Basic science

Anatomy

The primary function of the heart is to pump deoxygenated blood to the lungs and to return oxygenated blood to the rest of the body. The basic anatomy consists of:

- Pericardium (visceral and parietal): the fibrous sac containing the heart.
- Four cardiac chambers: the right and left atria and ventricles.
- Heart valves:
  - Two outflow valves: the aortic and pulmonary valves consist of three semi-lunar cusps.
  - Two atrioventricular (AV) valves: the mitral and tricuspid valves, which are attached by chordae tendinae to papillary muscles.
- Vascular system:
  - Great vessels: the pulmonary artery, pulmonary vein and aorta.
  - Three main coronary arteries: the left anterior descending (LAD) and circumflex (Cx) arteries, which originate from the left main stem (LMS) and the right coronary artery (RCA).
  - Venous system: the venous blood is drained via the great cardiac vein, small anterior cardiac vein and thesbian veins.
- Electrical conducting system, which consists of specialised cells that are able to depolarise spontaneously (*automaticity*) forming:
  - The sinoatrial (SA) node.
  - The atrioventricular node.
  - The Bundle of His (right and left) and terminal Purkinje fibres.

The foetal heart

A knowledge of basic cardiac embryology is helpful for understanding how lesions found in adult congenital heart disease develop.

Foetal atria and ventricles (Figure A)

The heart begins life as a primitive tube, which folds to produce early cardiac chambers: the sinus venosus, the primitive atrium, the ventricle and the bulbus cordis. Further separation of the chambers occurs as follows:

- A pair of septa, the *septum primum* and *septum secundum*, grow to separate the right and left atria. The septum primum fuses with the endocardial cushions, the septum secundum does not. The free edge of the septum primum and secundum form the *foramen ovale*.
- A muscular interventricular septum grows from the floor of the common ventricle to divide it into two chambers.

Foetal shunts (Figure B)

The lungs are bypassed in the foetal circulation by the following right to left shunts:

- *Foramen ovale:* oxygenated blood passes from the left umbilical vein to the right atrium via the *ductus venosus*. From the right atrium the blood is then shunted through the foramen ovale to the left atrium.
- *Ductus arteriosus:* the remaining oxygenated blood passes from the right atrium to the right ventricle and enters the pulmonary trunk. From here it passes via the ductus arteriosus directly to the aorta, bypassing the lungs.

Circulation changes at birth

As the newborn takes its first breath, the pulmonary vascular resistance drops and conversion from the foetal to adult circulation starts. The following changes occur:

- The *foramen ovale* closes by the mechanical effect of the reversal in pressure between the two atria, and forms the *fossa ovalis* in adult life.
- Changes in oxygen concentration of the blood and hormonal changes contribute to the closure of the *ductus arteriosus*.

Cardiology: Clinical Cases Uncovered. By T. Betts, J. Dwight and S. Bull. Published 2010 by Blackwell Publishing.

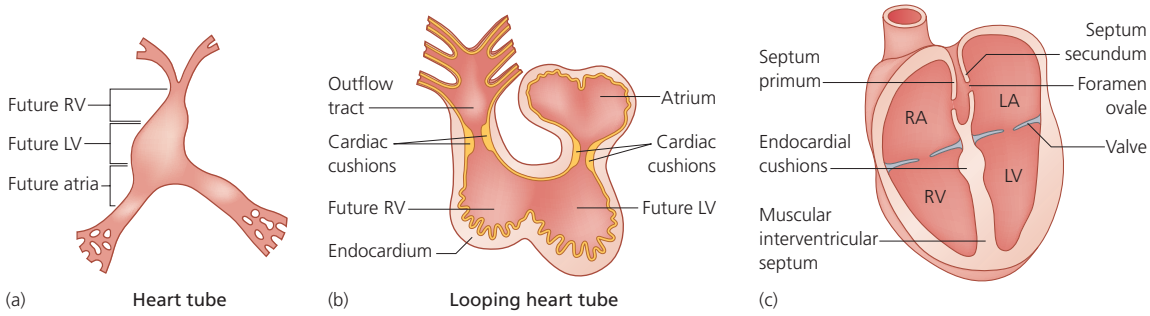


Figure A Development of the heart. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

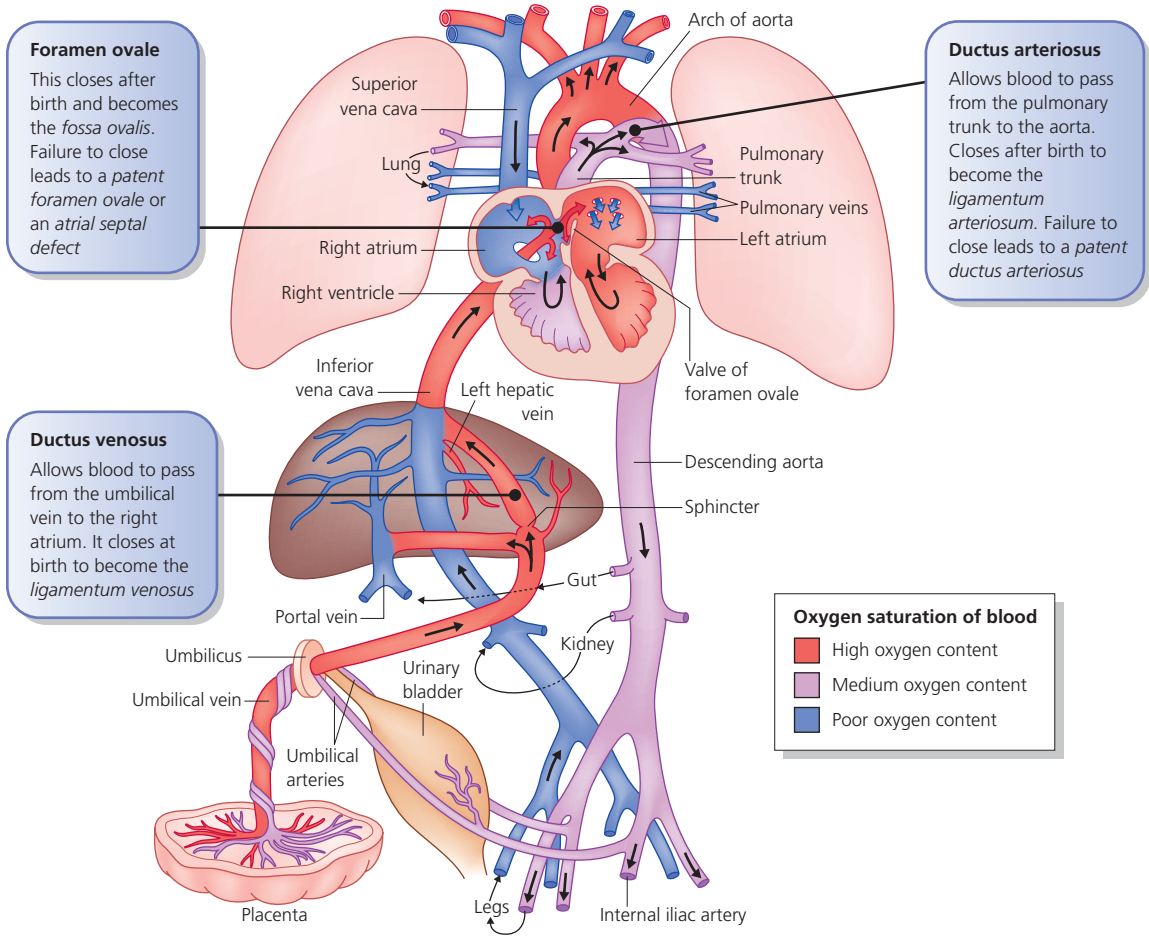


Figure B Foetal circulation and shunts.

