1 Cardiology

Questions

Answers can be found in the Cardiology Answers section at the end of this chapter.

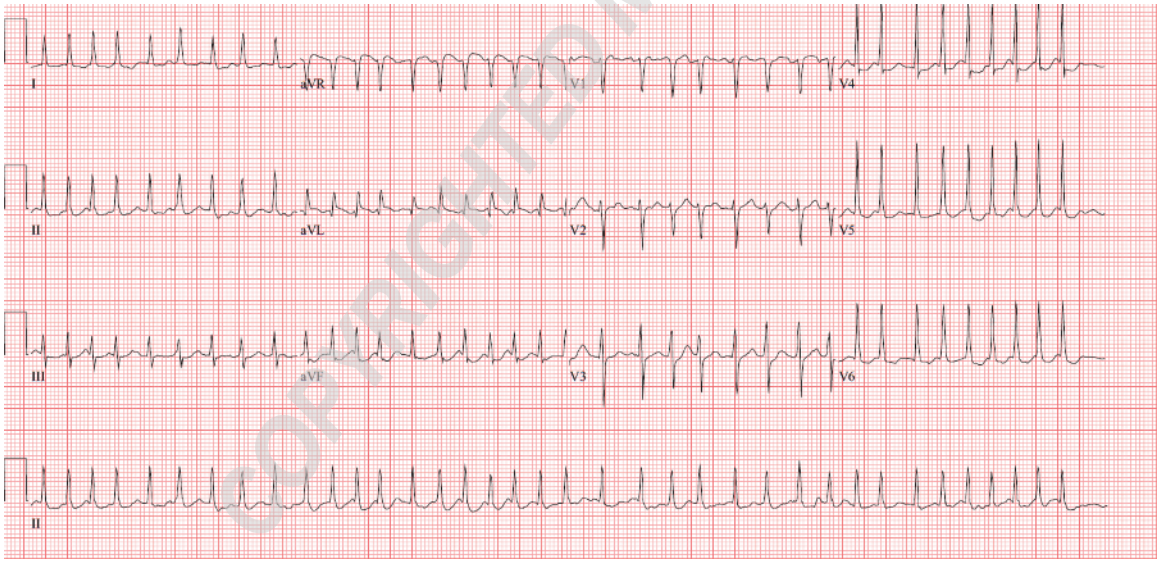
1. A 65-year-old accountant undergoes an abdominal ultrasound because of mildly abnormal liver function tests. The ultrasound reveals a few mobile gallstones and a 5 cm abdominal aortic aneurysm. He drinks three to four standard drinks of alcohol every day and is an ex-smoker. He is known to have hypertension and is taking irbesartan 150 mg daily. Blood pressure control is satisfactory with mean systolic BP of 130 mmHg.

What is your most appropriate course of action?

A. Abdominal CT with contrast immediately and suspension of driver's license.
B. Endovascular aneurysm repair immediately.
C. Follow up ultrasound in 6 months and continue driving.
D. Open surgical aneurysm repair immediately.

2. A 39-year-old man with a known atrial septal defect presents to emergency department with a 6-hour history of palpitations. His ECG is shown below:

Which one of the following signs is **UNLIKELY** to be present?



A. Fixed splitting of second heart sound.
B. Fourth heart sound.
C. Loud first heart sound.
D. Third heart sound.

How to Pass the FRACP Written Examination, First Edition. Jonathan Gleadle, Jordan Li, Danielle Wu, and Paul Kleinig.

3. Which of the following patient characteristics is **LEAST LIKELY** to increase an individual's susceptibility to anthracycline cardiomyopathy?

A. Age of 70 years.
B. Male sex.
C. Mediastinal radiotherapy.
D. Positive carrier status for *C282Y HFE* gene.

4. A 65-year-old-man presents with a three-month history of exertional dyspnoea. He is found to have aortic stenosis with a valve area of 0.9 cm^2 and a mean transvalvular pressure gradient of 15 mmHg. His left ventricle ejection fraction (LVEF) is 35%. A Dobutamine Stress Echocardiography (DSE) has been arranged which will provide all of the following information, **EXCEPT**:

A. Confirming the suitability for valve replacement.
B. Deciding the need for cardiac resynchronisation therapy.
C. Predicting prognosis post valve replacement.
D. Diagnosing low-flow, low-gradient aortic stenosis.

5. An 84-year-old man with severe aortic stenosis complains of shortness of breath after walking for 20 metres and a couple of episodes of unexplained collapse. He is independent with activities of daily living. His medical history includes hypertension, hyperlipidaemia, cholecystectomy, and hernia repair.

What is the most appropriate management approach?

A. Aortic valve balloon valvuloplasty.
B. Implantable cardioverter–defibrillator (ICD).
C. Surgical aortic valve replacement (SAVR).
D. Transcatheter aortic valve implantation (TAVI).

6. You see a 75-year-old woman with a new diagnosis of atrial fibrillation. Her CHA_2DS_2-VASc score is 4. She has a history of myocardial infarction four years ago, treated with percutaneous coronary intervention and a bare-metal stent inserted in the right coronary artery, and is currently on aspirin.

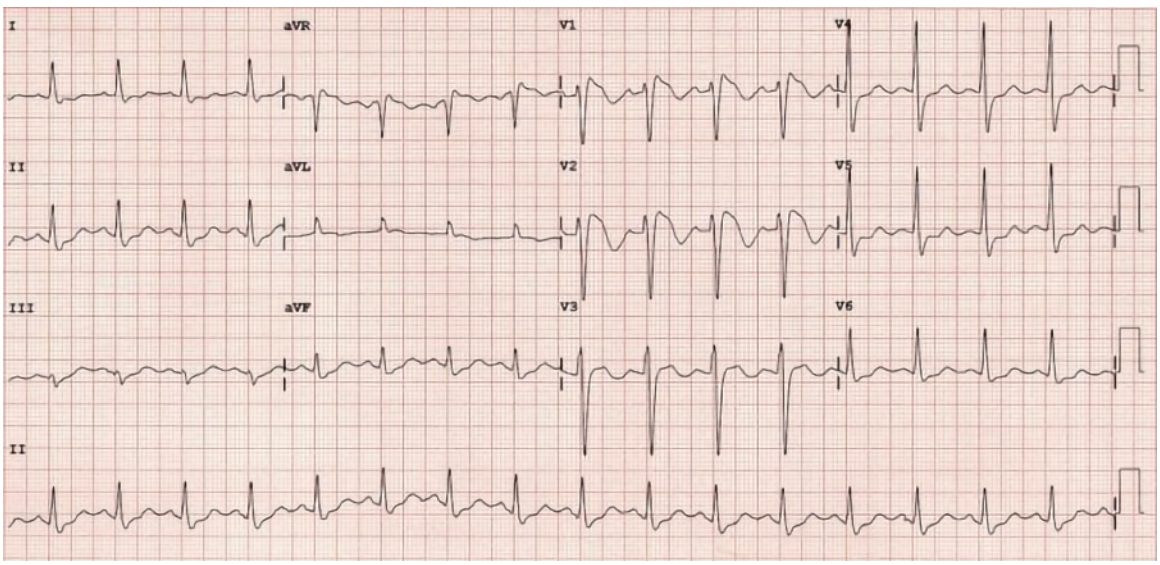
Which of the following options is the most appropriate regarding ongoing anti-thrombotic therapy?

A. Coronary angiogram to guide further therapy.
B. Rivaroxaban and clopidogrel.
C. Rivaroxaban and aspirin.
D. Rivaroxaban monotherapy.

7. Beta-blockers are recommended as first line therapy for stable angina. Their main mechanism of action is explained by:

A. Increased coronary artery blood flow.
B. Plaque stabilisation.
C. Reduction in blood pressure.
D. Reduction in myocardial oxygen demand.

8. What is the management strategy for a patient with the following ECG?



A. Amiodarone.
B. Beta-blocker.
C. Implantable cardioverter–defibrillator (ICD).
D. Pacemaker.

9. A 54-year-old man is admitted to hospital because of syncope. This is his third presentation with syncope due to severe postural hypotension over the past six months. He has developed chronic diarrhoea and lost 6 kg of body weight in the past six months. He has no significant past medical history. On examination, BP is 90/50 mmHg. HR is 86 bpm. There are no murmurs. Urinary analysis shows ++++ protein but no RBCs nor RBC casts. His investigation results are shown below. ECG shows sinus rhythm and low voltage in all leads. Echocardiogram reports moderate left ventricular hypertrophy, biatrial dilatation and grade 2 diastolic dysfunction.

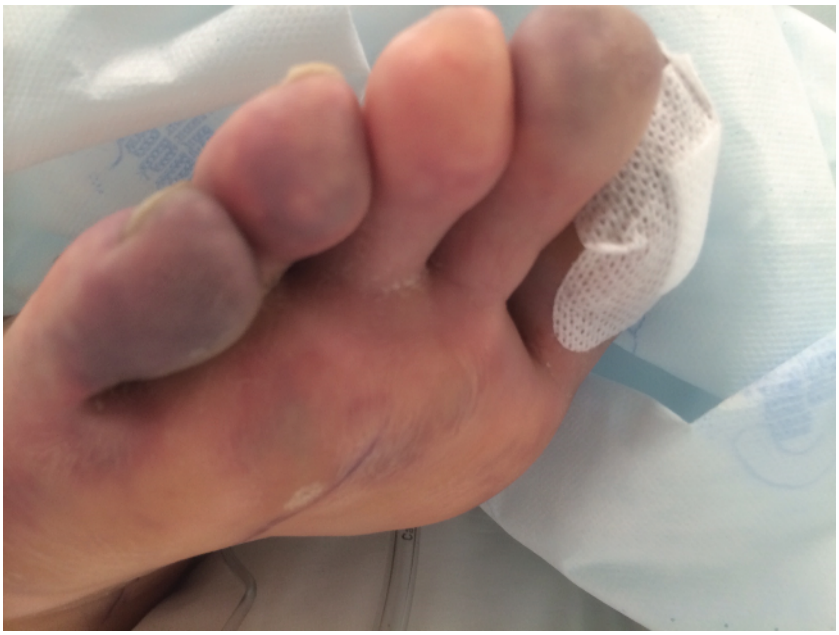
Tests	Results	Normal values
Haemoglobin	108 g/L	135–175
White blood cell	5.48 x 10^9/L	4.0–11.0
Platelet	206 x 10^9/L	150–450
Sodium	133 mmol/L	135–145
Potassium	4.3 mmol/L	3.5–5.2
Creatinine	156 µmol/L	60–110
Albumin	22 g/L	34–48
Globulin	42 g/L	21–41
Liver function tests	normal	
Troponin	<29 ng/L	0–29
N-terminal pro b-type Natriuretic Peptide (NT-proBNP)	1800 ng/L	0–124

What would you consider the most appropriate next investigation?
A. Cardiac MRI.
B. Coronary artery angiogram.
C. Holter monitor.
D. Implantable loop recorder.

10. Which one of the following increases cardiac output?
A. Atropine.
B. Acidosis.
C. Beta-blockers.
D. Hypertension.

11. A 72-year-old woman presents to emergency department after an episode of loss of consciousness. Which of the following clinical features, if present, **DO NOT** increase the likelihood that her loss of consciousness was due to cardiac syncope?
A. Breathlessness prior to the episode.
B. Cyanosis during the episode.
C. History of atrial fibrillation.
D. Significant injury as a result of loss of consciousness.

12. An 80-year-old man presents to emergency department with sudden onset of left-sided weakness two hours ago. His medical history includes hypertension, hypercholesterolaemia, and atrial fibrillation for which he is taking aspirin only. CT head shows acute right middle cerebral artery territory infarction. He is treated with thrombolysis followed by bridging low molecular weight heparin then a direct thrombin inhibitor. Two weeks later while in rehabilitation, he develops low grade fever, myalgia, painful feet (shown below), anaemia, and AKI.



The most likely diagnosis is:

A. Acute allergic reaction to direct thrombin inhibitor.
B. Antiphospholipid syndrome.
C. Cholesterol embolisation.
D. Multiple emboli due to inappropriate use of an oral direct thrombin inhibitor.

13. Which one of the following patients can be investigated appropriately with a computed tomography coronary angiography (CTCA)?

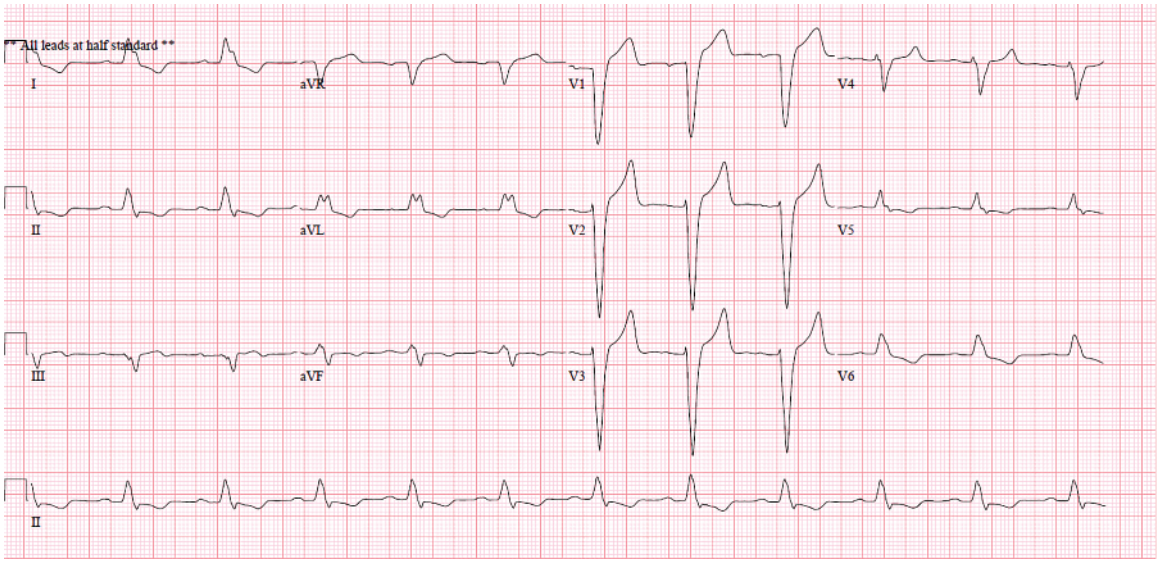
A. An asymptomatic patient with history of type 2 diabetes and normal renal function.
B. A patient with previous coronary stents presenting with chest pain and possible in-stent restenosis.
C. A patient presenting with central chest pain and rapid atrial fibrillation.
D. A patient with chest pain but with a low pre-test probability of coronary artery disease.

14. A 75-year-old man presents with transient weakness of his left arm. He is diagnosed with a transient ischaemic attack. He is known to have hypertension, type 2 diabetes, alcohol dependence, recent weight loss, and a low-grade fever. He undergoes a transthoracic echocardiogram which reveals a 12 mm mitral-valve vegetation.

Which one of the following statements is true?

A. Blood culture is negative in over 20% of cases of infectious endocarditis.
B. Cerebral complications are the most frequent extracardiac complications.
C. Majority (80%) of cases of infectious endocarditis develop in patients with known valvular disease.
D. Streptococci in the most common pathogen isolated.

