Medical Conditions

Aortic dissection

DEFINITION A condition where a tear in the aortic intima allows blood to surge into the aortic wall, causing a split between the inner and outer tunica media, and creating a false lumen.

AETIOLOGY Degenerative changes in the smooth muscle of the aortic media are the predisposing event. Common causes and predisposing factors are:

- hypertension;
- aortic atherosclerosis;
- connective tissue disease (e.g. SLE, Marfan's, Ehlers–Danlos);
- congenital cardiac abnormalities (e.g. aortic coarctation);
- aortitis (e.g. Takayasu's aortitis, tertiary syphilis);
- iatrogenic (e.g. during angiography or angioplasty);
- trauma;
- crack cocaine.

Stanford classification divides dissection into:

- *type A* with ascending aorta tear (most common);
- *type B* with descending aorta tear distal to the left subclavian artery.

Expansion of the false aneurysm may obstruct the subclavian, carotid, coeliac and renal arteries.

EPIDEMIOLOGY Most common in ♂ between 40 and 60 years.

HISTORY Sudden central 'tearing' pain, may radiate to the back (may mimic an MI). Aortic dissection can lead to occlusion of the aorta and its branches:

Carotid obstruction: Hemiparesis, dysphasia, blackout.
Coronary artery obstruction: Chest pain (angina or MI).
Subclavian obstruction: Ataxia, loss of consciousness.
Anterior spinal artery: Paraplegia.
Coeliac obstruction: Severe abdominal pain (ischaemic bowel).
Renal artery obstruction: Anuria, renal failure.

EXAMINATION Murmur on the back below left scapula, descending to abdomen.
Blood pressure (BP): Hypertension (BP discrepancy between arms of >20 mmHg), wide pulse pressure. If hypotensive may signify tamponade, check for pulsus paradoxus.
Aortic insufficiency: Collapsing pulse, early diastolic murmur over aortic area.
Unequal arm pulses.
There may be a palpable abdominal mass.

INVESTIGATIONS
Bloods: FBC, cross-match 10 units of blood, U&E (renal function), clotting.
CXR: Widened mediastinum, localized bulge in the aortic arch.
ECG: Often normal. Signs of left ventricular hypertrophy or inferior MI if dissection compromises the ostia of the right coronary artery.
CT-thorax: False lumen of dissection can be visualized.
Echocardiography: Transoesophageal is highly specific.
Cardiac catheterization and aortography.

MANAGEMENT
Acute: If suspected, CT-thorax should be performed urgently concurrent to resuscitation. Resuscitate and restore blood volume with blood products. Monitor pulse and BP in both arms, central venous pressure monitoring, urinary catheter. Best managed in ITU setting.

Aortic dissection (continued)

Type A dissection: Treated surgically. Emergency surgery because of the risk of cardiac tamponade. Affected aorta is replaced by a tube graft. Aortic valve may also be replaced.

Type B dissection: Can be treated medically, surgically or by endovascular stenting. Control BP and prevent further dissection with IV nitroprusside and/or IV labetalol (use calcium channel blocker if β-blocker contraindicated). Surgical repair may be appropriate for patients with intractable or recurrent pain, aortic expansion, end-organ ischemia or progression of dissection, and has similar outcome rates. Endovascular repair is a newer technique using endovascular stents and is available in some centres, although evidence of benefit is still lacking (ADSORB trial results pending).

COMPLICATIONS Aortic rupture, cardiac tamponade, pulmonary oedema, MI, syncope, cerebrovascular, renal, mesenteric or spinal ischaemia.

PROGNOSIS Untreated mortality: 30% at 24 h, 75% at 2 weeks.
Operative mortality of 5–10%. A further 10% have neurological sequelae.
Prognosis for type B better than type A.

Aortic regurgitation

DEFINITION Reflux of blood from aorta into left ventricle (LV) during diastole. Aortic regurgitation (AR) is also called aortic insufficiency.

AETIOLOGY

Aortic valve leaflet abnormalities or damage: Bicuspid aortic valve, infective endocarditis, rheumatic fever, trauma.

Aortic root/ascending aorta dilation: Systemic hypertension, aortic dissection, aortitis (e.g. syphilis, Takayasu's arteritis), arthritides (rheumatoid arthritis, seronegative arthritides), Marfan's syndrome, pseudoxanthoma elasticum, Ehlers–Danlos syndrome, osteogenesis imperfecta.

Reflux of blood into the LV during diastole results in left ventricular dilation and ↑ end-diastolic volume and ↑ stroke volume. The combination of ↑ stroke volume and low end-diastolic pressure in the aorta may explain the collapsing pulse and the wide pulse pressure. In acute AR, the LV cannot adapt to the rapid increase in end-diastolic volume caused by regurgitant blood.

EPIDEMIOLOGY Chronic AR often begins in the late 50s, documented most frequently in patients >80 years.

HISTORY Chronic AR: initially asymptomatic. Later, symptoms of heart failure: exertional dyspnoea, orthopnoea, fatigue. Occasionally angina.

Severe acute AR: sudden cardiovascular collapse.

Symptoms related to the aetiology, e.g. chest or back pain in patients with aortic dissection.

EXAMINATION Collapsing 'water-hammer' pulse and wide pulse pressure. Thrusting and heaving (volume-loaded) displaced apex beat.

Early diastolic murmur at lower left sternal edge, better heard with the patient sitting forward with the breath held in expiration. An ejection systolic murmur is often heard because of ↑ flow across the valve.

Austin Flint mid-diastolic murmur: Over the apex, from turbulent reflux hitting anterior cusp of the mitral valve and causing a physiological mitral stenosis.

Rare signs associated with a hyperdynamic pulse:

Quincke's sign: Visible pulsations on nail-bed.

de Musset's sign: Head nodding in time with pulse.

Becker's sign: Visible pulsations of the pupils and retinal arteries.

Müller's sign: Visible pulsation of the uvula.

Corrigan's sign: Visible pulsations in neck.

Traube's sign: 'Pistol shot' (systolic and diastolic sounds) heard on auscultation of the femoral arteries.

Duroziez's sign: A systolic and diastolic bruit heard on partial compression of femoral artery with a stethoscope.

Rosenbach's sign: Systolic pulsations of the liver.

Gerhard's sign: Systolic pulsations of the spleen.

Hill's sign: Popliteal cuff systolic pressure exceeding brachial pressure by >60 mmHg.

INVESTIGATIONS

CXR: Cardiomegaly. Dilation of the ascending aorta. Signs of pulmonary oedema may be seen with left heart failure.

ECG: May show signs of left ventricular hypertrophy (deep S wave in $V_{1–2}$, tall R wave in $V_{5–6}$, inverted T waves in I, aVL, $V_{5–6}$ and left-axis deviation).

Aortic regurgitation (continued)

Echocardiogram: 2D echo and M-mode may indicate the underlying cause (e.g. aortic root dilation, bicuspid aortic valve) or the effects of AR (left ventricular dilation/dysfunction and fluttering of the anterior mitral valve leaflet). Doppler echocardiography for detecting AR and assessing severity. Periodic (annual) follow-up echocardiogram for serial measurements of LV size and function.

Cardiac catheterization with angiography: If there is uncertainty about the functional state of the ventricle or the presence of coronary artery disease.

MANAGEMENT

Aortic valve replacement: In patients with symptoms of ventricular decompensation, or LV dysfunction: ejection fraction <50%, LV enlargement (end-systolic dimension >55 mm; end-diastolic dimension >75 mm).

Vasodilators (ACE inhibitor or nifedipine): In patients with LV systolic dysfunction (left ventricular ejection fraction (LVEF) <50%), or progressive LV dilatation. Vasodilators ↓ systemic vascular resistance and the afterload, i.e. the burden on the volume-loaded LV. Treat the complications (e.g. heart failure).

COMPLICATIONS Left ventricular failure and pulmonary oedema.

PROGNOSIS Chronic AR is often well tolerated for many years without symptoms.

Prognosis depends on the underlying aetiology. Acute AR caused by aortic dissection or infective endocarditis is fatal if not treated urgently.

Aortic stenosis

DEFINITION Narrowing of the left ventricular outflow at the level of the aortic valve.

AETIOLOGY

1. Stenosis secondary to rheumatic heart disease (commonest worldwide);
2. calcification of a congenital bicuspid aortic valve;
3. calcification/degeneration of a tricuspid aortic valve in the elderly.

EPIDEMIOLOGY Prevalence in ~3% of 75-year-olds. ♂ > ♀. Those with bicuspid aortic valve may present earlier (as young adults).

HISTORY May be asymptomatic initially.
Angina (because of ↑ oxygen demand of the hypertrophied ventricles).
Syncope or dizziness on exercise.
Symptoms of heart failure (e.g. dyspnoea).