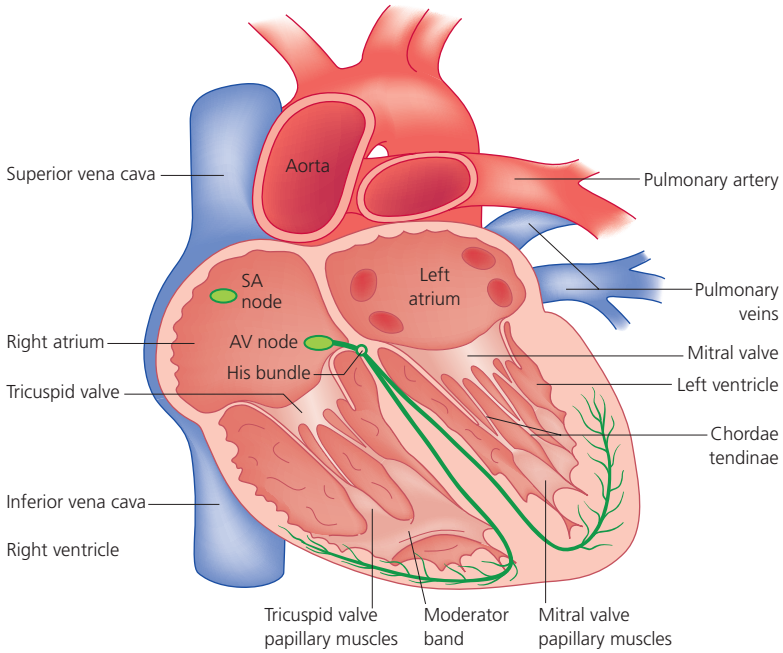


Figure C Adult heart. AV, atrioventricular; SA, sinoatrial.

The adult heart

Right atrium

This chamber is a low-pressure (0–7 mmHg), thin-walled receiving chamber for systemic and cardiac venous blood. It also contains the 'pacemaker' (SA node) and the AV node of the heart.

Right ventricle

This chamber receives the venous blood from the right atrium and ejects it into the pulmonary artery. Unlike the left ventricle, it is heavily trabeculated. It contains the moderator band, which contains part of the conduction system, and the papillary muscles of the tricuspid valve. The pressure in this chamber is 15–30 mmHg during systole.

Left atrium

This chamber receives oxygenated blood from the pulmonary veins. Clinically important structures are:

- *Pulmonary veins:* in normal hearts four pulmonary veins (two upper and two lower) drain oxygenated blood from the lungs into the left atrium.
- *Left atrial appendage:* a blind-ending sac related to the left atrium and a common site for thrombus formation in patients with atrial fibrillation.

The pressure in this chamber is slightly higher than in the right atrium (4–12 mmHg).

Left ventricle

This is a high-pressure (90–140 mmHg), thick-walled chamber, which reflects its greater contractile performance. It delivers oxygenated blood systemically. It contains the *mitral valve papillary muscles*. These are conical muscular projections from the walls of the left ventricle that attach to the chordae tendinae to support the two cusps of the mitral valve.

Vascular anatomy (Figure D)

Great vessels

- *Superior and inferior vena cava*: drain systemic deoxygenated venous blood into the right atrium.
- *Pulmonary artery:* carries *deoxygenated* blood to the lungs from the right ventricle. It has thinner walls than systemic arteries and subdivides many times into branches that carry blood to the network of 280 billion capillaries where it is oxygenated.

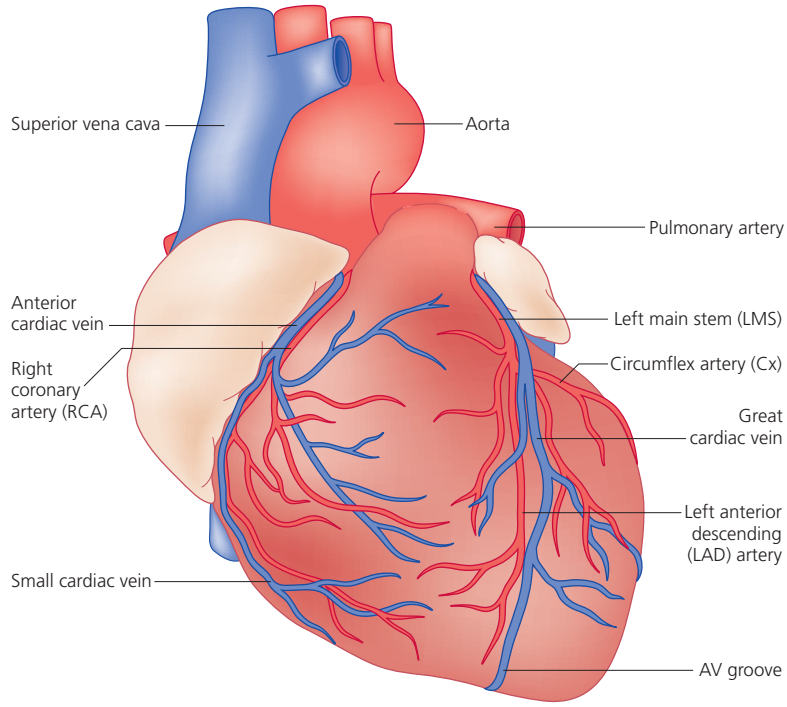


Figure D Vascular anatomy.

Box A Clinical reasons to know cardiac embryology

- A *patent foramen ovale (PFO)* is found in up to 20% of the population. The majority of people with a PFO have no symptoms. In some patients emboli form in the venous circulation and pass via the patent foramen into the systemic circulation, causing a stroke. (This is known as paradoxical embolus.) In such patients and some other selected groups, closure of the PFO is recommended. This can be done percutaneously.
- Failure of the *ductus arteriosus* to close after birth leads to the congenital heart defect *patent ductus arteriosus (PDA).* Surgical or percutaneous duct closure is recommended.
- Failure of the *interventricular septum* to fuse with the endocardial cushions gives rise to a *ventricular septal defect,* one of the most common congenital abnormalities.
- Failure of the atrial septum primum and septum secundum to fuse gives rise to the congenital defect known as *atrial septal defect.*

- *Pulmonary veins:* there are four draining oxygenated blood from the lungs into the left atrium.
- *Aorta:* carries oxygenated blood from the left ventricle to supply the rest of the body.

Arteries

Three main coronary arteries supply blood to the myocardium and arise from the sinuses of Valsalva above the semi-lunar cusps of the aortic valve. These are the RCA, the LAD and the Cx artery. The latter two arteries arise from the LMS.

- The RCA:
  - Arises from above the anterior cusp of the aortic valve.
  - Runs down the AV groove.
  - Supplies the *SA node,* the *AV node* and *right ventricle.*
  - Is 'dominant' in 85–90% of the population. It is called a 'right dominant system' when it gives rise to the *posterior descending artery* to supply the *inferior wall of the left ventricle* and *the inferior third of the interventricular septum.*

- The LMS arises from the left coronary cusp and bifurcates into the LAD and Cx coronary arteries.
- The LAD:
  - Arises from the LMS.
  - Runs down the AV groove.
  - Supplies the *anteroapical* aspects of the *left ventricle*, septum and part of the lateral wall.
- The Cx artery:
  - Arises from the LMS.
  - Runs down the posterior AV groove.
  - Supplies the *posterolateral* aspect of the *left ventricle*.
  - Gives rise to the posterior descending artery in 10–15% of patients (known as a 'left dominant' system).

Box B Clinical reasons to know the vascular system

Coronary arteries

To understand the main infarct sites, associated complications and prognosis of myocardial infarction:

- Acute occlusion of the LAD causes an anterolateral and anteroseptal territory infarct, which may result in extensive left ventricular impairment and increased morbidity and long-term mortality.
- Acute occlusion of the Cx causes a posterolateral territory infarct in a non-left dominant system.
- Acute occlusion of the RCA causes infarction of the inferior wall of the left and right ventricle and can lead to complete heart block because it supplies the SA and AV node.

Cardiac veins

- Great cardiac vein: drains blood from the left ventricle into the right atrium via the coronary sinus.
- Small anterior cardiac vein: drains blood from the right ventricle into the right atrium.
- Thebesian veins: drain remaining blood directly into the cardiac chambers.

Valve anatomy

The normal valve anatomy is demonstrated in Figure E.