15. A 75-year-old man presents with increasing dyspnoea. You note that he had four admissions in the past year due to decompensated CCF. He is known to have ischaemic heart disease with a drug eluting stent in two coronary arteries, insulin dependent type 2 diabetes, hypertension, stage 4 CKD with an eGFR of 26 ml/min/1.73 m^2 and severe smoking related COPD. His medications include aspirin, clopidogrel, perindopril, gliclazide, frusemide, digoxin, spironolactone, atorvastatin, formoterol/,budesonide inhaler, and tiotropium inhaler. Physical examination findings are consistent with decompensated CCF. His ECG is shown below. A dobutamine stress echocardiogram demonstrates no reversible ischaemic change but a large antero-apical area of akinesia. The ejection fraction is 30%.



The most effective treatment to reduce the frequency of readmissions and improve survival is:

A. Add a SGLT2 inhibitor.
B. Commence a beta-blocker.
C. Insert a biventricular pacemaker and defibrillator.
D. Insert a dual-chamber pacemaker.

16. Which one of the following medications is recommended in patients with type 2 diabetes with cardiovascular disease and inadequate glycaemic control despite metformin, to reduce the risk of cardiovascular events and hospitalisation for heart failure?

A. Dipeptidyl peptidase-4 (DPP-4) inhibitors.
B. Glucagon-like peptide-1 (GLP-1) analogues.
C. Sodium-glucose co-transporter-2 (SGLT2) inhibitors.
D. Thiazolidinediones.

17. A 70-year-old woman presents with a 2-month history of exertional dyspnoea for evaluation. Her medical history includes longstanding hypertension, obesity, OSA, and permanent AF. On examination, there are clinical signs consistent with CCF. Her troponin level is normal, but NT-pro BNP level is elevated. Her echocardiogram demonstrates a left ventricular ejection fraction of 55%, left ventricular end-diastolic volume index (LVEDI) <97 mL/m^2 and the ratio of mitral early diastolic inflow velocity to mitral early annular lengthening velocity (E/e')>15.

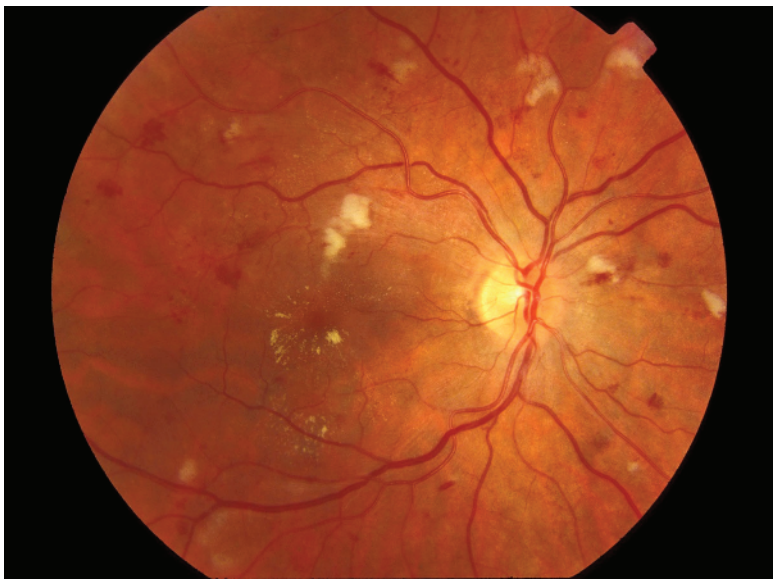
Which one of the following treatments will reduce mortality for this patient?

A. Angiotensin-converting enzyme inhibitor.
B. No pharmacological treatment proven to reduce mortality.
C. Phosphodiesterase-5 inhibitor.
D. Selective sinus node I_f sodium channel inhibitor.

18. In a 28-year-old man with known hypertrophic cardiomyopathy (HOCM) who has had one episode of syncope at work, which one of the following treatments should be instituted?

A. Amiodarone.
B. Anticoagulation.
C. Atenolol.
D. Implantable cardioverter–defibrillator.

19. A 56-year-old man presents to emergency department with severe headaches and blurred vision. He is known to have IgA nephropathy but has no regular follow up for this. His BP is 210/110 mmHg. You have performed a fundoscopic examination which is shown below:



What does the fundoscopy show?

A. Keith-Wagener (KG) grade 3 hypertensive retinopathy.
B. Keith-Wagener (KG) grade 4 hypertensive retinopathy.
C. Retinal artery occlusion.
D. Retinal vein occlusion.

20. Which one of the following patients **DOES NOT** have an indication for an implantable cardioverter defibrillator (ICD) implantation?

A. A 20-year-old man with congenital long QT syndrome with recurrent syncope who cannot tolerate beta-blockers.
B. A 65-year-old woman with haemodynamically unstable ventricular tachycardia not due to reversible causes.
C. A 59-year-old man with ischaemic cardiomyopathy with a myocardial infarction two weeks ago with left ventricular ejection fraction of 30%.
D. A 22-year-old asymptomatic female with hypertrophic cardiomyopathy, unexplained syncope but no family history of sudden cardiac death.

21. A 56-year-old man with a background history of type 2 diabetes, hypertension, and hyperlipidaemia presents with unexplained syncope and palpitations that happen around 6-monthly. His previous cardiac investigations, including repeated 12-lead ECG and 24-hour Holter monitoring, are normal.

What is the appropriate next step to investigate palpitations?

A. Three-day Holter monitoring.
B. Seven-day Holter monitoring.
C. Wearable device.
D. Implantable loop recorder.

22. Which one of the following lipid-lowering agents acts by blocking the inhibition of lysosomal degradation of low-density lipoprotein (LDL) receptors, thereby increasing the body's ability to sequester LDL?

A. Evolocumab.
B. Ezetimibe.
C. Mipomersen.
D. Rosuvastatin.

23. A 65-year-old man presents with dizziness and syncope. His ECG shows a QTc interval of 520 ms. Review of medical history, family history, medications, electrolytes, and echocardiogram did not find a reversible cause of the condition.

What is the first-line treatment for this patient?

A. Amiodarone.
B. Beta-blockers.
C. Implantable cardioverter defibrillator (ICD).
D. Left cardiac sympathetic denervation (LCSD).

24. A 25-year-old Aboriginal and Torres Strait Islander woman presents with a 3-month history of exertional dyspnoea. She has had an unproductive cough but no fevers, chest pain or other illnesses. She takes no medication. On examination, BP is 120/70 mmHg, HR 90/min and regular, there is a 2/6 diastolic murmur and a 3/6 systolic murmur, chest is clear, there is pitting oedema of both ankles. Echocardiogram reveals mitral stenosis with a mean transvalvular gradient of 14 mmHg and moderate mitral regurgitation. The left atrium is enlarged. There is normal biventricular size and function, as well as pulmonary hypertension with a pulmonary arterial pressure of 50 mmHg.

Which of the following is the most appropriate management?

A. Balloon mitral valvuloplasty.
B. Commence ACE inhibitor and repeat echocardiogram in 6 months.
C. Mitral valve open commissurotomy.
D. Mitral valve replacement.

25. A 65-year-old man suffers from ischaemic heart disease, chronic AF, insulin-dependent type 2 diabetes, stage 3 CKD, peripheral vascular disease with chronic claudication. He is taking multiple medications and is asking your advice about taking omega-3 fish oil supplements.

Which one of the following pieces of advice regarding omega-3 fish oil supplements for this patient is correct?

A. It is associated with a statistically significant reduction on all-cause mortality.
B. It has a beneficial effect on glycaemic control and increased fasting insulin levels.
C. It can improve walking distance, ankle brachial pressure index, and angiographic findings.
D. It can reduce serum triglycerides and raise HDL and LDL levels.

26. A 55-year-old man presents with repeated clinic blood pressure measurements of around 150/90 mmHg, after six months of therapy with perindopril, amlodipine, and hydrochlorothiazide at maximal doses. He is compliant with his medications and is engaging actively with lifestyle modifications.

Which one of the following additional agents is most likely to be beneficial?

A. Atenolol.
B. Doxazosin.
C. Hydralazine.
D. Spironolactone.

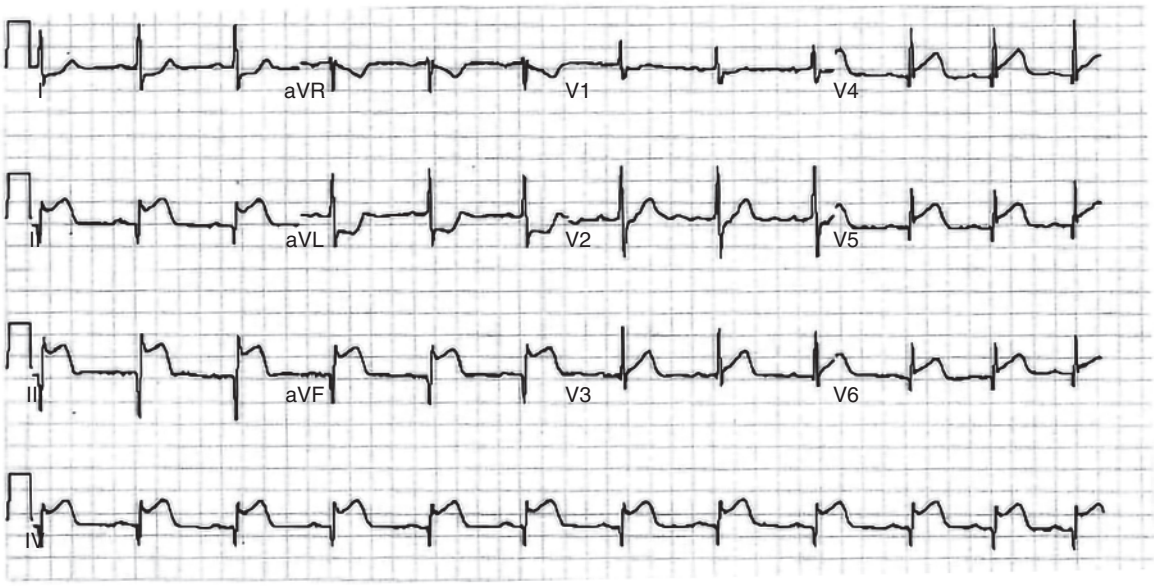
27. A 50-year-old woman with haemochromatosis presents with dyspnoea. She undergoes an echocardiogram. Which of the echocardiogram findings is most commonly seen in patients with early-stage restrictive cardiomyopathy?

A. Left ventricular dilatation with reduced left ventricular ejection fraction <45%.
B. Left ventricular outflow tract obstruction.
C. Normal ventricular size and systolic function with a restrictive ventricular filling pattern.
D. Regional wall motion abnormality in a non-coronary distribution.

28. Which of the following statements is correct regarding acute rheumatic fever (ARF) in Aboriginal and Torres Strait Islander (ATSI)?

A. It is usually associated with Group B streptococcal infection.
B. Secondary prophylaxis following rheumatic fever should be oral doxycycline.
C. The highest rates of ARF in ATSI are between the ages 34 to 45.
D. The major manifestations of ARF include carditis and chorea.

29. A 60-year-old woman presents with epigastric pain, nausea, vomiting, and shortness of breath. She has a HR of 58 bpm and a BP of 90/60 mmHg. Her ECG is shown below:



The occlusion of which coronary artery is likely to have produced this presentation.

A. Circumflex.
B. Left anterior descending.
C. Left marginal.
D. Right.

30. A 55-year-old man is referred by his GP because of bradycardia with a heart rate as low as 45 bpm at night. He has hypertension for which he is taking amlodipine 5 mg daily. An ECG performed today shows a sinus bradycardia 55/min and he is asymptomatic.

Which one of the following statements is correct?

A. Nocturnal bradycardia is an indication for permanent pacing.
B. Sinus node dysfunction is most likely due to ischaemic heart disease.
C. Sleep apnoea is not associated with nocturnal bradycardia.
D. There is no minimum heart rate or pause duration for which permanent pacing is recommended in sinus node dysfunction.

31. A 38-year-old woman is admitted to intensive care unit because of septic shock due to meningococcal septicaemia. She complains of increased dyspnoea on day 3 when she is discharged to the ward. She has a medical history of asthma and chronic back pain. She has been experiencing depressive symptoms since her husband passed away one year ago. Her ECG shows ST depression in the lateral leads. Initial Troponin I level is 54 ng/L [<29], N-terminal pro-B-type brain natriuretic peptide (NT-proBNP) level is 5400 ng/L [0–124]. Her echocardiogram shows ballooning of the left ventricular apex.

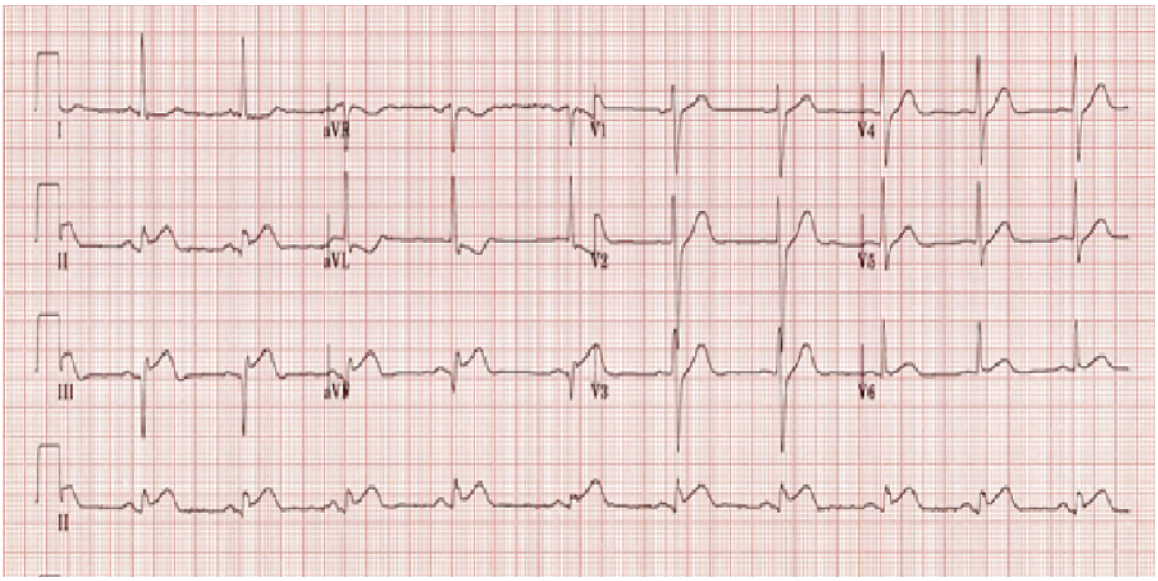
Which of the following medications will improve her survival at one year?

A. Angiotensin-receptor blockers.
B. Beta-blockers.
C. Calcium channel blockers.
D. Digitalis glycosides.

32. Which of the following statements is correct regarding transcatheter aortic valve implantation (TAVI) in inoperable and high-risk elderly patients?

A. Patients should be anticoagulated with a novel oral anticoagulant (NOAC) for 3 months post implantation.
B. Patients should be anticoagulated with dual antiplatelet therapy for 3 months post implantation.
C. Patients with asymptomatic severe aortic stenosis at intermediate surgical risk should be offered TAVI.
D. The need for permanent pacemaker insertion due to bradyarrhythmias post TAVI is about 30%.

33. A 62-year-old man presents with vague chest discomfort for 6 hours. He is known to have insulin-dependent type 2 diabetes, hypertension, hyperlipidaemia, stage 3A CKD with serum creatinine 150 μmol/L [60–110] and psoriatic arthritis treated with adalimumab. His ECG is shown below. His coronary artery angiography shows 50% stenosis of the left main, 75% stenosis of the left circumflex, 70% stenosis of the proximal left anterior descending artery, and 50% stenosis of the right coronary artery. Left ventricular systolic function is reduced with an ejection fraction of 40%.



Which one of the following is the best management option?

