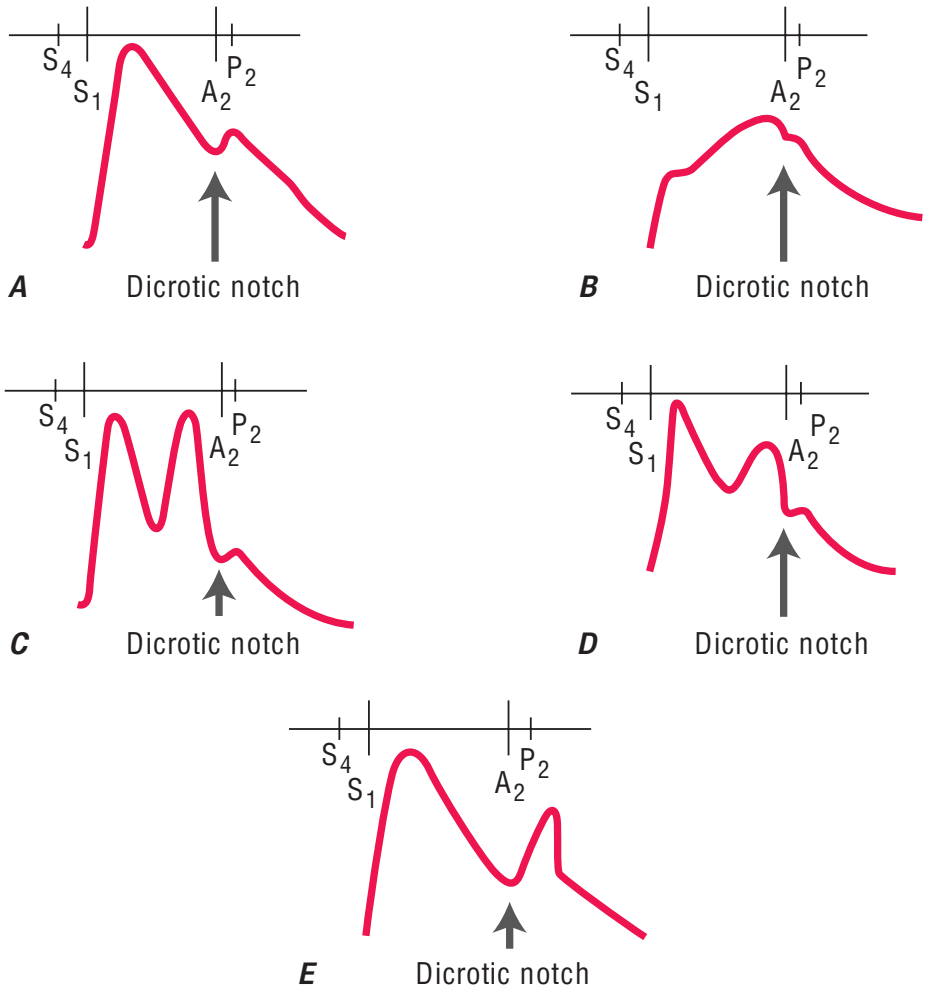
B. Aortic regurgitation

C. Post IABP insertion

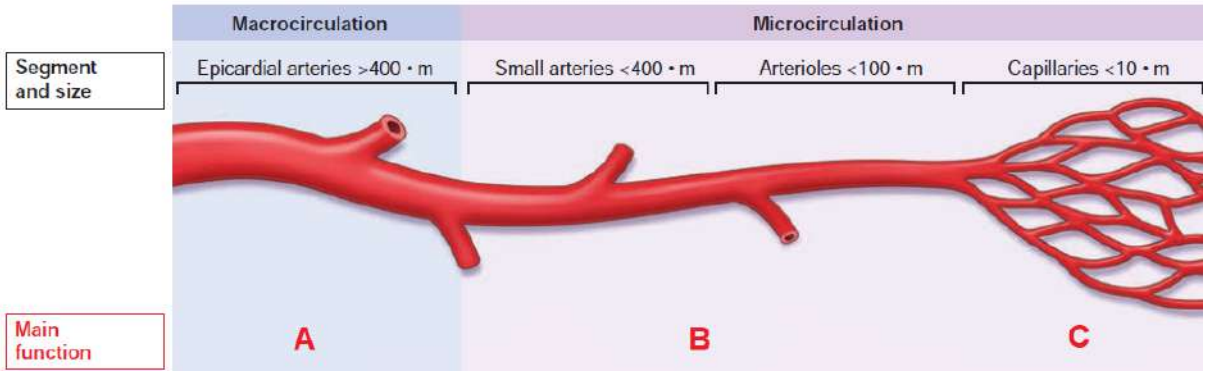
D. Typhoid fever

Answer: B(Page 1819)

Some patients with advanced AR may have a bifid or bisferiens pulse, in which two systolic peaks can be appreciated. A bifid pulse is also described in patients with hypertrophic obstructive cardiomyopathy (HOCM), with inscription of percussion and tidal waves. A bifid pulse is easily appreciated in patients on intraaortic balloon counterpulsation (IABP), in whom the second pulse is diastolic in timing.

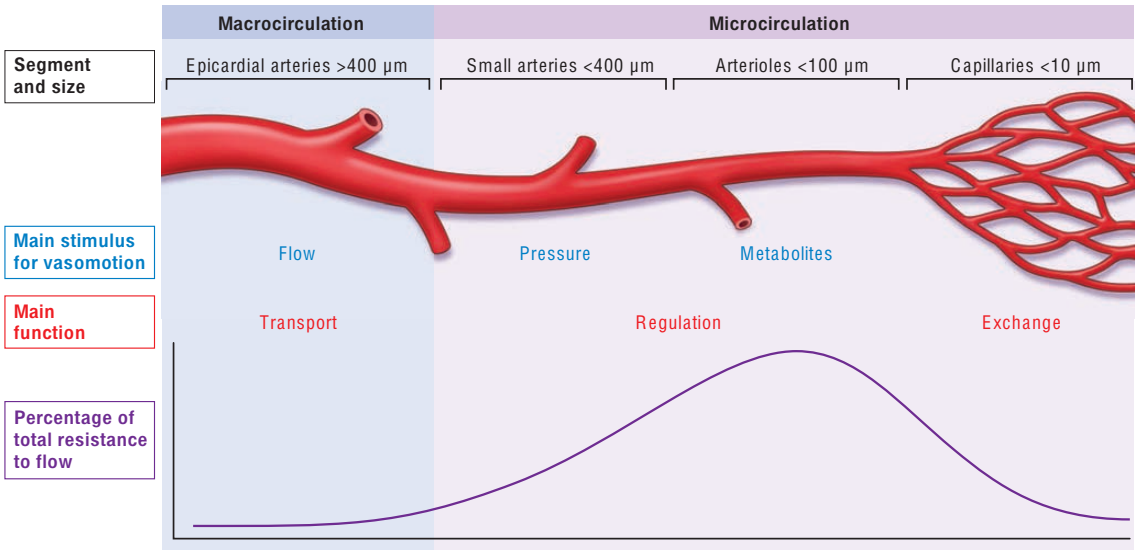


- 49. Macrocirculation and microcirculation across segments and sizes of the coronary arteries. The location and size of the arteries supplying blood to the heart is shown at the top. The main function of each of the arterial segments is shown – which is correct sequence?



1. A-Regulation, b-transport, c-exchange
2. A-Transport, b-regulation, c-exchange
3. A-pressure, b-flow, c-metabolites
4. A-flow, b-exchange, c-regulation

Answer: 2(Page 2031)



50. WHEN CORONARY DIAMETER REDUCED BY _____%, RESTING BLOOD FLOW TO HEART MAY BE REDUCED.

A. 50 %
B. 60 %
C. 70 %
D. 80 %

Answer:D(Page 2031)

Atherosclerosis develops at irregular rates in different segments of the epicardial coronary tree and leads eventually to segmental reduc- tions in cross-sectional area, i.e., plaque formation. There is also a predilection for atherosclerotic plaques to develop at sites of increased turbulence in coronary flow, such as at branch points in the epicardial arteries. When a stenosis reduces the diameter of an epicardial artery by 50%, there is a limitation of the ability to increase flow to meet increased myocardial demand. When the diameter is reduced by ~80%, blood flow at rest may be reduced, and further minor decreases in the stenotic orifice area can reduce coronary flow dramatically to cause myocardial ischemia at rest or with minimal stress.

51. NORMAL MYOCARDIUM PREFERENTIALLY UTILIZE _____ AS ENERGY SUBSTRATE.

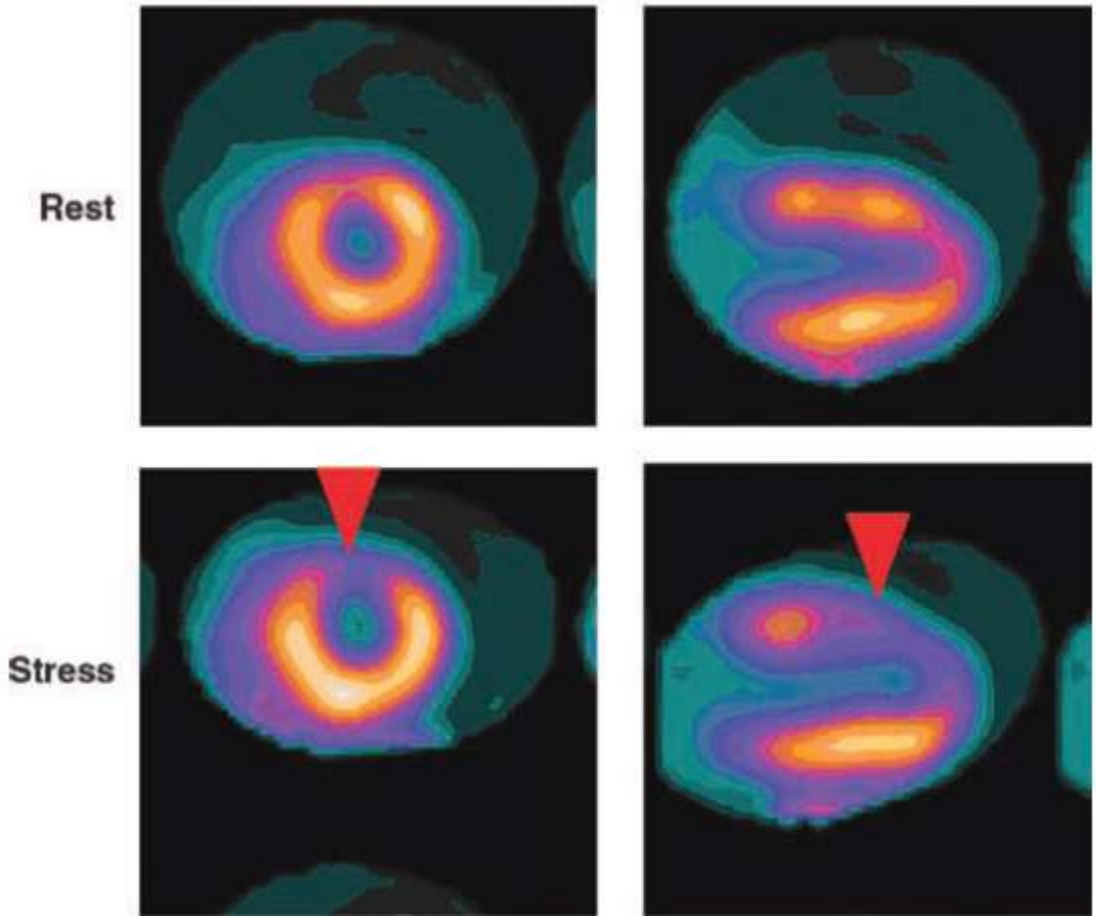
A. GLUCOSE
B. SUCROSE
C. FATTY ACID
D. LACTATE

Answer: C(Page 2032)

A wide range of abnormalities in cell metabolism, function, and structure underlie these mechanical disturbances during ischemia. The normal myocardium metabolizes fatty acids and glucose to carbon dioxide and water. With severe oxygen deprivation, fatty acids cannot be oxidized, and glucose is converted to lactate; intracellular pH is reduced, as are the myocardial stores of high-energy phosphates, i.e., ATP and creatine phosphate. Impaired cell membrane function leads to the leakage of potassium and the uptake of sodium by myocytes as well as an increase in cytosolic calcium. The severity and duration of the imbalance between myocardial oxygen supply and demand deter- mine whether the damage is reversible (≤20 min for total occlusion in the absence of collaterals) or permanent, with subsequent myocardial necrosis (>20 min).

52. STRESS AND REST MYOCARDIAL PERFUSION PET IMAGES OBTAINED WITH RUBIDIUM-82 IN A PATIENT WITH CHEST PAIN ON EXERTION. THE IMAGES DEMONSTRATE _____

A. REVERSIBLE DEFECT
B. FIXED DEFECT
C. NO DEFECT
D. SCAR



Answer:A(Page 1841 , 2036)

SPECT myocardial perfusion imaging is the most com- mon form of stress imaging tests for CAD eval- uation. The presence of a reversible myocardial perfusion defect is indicative of ischemia whereas a fixed perfusion defect generally reflects prior myocardial infarction . PET has advantages compared to SPECT, but it is not widely available and is more expensive and, thus, considered an emerging technology in clinical practice.

53. ……INHIBITS THE LATE INWARD SODIUM CURRENT (I_{Na})→ LIMITATION OF Na OVERLOAD OF ISCHEMIC MYOCYTES AND PREVENTION OF Ca^{2+} OVERLOAD VIA THE Na^{+}–Ca^{2+} EXCHANGER.

A. IVABRADINE
B. NICORANDIL
C. RANOLAZINE
D. PHENYTOIN

Answer: C(Page 2042)

Ranolazine, a piperazine derivative, may be useful for patients with chronic angina despite standard medical therapy . Its antianginal action is believed to occur via inhi- bition of the late inward sodium current (I_{Na}). The benefits of I_{Na} inhibition include limitation of the Na overload of ischemic myo- cytes and prevention of Ca^{2+} overload via the Na^{+}–Ca^{2+} exchanger. A dose of 500–1000 mg orally twice daily is usually well tolerated. Ranolazine is contraindicated in patients with hepatic impairment or with conditions or drugs associated with QT_c prolongation and when drugs that inhibit the CYP3A metabolic system (e.g., ketoco- nazole, diltiazem, verapamil, macrolide antibiotics, HIV protease inhibitors, and large quantities of grapefruit juice) are being used.