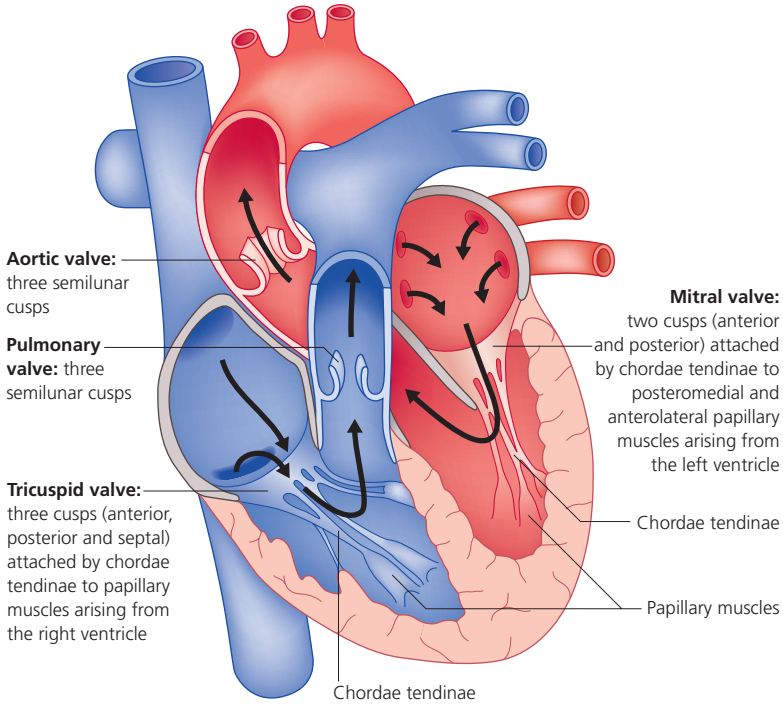


Figure E Valve anatomy.

Box C Clinical reasons to know valve structure and function

- 'Bicuspid' aortic valves have only two semi-lunar cusps and occur in 2% of the population. They are associated with coarctation of the aorta and can lead to development of early aortic stenosis (AS) or aortic regurgitation (AR).
- Dilatation of the aortic root and valve annulus can lead to AR due to failure of the aortic leaflets to coapt.
- The mitral valve may fail very suddenly if there is rupture of papillary muscle or chordae tendinae tethering the valve cusps (e.g. following an inferior or anterior myocardial infarction). This situation can be fatal.
- 'Functional' tricuspid regurgitation (TR) or mitral regurgitation (MR) occurs when there is dilatation of the right or left heart, respectively, resulting in failure of the valve leaflets to coapt due to stretching of the valve annulus.
- Valve lesions are a common cause of heart murmurs and can give rise to endocarditis.

Electrical conduction system anatomy (Figure F)
Specialised cardiac myocytes make up the cardiac electrical conducting system. It consists of the:

- SA node: this forms the 'pacemaker' of the heart and generates the electrical impulse. It consists of a collection of specialised cardiomyocytes in the right atrium with 'automaticity' (the ability to depolarise spontaneously and faster than other conducting tissue in the heart).
- AV node: located at the base of the right atrium, the AV node transmits the electrical impulse from the atria to the ventricles.
- The bundle of His descends from the AV node down the membranous interventricular septum. It is the only electrical connection between the atria and ventricles. It divides into left and right bundle branches.
- The bundle branches are specialised conducting fibres that conduct the impulse rapidly into the ventricular myocardium. The right bundle is a discrete structure. The left bundle further divides into:

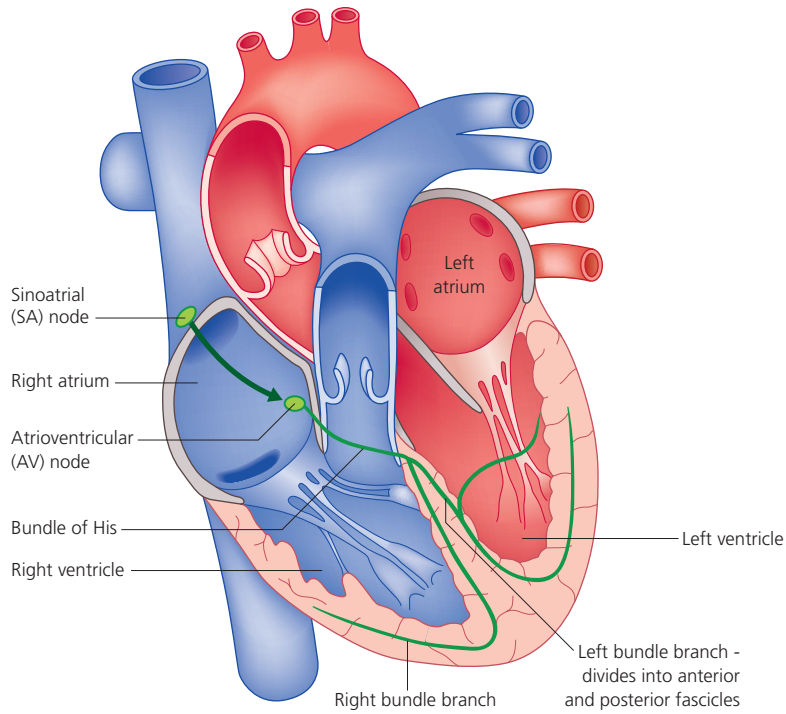


Figure F Electrical conduction system.

Box D Clinical reasons to know the conduction system

- Disease of the conduction system may result in changes in the ECG.
- Symptoms and prognosis of conducting tissue disease are determined by the level at which the conduction system is affected.
- The requirement for permanent and temporary pacing can be assessed from a knowledge of the severity of conducting tissue disease.

- A smaller anterior and larger posterior fascicle.
- Purkinje fibres: these distribute the impulse to the myocardial tissue.

Physiology

Cardiomyocytes

The myocardium is composed of specialised cardiac cells called *cardiomyocytes* that are characterised by:

- Electrical conduction: they are connected to each other via an *intercalated disc* containing *gap junctions*, which allow electrical conduction to neighbouring cells.
- Contraction: they can contract because of special contractile proteins that are arranged in a structural unit called a *sarcomere*, which consist of interlocking thin filaments (tropomysin) and thick filaments (myosin molecules).

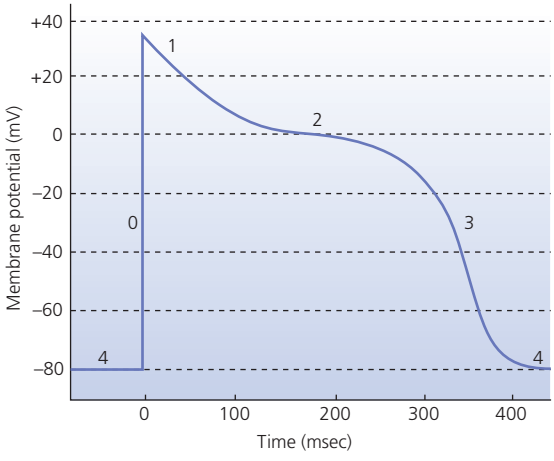


Figure G Action potential.

- Excitation–contraction coupling: describes the process by which an action potential triggers a cardiomyocyte to contract. The process is divided into the following phases:
  - Phase 0: depolarisation of the cell membrane caused by an increase in sodium channel conductance.
  - Phase 1: repolarisation caused by opening of potassium channels.
  - Phase 2: calcium influx delays repolarisation. Calcium binding causes sarcomere shortening.
  - Phase 3: repolarisation.
  - Phase 4: return to resting membrane potential.

KEY POINT

In heart failure there is decreased calcium influx into the cell and decreased affinity of the sarcomere proteins to bind calcium. This leads to reduced sarcomere shortening, impaired excitation–contraction coupling and a reduction in the force and effectiveness of the cardiac contraction in systole.

Cardiac cycle (Figure H)

The four phases of the cardiac cycle are:

- *Phase I: Isovolumetric contraction*. Tricuspid and mitral valves close as the ventricles contract. The aortic valve opens when left ventricular pressure exceeds aortic pressure.
- *Phase II: Ventricular ejection*. The aortic valve opens and blood is expelled into the aorta. The left ventricle begins to relax at the end of the T wave and the aortic valve closes after pressure falls.
- *Phase III: Isovolumetric relaxation*. All four cardiac valves are closed and the left ventricular pressure continues to fall.
- *Phase IV: Ventricular filling*. This occurs in two phases: 'rapid passive ventricular filling' happens when the mitral valve opens and 'active ventricular filling' happens at the end of diastole when the atrium contracts.