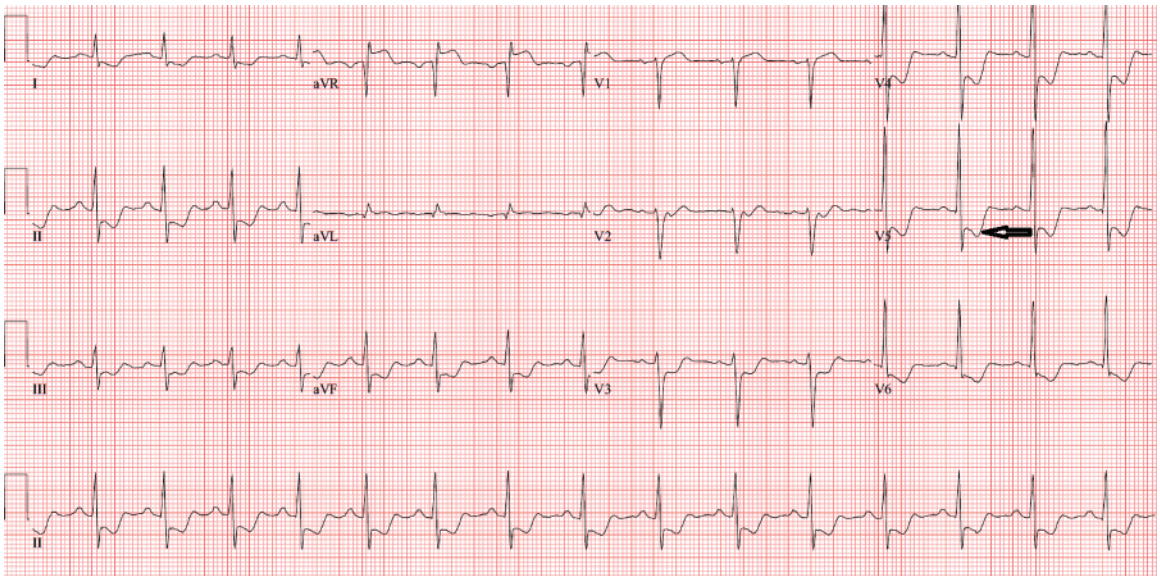
A. Coronary artery bypass graft surgery (CABG).
B. Infarct-related artery (IRA)-only revascularisation in primary PCI.
C. PCI plus biventricular pacemaker–defibrillator.
D. Percutaneous coronary intervention (PCI).

34. A 51-year-old woman presents to the emergency department with cellulitis of her left lower leg and epigastric discomfort after being on oral antibiotics for three days. She is otherwise well and has no other symptoms and ECG is normal. She is known to have autosomal dominant polycystic kidney disease with a serum creatinine of 86 μmol/L [60–110]. A serum high-sensitivity troponin (hs-cTn) is requested and the result is 40 ng/L [<29].

What is the best interpretation of this result in terms of the likelihood of acute coronary syndrome?

A. Likely because the specificity of hs-cTn is high.
B. Likely because the pre-test probability is high.
C. Unlikely because the specificity of hs-cTn is low.
D. Unlikely because the pre-test probability is low.

35. A 78-year-old woman is admitted to the Acute Medical Unit with severe community acquired pneumonia. Her BP is 90/60 mmHg and oxygen saturation is 90% on 4 L of oxygen. Her other medical history includes type 2 diabetes, stage 3B CKD, and hypertension. A central venous line is inserted because of difficult venous access. She complains of increased dyspnoea. A bedside ECG is taken and shown below. Troponin level is 289 ng/L [<29].



Which one of the following is the most likely diagnosis?

A. Type 1 myocardial infarction.
B. Type 2 myocardial infarction.
C. Type 3 myocardial infarction.
D. Type 4 myocardial infarction.

36. A 55-year-man presents with a 2-hour history of palpitations and chest discomfort. He had a similar episode one year ago. He is known to have ankylosing spondylitis, diet-controlled type 2 diabetes, and asthma. He uses a salbutamol inhaler two to three times a week. On examination, he is alert and orientated, BP is 110/60 mmHg, pulse rate is 150 bpm, SaO2 on room air is 95%. There is scattered expiratory wheeze. There is no heart murmur. His current ECG is shown in Figure 1.1A, while Figure 1.1B shows

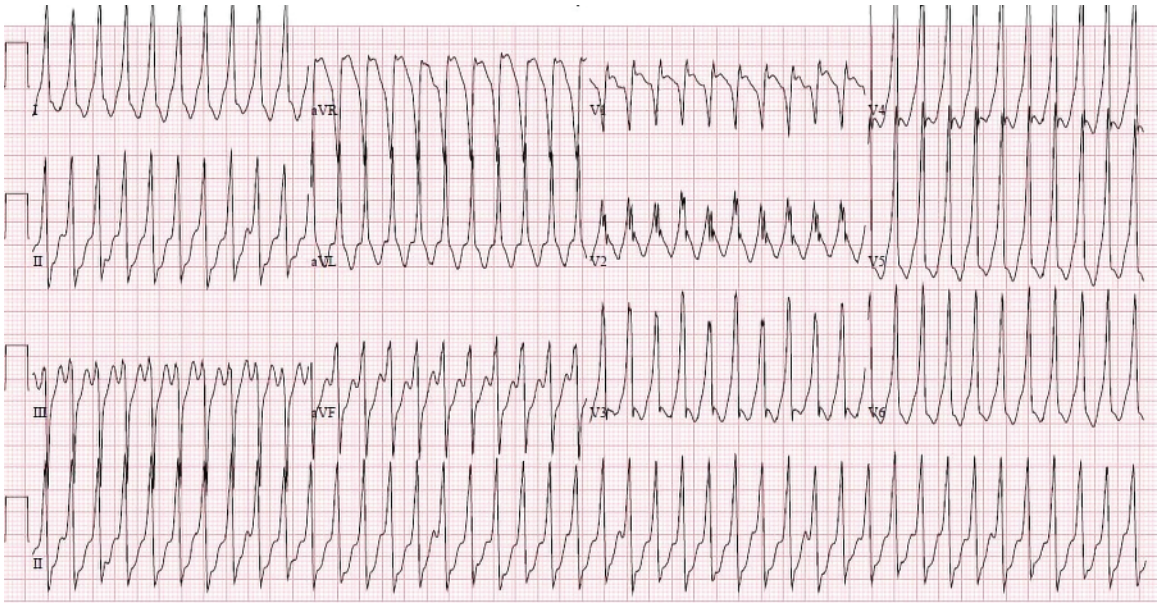


Figure 1.1A

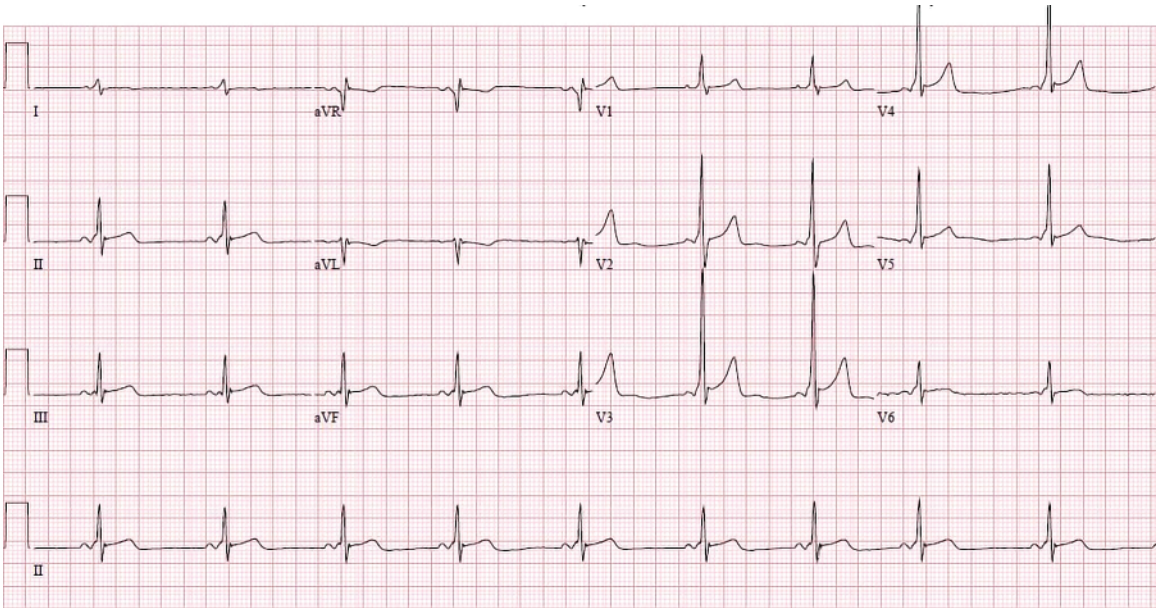


Figure 1.1B

an ECG taken 1-year ago during an infective exacerbation of asthma. His biochemistry results and troponin T are within normal reference range.

The most appropriate treatment for rate control is:

A. Intravenous adenosine.
B. Intravenous digoxin.
C. Intravenous flecainide.
D. Intravenous verapamil.

QUESTIONS (37–43) REFER TO THE FOLLOWING INFORMATION

Match the following blood pressure lowering agents to their mechanism of action.

A. Decreased renin secretion and decreased heart rate.
B. Decreased central synthesis of catecholamines.
C. Decreasing degradation of circulating natriuretic peptides.
D. Relaxation of smooth muscle through opening K_{ATP} channels.
E. Blocking the angiotensin I binding site on angiotensin converting enzyme (ACE).
F. Interfering with Ca^{2+} release on the sarcoplasmic reticulum.
G. Increased guanylyl cyclase activity.

37. Minoxidil

38. Sacubitril

39. Moxonidine

40. Captopril

41. Glyceryl trinitrate

42. Nebivolol

43. Hydralazine.

QUESTIONS (44–51) REFER TO THE FOLLOWING INFORMATION

Match the clinical features listed below to the most likely type of congenital heart disease or clinical syndrome found in patients.

A. Atrial septal defect (ASD).
B. Ventricular septal defect (VSD).
C. Patent ductus arteriosus (PDA).
D. Coarctation of the aorta (CoA).
E. Eisenmenger's syndrome.
F. Marfan's syndrome.
G. Tetralogy of Fallot.
H. Transposition of the great arteries.

44. Cyanosis, clubbing, polycythaemia, an elevated JVP with a dominant *a* wave pattern, right ventricular heave, palpable pulmonary component of the second heart sound (P2), loud P2, fourth heart sound, pulmonary ejection click, and pulmonary regurgitation on auscultation.

45. Cyanosis, clubbing, polycythaemia, right ventricular heave, a thrill at the left sternal edge, a single second heart sound (A2) and short pulmonary ejection murmur on auscultation, a boot shape heart, right ventricular enlargement, and decreased vascularity of lung vessels on CXR.

46. A better developed upper body in comparison with the lower limbs, radiofemoral delay, and hypertension in the upper limbs only, midsystolic murmur over the precordium and back, hypertensive changes in the fundi, a small aortic knuckle and rib notching on CXR.

47. Fixed splitting of the second heart sound (S2), pulmonary systolic ejection murmur (increasing on inspiration), signs of pulmonary hypertension.

48. A thrill and a harsh pansystolic murmur to the left sternal edge, mitral regurgitation.

49. Arachnodactyly, joint hypermobility, long, thin limbs, a long and narrow face, lens dislocation, blue sclerae, a high-arched palate, pectus excavatum, aortic regurgitation, mitral valve prolapse, kyphoscoliosis, and arm span exceeding overall height.

50. A continuous murmur along the left sternal border, ECG shows left ventricular hypertrophy, CXR shows increased pulmonary vasculature.

Answers

1. Answer: C

An aneurysm is an artery that has enlarged to greater than 1.5 times the expected diameter. In the infrarenal aorta, the threshold diameter is accepted as 3.0 cm. Abdominal aortic aneurysm (AAA) affects approximately 4–7% of men and 1–2% of women over the age of 65 years.

Medical therapy options remain limited and no aneurysm-specific pharmacotherapy is currently available. Medical management of AAA generally involves cardiovascular risk reduction, including antiplatelet, statin, and antihypertensive therapy. The best medical management is generally not intended to limit expansion or reduce the size of the AAA. However, managing cardiovascular risk factors is crucial for improving the overall survival of patients and the outcomes of future AAA repair.

Australian national driving regulations stipulate that untreated atherosclerotic aortic aneurysms >5.5 cm disqualify patients from an unconditional driver's licence, except with the approval of a treating vascular surgeon.

Patients with large aneurysms (men >5.5 cm; women >5.0 cm) should be considered for elective aneurysm repair. The optimal management of small AAAs (4.0–5.5 cm) has been clarified by several large, randomised control trials which demonstrate no long-term survival benefit with open or endovascular repair.

Most AAAs detected with screening or as an incidental finding are below the threshold for elective repair. The risk of AAA rupture increases with AAA diameter (Table 1.1). The surveillance interval for asymptomatic AAA also depends on AAA diameter (Table 1.2).

Table 1.1 Annual AAA rupture risk by diameter.

AAA diameter (cm)	Rupture risk (%/year)
3.0–3.9	0%
4.0–4.9	1%
5.0–5.9	1–10%
6.0–6.9	10–22%
>7.0	30–50%

Source: Based on Chuen J. Abdominal aortic aneurysm: An update AJGP 2018;47:252–56

Table 1.2 AAA surveillance intervals by diameter.

AAA diameter (cm)	Surveillance interval (months)
3.0–3.9	24
4.0–4.5	12
4.6–5.0	6
>5.0	3

Chuen J. Abdominal aortic aneurysm: An update AJGP 2018;47:252–56.
https://www1.racgp.org.au/ajgp/2018/may/aaa-an-update/

2. Answer: B

The ECG shows Atrial Fibrillation (AF) with a rapid ventricular response. A fourth heart sound (S4) is a late diastolic sound due to a high pressure atrial wave reflected back from a poorly compliant ventricle. It does not occur in patients with AF because S4 depends on effective atrial contraction. In patients with an atrial septal defect, there is fixed splitting of the second heart sound because there is equalisation of volume loads between the two atria occurring through the defect. A loud S1 can occur in AF due to reduced diastolic filling time so the mitral valve remains widely open at the end of diastole. S3 is a mid-diastolic sound. It can

occur in children and young people due to rapid diastolic filling. A pathologic S3 is present in patient with congestive heart failure (CCF) and other heart disorders due to reduced ventricular compliance and rapid diastolic filling.

Voin V, Oskouian R, Loukas M, Tubbs R. Auscultation of the heart. Clinical Anatomy. 2016;30(1):58–60. https://www.ncbi.nlm.nih.gov/pubmed/27576554

3. Answer: B

Anthracyclines such as doxorubicin and idarubicin exhibit an anti-cancer action through inhibition of topoisomerase (Top) 2α in cancer cells, and their toxicity is largely through inhibition of Top 2β in cardiac myocytes. Topoisomerases are highly expressed in proliferating eukaryotic cells and are involved in detangling and relegating aberrant DNA coils. By binding Top 2α in cancer cells' DNA, anthracyclines facilitate the cleavage function of topoisomerase, but not the religation, which leads to accumulation of double stranded DNA breaks and initiates programmed cell death pathways. Anthracyclines also cause mitochondrial dysfunction and excessive intracellular reactive oxygen species, which results in increased apoptosis. This process is promoted by high intracellular iron availability. Both of these pathways are thought to contribute to the death of cardiac myocytes, and result in cardiomyopathy.