TABLE 273-7 Antianginal Agents

AGENT	COMMON SIDE EFFECTS	CONTRAINDICATIONS	POTENTIAL DRUG INTERACTIONS
Agents That Have a Physiologic Effect			
Short-acting and long-acting nitrates	Headache, flushing, hypotension, syncope and postural hypotension, reflex tachycardia, methemoglobinemia	Hypertrophic obstructive cardiomyopathy	Phosphodiesterase type 5 inhibitors (sildenafil and similar agents), beta-adrenergic blockers, calcium channel blockers
Beta blockers	Fatigue, depression, bradycardia, heart block, bronchospasm, peripheral vasoconstriction, postural hypotension, impotence, masked signs of hypoglycemia	Low heart rate or heart conduction disorder, cardiogenic shock, asthma, severe peripheral vascular disease, decompensated heart failure, vasospastic angina; use with caution in patients with COPD (cardioselective beta blockers may be used if patient receives adequate treatment with long-acting beta agonists)	Heart rate–lowering calcium channel blockers, sinus node or AV conduction depressors
Calcium-channel blockers Heart rate–lowering agents	Bradycardia, heart conduction defect, low ejection fraction, constipation, gingival hyperplasia	Cardiogenic shock, severe aortic stenosis, obstructive cardiomyopathy	CYP3A4 substrates (digoxin, simvastatin, cyclosporine)
Dihydropyridine	Headache, ankle swelling fatigue, flushing, reflex tachycardia	Low heart rate or heart rhythm disorder, sick-sinus syndrome, congestive heart failure, low blood pressure	Agents with cardiodepressant effects (beta blockers, flecainide), CYP3A4 substrates
Agents That Affect Myocardial Metabolism			
Ranolazine	Dizziness, constipation, nausea, QT interval prolongation	Liver cirrhosis	CYP3A4 substrates (digoxin, simvastatin, cyclosporine), drugs that prolong the corrected QT interval

54. PATHOPHYSIOLOGY OF NSTE-ACS INCLUDES:

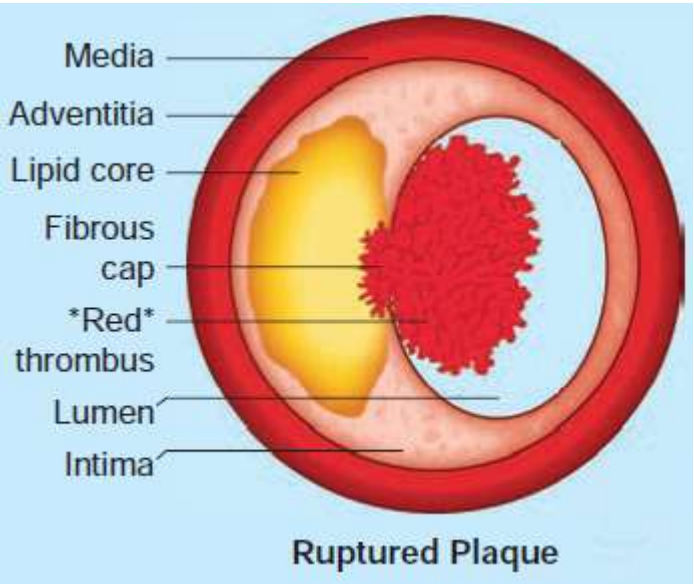
A. PLAQUE FISSURE WITH/WITHOUT INFLAMMATION
B. PLAQUE EROSION
C. EPICARDIAL OR MICROVASCULAR SPASM
D. ALL OF THE ABOVE

Answer: D(Page 2046)

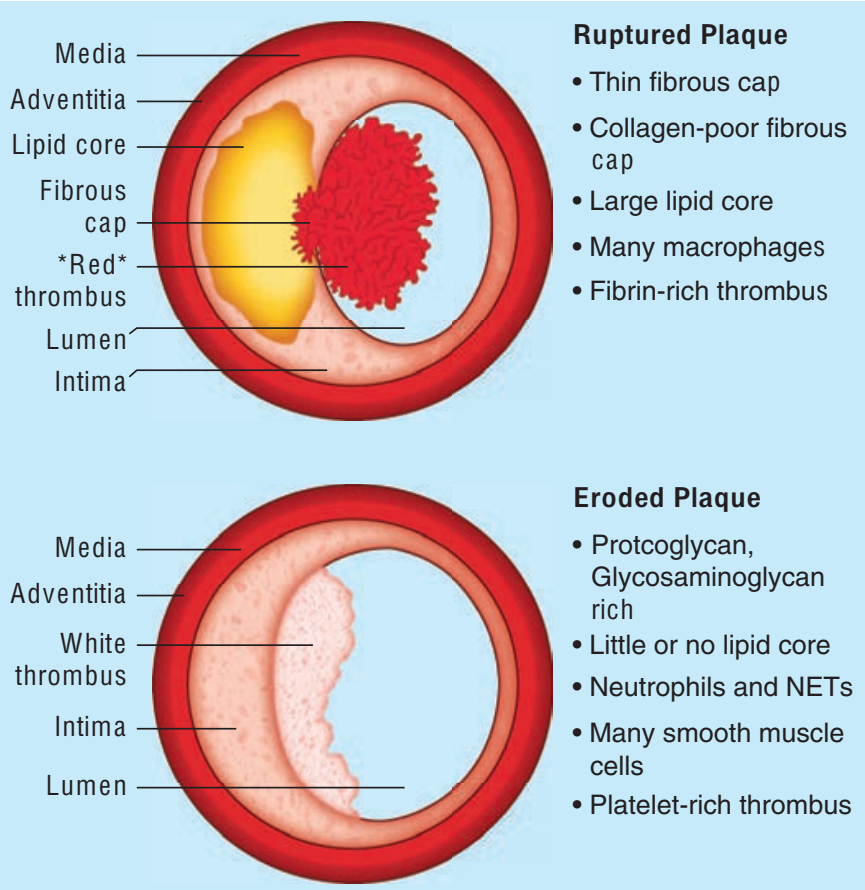
NSTE-ACS is caused by an imbalance between myocardial oxygen supply and demand resulting from one or more of three processes that lead to coronary arterial thrombosis: (1) plaque fissure with inflammation—the inflammatory response is reflected by an increased activity of effector T cells as part of an adaptive immunity dysregulation; (2) plaque fissure without inflammation; and (3) plaque erosion, which is present in at least one-third of ACS and is recognized with increasing frequency . The so-called “vulnerable plaques” responsible for ACS may show an eccentric stenosis with scalloped or overhanging edges and a narrow neck on coronary angiography. Such plaques usually are composed of a lipid-rich core with a thin fibrous cap. Patients with NSTE-ACS frequently have multiple such plaques that are at risk of disruption. A fourth process, without thrombosis, may be caused by epicardial or microvascular spasm or increased myocardial oxygen demand in the presence of fixed epicardial coronary obstruction.

55. WHICH OF THE FOLLOWING CHARACTERISTICS ARE ASSOCIATED WITH THIS EVENT?

A. THICK FIBROUS CAP, COLLAGEN RICH, NO OR LITTLE LIPID, PLATELET RICH THROMBUS
B. THIN FIBROUS CAP, COLLAGEN RICH, LESS LIPID, WHITE THROMBUS
C. THICK FIBROUS CAP, COLLAGEN POOR, LARGE LIPID CORE, RED THROMBUS
D. THIN FIBROUS CAP, COLLAGEN POOR, LARGE LIPID CORE, FIBRIN-RICH THROMBUS



Answer: D (Page 2047)



56. WHICH ORAL ANTIPLATELET IS REVERSIBLE INHIBITOR:

A. ASPIRIN
B. CLOPIDOGREL
C. TICAGRELOR
D. PRASUGREL

Answer: C(Page 2049,50)

Initial treatment should begin with the cyclooxygenase inhibitor aspirin with a dose of at least 162 mg of a rapidly acting preparation (oral non-enteric-coated or intravenous). Lower doses (75–100 mg/d) are recommended thereafter since they maintain efficacy while causing less bleeding. Contraindications are severe active bleeding and aspirin allergy.

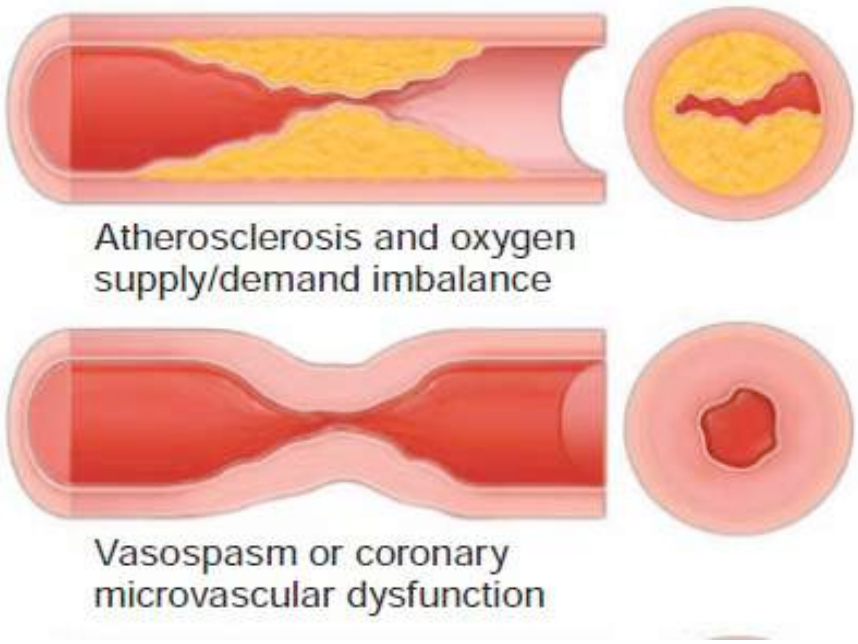
In the absence of a high risk for bleeding, patients with NSTE- ACS, irrespective of whether an invasive or conservative strategy (see below) is selected, should also receive a platelet $P2Y_{12}$ receptor blocker to inhibit platelet activation. There are now three oral and one intravenous $P2Y_{12}$ inhibitors to choose from. The thienopyri- dine clopidogrel is an inactive prodrug that is converted into an active metabolite that causes irreversible blockade of the platelet $P2Y_{12}$ receptor. The loading dose of clopidogrel is 600 mg, whereas the

maintenance dose is 75 mg daily. When clopidogrel is added to aspirin, so-called dual antiplatelet therapy (DAPT), in patients with NSTE-ACS, it confers a 20% relative reduction in cardiovascular death, MI, or stroke, compared to aspirin alone but is associated with a moderate (absolute 1%) increase in major bleeding

Two other $P2Y_{12}$ inhibitors have been shown to be superior to clopidogrel in preventing recurrent cardiac ischemic events but both increase bleeding. Prasugrel, also a thienopyridine, achieves a more rapid onset and higher level of irreversible platelet inhibition than clopidogrel. It has been approved for ACS patients following angiography when PCI is planned; it should be administered at loading dose of 60 mg followed by 10 mg/d. Compared to clopi- dogrel, prasugrel significantly reduces the combined risk of car- diovascular death, MI, stroke, and stent thrombosis but increases bleeding. Prasugrel is contraindicated in patients with prior stroke or transient ischemic attack or at high risk for bleeding.

Ticagrelor, a potent, *reversible* platelet $P2Y_{12}$ inhibitor, reduces the risk of cardiovascular death, total mortality, or MI compared to clopidogrel across a broad spectrum of patients with ACS. After a loading dose of 180 mg, 90 mg bid is administered as maintenance. Like prasugrel, ticagrelor increases the risk of bleeding. Unlike prasugrel, ticagrelor demonstrated benefit whether patients were managed conservatively or with an early invasive strategy (see below). Some patients may develop dyspnea soon after administra- tion, although the symptoms are often transient and infrequently serious.

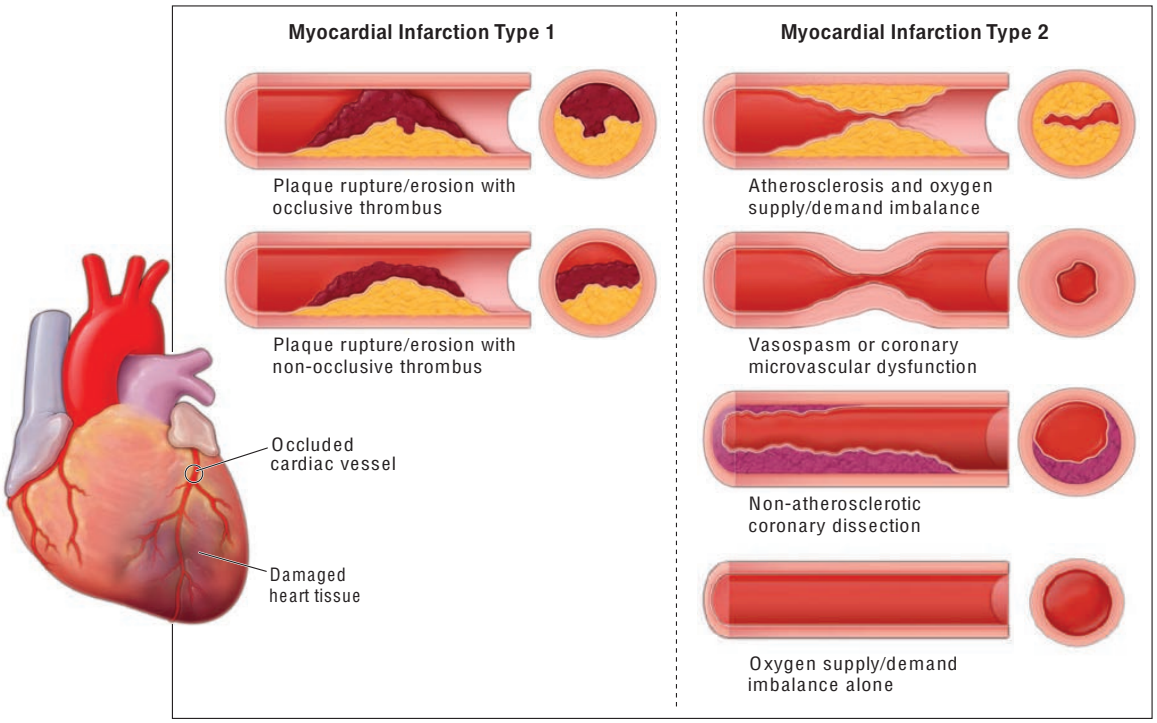
57. IMAGE REPRESENTS WHICH TYPE OF MI ?