Cardiac output

Cardiac output (L/min) is the product of stroke volume (L) and heart rate (bpm).

Cardiac output = stroke volume × heart rate.

The normal value in an average healthy human at rest is around 5 L/min. During exercise, cardiac output increases 4-to-6-fold.

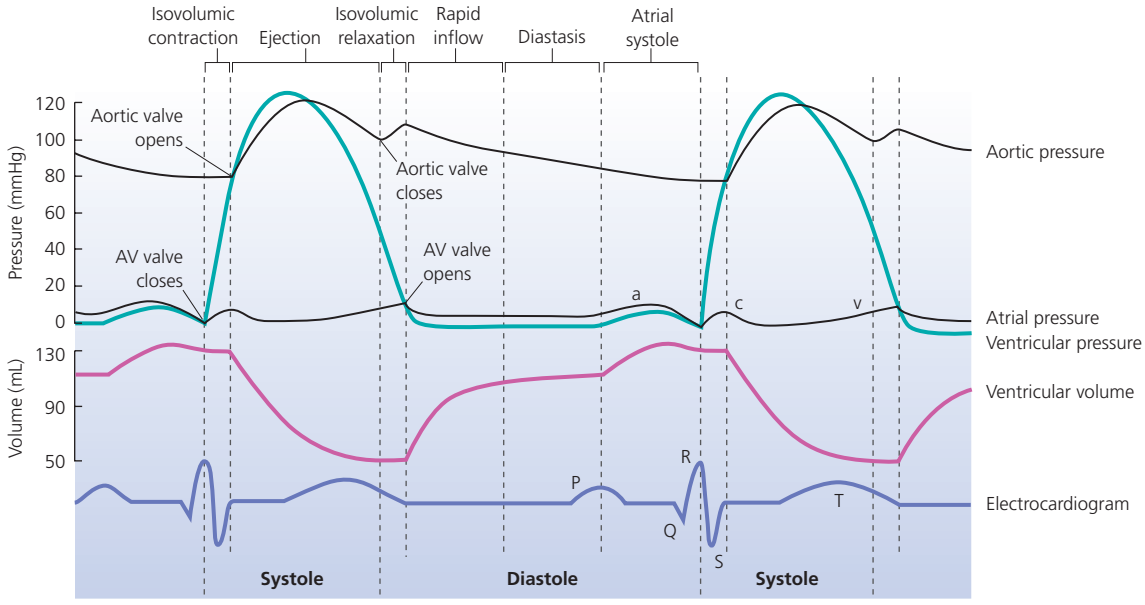


Figure H Cardiac cycle. AV, atrioventricular.

Cardiac output is influenced by:

- Preload: the filling pressure.
- Myocardial contractility.
- Afterload: this is also known as 'systemic vascular resistance'. This is the resistance to ejection of blood from the left ventricle. The majority of the resistance to flow comes from the small arterioles.

KEY POINT

Cardiac output can be measured invasively using a pulmonary catheter, thermodilution or, non-invasively, using an ultrasound Doppler probe. It is used to assess patients for cardiac transplantation and to monitor patients in cardiogenic shock.

Pathological processes affecting the cardiovascular system

Coronary artery disease

Coronary artery disease is one of the most common causes of death in the developed world. The atheromatous plaque underlines the pathophysiology of ischaemic heart disease (Figure I). Risk factors for development of atherosclerotic disease are described in Chapter 2.

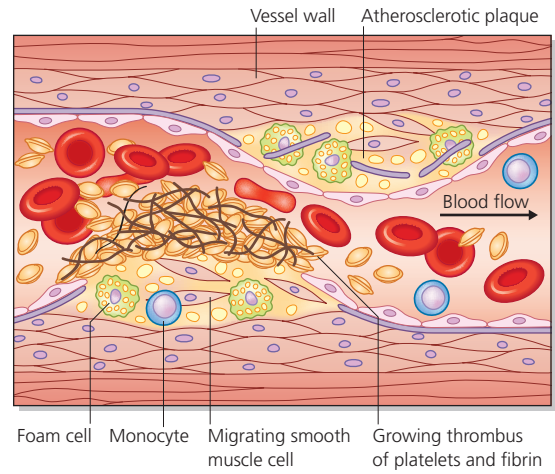


Figure I The atherosclerotic plaque.

Atherosclerotic plaque formation (Figure J)

This is a process of underlying and ongoing chronic inflammation:

- The process is triggered by lipid deposition and vascular wall injury, which leads to inflammation.
- Monocytes and leucocytes are recruited to the area of inflammation.
- 'Fatty streaks' are formed when inflammatory cells accumulate oxidised lipids to form macrophages and foam cells.

• Vascular smooth muscle cells are recruited to form a fibrous cap.
• The plaque expands further with ongoing inflammation and lipid deposition and encroaches on the vascular lumen.

Atherothrombosis formation

• Erosion or rupture of the fibrous cap overlying the atherosclerotic lesion may lead to thrombous formation.
• Thrombogenic material within the plaque is exposed, causing accumulation of platelets and formation of a thrombous within the vessel.
• The clinical course of the patient is largely determined by the nature and location of the thrombus formed on the atherosclerotic plaque.

Disruption of the atherosclerotic plaque

Erosion or rupture of the atherosclerotic plaque can result in three possible outcomes:

1. An acute coronary syndrome (ACS), where thrombus forms on the lesion causing obstruction to flow:
   a. Complete obstruction of the lumen by thrombus is associated with ST elevation on the ECG and characteristically gives rise to an ST elevation myocardial infarction (STEMI).
   b. Partial obstruction of the lumen by thrombus is associated with non-ST elevation ECG changes and either unstable angina or a non-ST elevation MI (non-STEMI).

The extent of myocardial damage is dependent upon the duration of occlusion of the infarct related vessel.
2. Resolution and healing of the plaque.
3. Plaque progression causing further occlusion of the vascular lumen and worsening of angina.

The reasons why some plaques rupture and others do not remain unclear.

Progression of the atherosclerotic plaque

When symptomatic this is associated with the development of angina. This is the syndrome of ischaemic chest pain occurring on exercise, which is associated with a mismatch between myocardial oxygen demand and supply.

Cardiac markers

Myocardial cell death can be recognised by the appearance in the blood of different proteins released into the circulation from the damaged myocytes. Cardiac

Box E Clinical situations: impact on cardiac output

Compensated

- *Sinus bradycardia*: cardiac output is maintained because the stroke volume is increased. There is a rise in pre-load (lower heart rate leads to increased filling time) and thus stroke volume is increased via the Frank-Starling relationship.
- *Rise in blood pressure*: the rise in systemic vascular resistance will initially reduce stroke volume. However the pre-load is increased because of incomplete left ventricular ejection leading to normalisation of cardiac output. These mechanisms maintain homeostasis.
- *Exercise and pregnancy:* cardiac output is increased in response to demand via an increase in venous return, stroke volume and heart rate.

Decompensated

High cardiac output states

These patients tend to have warm peripheries and a bounding pulse. Causes include:

- Anaemia.
- Thyrotoxicosis.
- Paget's disease.
- Sepsis.

Low cardiac output states

These patients have cool peripheries, low-volume pulse, prolonged capillary refill time and are hypotensive. Causes include:

- Hypovolaemia.
- Complete heart block.
- Tachyarrhythmia causing haemodynamic compromise.
- Poor left or right ventricular systolic function (e.g. ischaemic or dilated cardiomyopathy).
- Cardiac tamponade/ constrictive pericarditis
- Aortic stenosis (AS)
- Cardiogenic shock*

This carries a high mortality rate of 80% with treatment. It normally occurs due to pump failure following a massive myocardial infarction. The definition of cardiogenic shock is: 'a state of hypotension (with systolic blood pressure <90 mmHg) with reduced end-organ perfusion due to low cardiac output'.*

KEY POINT

STEMI is a medical emergency and requires prompt treatment with reperfusion therapy. Prompt primary percutaneous coronary intervention is the gold standard treatment for STEMI.

troponin is the preferred marker used to measure myocardial necrosis because of superior specificity to other cardiac markers. It has the following properties:

- Exists as a contractile protein, mainly bound as part of the actin/myosin complex.
- Exists in three forms, of which T and I are used clinically.
- Highly sensitive and specific.
- Measurement 12 hours after onset of pain has 100% sensitivity for myocardial infarction.
- Levels rise within 12 hours of myocardial injury, peak at 24 hours and remain elevated for up to 14 days.
- Other less-specific markers of myocardial injury that can be measured include myoglobin, creatine kinase and lactate dehydrogenase.
- Myocardial necrosis and elevated troponin levels can occur for reasons other than myocardial infarction.