Patient factors predictive of higher risk for anthracycline cardiomyopathy include lifetime cumulative dose, age less than 18 or over 65, female gender, renal failure, radiotherapy involving the heart, pre-existing cardiac disease, carbonyl reductase gene polymorphisms, and carrier status for haemochromatosis genes. Exposure to anthracycline chemotherapy also increases risk of cardiotoxicity with trastuzumab. Trials evaluating the pharmaceutical prevention of anthracycline related cardiomyopathy have found small but statistically significant differences in deterioration of left ventricular function for ACE inhibitors, beta-blockers, aldosterone antagonists and angiotensin 2 receptor blockers in predominantly low-exposure and low-risk patients. These results may or may not be clinically significant.

Trials that target those at highest risk of cardiomyopathy are most likely to yield more meaningful clinical results. For example, in one trial patients receiving high dose chemotherapy were randomised to receive placebo or ramipril should they have a troponin rise after their first cycle of chemotherapy. The results of the study found that the treatment arm had a roughly stable left ventricular ejection fraction over the trial period, but the placebo arm decreased by around 14%, which could translate to meaningful patient-centred outcomes. Dexrazoxane competitively binds Top 2β, preventing anthracycline binding, and very effectively prevents anthracycline cardiomyopathy. Previous concerns regarding increased risk of secondary cancers with dexrazoxane are being re-evaluated as the result of multiple influential trials, and it is now available for use in a number of settings.

Henriksen P. Anthracycline cardiotoxicity: an update on mechanisms, monitoring and prevention. Heart. 2017;104(12):971–977. https://heart.bmj.com/content/104/12/971.long

4. Answer: B

Most patients with symptomatic severe aortic stenosis (AS) have a valve area <1.0 cm^2 and/or a mean transvalvular pressure gradient >40 mmHg. Low gradient AS is defined as severe AS (valve area <1.0 cm^2) with a transvalvular pressure gradient <30 mmHg. Low gradient AS is seen in patients with left ventricular (LV) systolic dysfunction with reduced left ventricular ejection fraction (LVEF). A challenging clinical task in those with low gradient AS is differentiating who will survive and improve after aortic valve replacement surgery and those who will not. There is a subset of patients with AS and low transvalvular gradients who do not benefit from aortic valve replacement and are at considerable risk of operative death. These patients have a significant degree of LV dysfunction but only mild to moderate AS. They are considered to have pseudo-stenosis because their symptoms are primarily due to poor LV function, not significant valvular disease.

Dobutamine Stress Echocardiography (DSE) is useful to differentiate true aortic stenosis from pseudo-aortic stenosis. Patients with severe AS and secondary LV dysfunction that leads to a low transvalvular pressure gradient are considered to have true stenosis. In this setting, the severe stenotic lesion results in excessive afterload and a reduced LVEF, thereby producing both a markedly decreased stroke volume and low transvalvular pressure gradient. It is logical to assume that aortic valve replacement will be beneficial in these patients. In pseudostenosis, patients have a low transvalvular pressure gradient because of the combination of moderate AS and low cardiac output. The low output reduces the valve opening forces, resulting in limited mobility of a valve that is not severely diseased. The calculated valve area may mistakenly suggest severe stenosis because of valve area equation limitations when applied to low flow rate conditions. In contrast to true stenosis, surgical correction in these patients is unlikely to be beneficial.

There are no other clinical or haemodynamic variables that are helpful to stratify risk in such patients and determine the appropriate therapy. The aortic valve calcium score (determined by CT) has been associated with AS haemodynamic severity, progression rate, and clinical outcomes but not validated in patients with low ejection fraction. Patients with contractile reserve have a much better outcome after surgery.

Annabi M, Touboul E, Dahou A, et al. Dobutamine Stress Echocardiography for Management of Low-Flow, Low-Gradient Aortic Stenosis. Journal of the American College of Cardiology. 2018;71(5):475–485. https://www.sciencedirect.com/science/article/pii/S0735109717417785?via%3Dihub

5. Answer: D

There is an increased prevalence of AS in an ageing population. Severe AS is defined on transthoracic echocardiogram as a reduced aortic valve area (AVA) of < 1.0 cm^2, peak velocity of > 4 m/s and a mean gradient of greater than 40 mmHg across the valve. Patients with less severe aortic stenosis may remain asymptomatic for years. Once the symptoms of syncope, angina, or heart failure develop, average survival reduces rapidly, with an increased risk of sudden cardiac death. Until transcatheter aortic valve implantation (TAVI) became available, the options to treat severe AS include medical (palliative management), surgical aortic valve replacement (SAVR), or a Ross procedure (where a diseased aortic valve is replaced by a patient's own pulmonary valve). SAVR involves a midline sternotomy, general anaesthetic, cardiopulmonary bypass, typically requires 24 to 48 hours stay in an intensive care unit (ICU) and 10 to 14 days as an inpatient for post-op recovery and mobilisation, and an even longer hospital stay for rehabilitation in the frail and elderly. Patients who are greater than 75 years old have an increased morbidity and mortality associated with SAVR.

Since the advent of TAVI in 2002, elderly patients who were not suitable candidates for SAVR, can now be considered for TAVI if appropriate, provided they do not have severe COPD, debilitating stroke, active malignancy, and dementia indicating a survival of < 1 year.

The majority of TAVI procedures are performed with local anaesthetic and conscious sedation. In uncomplicated TAVI cases, patients can be mobilised 4 hours post-procedure and do not require ICU admission and likely to be discharged within 48 hours after telemetry monitoring to rule out conduction disturbance. Patients usually require dual-antiplatelets (aspirin and clopidogrel) for at least 3–6 months, with aspirin continued lifelong.

Adams H, Ashokkumar S, Newcomb A, et al. Contemporary review of severe aortic stenosis. Internal Medicine Journal. 2019;49(3):297–305. https://www.ncbi.nlm.nih.gov/pubmed/30091235

6. Answer: D

A randomised non-inferiority trial was terminated early due to safety concerns around the combination of rivaroxaban and antiplatelets in patients with coronary artery disease (CAD) and intervention (percutaneous coronary intervention or bypass grafting) more than one-year prior – or medically treated CAD. The primary efficacy measure of cardiovascular events or death from any cause was significantly lower in patients taking rivaroxaban alone compared to rivaroxaban plus aspirin. Bleeding was significantly lower in the monotherapy group, and the secondary outcome measure of death from any cause was significantly lower in the monotherapy group.

Yasuda S, Kaikita K, Akao M, Ako J, Matoba T, Nakamura M et al. Antithrombotic Therapy for Atrial Fibrillation with Stable Coronary Disease. New England Journal of Medicine. 2019;381(12):1103–1113. https://www.nejm.org/doi/full/10.1056/NEJMoa1904143

7. Answer: D

The anti-anginal action of β-blockers is predominantly through reduced heart rate, which results in a relative decrease in myocardial oxygen demand. This leads to decrease in angina symptoms. β-blockade also results in decreased heart contractility, decreased atrioventricular conduction and nodal refractiveness, and competitive catecholamine inhibition to prevent cardiac remodelling.

Angina results from myocardial ischemia, due to a mismatch between myocardial oxygen demand and supply. Heart rate is the major determinant of oxygen consumption, and increased heart rate precedes most episodes of angina. Other determinants of

myocardial oxygen demand include BP, myocardial wall tension, cardiac hypertrophy, and myocardial contractility. Coronary blood flow is the major determinant of myocardial oxygen supply, which is dependent on the pressure gradient across the coronary circuit, integrity of the coronary arteries, and oxygen carrying capacity of the blood. Typically, angina results from exercise or emotional stress precipitating further reduced coronary blood flow in patients with obstructive coronary artery disease (CAD). In a minority of patients, angina is secondary to functional alterations of coronary vessels, where coronary arteries can be angiographically normal, so called non-obstructive CAD.

Treatment for stable angina includes lifestyle changes such as weight loss, exercise, smoking cessation, aspirin, moderate to high intensity statin therapy, and antianginal therapy as listed below. Heart rate should be targeted <70/min and BP <120/85 mmHg, with β-blockers, calcium channel blockers, and/or nitrates, before considering newer agents.

1. Medications which reduce heart rate (reduce myocardial demand): β-blockers, ivabradine, non-dihydropyridine calcium antagonists.
2. Medications which induce coronary and vascular artery relaxation (increase myocardial supply): Dihydropyridine calcium channel blockers, nitrates, and nicorandil.
3. Medications which induce cellular tolerance to ischemia: Piperazine derivatives including trimetazidine, ranolazine.

Ohman E. Chronic Stable Angina. New England Journal of Medicine. 2016;374(12):1167–1176. https://www.nejm.org/doi/full/10.1056/NEJMcp1502240

8. Answer: C

Brugada syndrome (BS) is a rare inherited disease with ECG findings of coved type ST-segment elevation (≥ 2mm) followed by a negative T wave in (≥ 1) the right precordial leads in V1 to V3 in patients with a structurally normal heart in patients with Type I BS.

ECG of Type II BS: The ST segments also have a high take-off but the J amplitude of ≥ 2mV gives rise to a gradually descending ST elevation remaining ≥ 1mV above the baseline followed by a positive or biphasic T wave that results in a saddle back configuration. Type III BS ECG: Right precordial ST elevation of saddle-back type or coved type.

BS is genetically transmitted as an autosomal dominant syndrome with incomplete penetrance and has a male predominance. Mutations of several genes have been reported to be linked to BS, with mutations of SCN5A gene the most common.

Presenting clinical symptoms include syncope, seizures, agonal breathing at night due to polymorphic ventricular tachycardia (PVTs), ventricular fibrillation and sudden cardiac death (SCD). Lethal arrhythmias are mainly observed during the night or at rest during the day suggesting a likely association with bradycardia or vagal events. Clinical manifestations are also noted in patients with fever, which requires prompt management with antipyretics to minimise the risks of arrhythmias.

The mainstay of management currently is to prevent SCD associated with arrhythmias with an implantable cardioverter defibrillator (ICD). More recently, radiofrequency epicardial catheter ablation has been suggested as to be a new therapeutic option for patients with BS.

Larkin D. Brugada Syndrome • LITFL • ECG Library Diagnosis [Internet]. Life in the Fast Lane • LITFL • Medical Blog. 2019 [cited 8 September 2019]. Available from: https://litfl.com/brugada-syndrome-ecg-library/

Brugada J, Campuzano O, Arbelo E, Sarquella-Brugada G, Brugada R.et al. Present Status of Brugada Syndrome. Journal of the American College of Cardiology [Internet]. 2018 [cited 16 November 2020];72(9):1046–1059. Available from: https://www.sciencedirect.com/science/article/pii/S0735109718353622?via%3Dihub

9. Answer: A

This presentation is highly suspicious for cardiac amyloidosis (CA). His echocardiogram suggests cardiac involvement especially left ventricular hypertrophy in the context of hypotension and low voltage ECG, which could be further investigated with cardiac MRI.

Amyloidosis is a collection of diseases in which a protein-based infiltrate deposits in tissues as beta-pleated sheets. In >95% of CA, the main protein being deposited is light chain [termed light chain amyloidosis (AL)] and less commonly transthyretin [termed transthyretin amyloidosis (ATTR)].

AL is a plasma cell dyscrasia, where misfolded antibody light chain fragments can deposit systemically in any organ. The typical presentation is heart failure with preserved ejection fraction, with pathognomonic but infrequent presentations with macroglossia and/or periorbital purpura, and other symptoms dependent on organ involvement, commonly kidneys (proteinuria, especially

nephrotic range) gastrointestinal tract (diarrhoea, weight loss), and nervous system (peripheral neuropathy, carpal tunnel syndrome, orthostatic hypotension). AL presents in adults, with a median age of diagnosis of 63, and median untreated survival of 6 months if the initial presentation is heart failure. It is rare for patients to have clinical involvement of all of these organ systems. The diagnosis is often delayed. Physician should have a heightened clinical suspicion for amyloidosis in appropriate clinical context and should consider it until proven otherwise.

ATTR is due to monomerization and misfolding of the protein transthyretin, which is produced by the liver and functions as a transporter of thyroxine and retinol. There are two main subtypes of ATTR amyloidosis: (i) acquired wild type ATTR (ATTRwt, previously senile CA) which typically presents as heart failure or hypertrophic restrictive cardiomyopathy in a male >60, often preceded by carpal tunnel syndrome and/or spinal stenosis, and (ii) hereditary mutant variant (ATTRm) which can be due to >100 point mutations, and subsequently presents heterogeneously as polyneuropathy, cardiomyopathy, or mixed. ATTR prognosis is better than AL, with median survival of 4 years for ATTRwt and variable for ATTRm.

Echocardiogram is the first step in the CA diagnostic workup to identify patients likely to have the disease and prompting further workup. Typical features of CA on echocardiogram include:

- Left ventricular hypertrophy in the absence of secondary causes
- Mismatch between ECG finding of low voltage and echocardiogram finding of left ventricular hypertrophy
- Granular sparking appearance of left ventricular wall
- Apical sparing
- Diastolic dysfunction
- Biatrial dilatation and reduced left ventricular cavity dimensions

ECG hallmarks are low voltages. Further investigations include:

- sFLC ratio and serum/urine immunofixation (to investigate for AL).
- Cardiac imaging (non-invasive diagnosis)
  - Longitudinal strain imaging using 2D speckle tracking for 'apical sparing' pattern
  - Cardiac MRI for diffuse and subendocardial 'late gadolinium enhancement' pattern that does not respect coronary distributions (93% sensitive, 70% specific)
  - 99mTcPYP scintigraphy for grade 2 or 3 myocardial radiotracer uptake (100% specificity and positive predictive value, in absence of monoclonal gammopathy)
- Endomyocardial biopsy (diagnostic gold standard, 100% sensitive for CA).

Definitive diagnosis of amyloidosis requires biopsy (Table 1.3).

Table 1.3 Sensitivity of different organ biopsy in diagnosis of amyloidosis.

Organ	Sensitivity
Abdominal fat pad	70%
Bone marrow	50% to 60%
Clinically involved organ	99% to 100%
Rectum	70% to 85%

Donnelly J, Hanna M. Cardiac amyloidosis: An update on diagnosis and treatment. Cleveland Clinic Journal of Medicine. 2017;84(12 suppl 3):12–26.
https://www.ccjm.org/content/84/12_suppl_3/12.long

10. Answer: A

Cardiac output is defined by the amount of blood ejected by each ventricle in one minute. Cardiac output (litres/min) is the product of the stroke volume (litres/beat) and the heart rate (beats/min). There are four determinants of cardiac output, including heart rate, preload, afterload, and contractility.

Heart rate is affected by chronotropic factors. Sympathetic stimulation by noradrenaline, adrenaline, and medications such as atropine can increase heart rate. Activation of the parasympathetic system and chemicals, such as acetylcholine and adenosine, slows down the heart rate.

Preload is the amount of blood entering the ventricle during diastole, also known as the end-diastolic volume. It is influenced by blood volume, venous return, and atrial contraction.

Afterload occurs during systole when ventricle contracts and ejects blood out of the aorta and into the pulmonary trunk. Atherosclerosis, peripheral vasoconstriction, and hypertension increase afterload due to increased resistance.

Inotropic factors affect the contractility of the heart. Positive inotropes such as sympathetic stimulation by noradrenaline and dobutamine increases cardiac output. The parasympathetic system, acetylcholine, beta blockers, acidosis, and calcium channel blockers can have negative inotropic effect on the heart and reduce cardiac contractility.

Atropine increases heart rate, causing a higher cardiac output. Adenosine slows down heart rate hence reduces cardiac output. Hypertension increases the force against which the ventricles must pump in order to eject blood, hence reducing cardiac output. Beta blockers have negative inotrope effect and also slow down the heart rate, resulting in decreased cardiac output.