A. TYPE 1
B. TYPE 2
C. TYPE 3

D. NONE

Answer: B(Page 2058)



58. WHICH IF THESE IS NOT A CAUSE OF ST ELEVATION IN ECG?

A. HYPERKALEMIA

B. HYPERTHERMIA

C. HYPERCALCEMIA

D. TYPE IA ANTIARRHYTHMIC DRUGS

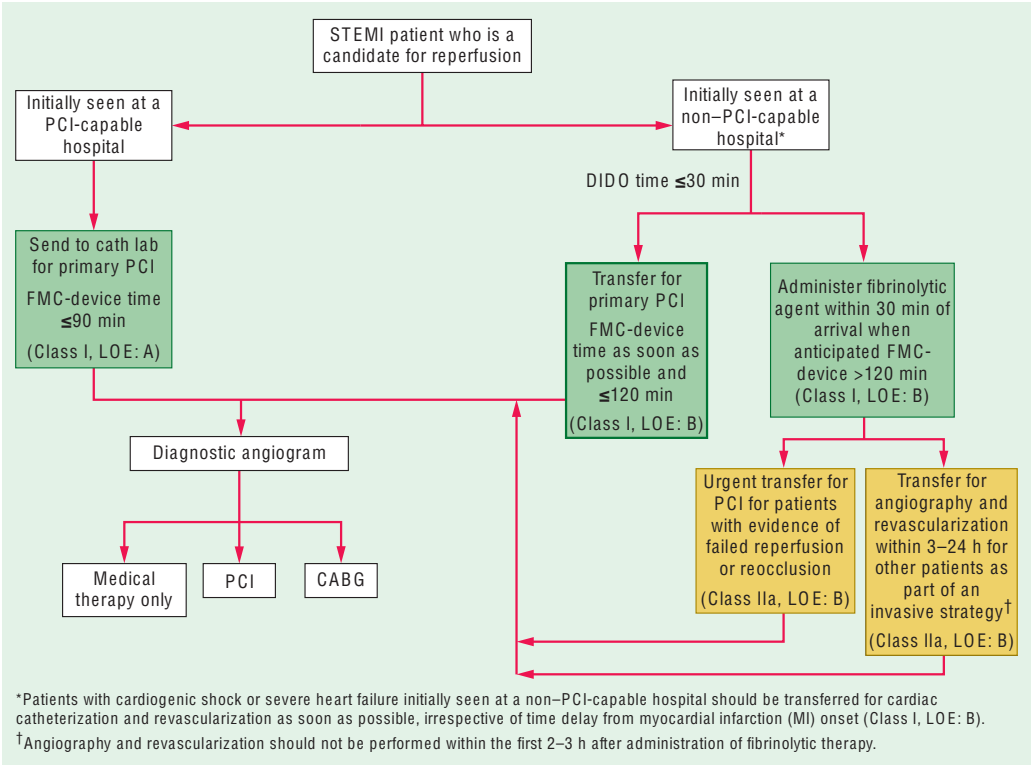
Answer: B(Page 1830)

TABLE 240-1 Differential Diagnosis of ST-Segment Elevations
Myocardial ischemia/infarction
Noninfarction, transmural ischemia (Prinzmetal's syndrome due to localized coronary spasm)
Acute myocardial infarction (especially due to epicardial coronary occlusion)
Takotsubo syndrome ("stress cardiomyopathy")
Postmyocardial infarction (ventricular aneurysm pattern)
Acute pericarditis
Normal variants (including benign "early repolarization" patterns)
Left ventricular hypertrophy/left bundle branch block[a]
Other (rarer)
Acute pulmonary embolism[a]
Brugada patterns (right bundle branch block–like morphology with ST elevations in right precordial leads)
Class 1C antiarrhythmic drugs[a]
DC cardioversion (transient)
Hypercalcemia[a]
Hyperkalemia[a]
Hypothermia (J [Osborn] waves)
Nonischemic myocardial injury
Myocarditis syndromes (infectious and non-infectious)
Tumor invading left ventricle
Trauma to ventricles

59. 56 YRS OLD MALE FROM REMOTE AREA PRESENTED WITH ACS-AWMI (WINDOW PERIOD 1 HR) AT LOCAL HOSPITAL. JOURNEY TO THE NEAREST PCI CAPABLE HOSPITAL IS 3 HRS FROM THIS LOCAL HOSPITAL BY ROAD. WHAT SHOULD BE YOUR STRATEGY?

A. GIVE DAPT, STATIN, HEPARIN AND MANAGE AT THE LOCAL HOSPITAL ONLY
B. IMMEDIATELY SHIFT TO PCI CAPABLE HOSPITAL
C. IMMEDIATELY THROMBOLYSE THE PATIENT AND SHIFT TO PCI CAPABLE HOSPITAL
D. GIVE DAPT, STATIN, HEPARIN AND AFTER 24 HRS SHIFT TO PCI CAPABLE HOSPITAL

Answer: C(Page 2058)



60. WHICH OF THE FOLLOWING FFR VALUE IS SIGNIFICANT ?

A. 0.90

B. 0.85

C. 0.75

D. 1.00

Answer: C(Page

Measurement of the fractional flow reserve provides a functional assessment of the stenosis and is more accurate in predicting long- term clinical outcome than imaging techniques. The fractional flow reserve is the ratio of the pressure in the coronary artery distal to the stenosis divided by the pressure in the artery proximal to the stenosis at maximal vasodilation. Fractional flow reserve is measured using a coronary pressure–sensor guidewire at rest and at maximal hyperemia following the infusion of adenosine. A fractional flow reserve of <0.80 indicates a hemodynamically significant stenosis that would benefit from intervention. The instantaneous wave-free ratio, which measures the gradient across the stenosis during the latter part of diastole, does not require the use of adenosine and may be preferred for some patients with asthma or documented allergy to adenosine. An instantaneous wave-free ratio of <0.89 is considered positive for ischemia. Resting gradients

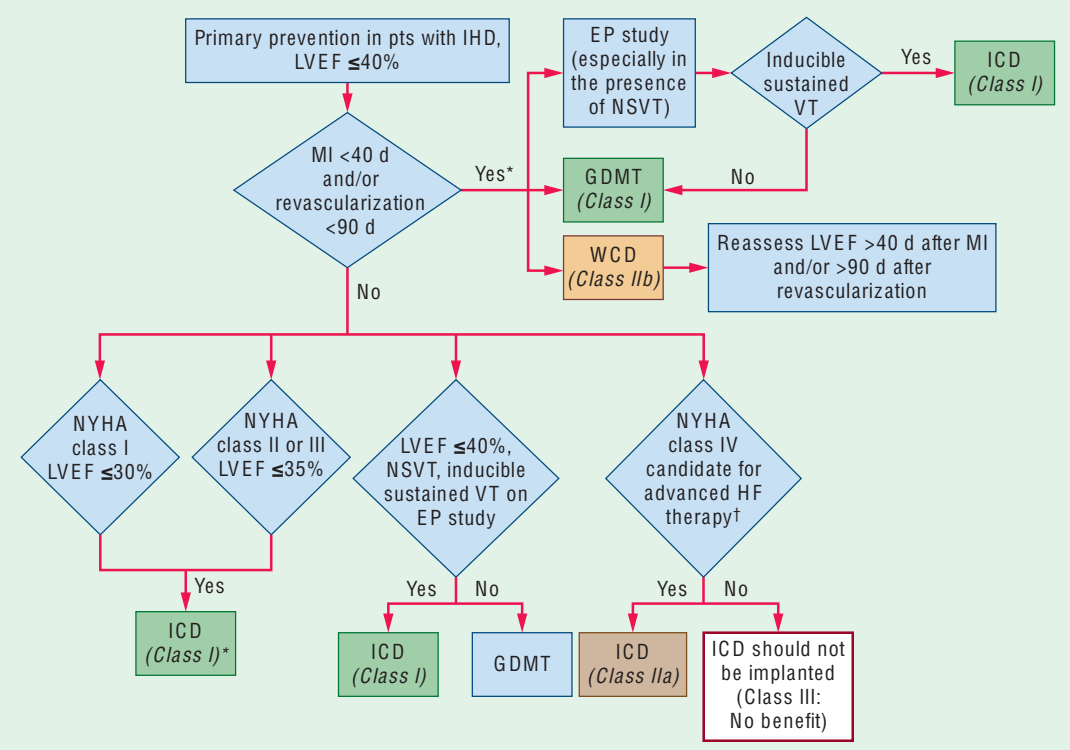
have also been shown to predict a signifi- cant stenosis. Using both pressure and velocity, an index of myocardial resistance can also be calculated. Studies have shown this to be an important predictor of outcome as well.

Microvascular dysfunction can be evaluated by assessing coronary flow reserve, the ratio between coronary blood flow at maximal hyper- emia and rest. Coronary flow reserve is measured using a Doppler wire– or pressure wire–based thermodilution technique in patients with unexplained chest pain or ischemia and no obstructive coronary artery disease. A coronary flow reserve <2.0 is considered abnormal.

61. 50 YRS OLD MALE HAD RECENT AWMI, S/P LAD PCI 4 MONTHS BEFORE, AND CURRENTLY ON NYHA II WITH LV EF 30% ON GDMT. WHICH OF THE FOLLOWING IS REQUIRED FOR PRIMARY PREVENTION OF SCD?

A. AMIODARONE
B. CRT-D
C. ICD
D. MEXILITINE

Answer: C(Page 2064)



62. WHICH OF THE FOLLOWING STATEMENTS REGARDIMG BUNDLE BRANCG BLOCK PATTERN IS NOT TRUE?

A. IN SUB- JECTS WITHOUT STRUCTURAL HEART DISEASE, RIGHT BUNDLE BRANCH BLOCK IS SEEN MORE COMMONLY THAN LEFT BUNDLE BRANCH BLOCK.

B. LEFT BUNDLE BRANCH BLOCK IS OFTEN A MARKER OF UNDERLYING CONDITIONS ASSOCIATED WITH INCREASED RISK OF CARDIOVASCULAR MORBIDITY AND MORTALITY RATES

C. LEFT POSTERIOR FASCICULAR BLOCK (QRS AXIS MORE RIGHTWARD THAN +110–120°) IS COMMON AS AN ISOLATED FINDING

D. PRIMARY REPOLARIZATION ABNOR- MALITIES ARE INDEPENDENT OF QRS CHANGES AND ARE RELATED INSTEAD TO ACTUAL ALTERATIONS IN THE ELECTRICAL PROPERTIES OF THE MYOCARDIAL FIBERS THEMSELVES

Answer:C(Page 1828)

In sub- jects without structural heart disease, right bundle branch block is seen more commonly than left bundle branch block. Right bundle branch block also occurs with heart disease, both congenital (e.g., atrial sep- tal defect) and acquired (e.g., valvular, ischemic). Left bundle branch block is often a marker of one of four underlying conditions associated with increased risk of cardiovascular morbidity and mortality rates: coronary heart disease (frequently with impaired left ventricular func- tion), hypertensive heart disease, aortic valve disease (including after transcatheter aortic valve replacement), and cardiomyopathy.

Bundle branch blocks and depolarization abnormalities secondary to artificial pacemakers not only affect ventricular depolarization (QRS) but also are characteristically associated with *secondary repo- larization* (ST-T) abnormalities. With bundle branch blocks, the T wave is typically opposite in polarity to the last deflection of the QRS

In contrast, *primary repolarization* abnor- malities are independent of QRS changes and are related instead to actual alterations in the electrical properties of the myocardial fibers themselves (e.g., in the resting membrane potential or action potential duration), not just to changes in the sequence of repolarization. Ische- mia, electrolyte imbalance, and drugs such as digitalis all cause such primary ST–T-wave changes

Left anterior fas- cicular block (QRS axis more negative than –45°) is probably the most common cause of marked left axis deviation in adults. In contrast, left posterior fascicular block (QRS axis more rightward than

+110–120°) is extremely rare as an isolated finding and requires exclusion of other factors causing right axis deviation mentioned earlier. Intraventricular conduction delays also can be caused by factors extrinsic (toxic) to the conduction system that slow ventricular conduction, particularly hyperkalemia or drugs (e.g., class 1 antiarrhythmic agents, tricyclic antidepressants, phenothiazines).

63. WHICH OF THE FOLLOWING IS NOT A CAUSE OF LOW VOLTAGE COMPLEX IN ECG?

A. MASSIVE PERICARDIAL EFFUSION

B. COPD

C. CARDIAC AMYLOIDOSIS

D.MULTIVALVULAR HEART DISEASE

Answer: D(Page 1830)

Many other factors are associated with ECG changes, particularly alterations in ventricular repolarization. T-wave flattening, minimal T-wave inversions, or slight ST-segment depression ("nonspecific ST–T-wave changes") may occur with a variety of electrolyte and acid- base disturbances, infectious or inflammatory processes, central ner- vous system disorders, endocrine abnormalities, many drugs, ischemia, hypoxia, and virtually any type of cardiopulmonary abnormality, in addition to physiologic changes (e.g., with posture or with meals). Low QRS voltage is arbitrarily defined as peak-to-trough QRS amplitudes of ≤5 mm in the six limb leads and/or ≤10 mm in the chest leads. Multiple factors may be responsible. Among the most serious include pericar- dial (or pleural effusions, chronic obstructive pulmonary disease, infiltrative cardiomyopathies, and anasarca.

64. WHICH OF THE FOLLOWING DOES NOT MEET THE CRITERIA OF CARDIAC CACHEXIA?

A.DECRAESED MUSCLE STRENGTH

B. DECREASED GLOBULIN

C. INCRASED INFLAMMATORY MARKERS

D. ANOREXIA

Answer : B(Page 1937)

TABLE 257-6 Definition of Cardiac Cachexia

Edema-free weight loss of at least 5% in 12 months or less in the presence of underlying illness (or a BMI <20 kg/m^2) and at least three of the following criteria:

- Decreased muscle strength (lowest tertile)
- Fatigue (physical and/or mental weariness resulting from exertion)
- Anorexia (limited food intake [<70% of usual] or poor appetite)
- Low fat-free BMI (lean tissue depletion by DEXA <5.45 in women and <7.25 in men)
- Abnormal biochemistry:
  - Increased inflammatory markers (CRP >5.0 mg/L, IL-6 >4.0 pg/mL)
  - Anemia (hemoglobin <12 g/dL)
  - Low serum albumin (<3.2 g/dL)

65. WHICH OF THE FOLLOWINBG DRUGS HAVE NOT SHOWN MORTALITY BENEFIT IN PATIENTS WITH HEART FAILURE?

A. SGLT2 INHIBITORS

B. ARNI

C. DIGOXIN

D. BETABLOCKERS

Answer: C(Page 1945)

Treatment of symptomatic HF has evolved from a renocen- tric (diuretics) and hemodynamic therapy model (digoxin, inotropic therapy) to an era of disease-modifying therapy with neurohormonal antagonism. In this

regard, RAAS blockers, beta-adrenergic receptor blockers, and most recently, SGLT2 inhibitors, form the pillars of pharmacotherapy and facilitate stabilization and even improvement in cardiac structure and function with consequent reduction in symp- toms, improvement in QOL, decreased burden of hospitalizations, and a decline in mortality from both pump failure and arrhythmic deaths

66. WHICH OF THIS IS NOT A TRIAL OF DRUGS IN HEART FAILURE WITH PRESERVED EJECTION FRACTION?

A. CHARM TRIAL

B. TOPCAT TRIAL

C.PEP-CHF TRIAL

D. PARADIGM-HF TRIAL

Answer: D(Page 1941)

Composite *angiotensin receptor-neprilysin inhibition* (ARNI) with sacubitril-valsartan reduced cardiovascular mortality, overall mortality, and HF hospitalization compared with enalapril among patients with HFrEF randomized in the PARADIGM-HF trial. The PARAGON-HF trial randomized 4822 patients with symptomatic HFpEF (LVEF ≥45%), elevated natriuretic peptides, and structural heart disease to treatment with either sacubitril-valsartan or valsartan with the novel composite primary endpoint of cardiovascular death and total hospi- talizations for HF. Although there was a 13% reduction in the rate of the primary composite endpoint in those allocated to sacubitril-valsartan, this result narrowly missed the margin for statistical significance in the primary statistical analysis ($p = .06$). Directional benefits in sec- ondary endpoints including QOL, NYHA class, and renal function favoring sacubitril-valsartan support a possible modest benefit of nepr- ilysin inhibition in this population, particularly among patients with lower (i.e., mildly reduced or mid-range) EF and women, subgroups who appeared to derive greater benefit.