Box F Myocardial infarction: definition and causes

The European Society of Cardiology definition of myocardial infarction is as follows: detection of a *rise* and *fall* of cardiac biomarkers (preferably troponin) *together* with evidence of myocardial ischaemia with at least one of the following:

- Symptoms of ischaemia.
- ECG changes indicative of new ischaemia (new ST changes, LBBB).
- Development of pathological Q waves on ECG.
- Imaging evidence of new loss of viable myocardium or new regional wall motion abnormality.

Atherosclerotic thrombosis causes the majority of myocardial infarcts. Other causes of myocardial infarction include:

- Embolus (vegetation from infective endocarditis).
- Spontaneous coronary artery dissection.
- Intense spasm of the coronary arteries (e.g. cocaine).
- Trauma.
- Aortic dissection.
- Iatrogenic due to coronary intervention.

Pathology affecting the great vessels

Aortic dissection

Aortic dissection is a tear in the aortic intima through which blood enters and strips the media from the adventitia. Anterograde blood flow may cause the dissection to extend down the length of the aorta affecting the coronary, renal or femoral arteries.

Prognosis and management depend on the location of the tear.

Type A dissections (arising in the ascending aorta)

- Are a medical emergency and require immediate surgery.
- Carry a high mortality rate.
- May cause aortic root dilatation, AR, pericardial effusions or myocardial infarction (if the dissection extends to the coronary arteries).

Type B dissections (arising in the descending aorta)

- Carry a lower mortality rate than type A dissections and can be managed medically.
- May cause symptoms due to vascular compromise of other areas (e.g. acute limb ischaemia – iliac vessels; renal failure – renal ischaemia; paraplegia – spinal artery occlusion; and abdominal pain – mesenteric ischaemia) as the dissection extends.

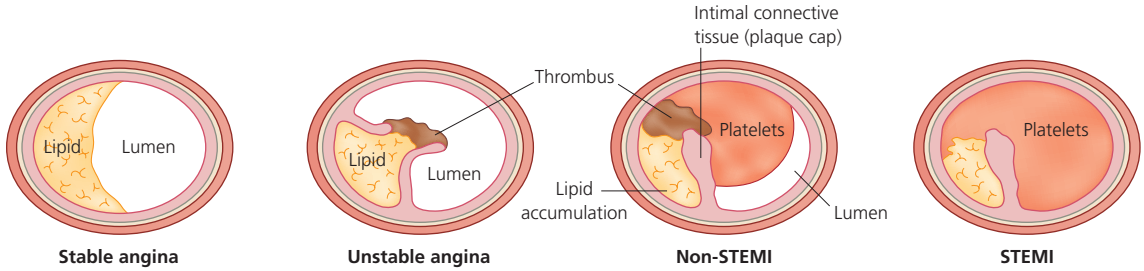
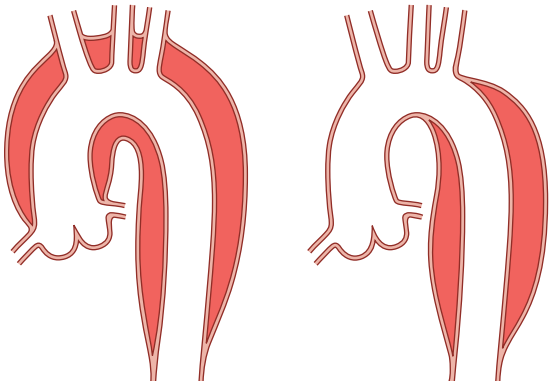


Figure J The fate of the atherosclerotic plaque. STEMI, ST-elevation myocardial infarction.



Type A aortic dissection Type B aortic dissection

Figure K Aortic dissection classification.

KEY POINT

Emergency surgery is essential for type A dissections, which carry a mortality rate of up to 5% per hour.

Pulmonary embolism

- Is commonly caused by venous thromboembolism to the pulmonary artery.
- Most frequently occurs secondary to deep vein thrombosis.
- Air, fat, amniotic fluid and tumour fragments in the pulmonary artery are also possible embolic causes.
- Common risk factors for development of pulmonary embolism include major surgery, lower limb fracture, malignancy, prolonged hospitalisation and previous thromboembolism.
- Patients may present with chest pain, shortness of breath, haemoptysis or (in the case of a massive pulmonary embolism) cardiac arrest.
- Prognosis depends on the size and underlying cause of the pulmonary embolism.

KEY POINT

Patients presenting with pulmonary venous thromboembolism with no risk factors may have an underlying thrombophilia or malignancy.

Pulmonary hypertension

- Pulmonary hypertension is defined as *an increase in pulmonary arterial pressure >25 mmHg at rest or >30 mmHg with exercise.*
- Primary pulmonary hypertension is the diagnosis in patients with pulmonary arterial hypertension of unexplained aetiology.
- Secondary hypertension can be due to a number of causes, including multiple pulmonary emboli, connective tissue disease, congenital AV shunts and chronic left ventricular failure.
- Patients present with shortness of breath and symptoms of right-sided heart failure.
- Prognosis for primary pulmonary hypertension without treatment is very poor.

Congenital heart disease

Congenital heart disease is uncommon, affecting less than 1% of live births. However, increased numbers of patients survive into adulthood.

Atrial septal defects (ASD)

- ASD is a common congenital defect, more frequent in females.
- Direct communication between the atria, results in shunting of blood from left to right.
- Many patients with ASD are asymptomatic but symptomatic patients present with shortness of breath, stroke or heart failure.
- Larger ASDs associated with major shunting should be closed surgically or percutaneously.
- If the ASD is large and remains untreated, Eisenmenger's syndrome may develop (see p. 12).

Ventricular septal defect (VSD)

- Most common congenital abnormality.
- Incomplete separation of the ventricles allows blood flow between the ventricles (usually left to right).
- The direction of the shunt is determined by the size of the VSD and pulmonary vascular resistance. Large shunts can lead to heart failure and pulmonary hypertension causing reversal of shunt from right to left (Eisenmenger's syndrome).
- Patients with a small VSD may be asymptomatic and can be managed conservatively.
- Larger VSDs may require surgical repair.

Coarctation

- Coarctation is narrowing of the aorta in the region of the ligamentum arteriosum most commonly distal to the left subclavian artery.

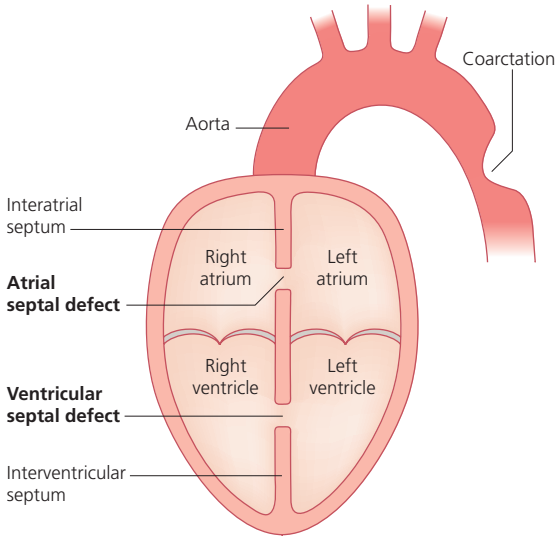


Figure L Congenital heart defects.

- 50% of patients with coarctation have a bicuspid aortic valve.
- Patients present with hypertension, radiofemoral delay, unequal upper limb pulses and a continuous murmur in the interscapular region.
- Surgical repair or stenting may be required if there is a significant gradient across the coarctation (>20 mmHg).

Cyanotic congenital heart disease

Cyanotic congenital heart disease is characterised by venous blood entering directly into the systemic circulation.