Acidosis reduces left ventricle contractility. However, this is compensated for by an increased heart rate and a reduced systemic vascular resistance, resulting in an increased cardiac output.

Vincent J. Understanding cardiac output. Critical Care. 2008;12(4):174.
https://www.ncbi.nlm.nih.gov/pmc/articles/PMC2575587/

11. Answer D

Episodic loss of consciousness is most commonly caused by cardiac syncope, reflex syncope, orthostatic syncope, and less commonly seizure. Clinical features present on history, examination and basic investigation findings can be useful in determining the likelihood of various causes, although no individual feature is diagnostic for a particular cause.

Patient features that increase the likelihood of cardiac syncope are increasing patient age, history of atrial fibrillation or flutter, history of cardiac failure, and history of severe structural heart disease. Some precipitating factors can decrease the likelihood of cardiac syncope, such as syncope in a warm place, during a medical procedure, or after using the toilet. Dyspnoea or chest pain prior to syncope are associated with higher likelihood of cardiac syncope, whereas cold sensitivity, headache, mood change, and abdominal discomfort decrease the likelihood of cardiac syncope. Witnessed cyanosis is associated with higher likelihood of cardiac syncope, whereas inability to remember details prior to the episode and mood changes after the episode are associated with lower likelihood of cardiac syncope. A combination of normal ECG and no history of cardiac disease is associated with a lower likelihood of cardiac syncope. Contrary to popular teaching, the presence or absence of injury does not predict for or against cardiac syncope.

Two clinical scores, Evaluation of Guidelines in SYncope Study (EGSYS) and vasovagal score), assign scores on the basis of a combination of clinical factors and have some predictive value over and above individual patient characteristics. EGSYS uses six patient variables and a score less than three has a likelihood ratio of cardiac syncope of 0.12–0.17. The vasovagal score has only been prospectively evaluated in one study but a cut-off score of less than –2 has likelihood ratio of over 8 for cardiac syncope and equal to or more than –2 has a likelihood ratio of 0.10, making this score a potentially good discriminator, if results can be duplicated.

Biomarkers are not yet utilised in widespread practice for the evaluation of syncope, but there is some evidence that high-sensitivity troponin and N-terminal pro-B-type natriuretic peptide may become useful in ruling out cardiac syncope.

Several clinical features have high specificity, but low sensitivity for the diagnosis of seizure. These include head turning during the event, unusual posturing during the event, urinary incontinence, tongue trauma, and the patient having no recall of witnessed unusual behaviours.

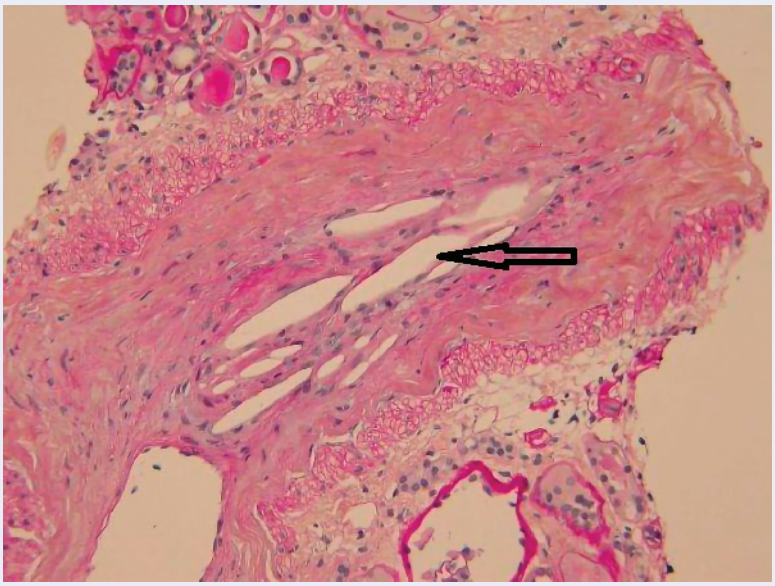
Albassam O, Redelmeier R, Shadowitz S, et al. Did This Patient Have Cardiac Syncope? JAMA. 2019;321(24):2448.
https://jamanetwork.com/journals/jama/article-abstract/2736568

12. Answer: C

This patient's clinical features, unexplained fever, myalgia, sudden appearance of several small, cool, cyanotic and painful areas of the toe, and AKI are consistent with cholesterol embolisation. Cholesterol embolism is caused by showers of cholesterol crystals from an atherosclerotic plaque that occludes small arteries. Embolisation can occur spontaneously or as an iatrogenic complication from an invasive vascular procedure (angiography or vascular surgery) and after anticoagulant therapy as a result of interference with the protective clot over ulcerative atheromatous plaques and thrombolytic therapy may lyse thrombi, including those covering atherosclerotic plaques.

Once in the circulation, cholesterol crystal emboli lodge in small arteries (150–200 um in diameter); which then cause an inflammatory reaction, intimal proliferation, and intravascular fibrosis leading to the narrowing or obliteration of the lumen and ischaemic changes. Peak incidence is usually two to four weeks after a procedure.

The true incidence of cholesterol embolisation is difficult to estimate. Retrospective autopsy (incidence 10–27%) or biopsy (incidence 1%) studies may include subclinical cases. The hallmark of the condition is the presence of needle-shaped empty spaces in histological sections as the lipids are dissolved by the techniques used to prepare the tissue for histological examination (See below).



Patients can be asymptomatic, or they can present with a distinct clinical syndrome, ranging from a cyanotic toe to a multiorgan systemic disease that can mimic other systemic diseases such as vasculitis. The distribution of end-organ damage depends on the anatomical location of the original atherosclerotic plaques and the extent of organ involvement. Laboratory investigations are non-specific. Eosinophilia appears to be the most common finding up to 80% of cases.

The presence of a triad of a precipitating event, AKI, and peripheral embolisation strongly suggests the diagnosis. The presence of other complications of atheroembolism, such as gastrointestinal bleeding and neurological involvement, should raise the suspicion level. To confirm diagnosis, a biopsy of the target organs is needed.

There is no effective treatment apart from symptomatic and supportive measures, including dialysis. Anticoagulants should be avoided if possible because they can potentiate the problem. Disagreement exists concerning steroid treatment. Recently, statins have been found to be associated with better renal outcome likely due to statin-induced plaque stabilisation and regression. Patients with cholesterol embolisation have a poor prognosis with one-year mortality rate ranging from 64–87%.

Kronzon I, Saric M. Cholesterol Embolization Syndrome. Circulation. 2010;122(6):631–641.
https://www.ncbi.nlm.nih.gov/pubmed/21993354

Scolari F, Ravani P. Atheroembolic renal disease. The Lancet. 2010;375(9726):1650–1660.
https://www.ncbi.nlm.nih.gov/pubmed/20381857

13. Answer: D

Computed tomography coronary angiography (CTCA) is an imaging test that has been shown in meta-analyses to have excellent sensitivity (98%) and good specificity (88%) for significant coronary artery disease (CAD) with stenosis >50%. Its high negative predictive values (96–100%) suggest CTCA is an excellent test for ruling out significant disease in patients with low-to-intermediate pretest probability of CAD. Current data does not support the use of CTCA in asymptomatic patients.

CTCA is not recommended in patients with previous coronary stents as the stents may produce artefact and make the results uninterpretable. Stent diameter <3 mm is thought to be unevaluable. However, very selective cases can be evaluated using CTCA, including large stents and simple left main stents. CTCA is not appropriate for patients with STEMI given the need for invasive coronary angiogram without any delay.

To avoid artefacts which may hamper interpretation of the results, the patient should be in sinus rhythm with a heart rate <65 beats/min, able to hold their breath for 10 seconds, able to tolerate beta blockers and nitrates (nitrates are given to dilate the coronary arteries by most centres), and able to hold their arms above the head during the scan. Previous contrast allergy must

be ruled out prior to CTCA. If the patient has significant renal impairment, CTCA may not be the best investigation for CAD due to the risk of contrast nephropathy.

Liew G, Feneley M, Worthley S. Appropriate indications for computed tomography coronary angiography. Medical Journal of Australia. 2012;196(4):246–249.
https://www.mja.com.au/journal/2012/196/4/appropriate-indications-computed-tomography-coronary-angiography

14. Answer: B

This patient has evidence of native mitralvalve infective endocarditis (IE) complicated by cerebral emboli. IE is the infection of the endocardial surface of the heart and may involve one or more heart valves, the mural endocardium, or a septal defect. The highest rates are observed among patients with prosthetic valves, intracardiac devices, unrepaired cyanotic congenital heart diseases, or a history of IE. However, more than 50% of cases are not associated with underlying valvular disease. Other risk factors include chronic rheumatic heart disease, age related degenerative valvular lesions, haemodialysis, and co-existing conditions such as diabetes, and intravenous drug use.

Streptococci and staphylococci account for 80% of cases of IE, with proportions varying according to valve (native vs prosthetic), source of infection, patient age, and coexisting conditions. Staphylococci are now the most common cause of IE and approximately 35%–60.5% of staphylococcal bacteraemia are complicated by IE.

Cases of IE in which a blood culture is negative (10% of cases) may be due to patients exposed to antibiotic agents before the diagnosis of IE or IE caused by fastidious microorganisms.

Cerebral complications are the most frequent and most severe extracardiac complications. Vegetations that are large, mobile, or in the mitral position and IE due to Staphylococcus aureus are associated with an increased risk of symptomatic embolism.

IE remains a diagnostic and therapeutic challenge. Identifying the causative microorganism is central to diagnosis and appropriate treatment; two or three blood cultures should routinely be drawn before antibiotic therapy is initiated. When IE is suspected, echocardiogram should be performed as soon as possible. However, the diagnosis of IE can never be excluded on the basis of negative echocardiogram findings, either from transthoracic echocardiogram or transesophageal echocardiogram.

Appropriate antibiotic treatment of IE is guided by Australian Therapeutic Antibiotic Guidelines. Approximately 15%–25% of patients with IE eventually require surgery. Indications for surgical intervention in patients with native valve IE are as follows:

- Congestive heart failure that is refractory to standard medical therapy
- Fungal infective endocarditis
- Persistent sepsis after 72 hours of appropriate antibiotic treatment
- Recurrent septic emboli, especially after 2 weeks of antibiotic treatment
- Rupture of an aneurysm of the sinus of Valsalva
- Conduction disturbances caused by a septal abscess
- Kissing infection of the anterior mitral leaflet in patients with IE of the aortic valve.

Hoen B, Duval X. Infective Endocarditis. New England Journal of Medicine. 2013;368(15):1425–1433.
https://www.ncbi.nlm.nih.gov/pubmed/23574121

15. Answer: C

According to the ACC/AHA guideline, a biventricular pacemaker and defibrillator should be offered to patients with NYHA class III or IV heart failure, an ejection fraction <35% and a QRS complex >0.12 second. Approximately 70% of patients' symptoms improve due to resynchronisation of the timing of the left and right ventricular contraction. Device treatment has been shown to improve mortality, ejection fraction, quality of life, and functional status, as well as reduce readmission.

A dual-chamber pacemaker does not provide symptomatic relief or protect patients from ventricular arrhythmias and sudden cardiac death. Recent clinical trials have demonstrated SGLT2 inhibitors reduces mortality and hospitalisation in patients with type 2 diabetes and established cardiovascular disease. However, it is currently contraindicated in patients with eGFR <30 ml/min/1.73 m^2. Adding a beta blocker in an elderly patient with severe airway disease and insulin dependent diabetes may need careful consideration and is not the most effective treatment for this patient.

Normand C, Linde C, Singh J, Dickstein K.Ref: Indications for Cardiac Resynchronization Therapy. A Comparison of the Major International Guidelines JACC: Heart Failure. 2018;6(Issue 4):308-316., April 2018
https://www.sciencedirect.com/science/article/pii/S2213177918301203

16. Answer: C

SGLT2 inhibitor (SGLT2i) works by inhibiting SGLT2 in the proximal convoluted tubule, to prevent reabsorption of glucose and facilitate its excretion in urine. As glucose is excreted in urine, its plasma levels fall leading to an improvement in glycaemic parameters. This mechanism of action of SGLT2i is dependent on blood glucose levels and has minimal potential for hypoglycaemia. SGLT2i's mode of action depends upon normal renal glomerular–tubular function and their efficacy is reduced in persons with renal impairment. SGLT2i are not prescribed on their own but can be used in combination with other diabetes medications. SGLT2i may also cause modest weight loss and reduce systolic blood pressure, which are beneficial for patients with heart failure.

Thiazolidinediones may cause water retention, weight gain, or worsen heart failure and are contraindicated in patients with New York Heart Association (NYHA) class II–IV heart failure. They should be used cautiously in patients with NYHA class I heart failure.

Atherton J, Sindone A, De Pasquale C, et al. National Heart Foundation of Australia and Cardiac Society of Australia and New Zealand: Australian clinical guidelines for the management of heart failure 2018. Medical Journal of Australia. 2018;209(8):363–369.
https://onlinelibrary.wiley.com/doi/abs/10.5694/mja18.00647?sid=nlm%3Apubmed

17. Answer: B

This patient's clinical presentation, risk factors, and echocardiogram findings are consistent with heart failure with preserved ejection fraction (HFpEF). No therapy has been proven to reduce mortality in patients with HFpEF.

HFpEF predominantly affects elderly (>65 years) hypertensive women. Other risk factors include obesity, coronary artery disease, diabetes, atrial fibrillation, and hyperlipidaemia. It is established that the prevalence of HFpEF among patients with heart failure averages 47% and its prevalence in the community is estimated to be 1.1% to 5.5% of the general population. The prevalence of HFpEF has increased over the last two decades, due to ageing population, increasing prevalence of risk factors, such as hypertension, diabetes, and increased survival.

The diagnosis of HFpEF can be challenging because the symptoms and signs are non-specific, hence it is a strictly clinical diagnosis. For patients presenting with heart failure and relatively normal LV ejection fraction (LVEF), valvular heart disease, infiltrative cardiomyopathies, pericardial disease, high-output heart failure, chronic pulmonary disease, and pulmonary arterial hypertension should be excluded. An elevated BNP or NT-proBNP on its own is insufficient for the diagnosis or exclusion of HFpEF.

Echocardiogram is the imaging modality of choice to establish the diagnosis of HFpEF by criteria; exclude valvular, right-sided, or pericardial disease; and assess for other differential diagnoses. The following are the echocardiographic criteria recommended by the European Society of Cardiology for the diagnosis of HFpEF:

- LVEF ≥50%
- LV end-diastolic volume index (LVEDI) <97 mL/m^2
- Raised LV filling pressure is indicated by a ratio of mitral early diastolic inflow velocity to mitral early annular lengthening velocity (E/e')>15.

Cardiac catheterisation is the gold standard for the diagnosis of HFpEF. Criteria for raised LV filling pressure include LV end-diastolic pressure >16 mmHg or a mean pulmonary capillary wedge pressure >12 mmHg.

In contrast to heart failure with reduced ejection fraction, there is limited clinical trial evidence guiding the treatment of HFpEF. At present, no therapy including ACE inhibitors, angiotensin receptor blockers, β-blockers and aldosterone antagonists has demonstrated mortality benefit in patients with HFpEF.