67. WHICH OF THIS NOVEL THERAPIES IN HEART FAILURE WITH REDUCED EJECTION FRACTION IS MYOSIN ACTIVATOR?

A.VERICIGUAT

B. OMAPATRILAT

C.OMECAMTIV MECARBIL

D. EMPAGLIFOZIN

Answer: C(Page 1949)

A novel approach to augmentation of cardiac output is to prolong ven- tricular systole without increasing myocardial contractility. As a selec- tive myosin activator, *omecamtiv mecarbil* prolongs the ejection period and increases fractional shortening without altering the force of con- traction as a consequence. This agent, distinct from inotropic agents, is not associated with an increase in myocardial oxygen demand. Importantly, the drug is available for oral, rather than intravenous, administration, enabling chronic use in the ambulatory setting. In the COSMIC-HF (Chronic Oral Study of Myosin Activation to Increase Contractility in Heart Failure) trial of 448 patients with chronic HF and left ventricular systolic dysfunction, treatment with omecamtiv mecarbil for 20 weeks was associated with significant improvements in cardiac function and indices of left ventricular remodeling, as well as reductions in natriuretic peptide levels. Notably, the safety profile was comparable to placebo, with no increase in cardiac adverse events despite a modest increase in cardiac troponins in patients allocated to omecamtiv mecarbil. These promising preliminary data fueled a larger clinical outcomes trial (GALACTIC-HF, in which 8256 patients with symptomatic chronic heart failure and ejection fraction of 35% or less were randomized to treatment with omecamtiv mecarbil (dosage 25-50 mg twice daily based on plasma levels) or placebo in addition to standard HF therapy. Over median follow up of 21.8 months, patients allocated to omecamtiv mecarbil experienced a 14% reduction in the primary composite endpoint of death from cardiovascular causes or first heart failure event (hospitalization or urgent visit for heart failure), an outcome driven principally by reduction in heart failure events (no measureable effect on CV death alone). benefit in patients with features of advanced HF (lower EF, higher **1949** natriuretic peptide levels, more severe symptoms) combined with a favorable safety and tolerability profile suggests a possible role for this agent in patients with

late-stage disease, though additional study is needed.

DRUG CLASS	GENERIC DRUG	MEAN DAILY DOSE IN CLINICAL TRIALS (mg)	INITIATION (mg)	TARGET DOSE (mg)
Angiotensin-Converting Enzyme Inhibitors				
	Lisinopril	4.5–33	2.5–5 qd	20–35 qd
	Enalapril	17	2.5 bid	10–20 bid
	Captopril	123	6.25 tid	50 tid
	Trandolapril	N/A	0.5–1 qd	4 qd
Angiotensin Receptor Blockers				
	Losartan	129	50 qd	150 qd
	Valsartan	254	40 bid	160 bid
	Candesartan	24	4–8 qd	32 qd
Aldosterone Antagonists				
	Eplerenone	42.6	25 qd	50 qd
	Spironolactone	26	12.5–25 qd	25–50 qd
Beta Blockers				
	Metoprolol succinate CR/XL	159	12.5–25 qd	200 qd
	Carvedilol	37	3.125 bid	25–50 bid
	Bisoprolol	8.6	1.25 qd	10 qd
Arteriovenous Vasodilators				
	Hydralazine isosorbide dinitrate	270/136	37.5/20 tid	75/40 tid
	Fixed-dose hydralazine/isosorbide dinitrate	143/76	37.5/20 qid	75/40 qid
Angiotensin Receptor-Neprilysin Inhibitor				
	Sacubitril-valsartan	375	100 bid	200 bid
Novel Therapies (Under Investigation)				
	Vericiguat (sGC stimulator)	9.2	2.5 qd	10 qd
	Dapagliflozin, Empagliflozin (SGLT-2 inhibitors)	10	10 qd	10 qd
	Omecamtiv mecarbil (myosin activator)	Not reported	25 bid	Up to 50 mg bid (based on plasma concentrations)

68. WHICH OF THE FOLLOWING STATEMENTS REGARDING ICD IMPLANTATION IN PATIENTS WITH HEART FAILURE FOR PREVENTION OF SUDDEN CARDIAC DEATH IS TRUE?

A. PATIENTS WITH NYHA CLASS III OR IV SYMPTOMS OF HF AND AN LVEF <35%, IRRESPECTIVE OF ETIOLOGY OF HF, ARE APPROPRIATE CANDIDATES FOR ICD PROPHYLACTIC THERAPY

B. IF A PATIENT WITH HFrEF MEETS THE QRS CRITERIA FOR CRT, ONLY CRT IMPLANTATION SHOULD BE CONSIDERED.

C. ICD CAN BE CONSIDERED AFTER REVASCULARISATION WITHIN 40 DAYS FOR PRIMARY PREVENTION

D. IN PATIENTS WITH A MYOCARDIAL INFARCTION AND OPTIMAL MEDICAL THERAPY WITH RESIDUAL LVEF ≤30% (EVEN WHEN ASYMPTOMATIC), PLACEMENT OF AN ICD IS APPROPRIATE

Answer: D(Page 1951)

Currently, patients with NYHA class II or III symptoms of HF and an LVEF <35%, irrespective of etiology of HF, are appropriate candidates for ICD prophylactic therapy. In patients with a myocardial infarction and optimal medical therapy with residual LVEF ≤30% (even when asymptomatic), placement of an ICD is appropriate. A recent Danish trial suggested that prophylactic ICD implantation in patients with symptomatic systolic HF not caused by coronary artery disease was not associated with a significantly lower long-term rate of death from any cause than was usual clinical care. In this trial, benefits were noted in those aged <60 years. In patients with a terminal illness and a predicted life span of <6 months or in those with NYHA class IV symptoms who are refractory to medications and who are not candidates for transplant, the risks of multiple ICD shocks must be carefully weighed against the survival benefits. If a patient meets the QRS criteria for CRT, combined CRT with ICD is often employed

TABLE 258-3 Principles of ICD Implantation for Primary Prevention of Sudden Death

PRINCIPLE	COMMENT
Arrhythmia–sudden death mismatch	Sudden death in heart failure patients is generally due to progressive LVD, not a focal arrhythmia substrate (except in patients with post-MI HF)
Diminishing returns with advanced disease	Intervention at early stages of HF most successful since sudden death diminishes as cause of death with advanced HF
Timing of benefits	LVEF should be evaluated on optimal medical therapy or after revascularization before ICD therapy is employed; no benefit to ICD implant within 40 days of an MI (unless for secondary prevention)
Estimation of benefits and prognosis	Patients and clinicians often overestimate benefits of ICDs; an ICD discharge is not equivalent to an episode of sudden death (some ventricular arrhythmias terminate spontaneously); appropriate ICD discharges are associated with a worse near-term prognosis

69. WHICH IS THE MOST COMMON MODE OF INHERITANCE OF FAMILIAL CARDIOMYOPATHIES?

A. AUTOSOMAL DOMINANT

B. AUTOSOMAL RECESSIVE

C. X-LINKED

D. NONE OF THE ABOVE

Answer: A(Page 1954)

Most familial cardiomyopathies are inherited in an autosomal dominant pattern, with occasional autosomal recessive, matrilineal (mitochondrial), and X-linked inheritance. Missense mutations with amino acid substitutions and truncating variants are the most common genetic abnormalities in cardiomyopathy. Expressed mutant proteins may interfere with function of the normal allele through a dominant negative mechanism. Mutations introducing a premature stop codon (nonsense) or shift in the reading frame (frame- shift) may create a truncated or unstable protein, the lack of which causes cardiomyopathy (haploinsufficiency). Deletions or duplications of an entire exon or gene are uncommon causes of cardiomyopathy, except for the dystrophinopathies.

70. DALLAS CRITERIA IN ENDOMYOCARDIAL BIOPSY OS USED. FOR THE DIAGNOSIS OF WHICH OF THE FOLLOWING DISEASES?

A. RESTRICTIVE CARDIOMYOPATHY

B. HYPERTROPHIC CARDIOMYOPATHY

C. MYOCARDITIS

D. PERICARDIAL DISEASES

Answer: C(Page 1960)

When biopsy is performed, the key Dallas criteria for myocarditis include lymphocytic infiltrate with evidence of myocyte necrosis and are neg- ative in 80–90% of patients with clinical myocarditis. Negative Dallas criteria can reflect sampling error or early resolution of lymphocytic infiltrates, but may also be influenced by the insensitivity of the test when inflammation results from cytokines and antibody-mediated injury. Routine histologic examination of endomyocardial biopsy rarely reveals a specific infective etiology, such as toxoplasmosis or cytomegalovirus subsets. Immunohistochemistry of myocardial biopsy samples is commonly used to identify active lymphocyte subtypes and may also detect upregulation of HLA antigens and the presence of complement components attributed to inflammation, but the specific- ity and significance of these findings are uncertain.

71. WHICH OF THE FOLLOWING IS NOT A FORM OF FULMINANT MYOCARDITIS?

A. GIANT CELL MYOCARDITIS

B. EOSINOPHILIC MYOCARDITIS

C. CHECKPOINT IMMUNE THERAPY

D. CHRONIC ACTIVE MYOCARDITIS

Answer: D(Page 1958)

Myocardial inflammation without obvious infection is seen in sarcoi- dosis and giant cell myocarditis, with checkpoint inhibitor therapy, in eosinophilic myocarditis, or in association with autoimmune diseases such as polymyositis and systemic lupus erythematosus. Fulminant myocarditis can result from viral infection, checkpoint inhibitor ther- apy, giant cell myocarditis, or necrotizing eosinophilic myocarditis, and is often complicated by recurrent arrhythmias. Early recognition of fulminant myocarditis is crucial as recovery to near-normal cardiac function can occur during aggressive circulatory support.

72. WHICH OF THE FOLLOWING STATEMENTS REGARDING METABOLIC CAUSES OF CARDIOMYOPATHY IS NOT TRUE?

A. INBORN DISORDERS OF METABOLISM MOST COMMONLY PRESENT WITH DCM.

B. PHEOCHROMOCYTOMA SHOULD BE CONSIDERED WHEN A PATIENT HAS HEART FAILURE AND VERY LABILE BLOOD PRESSURE AND HEART RATE, SOMETIMES WITH EPISODIC PALPITATIONS

C. THE MOST COMMON CURRENT REASON FOR THYROID ABNORMALITIES IN THE CARDIAC POPULATION IS THE TREATMENT OF TACHYARRHYTHMIAS WITH AMIODARONE

D. DIABETES IS A TYPICAL FACTOR IN HEART FAILURE WITH “PRESERVED” EJECTION FRACTION, ALONG WITH HYPERTENSION, ADVANCED AGE, AND FEMALE GENDER.

73. MATCH THE FOLLOWING FORMS OF CARDIOMYOPATHIES WITH THE GENES INVOLVED

1. ARVC

2. PERIPARTUM CARDIOMYOPATHY

3. LV NON COMPACTION

4. HYPERTROPHIC CARDIOMYOPATHY

I. MYH7

II. TAZ

III.TTN

IV.DESMOPLAKIN

A. 1-IV 2-II 3-III 4-I
B. 1-IV 2-III 3-II 4-1
C. 1-I 2-II 3-III 4-IV
D. 1-IV 2-I 3-II 4-III

74. WHICH OF THE FOLLOWING STATEMENTS REGARDING CARDIAC AMYLOIDOSIS IS TRUE?

A. CARDIAC INVOLEMENT IS MOST COMMONLY SEEN IN AA AMYLOIDOSIS

B. MOST COMMON CLINICAL PRESENTATION IS LEFT HEART FAILURE

C. LOW VOLTAGE COMPLEXES IS. MORE COMMON IN AL THAN ATTR AMYLOIDOSIS

D. ENDOMYOCARDIAL BIOPSY HAS LOW SENSITIVITY AND SPECIFICITY IN DIAGNOSIS.

Answer: C(Page 1967,68)

Conduction system disease and atrial fibrillation are common. Nephrotic syndrome is common in AL amyloid, which may also cause angina as the amy- loid encircles the coronary arteries. Because the ventricular cavity is diminished by amyloid infiltration, cardiac output may be very low with a modest ejection fraction reduction. Peripheral and autonomic neuropathy are common in both AL amyloidosis and ATTRm amyloi- dosis. A history of carpal tunnel syndrome is common in ATTRm and ATTRwt, often preceding cardiac symptoms by many years. ATTRwt is also associated with spinal stenosis.

Amyloidosis should be suspected when ventricular myocardium appears thick on imaging with low ECG voltage, but this mismatch is more common with AL than TTR amyloidosis. Atrial enlargement is prominent and diastolic dysfunction more severe than that of other causes of hypertrophy. Longitudinal strain is frequently more pre- served at the apex, creating a “bull’s-eye” pattern. MRI shows diffuse late gadolinium enhancement. Technetium-pyrophosphate scanning reliably highlights TTR amyloidosis but does not detect AL amyloid. ndomyocardial biopsy is virtually 100% reliable for the diagnosis of all amyloid due to the characteristic birefringence pattern of Congo red staining of the amyloid fibrils under polarized light, but immu- nohistochemistry may be necessary to confirm the amyloid type, as serum or urine electrophoresis may be misleading. Until recently, the therapy of amyloidosis was limited to the treatment of congestion and arrhythmias.