EXAMINATION
BP: Narrow pulse pressure.
Pulse: Slow-rising.
Palpation: Thrill in the aortic area (if severe). Forceful sustained thrusting undisplaced apex beat.
Auscultation: Harsh ejection systolic murmur at aortic area, radiating to the carotid artery and apex. Second heart sound (A2 component) may be softened or absent (because of calcification). A bicuspid valve may produce an ejection click.
Distinguish from aortic sclerosis[1] **and hypertrophic obstructive cardiomyopathy (HOCM).**[2]

INVESTIGATIONS
ECG: Signs of left ventricular hypertrophy (deep S wave in $V_{1–2}$, tall R wave in $V_{5–6}$, inverted T waves in I, aVL, $V_{5–6}$ and left-axis deviation), LBBB.
CXR: Post-stenotic enlargement of the ascending aorta, calcification of aortic valve.
Echocardiogram: Visualizes structural changes of the valves and level of stenosis (valvar, supravalvar or subvalvar). Estimation of aortic valve area and pressure gradient across the valve in systole and left ventricular function may be assessed.
Cardiac angiography: Allows differentiation from other causes of angina, and to assess for concomitant coronary artery disease (50% of patients with severe aortic stenosis have significant coronary artery disease).

MANAGEMENT General principle is surgical management unless contraindicated.
Surgical: Valve replacement is recommended if valve pressure difference is >50 mmHg in a symptomatic patient. If unfit for surgery, balloon dilation (valvoplasty) may be performed but this is considered to be a palliative procedure due to high rate of complications (e.g. MI, myocardial perforation, severe AR).
Medical: Manage left ventricular failure (*see* Cardiac failure); ACE inhibitors and vasodilators should be used very cautiously in aortic stenosis. Antibiotic prophylaxis against infective endocarditis.

[1]Aortic sclerosis is senile degeneration with no left ventricular outflow tract obstruction. The pulse character is normal, a thrill is not palpable and the ejection systolic murmur radiates only faintly. The S_2 heart sound is normal.

[2]HOCM: *see* Cardiomyopathy.

Aortic stenosis (continued)

COMPLICATIONS Arrhythmias, Stokes–Adams attacks, MI, left ventricular failure and sudden death.

PROGNOSIS Survival differs according to symptoms. In symptomatic patients with severe aortic stenosis causing left ventricular failure, average survival 50% at 18 months without surgery. Average survival with angina 5 years; syncope 3 years; dyspnoea 2 years.

Atrial fibrillation

DEFINITION Characterized by rapid, chaotic and ineffective atrial electrical conduction. Often subdivided into: 'permanent', 'persistent' and 'paroxysmal'.

AETIOLOGY There may be no identifiable cause ('lone' atrial fibrillation (AF)). Secondary causes lead to abnormal atrial electrical pathways that result in AF.

Systemic causes: Thyrotoxicosis, hypertension, pneumonia, alcohol.

Heart: Mitral valve disease, ischaemic heart disease, rheumatic heart disease, cardiomyopathy, pericarditis, sick sinus syndrome,[1] atrial myxoma.

Lung: Bronchial carcinoma, pulmonary embolism.

EPIDEMIOLOGY Very common in the elderly (~5% of those >65 years). May be paroxysmal.

HISTORY Often asymptomatic. Some patients experience palpitations or syncope. Symptoms of the cause of the AF.

EXAMINATION Irregularly irregular pulse, difference in apical beat and radial pulse. Look for thyroid disease and valvular heart disease.

INVESTIGATIONS

ECG: Uneven baseline (fibrillations) with absent P waves, irregular QRS complexes. If there is a saw-tooth baseline, consider if there is atrial flutter.[2]

Blood: Cardiac enzymes, TFT, lipid profile, U&E, Mg^{2+}, Ca^{2+} (risk of digoxin toxicity ↑ with hypokalaemia, hypomagnesaemia or hypercalcaemia).

Echocardiogram: To assess for mitral valve disease, left atrial dilation, left ventricular dysfunction or structural abnormalities.

MANAGEMENT Treat any reversible cause (e.g. thyrotoxicosis, chest infection).
Specific treatment strategy focuses on:

(1) **Rhythm control**[3]:

If the AF is >48 h from onset, anticoagulate (at least 3–4 weeks) before attempting cardioversion.

DC cardioversion: Synchronized DC shock (2 × 100 J, 1 × 200 J).

Chemical cardioversion: Flecainide (contraindicated if there is history of ischaemic heart disease) or amiodarone.

Prophylaxis against AF: Sotalol, amiodarone or flecainide. Also consider providing 'pill-in-the-pocket' strategy for suitable patients.

(2) **Rate control**[3]

Chronic 'permanent' AF: Ventricular rate control with digoxin, verapamil and/or β-blockers. Aim for rate of ~90/min..

[1]Sick sinus (or tachy–brady) syndrome: caused by ischaemia, infarction or fibrosis of the sinus node, presenting with bradycardia (caused by intermittent failure of sinus node depolarization), with ↑ P–P intervals (>2 s) and intermittent tachycardia (caused by ↑ ectopic pacemaker activity). Treated with pacemaker insertion for symptomatic bradycardia, anti-arrhythmics for tachycardia and anticoagulation to reduce risk of thromboembolism.

[2]Atrial flutter is characterized by atrial rate of 300 min^{-1} and ventricular rate of 150 min^{-1} (2:1 heart block as atrioventricular (AV) node conducts every second beat). ECG shows a regular 'saw-tooth' baseline (rate ~300 min^{-1}). Can be reversed by vagal manoeuvres, IV adenosine (with cardiac monitoring) or chemical cardioversion.

[3]The AFFIRM trial showed that rhythm control offers no survival advantage over rate control.

Atrial fibrillation (continued)

(3) **Stroke risk stratification**:
Low-risk patients can be managed with aspirin, and high-risk patients require anticoagulation with warfarin. Risk factors indicating high risk are previous thromboembolic event, age ≥75 years with hypertension, diabetes or vascular disease, and/or clinical evidence of valve disease, heart failure or impaired left ventricular function.

COMPLICATIONS Thromboembolism (e.g. embolic stroke ~4% risk per year, ↑ risk with left atrial enlargement or left ventricular dysfunction). Worsens any existing heart failure.

PROGNOSIS Chronic AF in a diseased heart does not usually return to sinus rhythm.

Cardiac Arrest[1]

DEFINITION Acute cessation of cardiac function.

AETIOLOGY Classical reversible causes of cardiac arrest are the four H's and four T's:

Hypoxia	Tamponade
Hypothermia	Tension pneumothorax
Hypovolaemia	Thromboembolism
Hypo- or hyperkalaemia	Toxins and other metabolic disorders (drugs, therapeutic agents, sepsis)

EPIDEMIOLOGY N/A.

HISTORY Management precedes or is concurrent to history (e.g. from witnesses).

EXAMINATION Unconscious, patient is not breathing, absent carotid pulses.

INVESTIGATIONS

Cardiac monitor: Classification of the rhythm directs management (see below).
Bloods: ABG, U&E, FBC, cross-match, clotting, toxicology screen, glucose.

MANAGEMENT

Safety: Approach any arrest scene with caution as the cause of the arrest may still pose a threat. Defibrillators and oxygen are hazards. Help should be summoned as soon as possible.

Basic life support[1]:

1. If the arrest is witnessed and monitored, consider giving a precordial thump if no defibrillator immediately available.
2. Clear and maintain *airway* with head tilt (if no spinal injury), jaw thrust and chin lift.
3. Assess *breathing* by look, listen and feel.
   If not breathing, give two effective breaths immediately.
4. Assess *circulation* at carotid pulse for 10 s.
   If absent, give 30 chest compressions at rate of $\sim$100 min^{-1}. Continue cycles of 30 compressions for every two breaths.
5. Proceed to advanced life support as soon as possible.

Advanced life support[1]:

1. Attach cardiac monitor and defibrillator.
2. Assess the rhythm:
   (A) If pulseless ventricular tachycardia (VT)[2] or ventricular fibrillation (VF)[3] ('shockable rhythm'):
   - Defibrillate once: 150–360 J biphasic, 360 J monophasic. (Ensure no one is touching patient or bed when defibrillating.)
   - Resume CPR immediately for 2 min, and then return to 2.
   - Administer adrenaline (1 mg IV) after second defibrillation and again every 3–5 min.
   - If 'shockable rhythm' persists after third shock, administer amiodarone 300 mg IV bolus (or lidocaine).

[1] Refer to latest UK Resuscitation Council guidelines.

[2] *VT*: >3 successive ventricular extrasystoles (broad QRS complexes: >120 ms) at a rate of >120 min^{-1}. Patients at high risk of recurrent VT should be considered for implantable defibrillator which has been shown to be more effective than amiodarone.

[3] *VF*: Irregular, rapid ventricular activation with no cardiac output.

Cardiac Arrest (continued)

(B) If pulseless electrical activity (PEA) or asystole:
- CPR for 2 min, and then return to 2.
- Administer adrenaline (1 mg IV) every 3–5 min.
- Atropine (3 mg IV, once only) if asystole or PEA with rate <60 min^{-1}.