Eisenmenger's syndrome

- Eisenmenger's syndrome is a pathophysiological condition resulting from adult congenital heart disease. VSD, ASD and patent ductus arteriosus (PDA) are responsible for 80% of cases of Eisenmenger's syndrome.
- Uncorrected left-to-right shunting leads to the development of pulmonary hypertension.
- When pulmonary hypertension develops, the right-sided pressures in the heart exceed systemic pressure and cause *reversal* of the shunt.
- Deoxygenated venous blood is thus shunted from the right to the left side of the heart and enters the systemic circulation, causing the patient to develop chronic cyanosis and clubbing.
- The shunt reversal and resulting clinical consequences are known as Eisenmenger's syndrome.

Transposition of the great arteries (Figure M)

- The aorta arises from the morphological right ventricle and the pulmonary artery arises from the morphological left ventricle.
- The majority of patients have an associated PDA (physiologically corrected transposition).
- Presents at birth as a profoundly cyanotic baby.
- Immediate surgical intervention is required shortly after birth.

Patent ductus arteriosus (PDA) (Figure N)

Normally, the PDA closes after birth under hormonal influences.

- Failure of the PDA to close leads to persistent left-to-right shunting between the aorta and pulmonary artery.
- Long-term left-to-right shunting leads to increased blood flow to the pulmonary circulation, which can lead to pulmonary hypertension and Eisenmenger's syndrome.
- Closure of PDA (surgically or percutaneously) is recommended in almost all cases.

Tetralogy of Fallot (Figure O)

Is the most common cause of cyanosis in infancy after the first year of life and the long-term outcome without surgical intervention is poor. The four features of tetralogy are:

1. Overriding aorta.
2. Ventricular septal defect.
3. Pulmonary stenosis.
4. Right ventricular hypertrophy.

Pathology of the conduction system

Arrhythmias

Arrhythmia is a disturbance of the electrical rhythm of the heart. It may be described using the following terms:

- Heart rate:
  - *Bradycardia*: slow (heart rate <60 bpm).
  - *Tachycardia*: fast (heart rate >100 bpm).
- Anatomy:
  - *Supraventricular*: arises above the ventricles.
  - *Ventricular tachycardia*: arises from the ventricles.
- Time course:
  - *Paroxysmal*: happens intermittently and stops spontaneously.
  - *Persistent*: does not stop spontaneously but normal sinus rhythm can be restored with some form

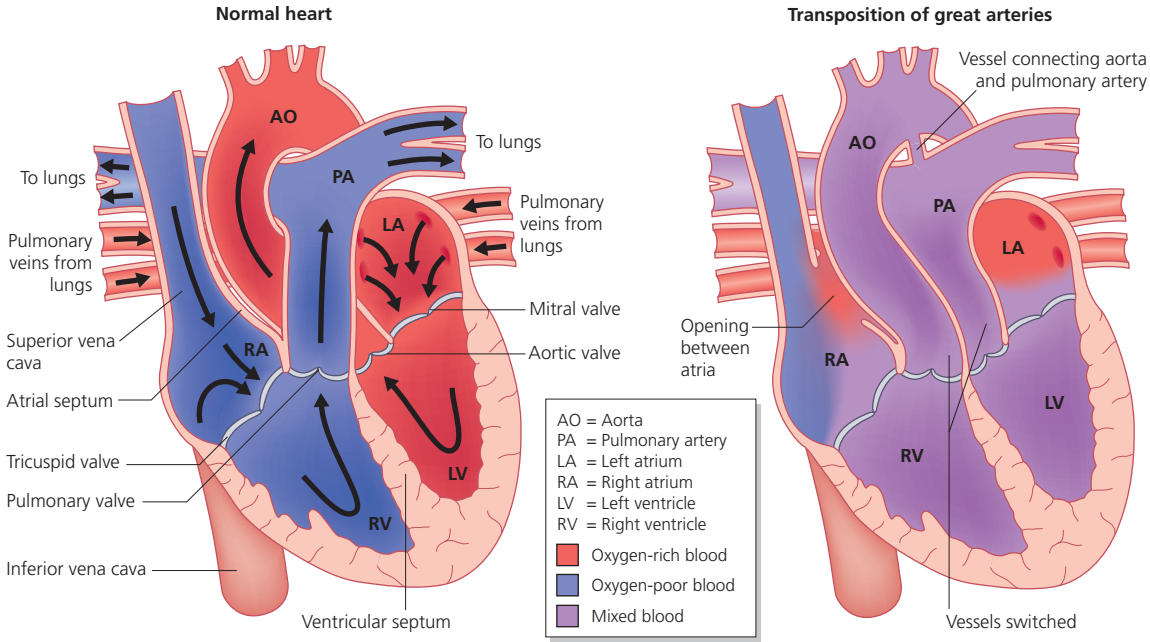


Figure M Transposition of the great arteries.

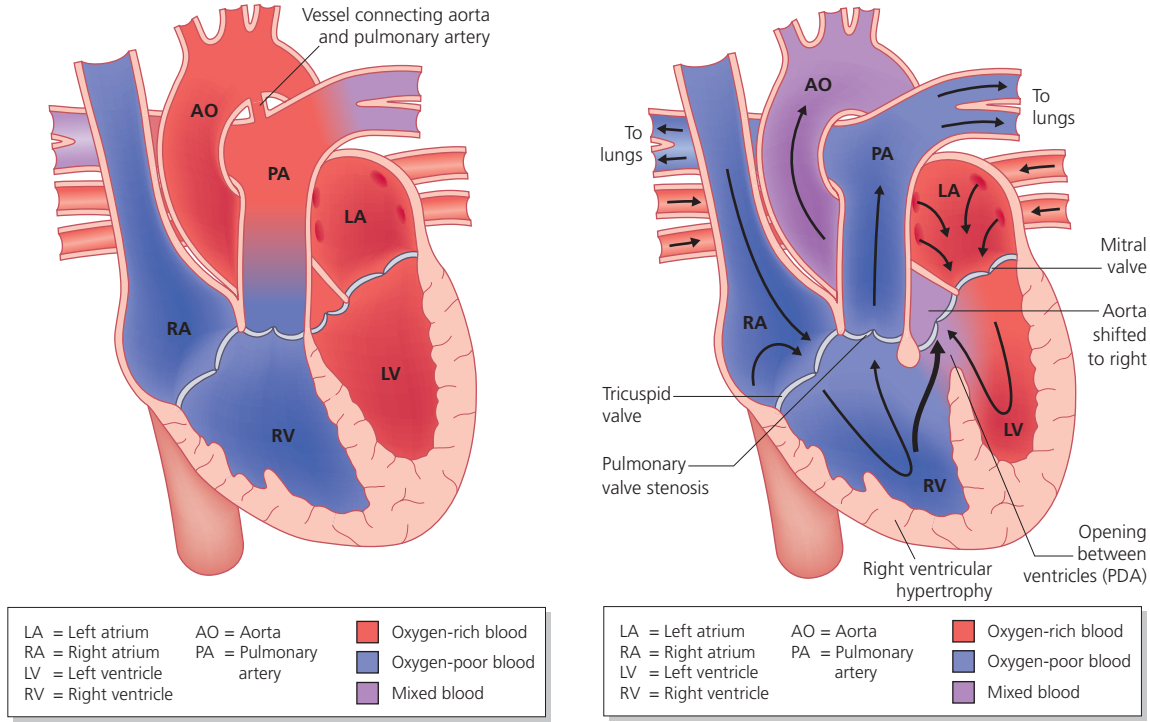


Figure N Patent ductus arteriosus (PDA).

Figure O Tetralogy of Fallot.

of treatment (drugs or direct current [DC] cardioversion).
  - *Permanent*: ongoing and sinus rhythm cannot be restored.
- Width of QRS complex on the surface ECG:
  - Narrow-complex tachycardias (QRS width ≤120 msec; 3 small squares).
  - Broad-complex tachycardias (QRS width >120 msec; 3 small squares).

Bradycardias

Mechanism

- Failure of electrical impulse generation from the SA node.
- Failure of electrical impulse conduction through the heart via AV node and Bundle of His.