75. WHICH OF THE FOLLOWING IS NOT A DEFINITE FACTOR FOR RISK STRATIFICATION IN HCM FOR SUDDEN DEATH?

A. HISTORY OF CARDIAC ARREST

B. SYNCOPE

C. ABNORMAL BLOOD PRESSURE RESPONSE

D. LATE GADOLINIUM ENHANCEMENT

Answer: D(Page 1972)

RISK FACTOR		SCREENING TECHNIQUE
History of cardiac arrest or spontaneous sustained ventricular tachycardia[a]		History
Syncope	Nonvagal, often with or after exertion	History
Family history of sudden cardiac death		Family history
Spontaneous nonsustained ventricular tachycardia	>3 beats at rate >120	Exercise or 24- to 48-h ambulatory recording
LV thickness >30 mm	Present in <10% of patients	Echocardiography
Abnormal blood pressure response to exercise[b]	Systolic blood pressure fall or failure to increase at peak exercise	Maximal upright exercise testing
Variables Utilized in the European Society of Calculator for Estimated Risk of Sudden Death		
LV outflow tract gradient	Peak gradient measured at rest or with the Valsalva maneuver, mmHg	Echocardiography
Left atrial diameter	Diameter measured in the parasternal long axis, mm	Echocardiography
LV thickness	Maximal wall thickness, mm	Echocardiography
Age		
Syncope, family history, nonsustained ventricular tachycardia	As above	As above
Emerging Risk Factors		
Late gadolinium enhancement	As a percentage of myocardial mass	Cardiac magnetic resonance imaging
Left ventricular apical aneurysm	Generally applicable to patients with apical hypertrophy	Echocardiography with contrast, cardiac magnetic resonance imaging

76. WHICH IS THE MOST COMMON VALVE LESION AMONG ADULT PATIENTS WITH CHRONIC VALVULAR HEART DISEASE ?

A. AORTIC STENOSIS

B. MITRAL STENOSIS

C. AORTIC REGURGITATION

D. MITRAL REGURGITATION

Answer: A(Page 1979)

Aortic stenosis (AS) is the most common valve lesion among adult patients with chronic valvular heart disease; the majority of adult patients with symptomatic, valvular AS are male.

77. WHICH OF THE FOLLOWING IS NOT A CLASS I INDICATION FOR SURGERY OR TAVR IN PATIENTS WITH SEVERE AS?

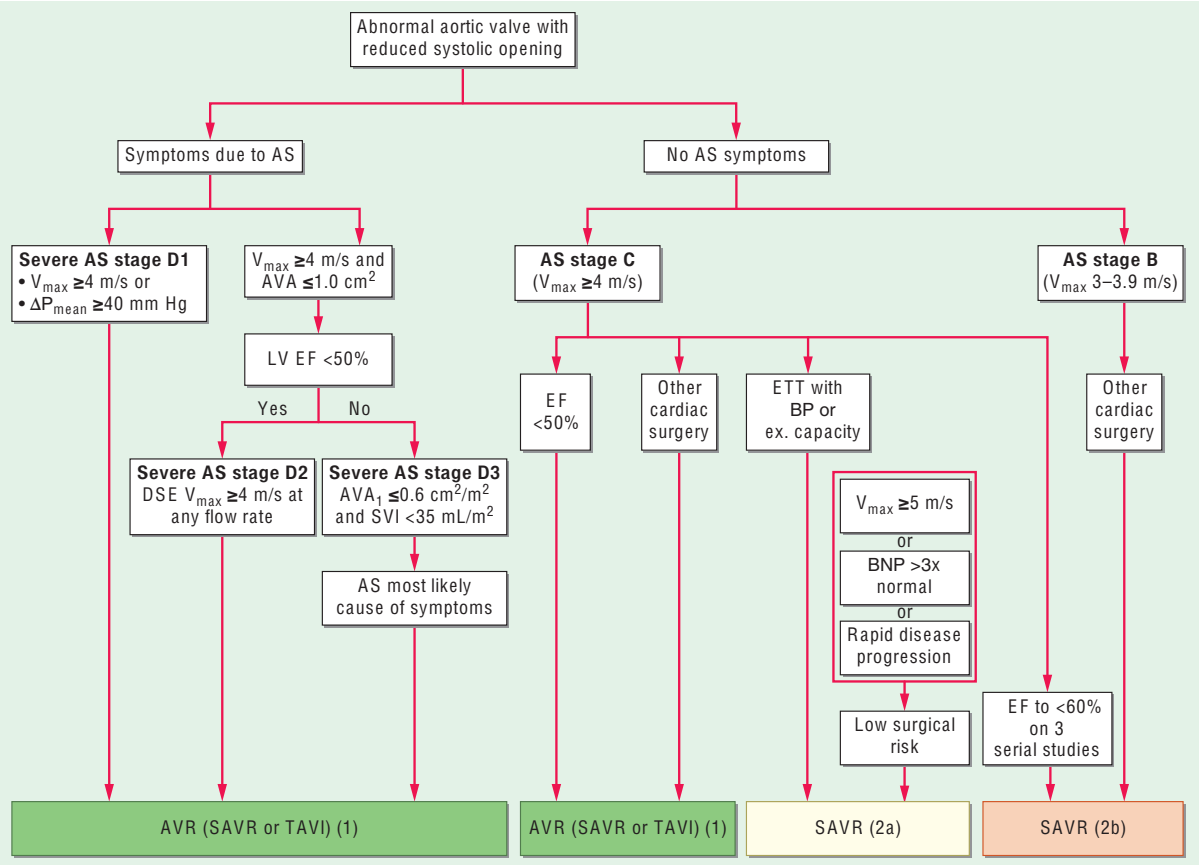
A. SYMPTOMATIC SEVERE AS

B. ASYMPTOMATIC SEVERE AS WITH EF LESS THAN 50%

C. ASYMPTOMATIC SEVERE AS UNDERGOING CABG

D. ASYMPTOMATIC SEVERE AS WITH RAPID DS PROGRESSION

Answer: D(Page 1983)



78. A 80 YEAR OLD MALE PRESENTS WITH SEVERE SYMPTOMATIC AS. HE HAS A HISTORY OF CKD ON HEMODIALYSIS AND KNOWN COPD AND HIGH FRAILITY INDEX. WHICH OF THE MODE OF TREATMENT WILL BE MOST PREFERED?

A. MEDICAL MANAGEMENT

B. SAVR

C. TAVR

D. BALLOON AORTIC VALVULOPLASTY

Answer:C(Page 1984)

TAVI is most frequently undertaken via the transfemoral route, although trans-LV apical, subclavian, carotid, and ascending aortic routes have been used. Aortic balloon valvuloplasty under rapid RV (or LV) pacing is performed as a first step to create an orifice of sufficient size for the prosthesis. Procedural success rates exceed 95% in appropriately selected patients. Valve performance characteristics are excellent over 5 years; longer-term durability assessment is ongoing. Outcomes achieved with this transformative technology have been very favorable and have allowed the extension of AVR to groups of patients previously considered at high or prohibitive risk for conven- tional surgery. Nevertheless, some prohibitive or high surgical risk patients are not candidates for this procedure because their comorbid- ity profile and frailty would make its undertaking inappropriate. The heart team is specifically charged with making challenging decisions of this nature. The use of these devices for treatment of patients with structural deterioration of bioprosthetic aortic valves (valve-in-valve TAVI), as an alternative to reoperative valve replacement, has increased sharply over the past 5 years.

79. WHICH OF THE FOLLOWING IS THE MOST SENSITIVE PERIPHRAL SIGN IN CHRONIC AORTIC REGURGITATION?

A. HILLS SIGN

B. TRAUBES SIGN

C. COLLAPSING PULSE

D. DUROZIEZS SIGN

Answer: D (Page 1988)

A rapidly rising “water-hammer” pulse, which col- lapses suddenly as arterial pressure falls rapidly during late systole and diastole (Corrigan’s pulse), and capillary pulsations, an alternate flush- ing and paling of the skin at the root of the nail while pressure is applied to the tip of the nail (Quincke’s pulse), are characteristic of chronic severe AR. A booming “pistol-shot” sound can be heard over the femo- ral arteries (Traube’s sign), and a to-and-fro murmur (Duroziez’s sign) is audible if the femoral artery is lightly compressed with a stethoscope

80. WHICH OF THE FOLLOWING IS NOT A CLASS I INDICATION OF SURGERY IN SEVERE AORTIC REGURGITATION?

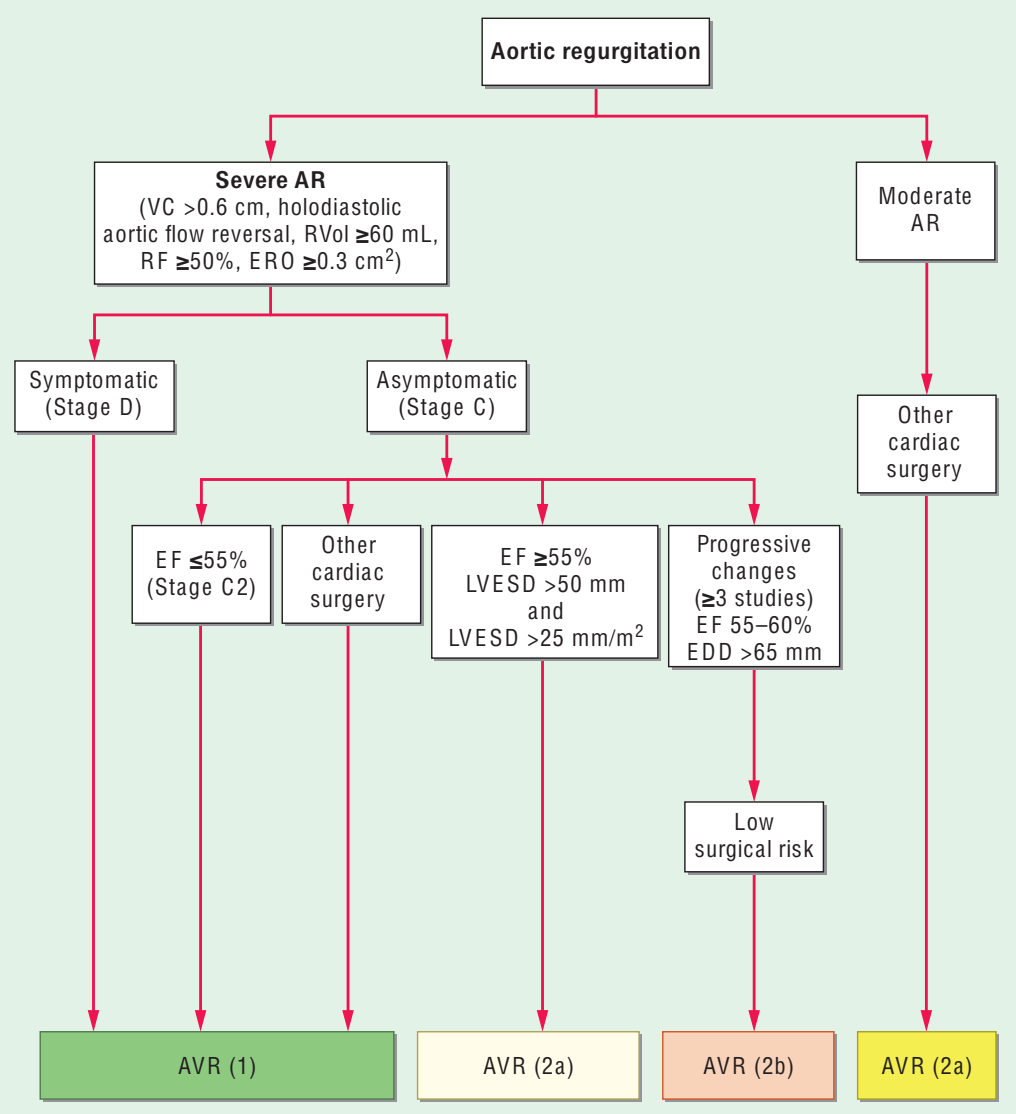
A. SYMPTOMATIC SEVERE AR

B. ASYMPTOMATIC AND EF LESS THAN 55 %

C. ASYMPTOMATIC AR PATIENTS UNDERGOING CABG

D. ASYMTOMATIC WITH ESD >5O MM AND EF >55 %

Answer: D(Page 1989)



81. WHICH OF THE FOLLOWING DRUGS ARE CONTRAINDICATED IN ACUTE SEVERE AR?

A. HYDRALAZINE

B. DIURETICS

C. BETA BLOCKERS

D. NITROPRUSSIDE

Answer: C(Page 1989)

Patients with acute severe AR may respond to intravenous diuretics and vasodilators (such as sodium nitroprusside), but stabilization is sually short-lived and operation is indicated urgently. Intra-aortic balloon counterpulsation is contraindicated. Beta blockers are best avoided so as not to reduce the CO further or slow the heart rate, thus allowing more time for diastolic filling of the LV. Surgery is the treatment of choice and is usually necessary within 24 h of diagnosis.

82. WHICH OF THE FOLLOWING DRUGS HAVE BEEN MOST STUDIED FOR BLOOD PRESSURE CONTROL IN ASYMPTOMATIC SEVERE AR?

A. BETABLOCKERS

B. NIFEDIPINE

C.HYDRALAZINE

D. LOSARTAN

Answer: B(Page 1989)

Medical treatment with diuretics and vasodilators (angiotensin-converting enzyme inhibitors, angiotensin receptor blockers [ARBs], dihydropyridine calcium channel blockers, or hydralazine) may be useful as a temporizing measure. The use of vasodilators to extend the compensated phase of chronic severe AR in asymptomatic patients before the onset of symptoms or the development of LV dysfunction is not useful, although these agents should be employed to treat hypertension (systolic blood pressure >140 mm Hg). It is often difficult to achieve adequate blood pressure control because of the increased stroke volume and enhanced LV ejection that accompany severe AR

Beta blockers and the ARB losartan may be useful to retard the rate of aortic root enlargement in young patients with Marfan's syndrome and aortic root dilation. A randomized controlled trial showed no differ- ence in efficacy between atenolol and losartan for this indication. Whether beta blockers or ARBs are useful in retarding the rate of growth of aortic aneurysms in other patient subsets (e.g., BAV disease with aortopathy, Takayasu's disease) has not been demon- strated. Beta blockers in patients with valvular AR were previously considered relatively contraindicated due to concern that slowing of the heart rate would allow more time for diastolic regurgitation. Observational reports, however, have suggested that beta blockers may provide functional benefit in some patients with chronic AR. Beta blockers can sometimes provide incremental blood pressure lowering in patients with chronic AR and hypertension. They can also lessen the sense of forceful heart action that many patients find uncomfortable. Patients with severe AR, particularly those with an associated aortopathy, should avoid isometric exercises.