(C) During CPR:
- Check electrodes, paddle positions and contacts.
- Secure airway (e.g. attempt endotracheal intubation, high-flow oxygen). Once airway secure, give continuous compressions and breaths.
- Consider magnesium, bicarbonate, external pacing.
- Stop CPR and check pulse only if change in rhythm or signs of life.

Treatment of reversible causes:
Hypothermia: Warm slowly.
Hypo- or hyperkalaemia: Correction of electrolytes.
Hypovolaemia: IV colloids, crystalloids or blood products.
Tamponade: Pericardiocentesis under xiphisternum up and leftwards.
Tension pneumothorax: Needle into second intercostal space, mid-clavicular line.
Thromboembolism: (*see* Pulmonary embolism, Myocardial infarction).
Toxins: (*see* drug formulary for antidotes).

COMPLICATIONS Irreversible hypoxic brain damage, death.

PROGNOSIS Resuscitation is less successful in the arrests that occur outside hospital. Duration of inadequate effective cardiac output is associated with poor prognosis.

Cardiac failure

DEFINITION Inability of the cardiac output to meet the body's demands despite normal venous pressures.

AETIOLOGY:

Low output (↓ cardiac output):

Left heart failure: Ischaemic heart disease, hypertension, cardiomyopathy, aortic valve disease, mitral regurgitation.

Right heart failure: Secondary to left heart failure, infarction, cardiomyopathy, pulmonary hypertension/embolus/valve disease, chronic lung disease, tricuspid regurgitation, constrictive pericarditis/pericardial tamponade.

Biventricular failure: Arrhythmia, cardiomyopathy (dilated or restrictive), myocarditis, drug toxicity.

High output (↑ demand): Anaemia, beriberi, pregnancy, Paget's disease, hyperthyroidism, arteriovenous malformation.

EPIDEMIOLOGY 10% of > 65-year-olds.

Left: Tachycardia, tachypnoea, displaced apex beat, bilateral basal crackles, third heart sound ('gallop' rhythm: rapid ventricular filling), pansystolic murmur (functional mitral regurgitation).

Acute LVF: Tachypnoea, cyanosis, tachycardia, peripheral shutdown, pulsus alternans, gallop rhythm, wheeze 'cardiac asthma', fine crackles throughout the lung.

Right: ↑ JVP, hepatomegaly, ascites, ankle/sacral pitting, oedema, signs of functional tricuspid regurgitation (*see* Tricuspid regurgitation).

HISTORY

Left (symptoms caused by pulmonary congestion): Dyspnoea (New York Heart Association classification):

1. no dyspnoea;
2. dyspnoea on ordinary activities;
3. dyspnoea on less than ordinary activities;
4. dyspnoea at rest.

Orthopnoea, paroxysmal nocturnal dyspnoea, fatigue.

Acute LVF: Dyspnoea, wheeze, cough and pink frothy sputum.

Right: Swollen ankles, fatigue, ↑ weight (resulting from oedema), ↓ exercise tolerance, anorexia, nausea.

INVESTIGATIONS

Blood: FBC, U&Es, LFTs, CRP, glucose, lipids, TFTs.

In acute LVF: ABG, troponin, brain natriuretic peptide (BNP). ↑ Plasma BNP suggests the diagnosis of cardiac failure. A low plasma BNP rules out cardiac failure (90% sensitivity).

CXR (in acute LVF): Cardiomegaly (heart >50 % of thoracic width), prominent upper lobe vessels, pleural effusion, interstitial oedema ('Kerley B lines'), perihilar shadowing ('bat's wings'), fluid in the fissures.

ECG: May be normal. May have ischaemic changes, arrhythmia, left ventricular hypertrophy (seen in hypertension).

Echocardiogram: To assess ventricular contraction. If left ventricular ejection fraction (LVEF) <40%: systolic dysfunction. Diastolic dysfunction: ↓ compliance leading to a restrictive filling defect.

Swan–Ganz catheter: Allows measurements of right atrial, right ventricular, pulmonary artery, pulmonary wedge and left ventricular end-diastolic pressures.

MANAGEMENT

Acute LVF: *Cardiogenic shock*: Severe cardiac failure with low BP requires the use of inotropes (e.g. dopamine, dobutamine) and should be managed in ITU.

Cardiac failure (continued)

Pulmonary oedema: Sit up patient, 60–100% O_2 and consider CPAP. Other first-line therapies are diamorphine (venodilator and anxiolytic effect), GTN infusion (↓ preload), IV furosemide if fluid overloaded (venodilator and later diuretic effect). Monitor BP, respiratory rate, sat. O_2, urine output, ECG. Treat the cause, e.g. myocardial infarction, arrhythmia.

Chronic LVF: Treat the cause, e.g. hypertension. Treat exacerbating factors, e.g. anaemia. The following drug therapies are evidence-based.[1]

ACE-inhibitors (e.g. enalapril, perindopril, ramipril): Inhibit intracardiac renin-angiotensin system which may contribute to myocardial hypertrophy and remodelling. ACE inhibitors slow progression of the heart failure and improve survival.

β-*Blockers* (bisprolol or carvedilol): Block the effects of chronically activated sympathetic system, slow progression of the heart failure and improve survival. The benefits of ACE inhibitors and β-blockers are additive.

Loop diuretics (e.g. furosemide) and dietary salt restriction to correct fluid overload.

Aldosterone antagonists (spironolactone or, if not tolerated, eplerenone) improve survival in patients with NYHA class III/IV symptoms and on standard therapy. Monitor K^+ (may cause hyperkalaemia). May be used to assist in the management of diuretic-induced hypokalaemia.

Angiotensin receptor blockers (ARB) (e.g. candesartan): May be added in patients with persistent symptoms despite ACE inhibitors and β-blockers. Monitor K^+ (may cause hyperkalaemia).

Hydralazine and a nitrate: May be added in patients (particularly in Afro-Caribbeans) with persistent symptoms despite therapy with an ACE inhibitor and β-blocker.

Digoxin: Positive inotrope, ↓ hospitalization, but does not improve survival.

n-3 *polyunsaturated fatty acids*: Provide a small beneficial advantage in terms of mortality.

Cardiac resynchronization therapy (CRT): Biventricular pacing improves symptoms and survival in patients with LVEF $\leq$ 35%, cardiac dyssynchrony (QRS > 120 msec) and moderate to severe symptoms despite optimal medical therapy. Most patients who meet these criteria are also candidates for an implantable *cardiac* defibrillator (ICD) and receive a combined device.

Avoid drugs that can adversely affect patients with heart failure due to systolic dysfunction, e.g. NSAIDs, non-dihydropyridine calcium channel blockers (i.e. diltiazem and verapamil).

COMPLICATIONS Respiratory failure, cardiogenic shock, death.

PROGNOSIS Fifty per cent of patients with severe heart failure die within 2 years.

[1]Important trials include: CIBIS-II and US-Carvedilol (role of β-blockers); SOLVD and CONSENSUS (role of ACE-inhibitors); DIG study (role of digoxin); CHARM-Added trial (value of adding an ARB to appropriate doses of an ACE inhibitor); RALES trial (benefit of spironolactone in severe heart failure); CARE-HF, COMPANION and MIRACLE trial (benefits of CRT); GISSI-HF trial (benefit of n-3 polyunsaturated fatty acids).

Cardiomyopathy

DEFINITION Primary disease of the myocardium. Cardiomyopathy may be dilated, hypertrophic or restrictive.

AETIOLOGY The majority are idiopathic.

Dilated: Post-viral myocarditis, alcohol, drugs (e.g. doxorubicin, cocaine), familial (~25% of idiopathic cases, usually autosomal dominant), thyrotoxicosis, haemochromatosis, peripartum.

Hypertrophic: Up to 50% of cases are genetic (autosomal dominant) with mutations in β-myosin, troponin T or α-tropomyosin (components of the contractile apparatus).

Restrictive: Amyloidosis, sarcoidosis, haemochromatosis.

EPIDEMIOLOGY Prevalence of dilated cardiomyopathy and hypertrophic cardiomyopathy is ~0.05–0.20%. Restrictive cardiomyopathy is rare.

HISTORY

Dilated: Symptoms of heart failure, arrhythmias, thromboembolism, family history of sudden death.

Hypertrophic: Usually none. Syncope, angina, arrhythmias, family history of sudden death.

Restrictive: Dyspnoea, fatigue, arrhythmias, ankle or abdominal swelling.

Enquire about family history of sudden death.

EXAMINATION

Dilated: ↑ JVP, displaced apex beat, functional mitral and tricuspid regurgitations, third heart sound.

Hypertrophic: Jerky carotid pulse, double apex beat, ejection systolic murmur.

Restrictive: ↑ JVP (Kussmaul's sign: further ↑ on inspiration), palpable apex beat, third heart sound, ascites, ankle oedema, hepatomegaly.

INVESTIGATIONS

CXR: May show cardiomegaly, and signs of heart failure.

ECG:

All types: Non-specific ST changes, conduction defects, arrhythmias

Hypertrophic: Left-axis deviation, signs of left ventricular hypertrophy (*see* Aortic stenosis), Q waves in inferior and lateral leads.