Managing comorbidities including hypertension, obesity, OSA, rate control for AF, and diabetes, are the mainstay of HFpEF treatment strategies. Diuretics can be used to reduce congestion and improve symptoms. Low-dose spironolactone is recommended to reduce hospital admission on the basis of the TOPCAT trial results. Sildenafil is an inhibitor of phosphodiesterase-5 that increases cGMP levels by blocking catabolism. Increased availability of cGMP could provide benefits for both vascular and myocardial remodelling, including attenuating hypertrophy, fibrosis, and impaired cardiac relaxation. In the RELAX trial, sildenafil did not improve 6 min walk distance or quality of life. Ivabradine is a selective sinus node If sodium channel inhibitor that reduces HR without affecting contractility. It can increase peak VO_2 and reduced exercise mitral early diastolic velocity/mitral annular velocity (E/e') ratio.

Harper A, Patel H, Lyon A.AR. Heart failure with preserved ejection fraction. Clinical Medicine.Clin Med (Lond). 2018;18(Suppl 2): s24–s29.
https://pubmed.ncbi.nlm.nih.gov/29700089/

18. Answer: D

Hypertrophic cardiomyopathy (HOCM) is a common monogenic cardiovascular disorder with a prevalence of 1 in 500. It is diagnosed with echocardiogram and MRI which shows a hypertrophic left ventricule without dilatation and it is not associated with another cardiac, systemic, metabolic, or syndromic disease. HOCM is diverse in clinical, phenotypic expression, and natural history. It is often underdiagnosed.

HOCM is inherited in an autosomal dominant pattern. It is associated with mutations (nucleotide sequence variants) in 11 or more genes encoding proteins of thick and thin myofilament contractile components of the cardiac sarcomere (Z-disk), with beta-myosin heavy chain and myosin-binding protein C genes most commonly involved.

HOCM is predominantly an obstructive disease, with 70% of patients having mechanical impedance to left ventricular outflow (gradients ≥30 mm Hg) at rest or with exertion. Left ventricular outflow obstruction is usually produced by mitral-valve systolic anterior motion and septal contact due to flow drag, causing complications such as mitral regurgitation, diastolic dysfunction, atrial fibrillation, congestive heart failure, and ventricular tachyarrhythmias.

HOCM is associated with increased sudden cardiac death associated with ventricular tachyarrhythmias, disorganised myocardial architecture, interstitial collagen deposition, and replacement scarring after myocyte death as a consequence of coronary microvascular mediated flow dysfunction and ischemia. Patients with a diagnosis of HOCM are often disqualified from participating in intensive competitive sports due to high risks of sudden cardiac death.

The American College of Cardiology and the American Heart Association (ACC–AHA) have developed an algorithm for risk stratification in patients with HOCM who would benefit from primary prevention of sudden cardiac death with implantable cardioverter–defibrillators (ICDs). ICDs should be considered in young and middle-aged patients whose clinical profiles include one or more major risk factors, include family history of HOCM-related sudden death, unexplained syncope, multiple, repetitive non-sustained VT, massive LVH (≥30 mm), left ventricle apical aneurysm, and extensive late gadolinium enhancement. If the level of risk remains uncertain, other clinical features such as left ventricular outflow obstruction, hypotensive response to exercise, can serve as mediating factors.

Management strategies for variable clinical manifestation of HOCM include ICDs to reduce the risk of sudden cardiac death, surgical myectomy (with alcohol septal ablation as a selective alternative) for permanent reversal of heart failure in patients with outflow obstruction, heart transplantation for patients with non-obstructive end-stage disease, and anticoagulant therapy to prevent embolic stroke caused by atrial fibrillation.

Maron B. Clinical Course and Management of Hypertrophic Cardiomyopathy. New England Journal of Medicine. 2018;379(7):655–668. https://www.ncbi.nlm.nih.gov/pubmed/30110588

19. Answer: B

This patient has papilloedema with almost complete obliteration of the margins of the optic disc and several hypertensive haemorrhages. Changes in hypertensive retinopathy include: Focal arteriolar narrowing and arterial venous nicking, flame or blot haemorrhages, microaneurysm, hard exudates, cotton wool spots, and papilloedema.

The Keith-Wagner grade is used to classify the severity of hypertensive retinopathy:

Grade 1: Isolated narrowing of the arterioles.

Grade 2: There is moderate to marked narrowing of retinal arterioles associated with a copper wire appearance or arteriovenous nicking.

Grade 3: Hypertensive retinal haemorrhage or exudates are present.

Grade 4: Papilloedema is present.

Modi P, Arsiwalla T. Hypertensive Retinopathy [Internet]. Statpearls.com. 2020 [cited 16 November 2020]. Available from: https://www.statpearls.com/articlelibrary/viewarticle/35600/

20. Answer: C

All of the above patients have a Class I indication for an ICD implantation except in the patient who just had a myocardial infarction two weeks ago. There is no survival benefit of early ICD implantation in patients with ischaemic cardiomyopathy. Evaluation of patients for ICD implantation should wait at least 40 days post-myocardial infarction and an ICD is then indicated in patients with left ventricular ejection fraction less than or equal to 35% and at least 90 days post revascularisation, with NYHA class II/III symptoms despite guideline directed medical therapy.

The current indications for ICD to prevent sudden cardiac death (SCD) are summarised in Table 1 in the following full text article based on the 2017 American Heart Association/American College of Cardiology/Heart Rhythm Society (AHA/ACC/HRS) guidelines for management of patients with ventricular arrhythmias and the 2015 European Society of Cardiology (ESC) guidelines for management of patients with ventricular arrhythmias.

Chieng D, Paul V, Denman R. Current Device Therapies for Sudden Cardiac Death Prevention – the ICD, Subcutaneous ICD and Wearable ICD. Heart, Lung and Circulation. 2019;28(1):65-75.
https://www.heartlungcirc.org/article/S1443-9506(18)31920-6/fulltext

21. Answer: D

This patient has infrequent palpitations which happen around once every 6 months. Three- or seven-day Holter monitoring may not capture arrythmias due to the infrequent events.

It is important to know the frequency of palpitations or syncopal events to decide what the best way is to investigate potential underlying arrhythmias.

The next preferred step of investigation for this patient is an implantable loop recorder due to the infrequent symptoms and the long duration between events.

It will be important to investigate underlying coronary artery disease in this patient with significant risk factors for cardiovascular disease due to his past medical history, which may contribute to his symptoms.

In patients with infrequent unexplained syncopal episodes, implantable loop recorders have high rates of diagnosing arrythmias compared to external loop recorders or physician follow-up. Implantable loop recorders are pen drive sized device inserted under the skin under local anaesthetic and are capable of recording cardiac events for up to three years. Implantable loop recorders can be considered in patients with cryptogenic stroke to detect paroxysmal or infrequent atrial fibrillation if the initial in-hospital or Holter monitoring are unable to detect atrial fibrillation, which is a risk factor for embolic stroke.

Wearable devices are gaining popularity in the recent years and may aid in detection of arrythmias such as atrial fibrillation and supraventricular tachycardia. However, in patients with syncopal episodes, they are unable to record these events due to potential loss of consciousness with syncope. More studies are required to see how to integrate smart devices to improve patient care.

Khalil C, Haddad F, Al Suwaidi J. Investigating palpitations: the role of Holter monitoring and loop recorders. BMJ. 2017; 358: j3123.
https://www.bmj.com/content/358/bmj.j3123.long

22. Answer: A

Evolocumab and alirocumab are monoclonal antibodies inhibiting Proprotein Convertase Sublitisin/Kexin Type-9 (PCSK9). PCSK9 inhibits low-density lipoproteins (LDL) receptor recycling, which in turn limits the ability for tissue to sequester LDL from the extracellular space. Individuals with activating mutations of the PCSK9 gene have high LDL and poor cardiac outcomes; conversely individuals with inactivating mutations of the PCSK9 gene have low LDL levels and relatively good cardiac outcomes. PCSK9 inhibitor therapy is administered by subcutaneous injection and is indicated where statin/combination therapy has failed to reach LDL targets. PCKS9 inhibitors have shown small but meaningful reductions in cardiovascular risk and mortality and have a greater protective effect for individuals with higher baseline LDL.

The primary action of statins (eg. rosuvastatin) is to decrease hepatocyte production of cholesterol by inhibition of HMG-CoA (3-hydroxy-3-methylglutaryl-coenzyme A) reductase, thereby increasing LDL receptor synthesis, and increasing LDL clearance. Ezetimibe is an azetidinone cholesterol absorption inhibitor, which acts by blocking NPC1L1 (Niemann-Pick C1-Like 1) a key brush-border transport protein. Mipomersen is an example of a newer advance in lipid-lowering therapy, which inhibits synthesis of apoB-100 and LDL, and can be considered for use in homozygous familial hypercholesterolaemia but hepatotoxicity is a significant potential adverse effect.

Burnett J, Hooper A. PCSK9 — A Journey to Cardiovascular Outcomes. New England Journal of Medicine. 2018;379(22):2161–2162.
https://www.nejm.org/doi/full/10.1056/NEJMe1813758

23. Answer: B

Long QT syndrome (LQT) is diagnosed by QT prolongation and T-wave abnormalities on an ECG. A QT interval is measured from the onset of the QRS complex to the end of the T wave. The corrected QT interval (QTc, corrected for heart rate) can be calculated as QTc = QT interval + square root of the RR interval. The latest European Society of Cardiology guideline suggests upper limits

of the QT interval of 480 ms on the ECG strip, regardless of gender. Presenting symptoms of LQT include dizziness, syncope, seizures, and in some cases cardiac arrest due to the presence of Torsades-de-Pointes which can degenerate to ventricular fibrillation causing sudden cardiac death. LQT can be congenital or acquired. LQT is associated with multiple risk factors, including a family history of LQT, ion channel abnormalities, structural heart disease, heart failure, advanced age, hypokalaemia, subarachnoid bleed, medications, diabetes, epilepsy, emotional stress, and physical stress, etc.

The aim of the treatment of LQT is to ensure patients are symptom free. The mainstay of management of LQT is beta blockers in both asymptomatic and symptomatic patients with LQT. Medication compliance is important to avoid life-threatening consequences. Unfortunately, cardiac events cannot be completely prevented by taking beta blockers.

Life-style modification such as avoiding strenuous exercises, emotional stress, physical stress, sleep disturbance, and medications that prolong the QT interval, should be implemented as much as possible.

ICD should be considered in symptomatic patients with syncope who are already on beta blockers, high risk patients with a very long QTc interval (> 550 ms), patients with T-wave alternans, or in patients who cannot tolerate beta blocker therapy. If there are reversible causes of LQT, an ICD is not indicated.

Left cardiac sympathetic denervation (LCSD) is a rarely performed but effective procedure for management of LQT in patients who cannot have beta blockers or ICD. LCSD procedure involves high thoracic left sympathectomy and ablation of the lower half of the stellate ganglion along with T2 to T4. It reduces noradrenaline release at the ventricles and increase the threshold for ventricular fibrillation.

Shah S, Park K, Alweis R. Long QT Syndrome: A Comprehensive Review of the Literature and Current Evidence. Current Problems in Cardiology. 2019;44(3):92–106.
https://www.sciencedirect.com/science/article/abs/pii/S0146280618300513?via%3Dihub

24. Answer: D

This patient is likely to have a history of rheumatic fever leading to mixed mitral valve disease. She is symptomatic with pulmonary hypertension which is the indication for intervention. The mortality for untreated symptomatic severe mitral stenosis is 85% over 10 years. Complications for untreated severe mitral stenosis include endocarditis, atrial fibrillation, stroke, pulmonary hypertension, and right heart failure. The clinical features of severe mitral stenosis are:

- Transmitral mean gradient >10 mmHg
- Enlarged left atrium
- Mitral valvular area <1.5cm^2
- Pulmonary hypertension.

In terms of intervention, percutaneous valvuloplasty should be considered if valve anatomy is amenable. Percutaneous valvuloplasty is less invasive and avoids anticoagulation which is an important consideration in an Aboriginal and Torres Strait Islander patient. Percutaneous valvuloplasty should not be performed in patient with moderate mitral regurgitation as the procedure will worsen mitral regurgitation. Recent emphasis on mitral valve conservation may lead to increased use of mitral valve open commissurotomy, wherein the surgeon, under direct vision, may be able to provide relief of obstruction in patients not suitable for balloon mitral valvuloplasty because of poor valve morphology. However, the percentage of patients that would likely have a bad outcome with balloon mitral valvuloplasty yet a good outcome with open commissurotomy is unknown but is probably small. When the valve can be conserved, it avoids the risks inherent to prosthetic valves and also avoids the need for anticoagulation for patients in sinus rhythm.

Putting these together, this patient has symptomatic, severe mitral stenosis with moderate mitral regurgitation and no other comorbidities therefore she should be considered for mitral valve replacement.

Carabello B. Modern Management of Mitral Stenosis. Circulation. 2005;112(3):432–437.
https://www.ahajournals.org/doi/10.1161/CIRCULATIONAHA.104.532498

25. Answer: D

Polyunsaturated fatty acids (PUFAs), such as omega-3 and omega-6 fatty acids, have multiple roles in membrane structure, lipid metabolism, coagulation, blood pressure, and inflammation. It has been suggested that regular supplementation of omega-3 fatty acids is linked to a reduction in cardiovascular disease and other beneficial effects.

According to a recent Cochrane review, increasing PUFA intake may slightly reduce the risk of cardiovascular events but has no effect on all-cause or cardiovascular disease mortality based on an extensive systematic review of RCTs conducted to date. The number need to treat to prevent one cardiovascular event is 63. The mechanism may be via serum triglycerides reduction.

Omega-3 PUFA supplementation in type 2 diabetes lowers triglycerides and VLDL cholesterol, but may raise LDL cholesterol (although results were nonsignificant in subgroups) and has no statistically significant effect on glycaemic control or fasting insulin levels.

Omega-3 fatty acids appear to have limited benefit in blood viscosity reduction in patients with intermittent claudication. However, there is no evidence of consistent improved clinical outcomes including quality of life, walking distance, ankle brachial pressure index, or angiographic findings. Supplementation may also cause adverse effects such as increased total and LDL cholesterol levels.

Direct evidence on the effect of omega-3 PUFA on incident dementia is lacking. The available trials showed no benefit of omega-3 PUFA supplementation on cognitive function in cognitively healthy older people.

Abdelhamid A, Martin N, Bridges C, et al. Polyunsaturated fatty acids for the primary and secondary prevention of cardiovascular disease. Cochrane Database of Systematic Reviews. 2018. https://www.cochranelibrary.com/cdsr/doi/10.1002/14651858.CD012345.pub3/full

26. Answer: D

Treatment resistant hypertension is defined as persistent hypertension despite adherent and optimal therapy with three different classes of antihypertensive medication including a diuretic, or any four agents. Cardiovascular and renal risk for such patients is two- to six fold higher than for patients with controlled hypertension. Medication compliance is an important factor in evaluating this patient. In addition to screening for secondary causes, which should be guided by history, examination, and laboratory findings, guidelines now suggest addition of a mineralocorticoid receptor antagonist (MRA) for this patient group. This is a result of multiple studies showing significant benefit for MRAs in this patient population. It is suggested that this may be due to high levels of undiagnosed hyperaldosteronism, but the benefits of MRA therapy, while predicted by presence of hyperaldosteronism, are not limited to this sub-population, and suggests of resistant hypertension is often indicative of a salt-retaining state.

Carey M, Whelton PK. Prevention, detection, evaluation and management of high blood pressure in adults: Synopsis of the 2017 American College of Cardiology/American Heart Association hypertension guideline. Ann Intern Med. 2018;168(5):351–8. https://www.acc.org/~/media/Non-Clinical/Files-PDFs-Excel-MS-Word-etc/Guidelines/2017/Guidelines_Made_Simple_2017_HBP.pdf

27. Answer: C

Restrictive cardiomyopathy (RCM) is a heterogeneous group of diseases with variable pathogenesis, clinical features, diagnostic criteria, management strategies and prognosis. It is the least common of the cardiomyopathies and is often underdiagnosed.