83. WHICH OF THE FOLLOWING REGARDING PATHOPHYSIOLOGY OF MITRAL STENOSIS IS NOT TRUE?

A. THE LV DIASTOLIC PRESSURE AND EJECTION FRACTION (EF) ARE NORMAL IN ISOLATED MS

B. IN PATIENTS WITH SEVERE MS (MITRAL VALVE ORIFICE 1–1.5 CM^2), THE CO IS NORMAL OR ALMOST SO AT REST

C. AT END STAGE, ORGANIC OBLITERATIVE CHANGES IN THE PULMONARY VASCULAR BED ARE SEEN

D. IN MS AND SINUS RHYTHM, THE ELEVATED LA AND PA WEDGE PRESSURES EXHIBIT A PROMINENT V WAVE PATTERN

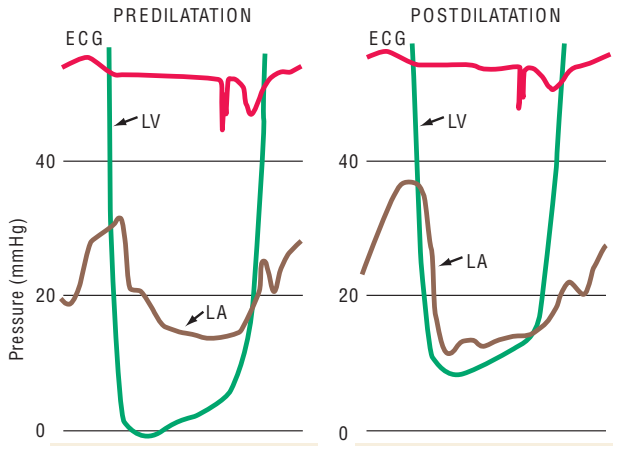
Answer: D(page 1991,92)

The LV diastolic pressure and ejection fraction (EF) are normal in isolated MS. In MS and sinus rhythm, the elevated LA and PA wedge pressures exhibit a prominent atrial contraction pattern (*a* wave) and a gradual pressure decline after the *v* wave and mitral valve opening (*y* descent). In severe MS and whenever pulmonary vascular resistance is significantly increased, the PA pressure (PAP) is elevated at rest and rises further during exercise, often causing secondary elevations of right ventricular (RV) end-diastolic pressure and volume.

In patients with severe MS (mitral valve orifice 1–1.5 cm^2), the CO is normal or almost so at rest, but rises subnormally during exertion. In patients with very severe MS (valve area <1 cm^2), particularly those in whom pulmonary vascular resistance is markedly elevated, the CO is subnormal at rest and may fail to rise or may even decline during activity.

The clinical and hemodynamic fea- tures of MS are influenced importantly by the level of the PAP. Pul- monary hypertension results from (1) passive backward transmission of the elevated LA pressure; (2) pulmonary arteriolar constriction (the so-called “second stenosis”), which presumably is triggered by LA and pulmonary venous hypertension (reactive pulmonary hypertension); (3) interstitial edema in the walls of the small pulmonary vessels; and (4) at end stage, organic obliterative changes in the pulmonary vascular bed.

84. THE BELOW IMAGES SHOWS RESULTS OF WHICH PROCEDURE?



A. PTMC
B. TAVR
C. BPV
D. BAV

Answer: A(Page 1994)

Successful commissurotomy is defined by a 50% reduction in the mean mitral valve gradient and a doubling of the mitral valve area. Successful commissurotomy, whether balloon or surgical, usually results in striking symptomatic and hemodynamic improvement and prolongs survival.

85. WHICH OF THE FOLLOWING STATEMENTS REGARDING TREATMENT OF MITRAL STENOSIS IS NOT TRUE?

A. ASYMPTOMATIC PTS WITH NEW ONSET AF AND PASP>50 MM HG ARE ELIGIBLE CANDIDATES FOR PTMC

B. NOACS CAN BE CONSIDERED FOR MODERATE TO SEVERE PATIENTS WITH MS AND ATRIAL FIBRILLATION

C. LEFT ATRIAL CLOT AND SEVERE COMMISURAL CALCIFICATION ARE CONTRAINDICATION FOR PTMC

D. OPERATIVE MORTALITY OF MITRAL VALVE REPLACEMENT AND CABG IS AROUND 9.6 %.

Answer: B(Page 1994)

Vitamin K antagonist therapy (such as warfarin) targeted to an international normalized ratio (INR) of 2–3 should be administered indefinitely to patients with MS who have AF, a history of thromboembolism, or demonstrated LA thrombus. The routine use of a vitamin K antag- onist in patients in sinus rhythm with LA enlargement (maximal dimension >5.5 cm) with or without spontaneous echo contrast is more controversial. As of this writing, non–vitamin K oral anti- coagulants (e.g., apixaban, rivaroxaban) have not been adequately studied in patients with moderate or severe rheumatic MS and, thus, are not recommended.

Unless there is a contraindication, mitral commissurotomy is indi- cated in symptomatic (New York Heart Association [NYHA] Func- tional Class II–IV) patients with isolated severe MS, whose effective orifice (valve area) is <~1 cm^2/m^2 body surface area, or <1.5 cm^2 in normal-sized adults. Mitral commissurotomy can be carried out either percutaneously or surgically. In PMBC a catheter is directed into the LA after transseptal punc- ture, and a single balloon is directed across the valve and inflated in the valvular orifice. Ideal patients have relatively pliable leaflets with little or no commissural calcium. In addition, the subvalvular structures should not be significantly scarred or thickened, and there should be no LA thrombus. Any associated MR should be of ≤2+/4+ severity

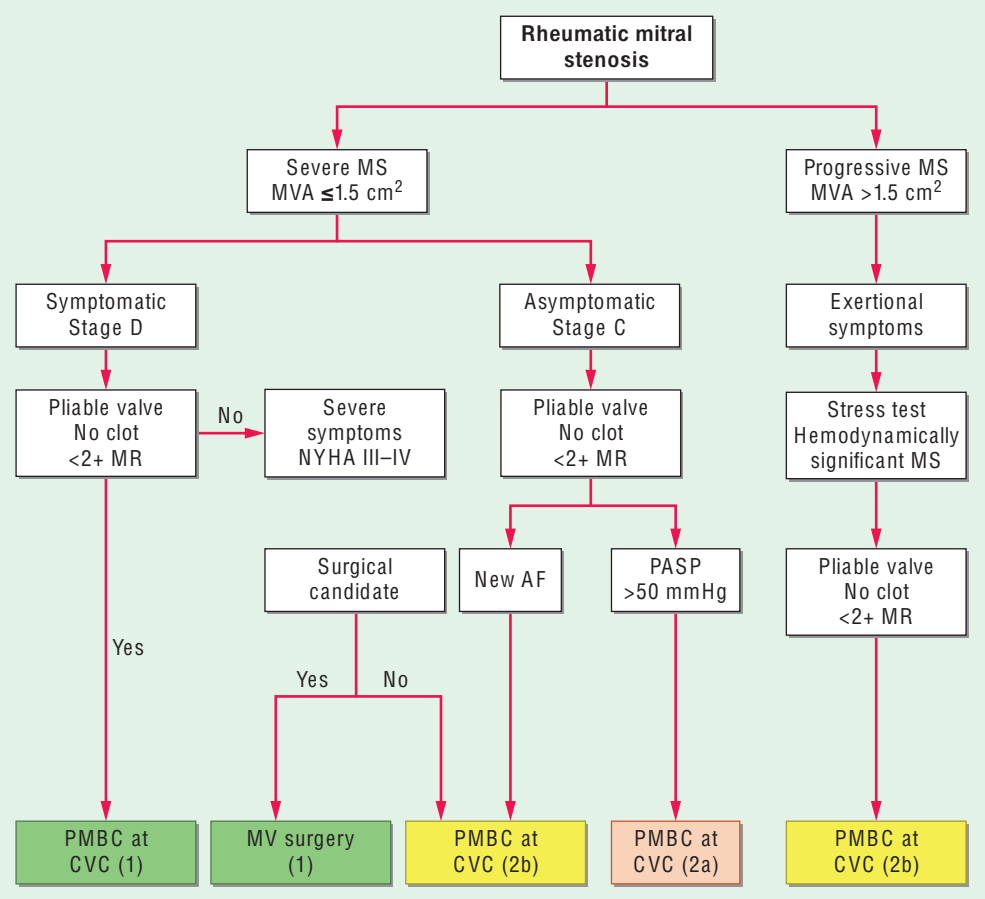


TABLE 263-2 Mortality Rates after Mitral Valve Surgery[a]

OPERATION	NUMBER	UNADJUSTED OPERATIVE MORTALITY (%)
MVR (isolated)	10,699	4.5
MVR + CAB	3509	9.6
MVRp	12,424	1.2
MVRp + CAB	4093	5.4

86. WHICH OF THE FOLLOWING CRITERIA FOR DIAGNOSIS OF SEVERE MR DOESNOT HOLD TRUE?

A. REGURGITANT VOL > 60 ML

B. REGURGITANT FRACTION MORE THAN 50 %

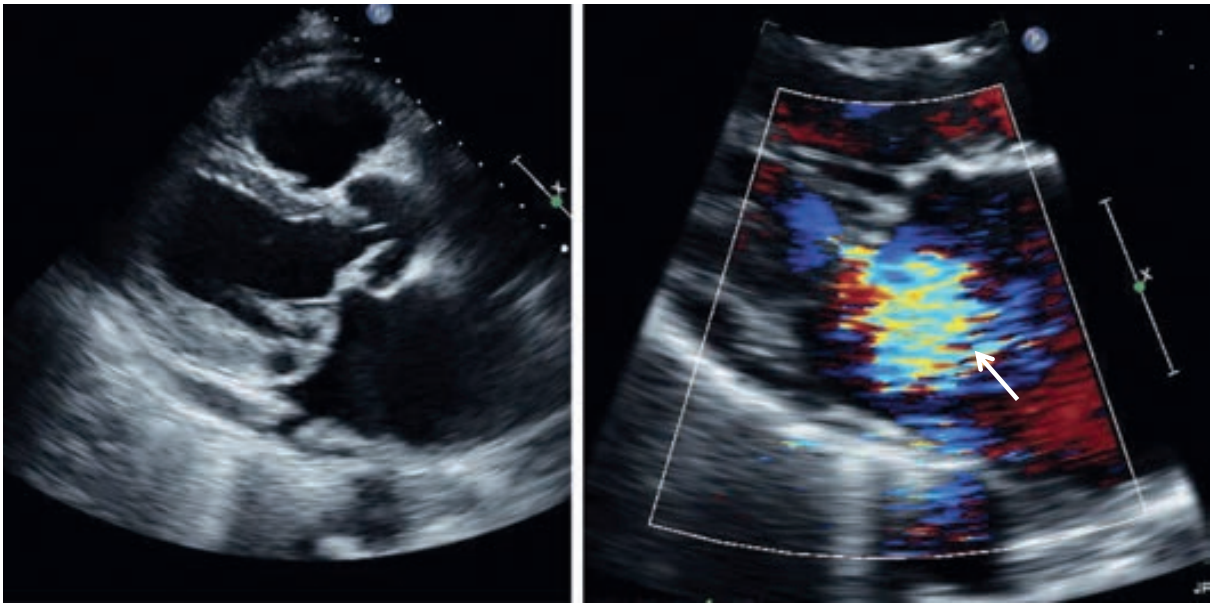
C. VENA CONTRACTA > 6 MM

D. EFFECTIVE REGURGITANT ORIFICE > 0.4 CM2

Answer: C(Page 1996,97)

Doppler imaging should demonstrate the width or area of the color flow MR jet within the LA, the duration and intensity of the continuous wave Doppler signal, the pulmonary venous flow contour, the early peak mitral inflow velocity, and quantitative measures of regurgitant volume, RF, and effective regurgitant orifice area. In addition, the PA pressures (PAPs) can be estimated from the TR jet velocity. TTE is also indicated to follow the ourse of patients with chronic MR and to provide rapid assessment for any clinical change. Chronic, severe MR is defined by a regurgitant volume ≥60 mL/beat, regurgitant fraction (RF) ≥50%, and effective regurgitant orifice area ≥0.40 cm^2.

87. THE IMAGE BELOW SHOWS WHICH OF THE FOLLOWINGF ETIOLOGY OF SEVERE MR



A. RHEUMATIC HEART DS
B. MITRAL VALVE PROLAPSE
C. FLAIL LEAFLET
D. MITRAL ANNULAR CALCIFICATION

ANSWER: B(PAGE 2001)

Transthoracic echocardiography (TTE) is particularly effective in identifying the abnormal position and prolapse of the mitral valve leaflets. A useful echocardiographic definition of MVP is systolic displacement (in the parasternal long axis view) of the belly of the mitral valve leaflets by at least 2 mm into the left atrium (LA) superior to the plane of the mitral annulus. There can be prolapse of one or both leaflets . Color flow and continuous wave Doppler imaging is helpful to evaluate the associated MR and provide estimates of severity. The jet lesion of MR due to MVP is most often eccentric, and assessment of the effective regurgitant orifice area and regurgitant volume can be difficult with standard techniques.

88. MITRAL ANNULAR DISJUNCTION ASSOCIATED WITH SUDDEN CARDIAC DEATH HAS BEEN REPORTED IN WHICH OF THE FOLLOWING DISEASES?

A. RHEUMATIC MS

B. CALCIFIC AS

C. MVP

D. RHEUMATIC MR

ANSWER : C(PAGE 2000)

Sudden death is a very rare complication and occurs most often in patients with severe MR and depressed left ventricle (LV) systolic function, although it can occur in individuals with normal LV size and function. A small subset of MVP patients with high-grade ventricular ectopy has been identified with pheno- typic features including electrocardiographic inferior-apical T-wave abnormalities, high-density premature ventricular complexes at rest, mitral annular disjunction (defined as abnormal atrial displacement of the mitral valve leaflet hinge point), and papillary muscle fibrosis on cardiac magnetic resonance imaging with late gadolinium enhance- ment. In addition, there may be an excess risk of sudden death among patients with a flail leaflet.

89. WHICH OF THE FOLLOWING STATEMENTS REGARDING SURGICAL CORRECTION OF MITRAL REGURGITATION IS TRUE?

A. SURGERY SHOULD BE RECOMMENDED FOR ASYMPTOMATIC PATIENTS WITH LV DYSFUNCTION CHARACTERIZED BY AN EF ≤60% OR AN LV END-SYSTOLIC DIMENSION (LV ESD) ≥50 MM

B. ISOLATED MITRAL VALVE REPAIR HAS A OPERATIVE MORTALITY OF 3 %.

C. MITRAL VALVE RELACEMENT IS PREFERRED OVER REPAIR IN PATIENTS WITH MVP OR FLAIL MITRAL LEAFLET

D. MITRACLIP IS CONSIDERED FOR TREATMENT OF SYMPTOMATIC PATIENTS AT PROHIBITIVE OR HIGH SURGICAL RISK WITH SEVERE PRIMARY MR DUE TO MVP

Answer: D(Page 1998, 2001

Surgery for chronic severe primary MR is indicated once symp- toms occur, especially if valve repair is feasible (Fig. 264-1). Surgery should also be recommended for asymptomatic patients with LV dysfunction characterized by an EF ≤60% or an LV end-systolic dimension (LV ESD) ≥40 mm. Other indications for early consider- ation of mitral valve repair in asymptomatic patients include a pro- gressive decrease in LVEF or increase in LV ESD on serial imaging as well as MV anatomy that would predict a >95% of a successful and durable repair in a low surgical risk patient .

Mitral valve repair is preferred over replacement in patients with MVP or flail mitral leaflet . Repair of isolated posterior leaflet prolapse is usually straightforward, but increasingly more complex pathol- ogies (e.g., anterior leaflet prolapse, bileaflet prolapse, Barlow’s deformity) require advanced skills.