Restrictive: Low voltage complexes.

Echocardiography:

Dilated: Dilated ventricles with 'global' hypokinesia.

Hypertrophic: Ventricular hypertrophy (disproportionate septal involvement)

Restrictive: Non-dilated non-hypertrophied ventricles. Atrial enlargement, preserved systolic function, diastolic dysfunction, granular or 'sparkling' appearance of myocardium in amyloidosis.

Cardiac catheterization: May be necessary for measurement of pressures.

Endomyocardial biopsy: May be helpful in restrictive cardiomyopathy.

Pedigree or genetic analysis: Rarely necessary.

MANAGEMENT

Dilated: Treat heart failure and arrhythmias. Consider implantable cardiac defibrillators (ICD) for recurrent VTs.

Hypertrophic: Treat arrhythmias with drugs, ICD for survivors of sudden death, reduce outflow tract gradients, pacemaker, surgery (e.g. septal myomectomy, septal ablation with ethanol). Screen family members with ECG or echocardiography.

Restrictive: No specific treatment. Manage the underlying cause.

Cardiac transplantation: May be considered in end-stage heart failure in all cardiomyopathy types.

Cardiomyopathy (continued)

COMPLICATIONS Heart failure, arrhythmias (atrial and ventricular).
Dilated and hypertrophic cardiomyopathy: Sudden death and embolism.
Hypertrophic: Infective endocarditis.

PROGNOSIS
Dilated: Depends on the aetiology, New York Heart Association functional class and ejection fraction.
Hypertrophic: Ventricular tachyarrhythmias are the major cause of sudden death.
Restrictive: Poor prognosis, many die within the first year after diagnosis.

Heart block

DEFINITION Impairment of the atrioventricular (AV) node impulse conduction, as represented by the interval between P wave and QRS complex.

First-degree AV block: Prolonged conduction through the AV node.

Second-degree AV block: Mobitz type I (Wenckebach): Progressive prolongation of AV node conduction culminating in one atrial impulse failing to be conducted through the AV node. The cycle then begins again.

Mobitz type II: Intermittent or regular failure of conduction through AV node. Also defined by the number of normal conductions per failed or abnormal one (e.g. 2 : 1 or 3 : 1).

Third-degree (complete) AV block: No relationship between atrial and ventricular contraction. Failure of conduction through the AV node leads to a ventricular contraction generated by a focus of depolarization within the ventricle (ventricular escape).

AETIOLOGY

MI or ischaemic heart disease (most common cause).
Infection (e.g. rheumatic fever, infective endocarditis).
Drugs (e.g. digoxin, β-blockers, Ca^{2+} channel antagonists).
Metabolic (e.g. hyperkalaemia, cholestatic jaundice, hypothermia).
Infiltration of conducting system (e.g. sarcoidosis, cardiac neoplasms, amyloidosis).
Degeneration of the conducting system.

EPIDEMIOLOGY Majority of the 250 000 pacemakers implanted annually are for heart block.

HISTORY

First degree: Asymptomatic.

Wenckebach: Usually asymptomatic.

Mobitz type II and third-degree block: May cause Stokes–Adams attacks (syncope caused by ventricular asystole). Other presentations include dizziness, palpitations, chest pain and heart failure.

EXAMINATION Often normal. Examine for signs of the cause.

Complete heart block: Slow large volume pulse; JVP may show 'cannon waves'.

Mobitz type II and third-degree block: Signs of a reduced cardiac output (e.g. hypotension, heart failure).

INVESTIGATIONS

ECG (consider ambulatory Holter or 24 h):

Type of heart block	ECG appearances
First degree	Prolonged PR interval (>0.2 s)
Mobitz type I (Wenckebach)	Progressively prolonged PR interval, culminating in a P wave that is not followed by a QRS. The pattern then begins again
Mobitz type II	Intermittently a P wave is not followed by a QRS. There may be a regular pattern of P waves not followed by a QRS (e.g. two P waves per QRS, indicating 2 : 1 block)
Third degree (complete)	No relationship between P waves and QRS complexes. If QRS initiated by focus in the bundle of His, the QRS is narrow. QRS initiated more distally are wide and slow rate (~30 beats/min)

Look for other signs of cause, e.g. ischaemia.

CXR: Cardiac enlargement, pulmonary oedema.

Blood: TFT, digoxin level, cardiac enzymes, troponin.

Echocardiogram: Wall motion abnormalities, aortic valve disease, vegetations.

Heart block (continued)

MANAGEMENT

Chronic block: Permanent pacemaker (PPM) insertion is recommended in patients with third-degree heart block, advanced Mobitz type II and symptomatic Mobitz type I.

Acute block (e.g. secondary to anterior MI): If associated with clinical deterioration, IV atropine and consider temporary (external) pacemaker.

COMPLICATIONS Asystole. Cardiac arrest. Heart failure. Complications of any pacemaker inserted.

PROGNOSIS Mobitz type II and third-degree block usually indicate serious underlying cardiac disease.

Hypertension

DEFINITION Defined as systolic BP >140 mmHg and/or diastolic BP >85 mmHg measured on three separate occasions. Malignant hypertension is defined as BP ≥200/130 mmHg.

AETIOLOGY

Primary:

- Essential or idiopathic hypertension (Commonest, >90% of cases).

Secondary:

- *Renal*: Renal artery stenosis, chronic glomerulonephritis, chronic pyelonephritis, polycystic kidney disease, chronic renal failure.
- *Endocrine*: Diabetes mellitus, hyperthyroidism, Cushing's syndrome, Conn's syndrome, hyperparathyroidism, phaeochromocytoma, congenital adrenal hyperplasia, acromegaly.
- *Cardiovascular*: Aortic coarctation, ↑ intravascular volume.
- *Drugs*: Sympathomimetics, corticosteroids, oral contraceptive pill.
- *Pregnancy*: Pre-eclampsia.

EPIDEMIOLOGY Very common. 10–20% of adults in the Western world.

HISTORY

Often asymptomatic.

Symptoms of complications (*see* Complications).

Symptoms of the cause.

Accelerated or malignant hypertension: Scotomas (visual field loss), blurred vision, headache, seizures, nausea, vomiting, acute heart failure.

EXAMINATION Measure on two to three different occasions before diagnosing hypertension and record lowest reading.

There may be loud second heart sound, fourth heart sound.

Examine for causes, e.g. radiofemoral delay (aortic coarctation), renal artery bruit (renal artery stenosis). Examine for end-organ damage, e.g. fundoscopy for retinopathy.

Keith–Wagner classification of retinopathy:

(I) 'silver wiring';
(II) as above, plus arteriovenous nipping;
(III) as above, plus flame haemorrhages and cotton wool exudates;
(IV) as above, plus papilloedema.

PATHOLOGY/PATHOGENESIS Fibrotic intimal thickening of the arteries, reduplication of elastic lamina and smooth muscle hypertrophy. Arteriolar wall layers replaced by pink hyaline material with luminal narrowing (hyaline arteriosclerosis).

INVESTIGATIONS

Blood: U&E, glucose, lipids.

Urine dipstick: Blood and protein.

ECG: May show signs of left ventricular hypertrophy (deep S wave in V_{1-2}, tall R wave in V_{5-6}, inverted T waves in I, aVL, V_{5-6}, left-axis deviation) or ischaemia.

Ambulatory BP monitoring (BP measured throughout the day): Excludes 'white coat' hypertension, allows monitoring of treatment response, assesses preservation of nocturnal dip.

Others: Especially in patients <35 years or other suspected secondary cases (see relevant topics).

MANAGEMENT Assessment and modification of other cardiovascular risk factors.

Conservative: Stop smoking, lose weight, ↓ alcohol, reduce dietary Na^+.

Hypertension (continued)

Investigate for secondary causes: Worthwhile in young patients, malignant hypertension or poor response to treatment.

Medical[1]: Treatment recommended for systolic BP $\geq$ 160 mmHg and/or diastolic BP $\geq$ 100 mmHg, or if evidence of end-organ damage. Other hypertension patients may still require treatment depending on other cardiac risk factors. Multiple drug therapies often necessary.

- *Thiazide diuretics (e.g. bendrofluamethiazide)*: Recommended first line, especially in >55-year-olds or black patients.
- *ACE inhibitors (e.g. ramipril) or angiotensin-II antagonist (e.g. losartan)*: First line in <55-year-olds, diabetic patients, heart failure or left ventricular dysfunction.
- *Ca^{2+} channel antagonists (e.g. amlodipine)*: Recommended first line, especially in >60-year-olds or black patients.
- β-*Blockers (e.g. atenolol)*: Not preferred initial therapy, but may be considered in younger patients. Avoid combining with thiazide diuretic to reduce patient risk of developing diabetes. May increase risk of heart failure.
- α-*Blockers (e.g. doxazosin)*: Fourth-line agent. May be useful for patients with prostatism.

Target BP:

- $\leq$140/85 mmHg (non-diabetic);
- $\leq$130/80 mmHg (diabetes without proteinuria);
- $\leq$125/75 mmHg (diabetes with proteinuria).