Causes (of conduction disease)

- Degenerative.
- Ischaemia, e.g. inferior myocardial infarction (the RCA supplies SA and AV nodes).
- Infiltrative: cardiac disease that cause infiltration of the conducting system (e.g. amyloid and sarcoidosis).
- Cardiac surgery.
- Antiarrhythmic drugs.

Presentation

- Blackouts.
- Breathlessness.
- Fatigue.
- Incidental finding.

Classification

- Sick sinus syndrome:
  - Incidence: common in the elderly.
  - Pathology: impaired SA node function. The SA node fails to generate and electrical impulse.
  - Symptoms: variable, ranging from no symptoms to blackouts.
  - ECG: sinus pauses.
  - Treatment: atrial pacing if symptomatic.
- First-degree AV block:
  - Incidence: common in the elderly.
  - Pathology: the AV node conducts sinus impulses more slowly to the ventricles.
  - Symptoms: most patients are asymptomatic.
  - ECG: shows a prolonged PR interval (>200 msec; 5 small squares).
  - Treatment: no treatment required unless associated with higher degrees of block.
- Second-degree AV block (Wenckebach or Mobitz type I):
  - Pathology: the AV node fatigues and conducts each successive impulse progressively more slowly until a beat is dropped.
  - Symptoms: most patients are asymptomatic. Some may complain of 'skipped beats'.
  - ECG: PR interval prolongs until an impulse fails to conduct to the ventricles and the node recovers.
  - Treatment: no treatment required unless associated with higher degrees of block.
- Second-degree AV block (Mobitz type II):
  - Pathology: intermittent failure of impulses to conduct from the AV node to the ventricles via the Bundle of His.
  - Symptoms: dizziness, breathlessness and syncope.
  - ECG: QRS complexes are dropped on a regular basis, e.g. 2:1, 3:1, etc.
  - Treatment: pacing indicated.
- Third-degree AV block (complete heart block):
  - Pathology: complete failure of AV nodal conduction.
  - Symptoms: syncope, breathlessness, dizziness.
  - ECG: complete dissociation between QRS complexes and P waves.
  - Treatment: pacing indicated.
- His–Purkinje disease:
  - Pathology: failure of conduction through the His–Purkinje system distal to the AV node. Conduction may fail through the right or left bundle branches or through the left anterior (associated with left-axis deviation) or posterior fascicles (associated with right-axis deviation).
  - ECG changes: variable. Right bundle branch block (RBBB), left bundle branch block (LBBB), or trifasicular block (RBBB, left-axis deviation and first-degree heart block)
  - RBBB and LBBB in isolation do not require pacing. Patients with trifasicular block should be considered for pacing.

Tachycardias

The two main mechanisms are:

- Automaticity: cells depolarise spontaneously. This is increased by sympathetic drive and decreased by parasympathetic drive.
- Re-entry (Figure T): occurs when there are two or more pathways of conduction within the heart that have different conduction properties (i.e. where there is an anatomical barrier such as scar tissue or pulmonary veins).

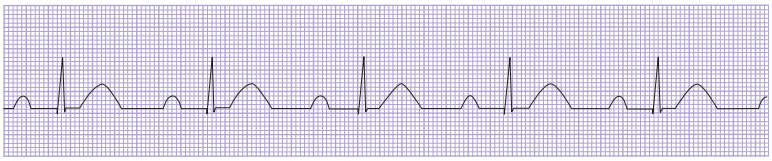


Figure P First-degree atrioventricular (AV) block. PR interval >200 mseconds.

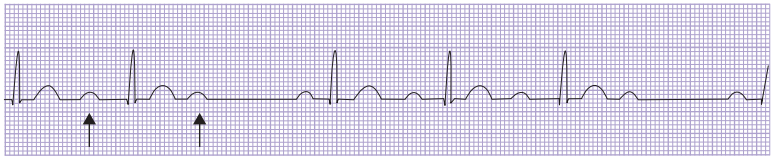


Figure Q Second-degree atrioventricular (AV) block Mobitz I. QRS complex dropped. Gradual prolongation of PR interval.

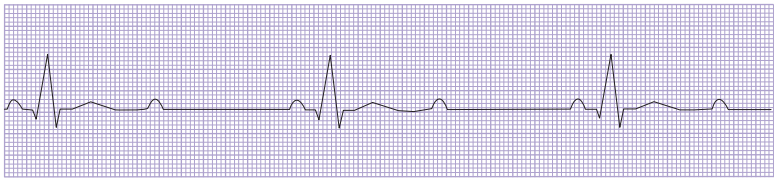


Figure R Second-degree atrioventricular (AV) block Mobitz II.

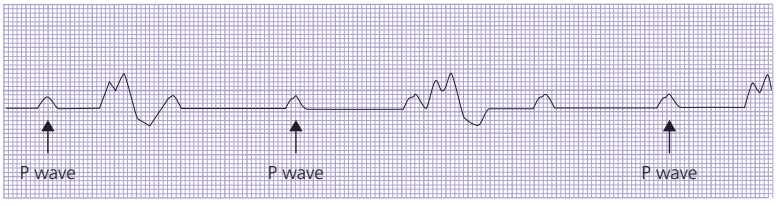


Figure S Third-degree atrioventricular (AV) block. Complete dissociation of P waves and QRS complexes.

• Patients with tachycardias of any description may present with palpitations and shortness of breath. If the tachycardia produces haemodynamic compromise (most commonly found with ventricular tachycardia and ventricular fibrillation) then patients may present with syncope or cardiac arrest.
• Tachycardias can be divided into:
  ○ Narrow complex (QRS ≤120 msec).
  ○ Broad complex (QRS >120 ms).

Narrow-complex tachycardias

Atrial fibrillation (Figure U)

• The most common arrhythmias presenting in hospital medicine and can be persistent, paroxysmal or permanent.
• Caused by *multiple* re-entry circuits in the atria.
• Triggered by ectopic beats originating in the pulmonary veins.
• Associated with enlarged or diseased atria.

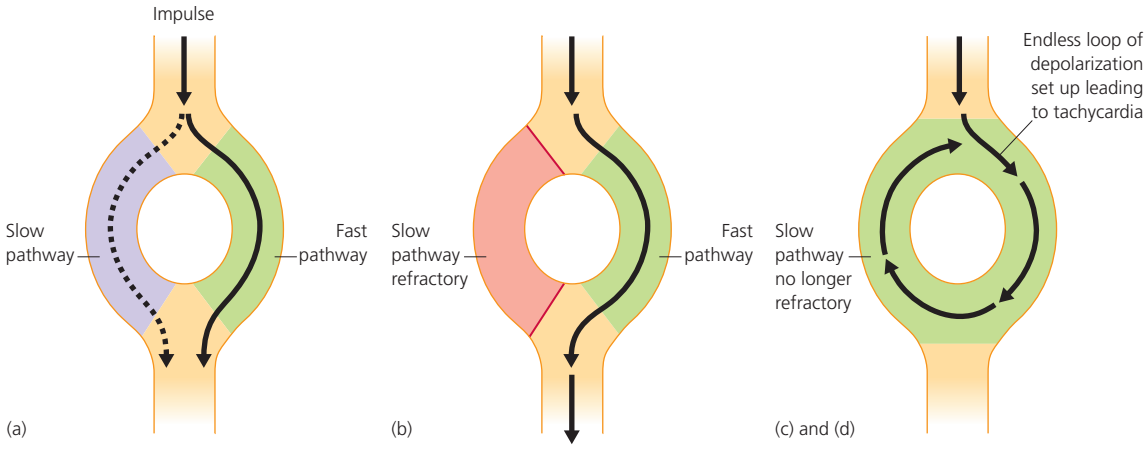


Figure T Tachycardiac re-entry mechanism. A, Conduction happens down both pathways in sinus rhythm. B, Premature early beat conduction may only be conducted down the fast pathway because the slow pathway remains refractory. C, The slow pathway is no longer refractory by the time the wave front reaches the distal aspect of the pathway and so conduction occurs down the slow pathway in the opposite direction. D, The wave front then travels down and re-enters the fast pathway setting up an endless loop of depolarisation and a tachycardia.