Transcatheter edge-to-edge repair (TEER) using a clip to grasp the anterior and posterior leaflets together can be considered for treatment of symptomatic patients at prohibitive or high surgical risk with severe primary MR due to MVP .Most often, the MR will be reduced in severity but not eliminated. Nevertheless, symptom status and indices of LV size and function can be improved with this approach

TABLE 264-2 Mortality Rates after Mitral Valve Surgery[a]

OPERATION	NUMBER	UNADJUSTED OPERATIVE MORTALITY (%)
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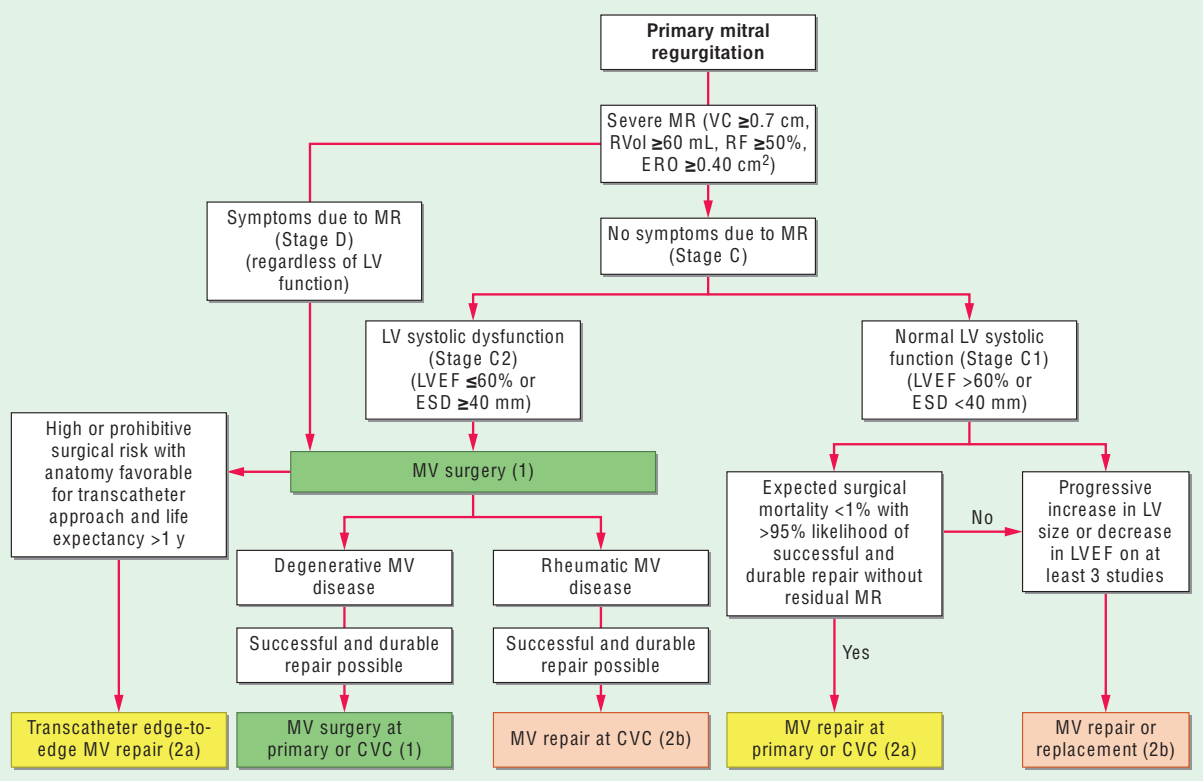


90. WHICH OF THIS IS NOT A CAUSE OF PRIMARY PULMONARY REGURGITATION?

A. CARCINOID

B. POST VALVOTOMY

C. ENDOCARDITIS

D. PULMONARY HYPERTENSION

Answer :D(Page 2004)

TABLE 267-1 Causes of Pulmonic Valve Disease

VALVE LESION	ETIOLOGIES
Pulmonic stenosis	Congenital Carcinoid Tumor Endocarditis
Pulmonic regurgitation	Primary valve disease Congenital Post-valvotomy Endocarditis Carcinoid Annular enlargement Pulmonary hypertension Idiopathic dilation Marfan syndrome

91. WHICH OF THE FOLLOWING CARRIES EXTREMELY HIGH RISK OF MATERNAL MORTALITY AND IS ABSOULUTE CONTRAINDICATION FOR CONTINUING PREGNANCY?

A. SYMPTOMATIC SEVERE MS OR AS

B. MODERATE LEFT VENTRICULAR IMPAIRMENT(EF 30-45%)

C. FONTAN CIRCULATION

D. MODERATE AORTIC DILATATION

Answer:A(Page 2010)

TABLE 269-2 Modified World Health Organization (mWHO) Classification of Heart Disease in Pregnancy

	mWHO I	mWHO II	mWHO II-III	mWHO III	mWHO IV
Diagnosis (if otherwise well and uncomplicated)	Small or mild • Pulmonary stenosis • Patent ductus arteriosus • Mitral valve prolapse Successfully repaired simple lesions (atrial or ventricular septal defect, patent ductus arteriosus, anomalous pulmonary venous drainage) Atrial or ventricular ectopic beats, isolated	Unoperated atrial or ventricular septal defect Repaired tetralogy of Fallot Most arrythmias (supraventricular arrhythmias) Turner syndrome without aortic dilatation	Mild left ventricular impairment (EF >45%) Hypertrophic cardiomyopathy Native or tissue valve disease not considered WHO I or IV (mild mitral stenosis, moderate aortic stenosis) Marfan or other HTAD syndrome without aortic dilatation Aorta <45 mm in bicuspid aortic valve pathology Repaired coarctation Atrioventricular septal defect	Moderate left ventricular impairment (EF 30–45%) Previous peripartum cardiomyopathy without any residual left ventricular impairment Mechanical valve Systemic right ventricle with good or mildly decreased ventricular function Fontan circulation Fontan circulation with good clinical course and without associated comorbidities Unrepaired cyanotic heart disease Other complex heart disease Moderate mitral stenosis Severe asymptomatic aortic stenosis Moderate aortic dilatation (40–45 mm in Marfan syndrome or other HTAD; 45–50 mm in bicuspid aortic valve, Turner syndrome ASI 20–25 mm/m^2, tetralogy of Fallot <50 mm) Ventricular tachycardia	Pulmonary arterial hypertension Severe systemic ventricular dysfunction (EF <30% or NYHA class III–IV) Previous peripartum cardiomyopathy with any residual left ventricular impairment Severe mitral stenosis Severe symptomatic aortic stenosis Systemic right ventricle with moderate or severely decreased ventricular function Severe aortic dilatation (>45 mm in Marfan syndrome or other HTAD, >50 mm in bicuspid aortic valve, Turner syndrome ASI >25 mm/m^2, tetralogy of Fallot >50 mm) Vascular Ehlers-Danlos Severe (re)coarctation Fontan with any complication
Risk	No detectable increased risk of maternal mortality and no/mild increased risk in morbidity	Small increased risk of maternal mortality or moderate increase in morbidity	Intermediate increased risk of maternal mortality or moderate to severe increase in morbidity	Significantly increased risk of maternal mortality or severe morbidity	Extremely high risk of maternal mortality or severe morbidity

92. DIGEORGE SYNDROME IS MOST COMMONLY ASSOCIATED WITH WHCIG CONGENITAL HEART DS?

A. ASD

B. TRANSPOSITION PHYSIOLOGY

C. CONOTRUNCAL LESIONS

D. TRUNCUS ARTERIOSUS

Answer: C (Page 2009)

Conotruncal defects are associated with a number of chromosomal abnormalities, most notably a deletion at chromosome 22q11 (DiGeorge syndrome). Echocardiographic clues to this association in patients with a conotruncal defect include an associated right aortic arch or aberrant subclavian artery. Many adults currently living with conotruncal defects may not have undergone test- ing for DiGeorge syndrome. This condition is important to recognize because a variety of psychiatric disorders and disabilities in cognitive function may be present and go untreated.

93. WHICH OF THE FOLLOWING ECG MARKERS IS ASSOCIATED WITH RISK OF SUDDEN CARDIAC DEATH AFTER A TOF SURGERY?

A. LBBB

B. PROLONGED PR INTERVAL

C. QRS MORE THAN 180 MSEC

D. RBBB

Answer: C(Page 2012,15)

A QRS duration on a resting ECG of 180 ms or more has been associated with increased risk of ventricular tachycardia and sudden death in this patient population

TABLE 269-4 Potential Sequelae of Repaired Tetralogy of Fallot
Right atrial dilation
Right ventricular dilation
Right ventricular dysfunction
Right ventricular outflow tract obstruction
Pulmonary regurgitation
Branch pulmonary artery stenosis
Tricuspid regurgitation
Residual ventricular septal defect
Left ventricular dysfunction
Aortic root dilation
Atrial arrhythmias
Ventricular arrhythmias
Sudden cardiac death

94. RASTELLI PROCEDURE IS DONE FOR WHICH OF THE FOLLOWING CCHD?

A. TOF

B. EISENMENGER SYNDROME

C. D -TGA

D. TAPVC

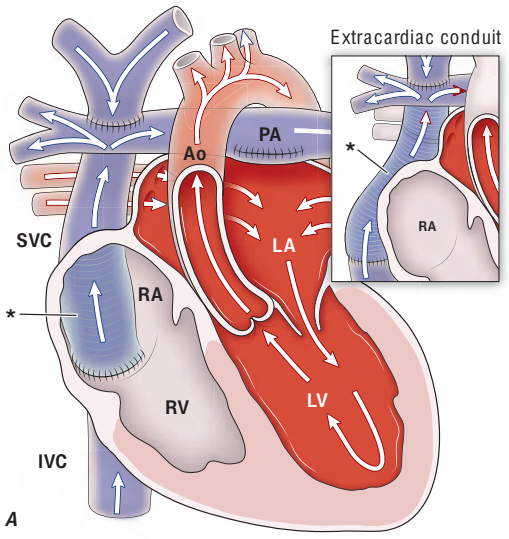
Answer: C(Page 2014)

A subset of patients with D-loop TGA, VSD, and PS may have undergone a Rastelli procedure. This intervention involves placing an RV-to-PA conduit and routing the LV to the aorta through the VSD, which results in relief of cyanosis and the benefit of a systemic LV.

TABLE 269-5 Long-Term Sequelae of D-Loop TGA Surgery

ATRIAL SWITCH	ARTERIAL SWITCH	RASTELLI PROCEDURE
Systemic venous baffle	Arterial anastomosis stenosis	Subaortic stenosis
Pulmonary venous baffle	Branch PA stenosis	RV-PA conduit obstruction
RV (systemic) dysfunction	Neo-aortic root dilation	Pulmonary regurgitation
Tricuspid regurgitation	Neo-aortic regurgitation	Ventricular dysfunction
Baffle leaks	Coronary artery stenosis	
LVOT obstruction (PS)	LV dysfunction	

95. IMAGE BELOW SHOWS WHICH CLASSICAL SURGERY FOR A CCHD



A. FONTAN SURGERY

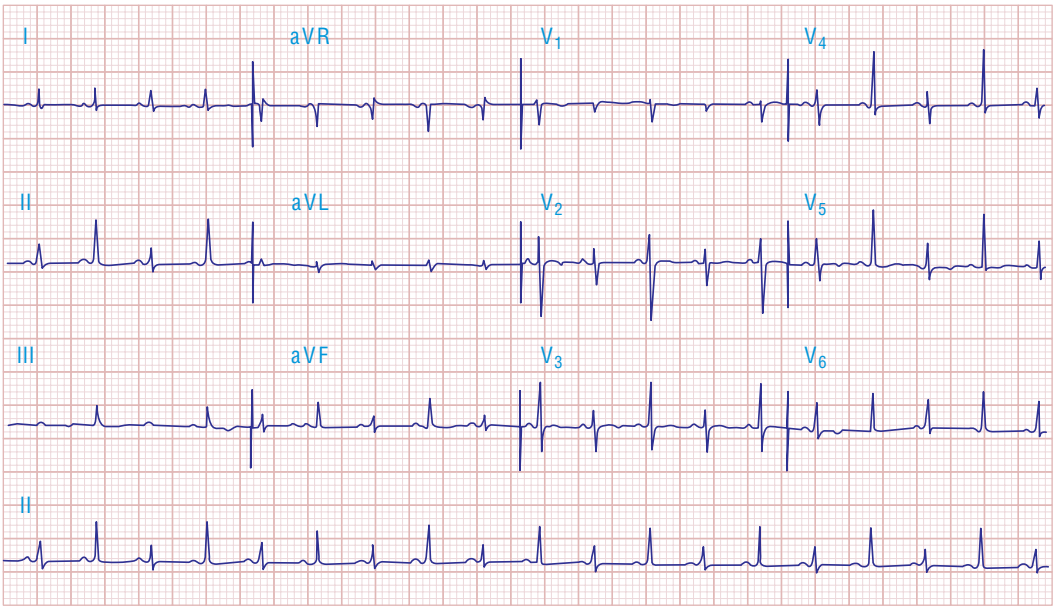
B. RASTELLI SURGERY

C. MODIFIED BT SHUNT
D. POTTS SHUNT

Answer: A(Page 2016)

The Fontan procedure utilizes the single ventri- cle to pump pulmonary venous (oxygenated) blood through the aorta to the body and allows for “passive” flow of systemic venous return of deoxygenated blood through surgically created connections to the lungs. Patients who have undergone a Fontan procedure are at risk for multiple comorbidities in adulthood, including atrial arrhythmias, heart failure, renal and hepatic dysfunction, and both venous

96. BELOW IS THE ECG OF A PATIENT PRESENTING WITH HYPOTENSION. WHICH OF THE FOLLOWING STATEMENT HOLDS TRUE?



A. BECKS TRIAD INCLUDES HYPOTENSION , MUFFLED HEART SOUNDS AND RAISED JVP WITH PROMINENT Y DESCENT
B. PULSUS PARADOX IS UNIVERSALLY SEEN IN PATIENTS WITH CARDIAC TAMPONADE AND CONSTRICTIVE PERICARDITIS
C. ECHOCARDIOGRAPHY SHOWS A EARLY DIASTOLIC COLLAPSE OF RIGHT VENTRICLE AND ATRIUM
D. RIGHT VENTRICULAR INFARCTION MAY RESEMBLE CLINICALLY WITH THIS CONDITION

Answer: D(Page 2021,22

The three principal features of tamponade (*Beck's triad*) are hypotension, soft or absent heart sounds, and jugular venous distention with a prominent *x* (early systolic) descent but an absent *y* (early diastolic) descent.