In patients with RCM, a restrictive ventricular filling pattern is noted secondary to ventricular stiffness. Biventricular size and normal or near-normal ventricular systolic function are usual in the early stages of the disease. Depending on the ventricle involved, patients may have signs of right or left heart failure, conduction disturbance or arrhythmias. Diagnosis of RCM is based on echocardiogram, cardiac MRI and in some cases endomyocardial biopsy.

RCM is categorised as infiltrative (amyloidosis, sarcoidosis, primary hyperoxaluria), storage disease (Fabry disease, Gaucher disease, hereditary haemochromatosis, glycogen storage disease, mucopolysaccharidosis type I and type II, Niemann-Pick disease), noninfiltrative (idiopathic, diabetic, scleroderma, myofibrillar myopathies, pseuxanthoma elasticum, sarcomeric protein disorders, Werner's syndrome), and endomyocardial (carcinoid heart disease, endomyocardial fibrosis including idiopathic, hypereosinophilic syndrome, chronic eosinophilic leukaemia, drug-induced secondary to serotonin, methysergide, ergotamine, mercurial agents, busulfan, anthracyclines), endocardial fibroelastosis, metastatic cancer, and radiation, etc.

Left ventricular outflow tract obstruction is often seen in patients with subaortic stenosis, bicuspid aortic valve, supravalvular aortic stenosis, coarctation of the aorta, and hypertrophic cardiomyopathy. Option C is often seen in patients with sarcoidosis and RCM. Option D is seen in patients with dilated cardiomyopathy, which is the most common cause of cardiomyopathy.

Treatment should be of the underlying cause(s) of RCM if possible. Diuretics should be used with caution for patients with fluid overload, as some patients with RCM rely on high filling pressure to maintain cardiac output. Excessive diuresis may cause reduced cardiac tissue perfusion. β-blockers and calcium channel blockers could be used to manage arrhythmias or increase filling time. However, careful monitoring of the response is required as some patients may be intolerant. Angiotensin-converting enzyme (ACE) inhibitors and angiotensin receptor blockers should be considered. However, currently there is no evidence that these agents may

be beneficial, and they may not be well tolerated by patients with RCM. Anticoagulation is required in patients with atrial fibrillation, mural thrombus, or evidence of systemic embolisation to prevent strokes. Left ventricular assist device may be beneficial in patients with advanced heart failure as a definitive therapy or as a bridge to cardiac transplant.

Muchtar E, Blauwet L, Gertz M. Restrictive Cardiomyopathy: Genetics, Pathogenesis, Clinical Manifestations, Diagnosis, and Therapy. Circulation Research. 2017;121(7):819–837. https://www.ncbi.nlm.nih.gov/pubmed/28912185

28. Answer: D

Acute rheumatic fever (ARF) and rheumatic heart disease (RHD) remain a significant cause of cardiovascular morbidity and mortality worldwide. In Australia, ARF and RHD disproportionately affect the ATSI population, with up to ×10 higher incidence, ×8 higher ARF hospitalisation rates, and ×20 higher mortality. The highest rates of ARF are in children aged 5–14 years old, and highest rates of RHD in adults aged 35–39, with an all-age RHD incidence up to 2% in ATSI populations in the Northern Territory.

All patients with suspected ARF should be hospitalised to enable appropriate diagnostic investigations including echocardiogram. In high risk groups there should be a lower threshold for diagnosis as listed below and high-risk groups are populations with an incidence of ARF >30/100 000 per year in 5–14 years old or incidence of RHD >2/1000 in all age groups.

ARF diagnosis requires evidence of a preceding group A streptococcus infection, and two major manifestations or one major manifestation and two minor manifestations listed below in low risk populations:

- Major manifestations:
  - Carditis (including subclinical evidence of rheumatic valve disease on echocardiograms)
  - Polyarthritis or aseptic monoarthritis or polyarthralgia
  - Chorea
  - Subcutaneous nodules
  - Erythema marginatum.
- Minor manifestations:
  - Fever
  - Polyarthralgia or aseptic monoarthritis
  - ESR ≥30 mm/hr or CRP ≥30 mg/L
  - Prolonged PR interval on ECG.

Treatment of ARF is benzathine penicillin G every four week, or every three week for high risk patients, for a minimum of 10 years after the last episode of ARF, or until aged 21 if no RHD or 35–40 if moderate–severe RHD.

ARF RHD Guideline [Internet]. Rheumatic Heart Disease Australia. 2019 [cited 19 August 2019]. Available from: https://www.rhdaustralia.org.au/arf-rhd-guideline

29. Answer: D

This patient's presentation and ECG changes are consistent with inferior and right ventricular (RV) myocardial infarction (MI). RV ischaemia complicates 30% to 50% of inferior MIs. Isolated RV myocardial infarction (RVMI) is rare. The coronary artery involved is usually an occluded right coronary artery (RCA). The proximal segment of the RCA supplies the sinoatrial (SA) node and the right atrial wall; the middle segment supplies the lateral and inferior right ventricle (RV); and the posterior portion of the left ventricle, the inferior septum, inferior left ventricular wall and atrioventricular (AV) node are perfused by the distal segment of the RCA. A few patients (10%) may have a right ventricle that is supplied by the circumflex artery.

Although the RVMI often shows good long-term recovery, in the short term RVMI has a worse prognosis to uncomplicated inferior MI, with haemodynamic and electrophysiologic complications increasing in-hospital morbidity and mortality. Acute RV shock has an equally high mortality to left ventricular (LV) shock.

Clinically, the triad of hypotension, elevated jugular venous pressure (JVP), and clear lung fields should raise the possibility of RVMI in patients with acute inferior MI. The classic 12 lead ECG provides information on the LV, but limited information on the electrical activity of the RV. Only lead V1 provides a partial view of the RV free wall. Right precordial leads are obtained by placing the precordial electrodes over the right chest in positions mirroring their usual arrangement. The presence of acute ST segment elevation, Q waves or both in the right precordial leads (V3R to V6R), is highly reliable in the diagnosis of RVMI. ST segment elevation 0.1 mV in the right precordial leads, especially V4R, is observed in 60–90% of patients with acute RVMI. ST elevation from the RV free wall may also be detected by ST elevation in lead III being more than that in lead II or by reciprocal ST depression in leads I and aVL. Echocardiogram should be performed in patient with or suspected RVMI. It may show RV dysfunction.

Additional features of RV involvement include paradoxical septal motion due to increased RV end diastolic pressure, tricuspid regurgitation, and increased right heart pressure.

Cardiac MRI (CMR) can directly evaluate RV size, mass, morphology, and function in an accurate and reproducible manner. CMR is now considered the gold standard for non-invasive assessment of RV function, particularly as it provides additional information on RV anatomy and myocardial mass.

It is important to recognise and diagnose RVMI, as the treatment is different to LVMI and inferior MI. Please see the following principles of the RVMI management.

1. Reperfusion therapy
   - Primary percutaneous coronary intervention preferable to thrombolysis, this should be performed as early as possible to preserve right heart function.
2. Optimise RV preload
   - Avoid morphine, diuretics, β-blockers, nitrates, ACE inhibitor
   - Trial of judicious fluid administration in the absence of pulmonary oedema
   - Consider intravenous fluid therapy to increase right sided preload in the absence of pulmonary oedema.
3. Reduce RV afterload
   - Inotropes, pulmonary vasodilators (nitric oxide, prostacycline)
   - Intra-aortic balloon pump.
4. Maintain chronotropic competence and atrioventricular synchrony
   - Avoid β-blockers in patients with proximal right coronary artery occlusion
   - Consider dual-chamber temporary pacing.

Kakouros N, Cokkinos D. Right ventricular myocardial infarction: pathophysiology, diagnosis, and management. Postgraduate Medical Journal. 2010;86(1022):719–728. https://www.ncbi.nlm.nih.gov/pubmed/20956396

30. Answer: D

Permanent pacing for sinus node dysfunction is only indicated in patients with symptoms directly attributable to bradycardia, irrespective of minimum heart rate or pause duration.

Sinus node dysfunction is most often related to age-dependent progressive fibrosis of the sinus nodal tissue and surrounding atrial myocardium. It may lead to abnormalities of the sinus node, and atrial impulse formation and propagation, which will therefore result in various bradycardic or pause-related syndromes. Less common causes include acute myocardial ischemia, atrial tachyarrhythmias, electrolyte abnormalities, hypothyroidism, medications, infections, and metabolic abnormalities. Evaluation for these potentially treatable or reversible causes can be performed non-urgently in most cases.

Nocturnal bradycardias should prompt consideration of screening for sleep apnoea. Nocturnal bradycardias are common in patient with sleep apnoea. Treatment of sleep apnoea not only reduces the frequency of nocturnal bradycardias but also might offer cardiovascular benefits. The presence of nocturnal bradycardia is not in itself an indication for permanent pacing.

Kusumoto F, Schoenfeld M, Barrett C, et al. 2018 ACC/AHA/HRS Guideline on the Evaluation and Management of Patients with Bradycardia and Cardiac Conduction Delay. Circulation. 2018. https://www.ahajournals.org/doi/abs/10.1161/CIR.0000000000000628

31. Answer: A

This patient has classical clinical features of Takotsubo (stress) cardiomyopathy which was first described in 1990. Its characteristic finding is the left ventricular apex ballooning. Presenting symptoms include chest pain, dyspnoea, and syncope and can be similar to those in patients with acute coronary syndrome (ACS). 80% of patients have elevated troponin levels and 80% of patients have ischaemic changes on ECG and most have elevated levels of NT-pro BNP.

Diagnosis of Takotsubo cardiomyopathy does not preclude the diagnosis of ACS. Up to 15% of patients with Takotsubo cardiomyopathy have concurrent coronary artery disease on coronary angiography. Diagnostic criteria for Takotsubo cardiomyopathy includes the presence of a transient abnormality in left ventricular wall motion beyond a single epicardial coronary artery perfusion territory, the absence of obstructive coronary artery disease or angiographic evidence of acute plaque rupture, the presence of new ECG abnormalities or elevation in cardiac troponin levels, and the absence of pheochromocytoma and myocarditis. There are four types of Takotsubo cardiomyopathy: apical type (in most patients) followed by the midventricular type, the basal type, and the focal type.

Takotsubo cardiomyopathy has a higher incidence in patients with a past medical history of neurological and psychiatric disorders, such as epilepsy, stroke, subarachnoid haemorrhage, electroconvulsive therapy, anxiety, and depression. Previous studies suggest Takotsubo cardiomyopathy is associated with emotional triggers. Subsequent studies have found the condition may also occur with physical triggers or even without preceding triggers. Takotsubo cardiomyopathy has a female predominance.

Takotsubo cardiomyopathy was once thought a benign disease with transient systolic and diastolic left ventricular dysfunction with a variety of wall-motion abnormalities. However, rates of in-hospital shock and death were similar in patients with Takotsubo cardiomyopathy and ACS. Other complications such as ventricular tachycardia, ventricular aneurysm, and ventricular rupture have been reported. The rate of major adverse cardiac and cerebrovascular events is 10% per patient-year, and the rate of death is 5.6% per patient-year, during long-term follow-up.

Angiotensin-converting enzyme (ACE) inhibitors or angiotensin-receptor blockers have been shown to improve survival at one year. Beta-blockers showed no survival benefits at one year following diagnosis of Takotsubo cardiomyopathy.

Templin C, Ghadri J, Diekmann J, et al Clinical Features and Outcomes of Takotsubo (Stress) Cardiomyopathy. New England Journal of Medicine. 2015;373(10):929–938.
https://www.ncbi.nlm.nih.gov/pubmed/26332547

32. Answer: B

TAVI valves are bioprosthetic, so the recommendation is for dual antiplatelet therapy with aspirin and clopidogrel for 3–6 months, with aspirin continued lifelong.

Severe aortic stenosis (AS) is the most common form of valvular heart disease in the developed world, affecting 7% of adults older than 65 years. Most patients with early AS are asymptomatic. Currently, there is no evidence to support surgical aortic valve replacement (SAVR) or TAVI in asymptomatic patients, even with severe AS. However, once symptoms of angina, syncope, or heart failure develop, there is significantly increased mortality if untreated. SAVR remains the preferred treatment for low risk patients. For intermediate and high- risk patients, TAVI in suitable candidates is at least non-inferior.

According to the largest trial comparing TAVI to SAVR, major complications include:

- Major vascular complication rate (7.9% TAVI vs 5% SAVR).
- Major stroke rate (3.2% TAVI vs 4.3% SAVR).
- Heart block (secondary to direct compression of the bioprosthetic valve on the conduction tissue) requiring pacemaker insertion (8.5% TAVI vs 6.9% SAVR).
- Acute kidney injury (1.3% TAVI vs 3% SAVR).
- Other: Paravalvular aortic regurgitation (<5%), infectiveInfective endocarditis, catastrophic complications: coronary obstruction, aortic dissection, cardiac perforation (0.2–1.1%).

Adams H, Ashokkumar S, Newcomb A, et al. Contemporary review of severe aortic stenosis. Internal Medicine Journal. 2019;49(3):297–305.
https://onlinelibrary.wiley.com/doi/abs/10.1111/imj.14071

33. Answer: A

This patient has an inferolateral STEMI due to multivessel coronary disease. CABG is indicated in patients with severe left main coronary artery disease or triple vessel disease. CABG offers improved survival and quality of life for patients with more extensive coronary disease with reduced left ventricular systolic dysfunction and remodelling. Furthermore, patients with diabetes and multivessel disease have been shown to benefit from CABG.

Percutaneous coronary intervention (PCI) may be considered for patients with multivessel coronary artery disease who are not a candidate for open heart surgery due to comorbidities and poor functional status. PCI even with biventricular pacemaker–defibrillator is not the best option for this patient. He is young; stage 3A CKD and treatment with adalimumab are not contraindications for CABG.

Infarct-related artery (IRA) or culprit-only revascularisation in primary PCI is associated with higher rate of long-term major adverse cardiac events (MACE) compared with multivessel treatment. Patients scheduled for staged revascularisation experienced a similar rate of MACE to patients undergoing complete simultaneous treatment of non-IRA.

Pineda AM, Carvalho N, Gowani SA, Desouza KA, Santana O, Mihos CG, Stone GW, Beohar N. Managing Multivessel Coronary Artery Disease in Patients With ST-Elevation Myocardial Infarction: A Comprehensive Review. Cardiol Rev. 2017 Jul/Aug;25(4):179–188.
https://pubmed.ncbi.nlm.nih.gov/27124268/

34. Answer: D

Troponin is a protein complex of three subunits (T, I, and C) that are involved in the contractile process of skeletal and cardiac muscle. Both cardiac and skeletal muscle express troponin C; whereas troponin T and I are cardiac-specific. Cardiac troponin (cTn) is integral to the diagnosis of acute coronary syndrome (ACS), but is elevated in many patients without ACS.

Conditions associated with non-ACS cTn elevations include:

- Tachyarrhythmias
- Congestive heart failure
- Malignant hypertension
- Sepsis
- Myocarditis
- Valvular heart disease (aortic stenosis)
- Aortic dissection
- Pulmonary embolism, pulmonary hypertension
- Chronic kidney disease
- Acute ischaemic or haemorrhagic stroke
- Cardiac contusion or cardiac procedures (CABG, PCI, ablation, pacing, cardioversion, or endomyocardial biopsy)
- Infiltrative diseases (amyloidosis, hemochromatosis, sarcoidosis)
- Myocardial drug toxicity or poisoning (doxorubicin, trastuzumab, snake venoms)
- Rhabdomyolysis.