Severe hypertension (diastolic BP > 140 mmHg): Atenolol or nifedipine.

Acute malignant hypertension: IV β-blocker, labetolol or hydralazine sodium nitroprusside. Avoid very rapid lowering which can cause cerebral infarction.

COMPLICATIONS Heart failure, coronary artery disease and MI, CVA, peripheral vascular disease, emboli, retinopathy, renal failure, hypertensive encephalopathy, posterior reversible encephalopathy syndrome (PRES), malignant hypertension.

PROGNOSIS Good, if BP controlled. Uncontrolled hypertension linked with increased mortality (6× stroke risk and 3× cardiac death risk). Treatment reduces incidence of renal damage, stroke and heart failure.

[1]See NICE guidelines 2006 (available at http://www.nice.org.uk/nicemedia/pdf/CG034NICEguideline.pdf).

Ischaemic heart disease

DEFINITION Characterized by ↓ blood supply (ischaemia) to the heart muscle resulting in chest pain (angina pectoris). May present as 'stable angina' or 'acute coronary syndrome' (ACS).

ACS can be further subdivided into unstable angina (no cardiac injury), non-ST-elevation myocardial infarction (NSTEMI) or ST-elevation MI (STEMI, transmural infarction). MI refers to cardiac muscle necrosis resulting from ischaemia.

AETIOLOGY Angina pectoris occurs when myocardial oxygen demand exceeds oxygen supply. The most common cause is atherosclerosis. Other causes of coronary artery narrowing such as spasm (e.g. from cocaine), arteritis and emboli are rare.

MI is caused by sudden occlusion of a coronary artery due to rupture of an atheromatous plaque and thrombus formation.

Atherosclerosis: Endothelial injury is followed by migration of monocytes into subendothelial space and differentiation into macrophages. Macrophages accumulate LDL lipids insudated in the subendothelium and become foam cells. They release growth factors, which stimulate smooth muscle proliferation, production of collagen and proteoglycans. This leads to the formation of an atheromatous plaque.

ASSOCIATIONS/RISK FACTORS Male, diabetes mellitus, family history, hypertension, hyperlipidaemia, smoking, previous history.

EPIDEMIOLOGY Common, prevalence is >2%. More common in males. Annual incidence of MI in the United Kingdom is ~5 in 1000.

HISTORY

ACS: Chest pain or discomfort of acute onset. Central heavy tight 'gripping' pain that radiates to arms (usually left), neck, jaw or epigastrium. Occurring at rest, ↑ severity and frequency of previously stable angina.

May be associated with breathlessness, sweating, nausea and vomiting.

May be silent in elderly or in patients with diabetes.

Stable angina: Brought on by exertion and relieved by rest.

EXAMINATION

Stable angina: Look for signs of risk factors.

ACS: May have no clinical signs. Pale, sweating, restless, low-grade pyrexia. Check both radial pulses for aortic dissectio.

Arrhythmias, disturbances of BP. New heart murmurs (e.g. pansystolic murmur of mitral regurgitation from papillary muscle rupture or ventricular septal defect).

Signs of complications, i.e. acute heart failure, cardiogenic shock (hypotension, cold peripheries, oliguria).

INVESTIGATIONS

Blood: FBC, U&Es, CRP, glucose, lipid profile, cardiac enzymes: CK-MB and troponin-T or I[1] (sensitive marker of cardiac injury, ↑ after 12 h), amylase (pancreatitis may mimic MI), TFTs. AST and LDH ↑ after 24 and 48 h, respectively; occasionally used only to make retrospective diagnosis.

ECG:

Unstable angina or NSTEMI: May show ST depression, T-wave inversion (Q waves in these patients may indicate old MIs).

ST-elevation (Q-wave) MI: Hyperacute T waves, ST elevation (>1 mm in limb leads, >2 mm in chest leads), new-onset LBBB. Later: T inversion (hours) and Q waves (days).

[1]Causes of ↑ troponin other than acute thrombotic coronary artery occlusion include sepsis, tachycardia, myocarditis/pericarditis, pulmonary embolism, cardiac failure, renal failure, stroke and cardiac contusion.

Ischaemic heart disease (continued)

Location of infarct	Changes in leads
Inferior wall	II, III, aVF
Anterior wall	Septum (V_1–V_2), apex (V_3–V_4), anterolateral wall (V_5–V_6)
Lateral wall	I, aVL
Posterior infarct	Tall R wave and ST depression in V_1–V_3

CXR: To look for signs of heart failure. Useful to look for differentials (e.g. aortic dissection).

Exercise ECG testing (treadmill test)[2]: To determine prognosis and management.

Indications: In patients with troponin-negative ACS, or stable angina with an intermediate or high pretest probability of coronary heart disease (based on the characteristics of their chest pain, cardiac risk factors, age and gender). Patients should not be on digoxin (associated with a false-positive result).

Positive test: Defined as $\geq$ 1 mm horizontal or downsloping ST-segment depression measured at 80 ms after the end of the QRS complex (sensitivity: 60% and specificity: 90%).

Failed test: Failure to achieve at least 85% of the predicted maximal heart rate (220 – age) and otherwise negative (i.e. absence of chest discomfort or ECG findings). β-Blockers ↓ the maximal heart rate that is achieved and may be stopped prior to the test.

Resting ECG abnormalities (e.g. preexcitation syndrome, >1mm of ST depression, LBBB or paced ventricular rhythm) interfere with interpretation.

Radionuclide myocardial perfusion imaging (rMPI): Uses Tc-99m sestamibi or tetrofosmin. Can be performed under stress (exercise or pharmacological) or at rest. Stress testing would show low uptake in ischaemic myocardium. Rest testing can be used in patient with ACS, no previous MI but non-diagnostic troponin and ECGs.

Echocardiogram: Measure LVEF (early measurements may be misleading because of myocardial stunning). Exercise (or dobutamine stress) echocardiography may detect inducible wall motion abnormalities.

Pharmacologic stress testing: For patients who cannot exercise or if the exercise test is inconclusive. Pharmacological agents such as dipyridamole, adenosine or dobutamine can be used to induce a tachycardia. Various imaging modalities (e.g. rMPI, echocardiography) can be used to detect ischaemic myocardium. Dipyridamole and adenosine are contraindicated in AV block and reactive airway disease.

Cardiac catheterization/angiography: In ACS with positive troponin or TIMI score 5–7, or if high risk on stress testing (using Duke treadmill score based on exercise time, maximum ST-segment deviation and exercise angina).

Coronary calcium scoring using specialized CT (if available): may have a role in outpatients with atypical chest pain or in acute chest pain that is not clearly due to ischaemia (absence of CAC excludes obstructive coronary artery disease).

MANAGEMENT

Stable angina:

- *Minimize cardiac risk factors*: Control BP, hyperlipidaemia and diabetes. Provide advice on smoking, exercise, weight loss and low-fat diet. All patients should receive aspirin (75 mg/day) unless contraindicated.

[2]Contraindications to exercise ECG testing: severe aortic stenosis, uncontrolled arrhythmia, hypertension or heart failure. Stop if breathlessness or chest pain occurs, patient feels faint, arrhythmia, development of left bundle branch block, ST-segment elevation or depression >2 mm, fall in BP, BP > 230 mmHg, or if 90% maximal heart rate for age is achieved.

- *Immediate symptom relief*: GTN as a spray or sublingually.
- *Long-term treatment*: β-Blockers, e.g. atenolol, unless contraindicated (contraindications include acute heart failure, cardiogenic shock, bradycardia, heart block, asthma), calcium channel blockers (e.g. verapamil, diltiazem), nitrates (e.g. isosorbide dinitrate). Dual therapy may be indicated if monotherapy is ineffective.
- *Percutaneous coronary intervention (PCI)*: For localized areas of stenosis, in patients with angina not controlled despite maximal tolerable medical therapy. Restenosis rate is ~25% at 6 months but drug-eluting coronary stents ↓ restenosis rates (release an anti-restenotic drug, e.g. sirolimus or paclitaxel).
- *Coronary artery bypass graft (CABG)*: For more severe cases (three-vessel disease). The rates of MI and overall survival are generally similar between PCI and CABG.

Unstable angina/NSTEMI[3]:
- Admit to coronary care unit (CCU), oxygen, IV access, monitor vital signs and serial ECG.
- Analgesia: GTN (initially sublingual, IV infusion if persistent chest pain), morphine sulphate/diamorphine, antiemetic (metoclopramide).
- Aspirin (loading): 300 mg chewed. Maintenance: 75 mg, indefinite.
- Clopidogrel (loading): 300 mg. Maintenance: 75 mg for at least 1 year if troponin positive or high risk.
- Low-molecular-weight heparin (e.g. enoxaparin or dalteparin).
- β-Blocker (e.g. metoprolol) if not contraindicated.
- Glucose–insulin infusion if blood glucose >11 mmol/L.
- Consider glycoprotein (GP) IIb/IIIa inhibitors, e.g. tirofiban (initiated on presentation and continued for 48–72 h or until PCI) in patients:
  - undergoing PCI; *or*
  - at high risk for further cardiac events (troponin positive, TIMI risk score ≥4, continuing ischaemia or other high-risk features).
- If little improvement, consider urgent angiography ± revascularization.