Paradoxical pulse also occurs in approximately one-third of patients with constrictive pericarditis (see below), and in some cases of hypovolemic shock, acute and chronic obstructive airway disease, and pulmonary embolism. Right ventricular infarction may resemble cardiac tamponade with hypotension, elevated jugular venous pressure, an absent *y* descent in the jugular venous pulse, and occasionally, a paradoxical pulse

In tamponade, there is late diastolic inward motion (collapse) of the right ventricular free wall and the right atrium.

97. WHICH OF THE FOLLOWING IS NOT USED IN THE TREATMENT OF RECURRENT AND RELAPSING PERICARDITIS?

A. STEROIDS

B. ANAKINRA

C. AZATHIOPRENE

D. IVIG

Answer: D(Page 2021)

In patients with multiple, frequent, and disabling recurrences that continue for >2 years, are not prevented by continuing colchic- ine and other NSAIDs, and are not controlled by glucocorticoids, treatment with azathioprine or anakinra (an interleukin 1β receptor antagonist) has been reported to be of benefit. Rarely, pericardial stripping may be necessary; however, this procedure may not always terminate the recurrences.

98. WHICH OF THE FOOLWING ARE NOT ASSOCIATED WITH CARDIAC MYXOMAS?

A. CARNEYS COMPLEX

B. LAMB SYNDROME

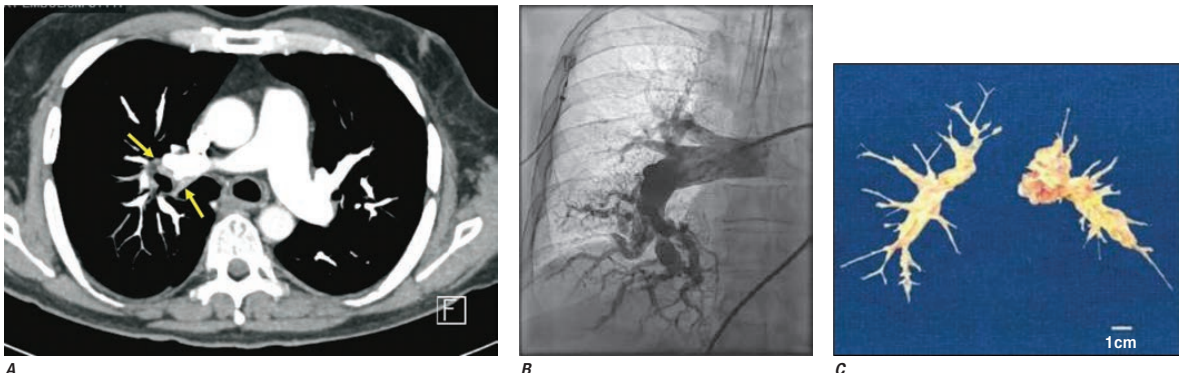
C. NAME SYNDROME

D. MUTATION IN TSC1/2

Answer: D(Page 2026)

Approximately 90% of myxomas are sporadic; the remainder are familial with autosomal dominant transmission. The familial variety often occurs as part of a syndrome complex (Carney complex) that includes (1) myxomas (cardiac, skin, and/or breast), (2) lentigines and/or pigmented nevi, and (3) endocrine overactivity (primary nodular adrenal cortical disease with or without Cushing's syndrome, testicular tumors, and/or pituitary adenomas with gigantism or acromegaly). Certain constellations of findings have been referred to as the *NAME* syndrome (*n*evi, *a*trial myxoma, *m*yxoid neurofibroma, and *e*phelides) or the *LAMB* syn- drome (*l*entigines, *a*trial *m*yxoma, and *b*lue nevi), although these syndromes probably represent subsets of the Carney complex. The genetic basis of this complex has not been elucidated completely; however, inactivating mutations in the tumor-suppressor gene *PRKAR1A*, which encodes the protein kinase A type I-α regulatory subunit, have been identified in ~70% of patients with Carney complex.

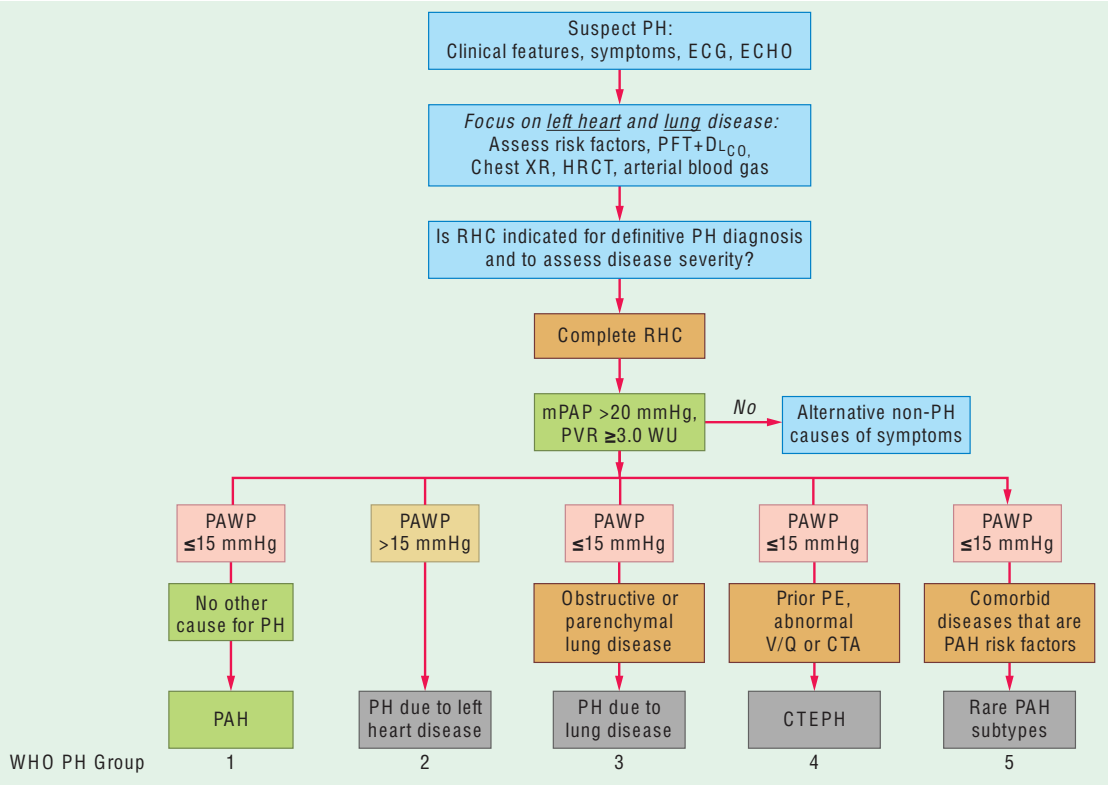
99. BELOW IMAGES FITS INTO WHICH GRADE OF PULMONARY HYPERTENSION?



A. GROUP 1
B. GROUP2
C. GROUP3
D. GROUP4

Answer: D(Page 2125,2126)

The development of PH after chronic thromboembolic obstruction of the pulmonary arteries, termed CTEPH, is well described. The incidence of CTEPH following a sin- gle pulmonary embolic event is difficult to determine accurately, but probably is between 3 and 7% of patients. Importantly, 25% of patients with CTEPH have no history of clinical venous thromboembolism, suggesting that CTEPH may develop following a subclinical pulmo- nary embolism or through a diverse range of mechanisms. Obstruction of the proximal pulmonary vasculature due to webbing, stricture, or focal fibrotic occlusion signifies proximal vessel involvement. Distal pulmonary arterioles remodel by luminal narrowing or obliteration. Approximately 10–15% of patients will develop a disease very similar clinically and pathologically to PAH after resection of the proximal thrombus



100. WHICH OF THE FOLLOWING DRUGS USED FOR TREATMENT OF PAH ACTS AS SOLUBLE GUANYL CYCLASE STIMULATOR?

A. MACITENTAN

B. TADALAFIL

C. RIOCIGUAT

D. SELEXIPAG

Answer: C(Page 2129)

Riociguat increases bioactive cGMP by (1) stabilizing the molecular interaction between $NO^{\bullet}$ and sGC, and (2) directly stimulating sGC independent of $NO^{\bullet}$ bioavailability. Riociguat significantly improved exercise capacity, pulmonary hemodynamics, WHO FC, and time to clinical worsening in patients with PAH and is the sole approved pharmacotherapy for CTEPH patients for whom surgical pulmonary endarterectomy is ineffective or contraindicated.

TABLE 283-2 FDA-Approved Therapies for the Treatment of Pulmonary Arterial Hypertension (PAH)

GENERIC NAME	ROUTE OF ADMINISTRATION	DRUG CLASS	INDICATION
Epoprostenol	IV	Prostacyclin derivative	Treatment of PAH to improve exercise capacity
Iloprost	Inhaled	Prostacyclin derivative	Treatment of PAH to improve a composite endpoint consisting of exercise tolerance, symptoms (NYHA class), and lack of deterioration
Treprostinil	IV or SC	Prostacyclin derivative	Treatment of PAH to diminish symptoms associated with exercise
Treprostinil	Inhaled	Prostacyclin derivative	Treatment of PAH to improve exercise ability
Treprostinil	Oral	Prostacyclin derivative	Treatment of PAH to improve exercise ability
Selexipeg	Oral	Selective IP receptor agonist	Treatment of PAH to improve a composite endpoint lack of clinical deterioration
Bosentan	Oral	Endothelin receptor antagonist	Treatment of PAH to improve exercise capacity and to decrease clinical worsening
Ambrisentan	Oral	Endothelin receptor antagonist	Treatment of PAH to improve exercise capacity and delay clinical worsening
Macitentan	Oral	Endothelin receptor antagonist	Treatment of PAH to improve a composite endpoint of delay of clinical worsening
Sildenafil	Oral or IV	PDE5 inhibitor	Treatment of PAH to improve exercise capacity and delay clinical worsening
Tadalafil	Oral	PDE5 inhibitor	Treatment of PAH to improve exercise ability
Riociguat	Oral	Soluble guanylyl cyclase stimulator	Treatment of PAH to improve exercise ability

101. WHICH OF THIS FACTORS FAVOUR REVASCULARISATION IN PATIENT WITH RENAL ARTERY STENOSIS?

A. RECUURENT CCF IN APATIENT IN WHOM LV DYSFUNCTION CANNOT FULLY EXPLAIN THE CAUSE

B. DECLINE IN GFR DURING THERAPY WITH ACEI/ARBS

C. FAILURE TO ATTAIN TARGET BLOOD PRESSURE ON OMT

D. CONCOMITANT RENAL PARENCHYMAL DS THAT CAUSE PROGRESSIVE RENAL DYSFUNCTION

Answer:D(Page 2090)

TABLE 278-2 Clinical Factors That Determine the Role of Revascularization in Addition to Medical Therapy for Renal Artery Stenosis

Factors Favoring Medical Therapy with Revascularization for Renal Artery Stenosis
• Progressive decline in GFR during treatment of systemic hypertension
• Failure to achieve adequate blood pressure control with optimal medical therapy (medical failure)
• Rapid or recurrent decline in the GFR in association with a reduction in systemic pressure
• Decline in the GFR during therapy with ACE inhibitors or ARBs
• Recurrent congestive heart failure in a patient in whom left ventricular dysfunction does not fully explain the cause
Factors Favoring Medical Therapy and Surveillance of Renal Artery Disease
• Controlled blood pressure with stable renal function (e.g., stable renal insufficiency)
• Stable renal artery stenosis without progression on surveillance studies (e.g., serial duplex ultrasound)
• Advanced age and/or limited life expectancy
• Extensive comorbidity that make revascularization too risky
• High risk for or previous experience with atheroembolic disease
• Other concomitant renal parenchymal diseases that cause progressive renal dysfunction (e.g., interstitial nephritis, diabetic nephropathy), particularly with proteinuria

102. WHICH OF THE FOLLIOWNG IS NOT A CAUSE OF HYPETRENSION WITH HYPOKALEMIA?

A. RENAL ARTERY STENOSIS

B. CONNS SYNDROME

C.CUSHINGS SYNDROME

D. PHEOCHROMOCYTOMA

Answer: D

103. WHICH OF THE FOLLOWING RARE MENDELIAN FORMS OF HYPERTENSION IS ASSOCIATED WITH HYPERKALEMIA?

A. GORDONS SYNDROME

B. LIDDLES SYNDROME

C.APPARENT MINERALOCORTICOID EXCESS SYNDROME

D. GLUCOCORTICOID REMEDIABLE HYPERALDOSTERONISM

Answer: A(Page 2080)

TABLE 277-4 Rare Mendelian Forms of Hypertension

DISEASE	PHENOTYPE	GENETIC CAUSE
Glucocorticoid-remediable hyperaldosteronism	Autosomal dominant Absent or mild hypokalemia	Chimeric 11 -hydroxylase/aldosterone gene on chromosome 8
17 -Hydroxylase deficiency	Autosomal recessive Males: pseudohermaphroditism Females: primary amenorrhea, absent secondary sexual characteristics	Random mutations of the *CYP17* gene on chromosome 10
11 -Hydroxylase deficiency	Autosomal recessive Masculinization	Mutations of the *CYP11B1* gene on chromosome 8q21-q22
11 -Hydroxysteroid dehydrogenase deficiency (apparent mineralocorticoid excess syndrome)	Autosomal recessive Hypokalemia, low renin, low aldosterone	Mutations in the 11 -hydroxysteroid dehydrogenase gene
Liddle's syndrome	Autosomal dominant Hypokalemia, low renin, low aldosterone	Mutation subunits of the epithelial sodium channel *SCNN1B* and *SCNN1C* genes
Pseudohypoaldosteronism type II (Gordon's syndrome)	Autosomal dominant Hyperkalemia, normal glomerular filtration rate	Linkage to chromosomes 1q31-q42 and 17p11-q21
Hypertension exacerbated in pregnancy	Autosomal dominant Severe hypertension in early pregnancy	Missense mutation with substitution of leucine for serine at codon 810 (MR_{L810})
Polycystic kidney disease	Autosomal dominant Large cystic kidneys, renal failure, liver cysts, cerebral aneurysms, valvular heart disease	Mutations in the *PKD1* gene on chromosome 16 and *PKD2* gene on chromosome 4
Pheochromocytoma	Autosomal dominant (a) Multiple endocrine neoplasia, type 2A Medullary thyroid carcinoma, hyperparathyroidism (b) Multiple endocrine neoplasia, type 2B Medullary thyroid carcinoma, mucosal neuromas, thickened corneal nerves, alimentary ganglioneuromatoses, marfanoid habitus (c) von Hippel-Lindau disease Retinal angiomas, hemangioblastomas of the cerebellum and spinal cord, renal cell carcinoma (d) Neurofibromatosis type 1 Multiple neurofibromas, café-au-lait spots	 (a) Mutations in the *RET* protooncogene (b) Mutations in the *RET* protooncogene (c) Mutations in the *VHL* tumor-suppressor gene (d) Mutations in the *NF1* tumor-suppressor gene