As cTn can be detected among healthy adults, there are guidelines regarding what is considered an 'elevated' level. The joint European/American College of Cardiology guidelines define a clinically relevant increase in cTn levels as a level that exceeds the 99th percentile of a normal reference population. However, using a statistical cut-off means that some normal individuals will have a value above this cut-off, and because other clinical causes can cause an elevation, cTn should be interpreted in the context of pre-test probability of ACS. cTn concentrations are elevated in one in eight patients in the emergency department (ED).

High-sensitivity assays can accurately detect cTn at lower levels than older-generation assays, giving them higher sensitivity for the detection of ACS at presentation, which means that the time interval to the second measurement of high-sensitivity cTn (hs-cTn) can be significantly shortened, thereby reducing the time to diagnosis and improving efficiency in the ED. There is concern that hs-cTn may have lower diagnostic accuracy in patients with low pre-test probability for ACS. Concern of misinterpretation of these hs-cTn elevations as ACS and patient harm associated with potential unnecessary therapies such as anticoagulation and coronary angiography has led some experts to recommend withholding hs-cTn testing in patients with a low pre-test probability for ACS.

However, practice guidelines also highlight that ACS frequently presents with atypical symptoms especially in the elderly and patients with diabetes. It recommends scrutiny for ACS with ECG and hs-cTn. These divergent recommendations result in uncertainty in clinical practice regarding hs-cTn testing in patients with low pre-test probability for ACS. A recent retrospective analysis reported low specificity for hs-cTn to diagnose MI when grouping ED patients with suspected MI together with patients with acute heart failure and patients with documented PE. Hence, it is very important to highlight that diagnostic testing with hs-cTn should be applied to the correct population, at the optimal time and in the appropriate clinical context. This patient has very low pretest probability because her epigastric discomfort is nonspecific; her renal function is normal, and there are no other cardiovascular risk factors.

Twerenbold R, Boeddinghaus J, Nestelberger T, Wildi K, Rubini Gimenez M, Badertscher P et al. Ref: JACC 70(8):996–1012. Clinical Use of High-Sensitivity Cardiac Troponin in Patients With Suspected Myocardial Infarction. Twerenbold et al. 2017;70(8):996–1012.
https://pubmed.ncbi.nlm.nih.gov/28818210/

35. Answer: B

This elderly woman is septic due to severe pneumonia. She has multiple cardiovascular risk factors. Her ECG shows ischaemic change, and the troponin level is elevated. This is a typical case of type 2 myocardial infarction (MI).

MI can be classified into various types, based on pathological, clinical, and prognostic differences, along with different treatment strategies:

Type 1: Spontaneous myocardial infarction

Spontaneous MI related to atherosclerotic plaque rupture, ulceration, erosion, or dissection with resulting intraluminal thrombus in one or more of the coronary arteries leading to decreased myocardial blood flow or distal platelet emboli with ensuing myocyte necrosis. The patient may have underlying severe coronary artery disease (CAD) but on occasion non-obstructive or no CAD.

Type 2: Myocardial infarction secondary to an ischemic imbalance
In instances of myocardial injury with necrosis where a condition other than CAD contributes to an imbalance between myocardial oxygen supply and/or demand, e.g. coronary endothelial dysfunction, coronary artery spasm, coronary embolism, tachy-/brady-arrhythmias, anaemia, respiratory failure, hypotension.
Type 3: Myocardial infarction resulting in death when biomarker values are unavailable.
Type 4a: Myocardial infarction related to percutaneous coronary intervention (PCI).
Type 4b: Myocardial infarction related to stent thrombosis.
Type 5: Myocardial infarction related to coronary artery bypass grafting (CABG).
Patients with type 2 MI are frequently encountered in clinical practice and may be more common than type 1 MI. Diagnostic criteria for type 2 MI include:

Detection of a rise and/or fall of troponin values with at least one value above the 99th percentile, and evidence of an imbalance between myocardial oxygen supply and demand unrelated to acute coronary atherothrombosis, requiring at least one of the following:

- Symptoms of acute myocardial ischaemia
- New ischaemic ECG changes
- Development of pathological Q waves
- Imaging evidence of new loss of viable myocardium or new regional wall motion abnormality in a pattern consistent with an ischaemic aetiology.

The diagnosis of type 2 MI is associated with a poor prognosis: less than 40% of patients will live 5 years past their diagnosis. In contrast, 65% of patients with type 1 MI will survive for 5 years. This is because type 2 MI typically occurs among older patients with multiple comorbidities and is identified in the context of hemodynamic instability, including shock, tachycardia, respiratory failure, gastrointestinal bleeding, decompensated heart failure, or recent surgery. Moreover, in contrast to type 1 MI that has a clear set of guideline-based recommendations for treatment, management of type 2 MI remains uncertain.

Although the benefits of antiplatelet agents, β-blockers, and statins have been demonstrated among patients with type 1 MI, the utility of these medications among patients with type 2 MI remains uncertain. Currently, patients with type 2 MI are less likely to be discharged while taking these cardioprotective agents.

Reported prevalence of CAD among patients with type 2 MI varies with study design, ranging from 36% to 78%. About 30% of patients with type 2 MI experiencing major adverse cardiovascular events at 5 years, it is plausible that coronary revascularisation could be beneficial in those with obstructive CAD.

Thygesen K, Alpert J, Jaffe A, et al. Fourth universal definition of myocardial infarction (2018). European Heart Journal. 2018;40(3):237–269.
https://academic.oup.com/eurheartj/article/40/3/237/5079081

36. Answer: C

This patient's ECG, while in sinus rhythm 1-year ago shows classic pattern of pre-excitation – Wolff-Parkinson-White (WPW) syndrome. The ECG features a short PR interval; a slurred, thickened initial upstroke of the QRS complex, which is termed a delta wave; and a slight widening of the QRS deflexion with increased ventricular activation time. He presents with symptomatic pre-excited atrial fibrillation (AF) with a rapid ventricular response requiring urgent treatment.

AF is a medical emergency when rapid antegrade conduction over an accessory pathway occurs in WPW syndrome. This is because the normal rate-limiting effects of the atrioventricular (AV) node are bypassed, and the resultant excessive ventricular rates may lead to ventricular fibrillation and sudden death. The incidence of paroxysmal AF is between 10% and 38% in patients with WPW syndrome.

Urgent treatment for patients with WPW syndrome presenting with a tachydysrhythmia who are haemodynamically unstable, regardless of the QRS duration or regularity is direct current cardioversion. The electrical shock depolarises all excitable myocardium, lengthens refractoriness, interrupts re-entrant circuits, discharges foci, and establishes electrical homogeneity that terminates re-entry. Embolic episodes occur in 1–3% of the patients converted from AF to sinus rhythm if the episodes are longer than 48 hours. In those patients, anticoagulation must be addressed prior to cardioversion, with consideration of a transesophageal echocardiogram to exclude left atrial thrombus.

The acute treatment of pre-excited AF WPW syndrome in a haemodynamically stable patient requires a rapid acting drug that can be given intravenously and can lengthen antegrade refractoriness and slow conduction in both the AV node/His-Purkinje system and the accessory pathway. The class IC antiarrhythmic drugs such as flecainide which prolong the refractory period of the accessory connection are effective in this setting. Amiodarone, a class III antiarrhythmic drug, may also be used. AV node

blockers (β-blockers, non-dihydropyridine calcium channel blockers and digoxin) are contraindicated for pre-excited AF because inhibition of AV node conduction can increase pre-excitation and lead to an increase in the ventricular rate and ventricular fibrillation. Intravenous adenosine is also contraindicated because it causes an effect similar to verapamil and can precipitate ventricular fibrillation. Catheter ablation is the recommended treatment for the long-term therapy of pre-excited AF.

Svendsen J, Dagres N, Dobreanu D, et al. Current strategy for treatment of patients with Wolff-Parkinson-White syndrome and asymptomatic preexcitation in Europe: European Heart Rhythm Association survey. Europace. 2013;15(5):750–753.
https://academic.oup.com/europace/article/15/5/750/675642

37. Answer: D

Minoxidil is metabolised to an active sulfate metabolite, which antagonises the effect of ATP on K_{ATP} channels. The cell is thus hyperpolarised, which deactivates voltage-dependant calcium channels. The net effect of this action is smooth muscle relaxation. Undesirable side effects of K_{ATP} channel blockade include hirsutism and marked salt and water retention. Therefore, minoxidil is usually co-administered with a loop diuretic.

38. Answer: C

Sacubitril (through its active metabolite sacubitilat) inhibits neprolysin, which in turn increases circulating levels of natriuretic peptides, thus decreasing extracellular volume through induction of renal sodium excretion. In combination with valsartan, sacubitril shows efficacy in treating symptomatic patients with heart failure with reduced ejection fraction. Neprolysin is involved in the degradation of other peptides including bradykinin which elicited adverse effects of renal failure, angioedema, and hyperkalaemia in the randomised controlled trial.

39. Answer: B

Moxonidine antagonises the central control of sympathetically mediated vasoconstriction by stimulating the imidazoline (I_1) receptor present in the brainstem, which in turn decreases central catecholamine synthesis. Moxonidine and clonidine act at α_2 receptors as well, however moxonidine has much higher affinity for the I_1 receptor.

40. Answer: E

Captopril is the archetypal ACE inhibitor that decreases the production of angiotensin II by competitively adhering to the binding site for angiotensin I on ACE. ACE is a membrane bound enzyme predominantly present on vascular endothelium which is most extensively, but not exclusively, expressed in the lung. Angiotensin I undergoes conformational change when interacting with ACE to produce angiotensin II. Angiotensin II has multiple effects which increase blood pressure including proximal tubular absorption of sodium, increased secretion of aldosterone, increased noradrenaline release, and growth of cardiac and vascular cells. ACE is also involved in the degradation of bradykinin, consequently angioedema is a well-known side effect.

41. Answer: G

Some drugs induce relaxation of vascular smooth muscle by increasing cellular concentration of either cyclic adenosine monophosphate (cAMP) or cyclic guanine monophosphate (cGMP). Nitrates are reduced to nitric oxide by a variety of mechanisms. Nitric oxide activates guanylyl cyclase, which in turn increases cGMP production from guanosine triphosphate, which stimulates dephosphorylation of myosin, leading to vasodilation.

42. Answer: A

Beta receptor blockers decrease blood pressure by two main mechanisms. The first is by decreasing cardiac output by blockade of cardiac β_1-receptors, the second is by blocking renal β_1-receptors, resulting in decreased renin secretion.

43. Answer: F

Hydralazine is an arterial and arteriolar vasodilator that directly causes a fall in systemic vascular resistance by interfering with inositol triphosphate's effects on the sarcoplasmic reticulum, which decreases calcium release, resulting in less smooth muscle contraction. The fall in blood pressure is usually accompanied by reflex tachycardia and increased cardiac output. Hydralazine causes drug-induced lupus, which can limit its use in the medium to long term.

Rang H, Ritter J, Flower R, et al. Rang and Dale's pharmacology. 9th ed. Edinburgh: Elsevier; 2019.
https://www.elsevierhealth.com.au/rang-dales-pharmacology-9780702074486.html

44. Answer: E

All of the signs suggest pulmonary hypertension, pointing to Eisenmenger's syndrome, which may be found in older adults who have had a reversal of shunt from right-to-left before open-heart surgery is available. It is the most advanced form of pulmonary arterial hypertension due to elevated pulmonary vascular resistance causing right-to-left intracardiac shunt or great artery shunting, leading to systemic arterial desaturation.

45. Answer: G

Tetralogy of Fallot is associated with a combination of four clinical features:

1. VSD
2. Right ventricular outflow obstruction
3. Overriding aorta
4. Right ventricular hypertrophy

Although the long-term survival has improved for patients with repaired tetralogy of Fallot, residual haemodynamic and electrophysiological sequalae are common in adults. These groups of patients may have symptoms of arrythmias, heart failure, exercise intolerance, and death in early adulthood. Implantable cardioverter–defibrillators (ICDs) as a primary intervention should be considered in patients who meet standard qualifying criteria (i.e. LV ejection fraction ≤35% with NYHA class II or III symptoms).

46. Answer: D

The most common site for coarctation (CoA) is just distal to the origin of the left subclavian artery. An ECG may show signs of systolic overload, including left ventricular hypertrophy.

Sometimes the diagnosis can be easily missed if the lower limb blood pressure is not routinely measured. The long-term complications of CoA are generally related to chronic upper body systemic hypertension. Complications in patients with repaired CoA include recoarctation of the aorta, aneurysm, pseudoaneurysm, and dissection; thus, patients will require ongoing monitoring after operation. CoA is also known to be associated with Turner's syndrome.

47. Answer: A

There are two common types of ASD in adults: ostium secundum (most common) and ostium primum. These patients characteristically exhibit a left-to-right intracardiac shunt which may lead to right heart enlargement and, in a minority of the patients, pulmonary arterial hypertension (PAH). ECG findings may demonstrate right-axis deviation, right bundle branch block pattern, right ventricular hypertrophy from systolic overload and CXR findings may show an enlarged right atrium and ventricle, increased pulmonary vasculature, a dilated main pulmonary artery, and a small aortic knob. Almost all ASDs need to be closed surgically or, if the patient is suitable, with a percutaneous closure device when a pulmonary-to-systemic blood flow (shunt) ratio (Qp/Qs) >1.5:1. It is important to evaluate severe PAH prior to operation since closure of ASDs is contraindicated in this group of patients.

ASD due to ostium primum is caused by an endocardial cushion defect adjacent to the atrioventricular valves. In addition to the aforementioned signs in patients with ostium secundum, there is also associated mitral regurgitation, tricuspid regurgitation or VSD. ECG findings should show left-axis deviation, right bundle branch block and sometimes a prolonged PR interval. The condition is associated with Down's syndrome and Holt-Oram syndrome.

48. Answer: B

VSD in adults are usually small or large with Eisermenger's syndrome. Small restrictive defects may be monitored conservatively without the need for operation. ECG and CXR findings may demonstrate left ventricular hypertrophy. Closure of the VSD is indicated when the left-to-right shunt is moderate to large when the pulmonary-to-systemic flow is >1.5 to 1. Eisermenger's syndrome and severe PAH are contraindications to operative intervention due to significantly increased surgical risks. VSD is associated with Down syndrome.

49. Answer: F

A Marfanoid habitus is suggestive of a diagnosis of Marfan's syndrome. Patients with Marfan's syndrome are at an increased risk of experiencing progressive aortic root dilatation and aortic dissection. Serial echocardiograms are required to monitor the size of the aortic root over time. A slit-lamp examination may be required to diagnose lens dislocation. Patients with lens dislocation may have had lens replacement surgery in the past.

50. Answer: C

The ductus arteriosus usually closes shortly after birth. Failure of the ductus to close in the early weeks of life results in a PDA, which allows blood to flow between the aorta and the pulmonary artery, increasing blood flow in the pulmonary circulation.

Patients with a large PDA can exhibit signs and symptoms of pulmonary hypertension and heart failure. Patients with a small PDA are often asymptomatic but may develop infective endocarditis. PDAs are most commonly closed using catheter-based or surgical intervention. An enlarged left ventricle and calcification of the duct may be evidenced on CXR.

Stout K, Daniels C, Aboulhosn J, et al. 2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease: A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. Circulation. 2019;139(14).
https://ahajournals.org/doi/pdf/10.1161/CIR.0000000000000603