STEMI:
- Admit to CCU, oxygen, IV access, monitor vital signs and serial ECG.
- Analgesia: GTN (initially sublingual, IV infusion if persistent chest pain), morphine sulphate/diamorphine, antiemetic (metoclopramide)
- Aspirin (loading): 300 mg chewed. Maintenance: 75 mg, indefinite.
- Clopidogrel (loading): 600 mg if patient going to primary PCI, 300 mg if undergoing thrombolysis and ≤75 years of age, 75 mg if undergoing thrombolysis and >75 years of age. Maintenance: 75 mg, for at least 1 year.
- β-Blocker (e.g. metoprolol) if not contraindicated.
- If undergoing primary PCI: IV heparin (plus GP IIb/IIIa inhibitor) or bivalirudin (an antithrombin). If undergoing thrombolysis with recombinant tissue plasminogen activator (rtPA): IV heparin.
- Glucose–insulin infusion if blood glucose >11 mmol/L.

[3]Evidence-based medicine for management of ACS:
- ISIS-1: beta-blockers.
- ISIS-2: aspirin and streptokinase.
- ISIS-4: ACE inhibitors.
- PROVE IT trial: initial high dose statin therapy.
- DIGAMI: initial insulin–glucose infusion and insulin for 3 months in all diabetic patients.
- CURE: combined clopidogrel and aspirin in NSTEMI.
- CLARITY: combined clopidogrel, aspirin and thrombolysis in STEMI
- PURSUIT, PRISM, EPIC, CAPTURE = GP-IIb/IIIa inhibitors in NSTEMI
- HORIZONS AMI trial: Bivalirudin (direct thrombin inhibitor) superiority compared with combined heparin and GP-IIb/IIIa inhibitors in STEMI.

Ischaemic heart disease (continued)

- *Primary PCI*[4] with goal <90 min if available *or thrombolysis* (see below) with goal of 30 min.
- *Thrombolysis*: Using fibrinolytics such as streptokinase or rtPA (alteplase, reteplase, tenecteplase) if within 12 h of chest pain with ECG changes (ST elevation, new-onset LBBB or posterior infarction) and not contraindicated.[5] *Rescue PCI*: If continued pain or ST elevation after thrombolysis (<50% ↓ of the initial ST-segment elevation on a follow-up ECG 60–90 min after fibrinolytic therapy).

Secondary Prevention: Antiplatelet agents (aspirin and clopidogrel), ACE inhibitors, β-blockers and statins. Control other risk factors (smoking, diabetes, hypertension).

Advice: Not to drive for 1 month following MI. Education by cardiac rehabilitation team: lifestyle changes (e.g. exercise, stop smoking, changing diet).

CABG for patients with left main stem or three-vessel disease.

COMPLICATIONS At risk of MI and other vascular diseases (e.g. stroke, peripheral vascular disease). Cardiac injury can lead secondarily to heart failure and arrhythmias.

Early complications of MI (24–72 h): Death, cardiogenic shock, heart failure, ventricular arrhythmias, heart block, pericarditis, myocardial rupture, thromboembolism.

Late complications of MI: Ventricular wall (or septum) rupture, valvular regurgitation, ventricular aneurysms, tamponade, Dressler's syndrome (pericarditis), thromboembolism.

PROGNOSIS

ACS: TIMI score (range 0–7) can be used for risk stratification (high scores are associated with high risk of cardiac events within 30 days), consists of:

(1) >65 years;
(2) known coronary artery disease;
(3) aspirin in last 7 days;
(4) severe angina (>2 episodes in 24 h);
(5) ST deviation >1 mm;
(6) elevated troponin levels;
(7) >3 coronary artery disease risk factors (hypertension, hyperlipidaemia, family history, diabetes, smoking).

Killip classification of acute MI:

Class I: no evidence of heart failure.
Class II: mild to moderate heart failure (S_3, crepitations <one-half way up the posterior lung fields, or jugular venous distension).
Class III: overt pulmonary oedema.
Class IV: cardiogenic shock.

The higher the Killip class on presentation, the greater the subsequent mortality.

[4]Other indications of primary PCI: typical and persistent symptoms and new or presumably new left bundle branch block; patients presenting 12–24 h after symptom onset, with persistent ischaemic symptoms, severe heart failure, haemodynamic or electrical instability.

[5]*Absolute contraindications to thrombolysis*: History of intracranial haemorrhage/vascular lesion/neoplasm, ischaemic stroke within 3 months, suspected aortic dissection, active bleeding or bleeding diathesis, significant closed-head or facial trauma within 3 months. *Relative contraindications*: Systolic BP > 180 mmHg, ischaemic stroke >3 months previously, dementia, traumatic or cardiopulmonary resuscitation >10 min, major surgery (within <3 weeks), recent internal bleeding (within 2–4 weeks), non-compressible vascular puncture, pregnancy, active peptic ulcer, current use of anticoagulants, for streptokinase: prior exposure or allergic reaction.

Mitral regurgitation

DEFINITION Retrograde flow of blood from LV to left atrium during systole.

AETIOLOGY Mitral valve damage or dysfunction:
- rheumatic heart disease (most common);
- infective endocarditis;
- mitral valve prolapse (prolapse of mitral valve leaflets into the left atrium during systole);
- papillary muscle rupture or dysfunction (secondary to ischaemic heart disease or cardiomyopathy);
- chordal rupture and floppy mitral valve associated with connective tissue diseases (e.g. pseudoxanthoma elasticum[1], osteogenesis imperfecta,[2] Ehlers–Danlos syndrome,[3] Marfan syndromes, SLE).

Functional mitral regurgitation may be secondary to left ventricular dilation.

EPIDEMIOLOGY Affects ~5% of adults. Mitral valve prolapse is more common in young females.

HISTORY

Acute MR: May present with symptoms of left ventricular failure.

Chronic MR: May be asymptomatic or present with exertional dyspnoea, palpitations if in AF and fatigue.

Mitral valve prolapse: Asymptomatic or atypical chest pain or palpitations.

EXAMINATION Pulse may be normal or irregularly irregular (AF).

Apex beat may be laterally displaced and thrusting (left ventricular dilation).

Pansystolic murmur, loudest at apex, radiating to axilla (palpable as a thrill). S_1 is soft; S_3 may be heard (rapid ventricular filling in early diastole).

Signs of left ventricular failure in acute mitral regurgitation.

Mitral valve prolapse: Mid-systolic click and late systolic murmur. The click moves towards the first heart sound on standing and moves away on lying down.

INVESTIGATIONS

ECG: Normal or may show AF or broad bifid *p* wave (*p* mitrale) indicating delayed activation of left atrium due to left atrial enlargement.

CXR: Acute mitral regurgitation may produce signs of left ventricular failure. Chronic mitral regurgitation shows left atrial enlargement, cardiomegaly (caused by left ventricular dilation) or mitral valve calcification in rheumatic cases.

Echocardiography: Every 6–12 months for moderate–severe MR to assess the LV ejection fraction and end-systolic dimension.

MANAGEMENT

Surgical: Indicated in patients with chronic MR with symptoms or LV enlargement or dysfunction, pulmonary hypertension, or new-onset AF.

[1]**Pseudoxanthoma elasticum**: Collagen/elastic tissue disorder characterized by yellow papules on the skin in the flexures, ischaemic heart disease, GI bleeding and angioid streaks on fundoscopy (caused by degeneration of Bruch's membrane).

[2]**Osteogenesis imperfecta**: Group of autosomally dominant inherited disorders caused by mutations in type I collagen gene causing brittle, deformed bones, blue sclerae, hypermobile joints and heart valve disorders.

[3]**Ehlers–Danlos syndrome**: A group of genetic disorders due to defects in type III collagen resulting in joint hypermobility and skin hyperelasticity and fragility. Spontaneous rupture of arteries is the most serious cardiovascular complication.

Mitral regurgitation (continued)

Mitral valve repair: May occasionally be feasible.

Mitral valve replacement: Mechanical valve with lifelong anticoagulation in patients <65 years with long-standing AF. Bioprosthetic valve in patients with a contraindication to warfarin or ≥65 years of age (since limited durability of bioprosthetic valve is less of an issue). The choice of valve in patients <65 years who are in sinus rhythm relies on patient preference. Discuss the risks of warfarin compared to the likelihood of repeat valve replacement in the future.

Medical:

Functional MR due to LV dysfunction: ACE inhibitors and/or angiotensin II receptor blockers. Optimal medical therapy of heart failure. In appropriate patients, CRT.

Mitral valve prolapse and symptomatic patients with primary MR (e.g. rheumatic or myxomatous): ↓ systolic pressure (β-blockers, diuretic).

Treatment of atrial fibrillation (See Atrial Fibrillation).

Anticoagulation with warfarin (target INR: 2–3): In patients with AF, history of systemic embolism or rheumatic mitral valve disease with left atrial thrombus. Antibiotic prophylaxis is no longer recommended in the absence of prosthetic repair or replacement for those undergoing dental or other invasive procedures.

Advice: Patients with severe MR, LV enlargement, pulmonary hypertension or ↓ left ventricular systolic function at rest should not participate in any competitive sports. Patients treated with long-term anticoagulation therapy should not engage in contact sports.

COMPLICATIONS Left atrial enlargement and AF (and resultant systemic embolism), pulmonary oedema, pulmonary hypertension, right heart failure, infective endocarditis.

PROGNOSIS Prognosis depends on aetiology, severity and state of left ventricular function. Acute mitral regurgitation resulting from rupture of a cusp or papillary muscle has a poor prognosis. Rheumatic mitral regurgitation may slowly deteriorate over 10–20 years.

Mitral stenosis

DEFINITION Mitral valve narrowing causing obstruction to blood flow from the left atrium to the ventricle.

AETIOLOGY Most common cause is rheumatic heart disease (90%).
Rarer causes are congenital mitral stenosis, SLE, rheumatoid arthritis, endocarditis and atrial myxoma (rare cardiac tumour).

EPIDEMIOLOGY Incidence is declining in industrialized countries because of declining incidence of rheumatic fever.

HISTORY May be asymptomatic.
Presents with fatigue, shortness of breath on exertion or lying down (orthopnoea). Palpitations (related to AF).
Rare symptoms: Cough, haemoptysis, hoarseness caused by compression of the left laryngeal nerve by an enlarged left atrium.

EXAMINATION May have peripheral or facial cyanosis (malar flush).
Pulse: May be 'thready' or irregularly irregular (AF).
Palpation: Apex beat is undisplaced and tapping. Parasternal heave (right ventricular hypertrophy and pulmonary hypertension)
Auscultation: Loud first heart sound with opening snap.
Mid-diastolic murmur (presystolic accentuation if in sinus rhythm).
Evidence of pulmonary oedema on lung auscultation (if decompensated).

INVESTIGATIONS
ECG: May be normal, broad bifid p wave (p mitrale) caused by left atrial hypertrophy, AF or evidence of right ventricular hypertrophy in cases of severe pulmonary hypertension.
CXR: Left atrial enlargement, cardiac enlargement, pulmonary congestion; mitral valve may be calcified in rheumatic cases.
Echocardiography: To assess functional and structural impairments. Transoesophageal gives better valve visualization.
Cardiac catheterization: Measures severity of heart failure.

MANAGEMENT
Medical: Anticoagulation for AF. Treat dyspnoea and heart failure with diuretics. Antibiotic cover for dental/invasive procedures. Cardioversion of AF may be considered.
Surgical: Mitral valvuloplasty, valvotomy or replacement if severe.

COMPLICATIONS AF and systemic embolism, pulmonary oedema, pulmonary hypertension and right heart failure, infective endocarditis.

PROGNOSIS Significantly worse if pulmonary hypertension or right heart failure develops.

Myocarditis

DEFINITION Acute inflammation and necrosis of cardiac muscle (myocardium).

AETIOLOGY Usually unknown (idiopathic).

Infection:

Viruses: e.g. Coxsackie B, echovirus, EBV, CMV, adenovirus, influenza.
Bacterial: e.g. post-streptococcal, tuberculosis, diphtheria, Lyme disease.
Fungal: e.g. candidiasis.
Protozoal: e.g. trypanosomiasis (Chagas disease).
Helminths: e.g. trichinosis.

Non-infective: Systemic disorders (e.g. SLE, sarcoidosis, polymyositis), hypersensitivity myocarditis (e.g. sulphonamides).
Drugs: Chemotherapy agents (e.g. doxorubicin, streptomycin)
Others: Cocaine abuse, heavy metals, radiation.

EPIDEMIOLOGY True incidence is unknown, as many cases are not detected at the time of acute illness. Coxsackie B virus is a common cause in Europe and the USA. Chagas disease is a common cause in South America.

HISTORY Prodromal 'flu-like' illness, fever, malaise, fatigue, lethargy.
Breathlessness (pericardial effusion/myocardial dysfunction).
Palpitations.
Sharp chest pain (suggesting associated pericarditis).

EXAMINATION Signs of concurrent pericarditis or complications: heart failure, arrhythmia.

INVESTIGATIONS

Blood: FBC (↑ WCC in infective causes), U&E, ↑ ESR or CRP, cardiac enzymes (may be ↑). To identify the cause (viral or bacterial serology, antistreptolysin O titre, ANA, serum ACE, TFT).
ECG: Non-specific T wave and ST changes, widespread saddle-shaped ST elevation in pericarditis.
CXR: May be normal or show cardiomegaly with or without pulmonary oedema.
Pericardial fluid drainage: Measure glucose, protein, cytology, culture and sensitivity.
Echocardiography: Assesses systolic/diastolic function, wall motion abnormalities, pericardial effusion.
Myocardial biopsy: Rarely required (result does not influence management).

MANAGEMENT

Supportive: Bed rest, treatment of complications (heart failure, arrhythmias), pericardial drainage for compromising pericardial effusion.
Steroids and immunosuppressants have been used in severe cases but are of unproven benefit.
Surgical: Cardiac transplantation for severe cases.

COMPLICATIONS Severe cases can lead to chronic inflammation, cardiac failure. Resolution of inflammation with different degrees of residual dilated cardiomyopathy, arrhythmias and death.

PROGNOSIS Usually mild and self-limiting. Recovery is variable in patients with severe acute myocarditis.

Pericarditis

DEFINITION Inflammation of the pericardium, may be acute, subacute or chronic.

AETIOLOGY
- Idiopathic;
- infective (commonly, coxsackie B, echovirus, mumps virus, streptococci, fungi, staphylococci, TB);
- connective tissue disease (e.g. sarcoid, SLE, scleroderma);
- post-myocardial infarction (24–72 h) in up to 20% of patients;
- Dressler's syndrome (weeks to months after acute MI);
- malignancy (lung, breast, lymphoma, leukaemia, melanoma);
- metabolic (myxoedema, uraemia);
- radiotherapy;
- thoracic surgery;
- drugs (e.g. hydralazine, isoniazid).

EPIDEMIOLOGY Uncommon. The clinical incidence is <1 in 100 hospital admissions. More common in males.

HISTORY

Chest pain: Sharp and central, which may radiate to neck or shoulders. Aggravated by coughing, deep inspiration and lying flat. Relieved by sitting forward.

Dyspnoea, nausea.

EXAMINATION Fever, pericardial friction rub (best heard lower left sternal edge, with patient leaning forward in expiration), heart sounds may be faint in the presence of an effusion.

Cardiac tamponade: ↑ JVP, ↓ BP and muffled heart sounds (Beck's triad). Tachycardia, pulsus paradoxus (reduced systolic BP by >10 mmHg on inspiration).

Constrictive pericarditis (chronic): ↑ JVP with inspiration (Kussmaul's sign), pulsus paradoxus, hepatomegaly, ascites, oedema, pericardial knock (rapid ventricular filling), AF.

INVESTIGATIONS

ECG: Widespread ST elevation that is saddle-shaped.

Echocardiogram: For assessment of pericardial effusion and cardiac function.

Blood: FBC, U&E, ESR, CRP, cardiac enzymes (usually normal). Where appropriate: blood cultures, ASO titres, ANA, rheumatoid factor, TFT, Mantoux test, viral serology.

CXR: Usually normal (globular heart shadow if >250 mL effusion). Pericardial calcification can be seen in constrictive pericarditis (best seen on lateral CXR or CT).

MANAGEMENT

Acute: Cardiac tamponade treated by emergency pericardiocentesis.

Medical: Treat the underlying cause, NSAIDs for relief of pain and fever.

Recurrent: Low-dose steroids, immunosuppressants or colchicine.

Surgical: Surgical excision of the pericardium (pericardiectomy) in constrictive pericarditis.

COMPLICATIONS Pericardial effusion, cardiac tamponade, cardiac arrythmias.

PROGNOSIS Depends on underlying cause. Good prognosis in viral cases (recovery within ~2 weeks), poor in malignant pericarditis. Pericarditis may be recurrent (particularly in those caused by thoracic surgery).

Pulmonary hypertension

DEFINITION A consistently increased pulmonary arterial pressure (>20 mmHg) under resting conditions.

AETIOLOGY

Primary: Idiopathic.

Secondary: Left heart disease (mitral valve disease, left ventricular failure, left atrial myxoma/thrombosis), chronic lung disease (COPD), recurrent pulmonary emboli, ↑ pulmonary blood flow (ASD, VSD, patent ductus arteriosus), connective tissue disease (e.g. SLE, systemic sclerosis), drugs (e.g. amiodarone).

EPIDEMIOLOGY Primary pulmonary hypertension is usually seen in young females.

HISTORY Dyspnoea (on exertion), chest pain, syncope, tiredness.
Symptoms of the underlying cause (e.g. chronic cough).

EXAMINATION ↑ JVP (Prominent a wave in the JVP waveform).

Palpation: Left parasternal heave (right ventricular hypertrophy).

Auscultation: Loud pulmonary component of S_2 (S_3/S_4 may be heard), an early diastolic murmur (Graham–Steell murmur) caused by pulmonary regurgitation may be present, if tricuspid regurgitation develops (large cv wave and pansystolic murmur).

Signs of the underlying condition, or right heart failure in severe cases.

INVESTIGATIONS

CXR: Cardiomegaly (right ventricular enlargement, right atrial dilation), prominent main pulmonary arteries (which taper rapidly), signs of the cause (e.g. COPD, calcified mitral valves).

ECG: Right ventricular hypertrophy (right-axis deviation, prominent R wave in V_1, T inversion in V_1, V_2), right atrial enlargement (peaked P wave in II, called 'P pulmonale'); limb leads exhibit low voltage (R < 5 mm) in COPD.

Echocardiography: To visualize right ventricular hypertrophy or dilation and possible underlying cause.

Lung function tests: To assess for chronic lung disease.

VQ scan: To assess for pulmonary embolism.

Cardiac catheterization: To assess severity, right heart pressures and response to vasodilators.

High resolution CT-thorax: Images pulmonary arteries and to diagnose lung disease.

Lung biopsy: Assesses structural lung changes.

MANAGEMENT

Medical: Treat secondary cause. For primary pulmonary hypertension, consider:

- anticoagulation (warfarin);
- calcium channel antagonists (e.g. verapamil, nifedipine);
- prostacycline analogues (e.g. iloprost);
- endothelial receptor antagonist (bosentan) blocks vasoconstriction, but teratogenic, so only indicated in NYHA III or IV patients;
- phosphodiesterase inhibitor (e.g. sildenafil) promotes pulmonary smooth muscle relaxation reducing pulmonary hypertension.

Surgical: Heart and lung transplantation may be an option for younger patients.

COMPLICATIONS Right heart failure, arrhythmias (AF, VT, VF), sudden death.

PROGNOSIS Chronic and incurable with unpredictable survival rate. Length of survival has improved to up to 15–20 years.

Rheumatic fever

DEFINITION An inflammatory multisystem disorder, occurring following group A β-haemolytic streptococci (GAS) infection.

AETIOLOGY The pathogenic mechanisms remain incompletely understood. Streptococcal pharyngeal infection is required, and genetic susceptibility may be present. Molecular mimicry is thought to play an important role in the initiation of the tissue injury (antibodies directed against GAS antigens cross-react with host antigens).

EPIDEMIOLOGY Peak incidence: between 5 and 15 years. More common in the Far East, Middle East, eastern Europe and South America. The mean incidence is 19/100 000. Despite ↓ incidence over time in the West, the incidence rates remain relatively high in non-Western countries.

HISTORY 2–5 weeks after GAS infection.
General: Fever, malaise, anorexia. *Joints*: Painful, swollen, ↓ movement/function. *Cardiac*: Breathlessness, chest pain, palpitations.

EXAMINATION
Duckett Jones criteria: Positive diagnosis if at least two major criteria, or one major plus two minor criteria are present.
Major criteria:
- Arthritis: Migratory or fleeting polyarthritis with swelling, redness and tenderness of large joints.
- Carditis: New murmur, e.g. Carey Coombs murmur (mid-diastolic murmur due to mitral valvulitis). Pericarditis, pericardial effusion or rub, cardiomegaly, cardiac failure, ECG changes (*see* Investigations).
- Chorea (Sydenham's): Rapid, involuntary, irregular movements with flowing or dancing quality. May be accompanied by slurred speech. More common in females.
- Nodules: Small firm painless subcutaneous nodules seen on extensor surfaces, joints and tendons.
- *Erythema marginatum* (20% cases): Transient erythematous rash with raised edges, seen on trunk and proximal limbs. They may form crescent- or ring-shaped patches.

Minor criteria:
- Pyrexia;
- previous rheumatic fever;
- arthralgia (only if arthritis is not present as major criteria);
- recent streptococcal infection (supported by positive throat cultures or ↑ antistreptolysin O titre);
- ↑ inflammatory markers (ESR, CRP or WCC);
- ↑ PR and QT intervals on ECG (only if carditis not present as major criteria).

INVESTIGATIONS
Blood: FBC (↑ WCC), ESR/CRP (↑), ↑ or rising antistreptolysin O titre.
Throat swab: Culture for GAS, rapid streptococcal antigen test.
ECG: Saddle-shaped ST elevation and PR segment depression (features of pericarditis), arrhythmias.
Echocardiogram: Pericardial effusion, myocardial thickening or dysfunction, valvular dysfunction.

MANAGEMENT
Strict bed rest: (∼ 4 weeks) Gradual mobilization with clinical improvement.

Rheumatic fever (continued)

Anti-inflammatory drugs: High-dose aspirin or, for more severe carditis, consider corticosteroids.

Antibiotics: Eradicate residual streptococcal infection using oral penicillin V for 10 days. Long-term antibiotics to prevent recurrence is necessary (e.g. long-acting benzathine penicillin G intramuscularly every 4 weeks, switch to oral prophylaxis in young adulthood). Prophylaxis in the setting of carditis should continue usually for 10 years. The risk for GAS exposure and severity of valvular disease should then be reviewed.

Carditis: Treat heart failure if present.

Chorea: May be controlled with diazepam or haloperidol.

Valve surgery: If valvular disease cannot be managed with medical therapy alone.

COMPLICATIONS Recurrence, may be precipitated by streptococcal infection, more common in patients with residual cardiac damage. Chronic rheumatic valvular disease: scarring, deformation and dysfunction (usually mitral or aortic) after 10–20 years; more common in those with carditis as part of acute rheumatic fever.

PROGNOSIS Acute rheumatic fever may last up to 3 months if untreated. ♀ more likely to develop mitral stenosis.

Note: Post-streptococcal reactive arthritis is a reactive arthritis that occurs ~1–2 weeks after GAS pharyngitis. Patients have marked arthritis with poor response to aspirin/NSAIDs, tenosynovitis, renal abnormalities and lower ESR and CRP than acute rheumatic fever. Secondary prophylaxis is given for up to 1 year and discontinued, unless there is evidence of valvular disease.

Tricuspid regurgitation

DEFINITION Tricuspid regurgitation is backflow of blood from the right ventricle to the right atrium during systole.

AETIOLOGY

Congenital: Ebstein anomaly (malpositioned tricuspid valve), cleft valve in ostium primum defect.
Functional: Consequence of right ventricular dilation (e.g. in pulmonary hypertension), valve prolapse.
Rheumatic heart disease: Associated with other valvular disease.
Infective endocarditis: Common in IV drug users. Usually staphylococcal.
Other: Carcinoid syndrome, trauma, cirrhosis (long-standing), iatrogenic (e.g. radiotherapy to the thorax).

EPIDEMIOLOGY The epidemiology differs with various causes. Infective endocarditis probably most common cause.

HISTORY Fatigue, breathlessness, palpitations, headaches, nausea, anorexia, epigastric pain made worse by exercise, jaundice, lower limb swelling.

EXAMINATION

Pulse: May be irregularly irregular due to AF (may occur with right atrial enlargement).
Inspection: ↑ JVP with giant *v* waves which may oscillate the earlobe. This is caused by transmission of right ventricular pressure to the great veins. There may be giant *a* wave, if the patient is in sinus rhythm.
Palpation: Parasternal heave.
Auscultation: Pansystolic murmur heard best at the lower left sternal edge, louder on inspiration (Carvallo sign). Loud P2 component of second heart sound.
Chest: Pleural effusion. Causes of pulmonary hypertension (e.g. emphysema).
Abdomen: Palpable liver (tender, smooth, pulsatile), ascites.
Legs: Pitting oedema.

INVESTIGATIONS

Blood: FBC, LFT, cardiac enzymes, blood cultures.
ECG: Tall P wave (right atrial hypertrophy) if in sinus rhythm. Changes indicative of other cardiac disease.
CXR: Right-sided enlargement of cardiac shadow.
Echocardiography: Extent of regurgitation estimated by colour flow Doppler. May be able to detect tricuspid valve abnormality (e.g. prolapse), right ventricular dilation.
Right heart catheterization: Rarely necessary but may be considered to assess pulmonary artery pressure.

MANAGEMENT

Medical: Treat the underlying condition, e.g. infective endocarditis or functional regurgitation caused by pulmonary hypertension. Diuretics may be given for fluid retention.
Surgery: Annuloplasty, plication or, rarely, replacement. Repair of the valve only in very severe tricuspid regurgitation, when the required doses of diuretics are large enough to cause metabolic consequences. Surgical removal of the valve may be required to eradicate the source of infection in IV drug users with infective endocarditis.

COMPLICATIONS Heart failure, hepatic fibrosis.

PROGNOSIS Prognosis varies depending on the underlying cause.