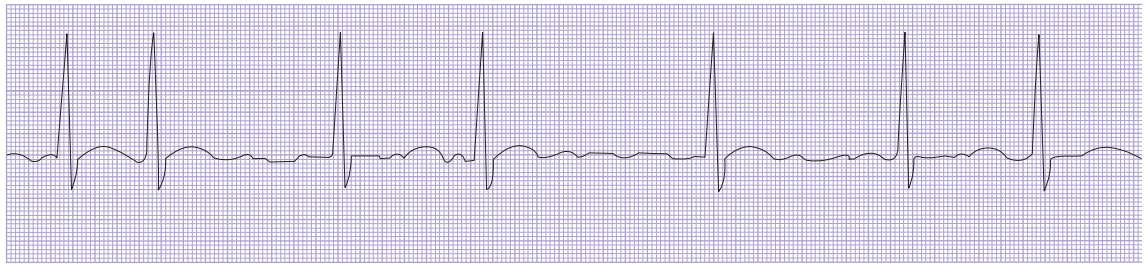


Figure U Atrial fibrillation.

• ECG is characterised by an irregularly irregular ventricular rhythm and by the absence of discrete P waves.

Atrial flutter (Figure V)

• Results from *one single* large re-entry circuit within the *right atrium.*

• Flutter circuit is centred around the tricuspid valve ring and strip of tissue in this vicinity can be targeted with ablation for curative treatment.

• The AV node limits the ventricular response to the atrial flutter. The atria normally depolarize at 300 bpm. Often a 2:1 or 4:1 block occurs, resulting in a ventricular rate of 150 or 75 bpm.

• ECG is characterised by a saw-tooth baseline.

Supraventricular tachycardias (SVTs) (Figure W)

• Can be divided into atrioventricular re-entrant (AVRT) and atrioventricular nodal re-entrant tachycardias (AVNRT).

• AVRT occurs when there is an accessory pathway between the atria and ventricles resulting in a re-entry circuit.

• AVNRT: there are two conduction pathways *within* the AV node and re-entrant tachycardia can be set up *within the node itself.*

Broad-complex tachycardias

Ventricular tachycardias

The underlying pathology is automatic activity or re-entry caused by:

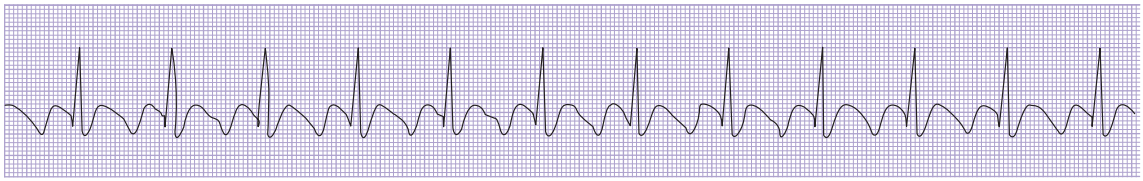


Figure V Atrial flutter. Saw tooth baseline with 2:1 block.

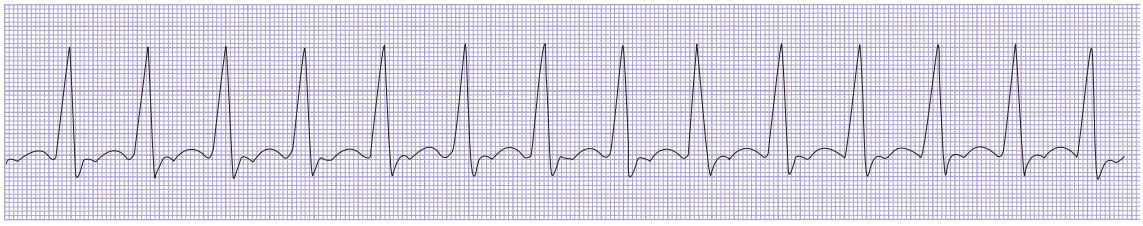


Figure W Supraventricular tachycardia.

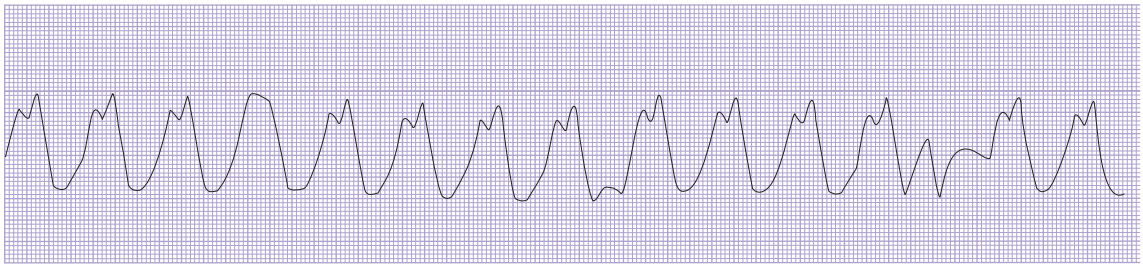


Figure X Ventricular tachycardia.

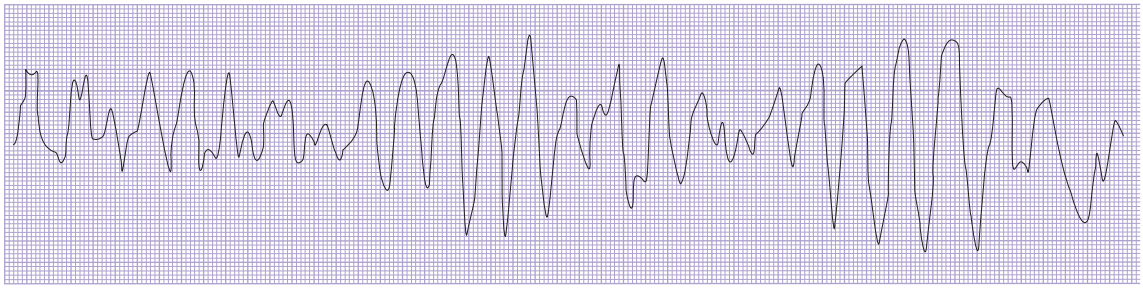


Figure Y Ventricular fibrillation.

- Ischaemic heart disease: re-entrant VT occurs around the infarct scar.
- Cardiomyopathies: can affect the His–Purkinje system and lead to re-entry.
- Normal heart VT: some patients have a structurally normal heart and present with VT during exercise. The mechanism is automatic and catecholamine driven. These patients have a good prognosis.
- Ion-channel defects: mutations affecting genes for cardiac ion channels can lead to unusual forms of VT. Examples include long QT syndrome (LQTS) and Brugada syndrome.
- Ventricular rate may range from 100 to 300 bpm with symptoms ranging from mild chest pain to complete cardiovascular collapse and cardiac arrest.

Ventricular fibrillation

The underlying mechanism is re-entry and automaticity.

- Results when multiple sites in the ventricles fire impulses rapidly in an uncoordinated fashion.
- Common mode of death in patients with ischaemic heart disease.
- Death follows within a few minutes, unless a normal rhythm is restored with immediate defibrillation.

Hypertension

- Hypertension is diagnosed after three successive measurements of a systolic blood pressure >140 mmHg or diastolic blood pressure >90 mmHg.
- Hypertension is a major risk factor for cardiovascular morbidity.
- 95% of cases are described as 'essential hypertension'.
- 5% of cases are described as 'secondary hypertension' and may be reversed with treatment of the underlying condition (see Box G).
- Malignant hypertension occurs in <1% of hypertensive patients (systolic blood pressure >200 mmHg and/or diastolic blood pressure >130 mmHg) together with grade 3 and 4 retinopathy and is a medical emergency with a mortality of 90% in 1 year.
- Long-term consequences of untreated hypertension include end-organs damage leading to heart failure, renal disease, vascular disease and hypertensive retinopathy.

Valvular heart disease

Valvular heart disease is commonly seen in cardiology outpatient clinics. Many patients are asymptomatic and are regularly monitored for signs and symptoms of deterioration. Assessment includes history, examination and echocardiography.

Aortic stenosis (AS)

Epidemiology

- Commonest valve lesion in the UK.

Aetiology

- Degenerative calcific disease is the most frequent cause in the elderly.
- Bicuspid aortic valves.
- Rheumatic valve disease.
- Supravalvular: above the valve (e.g. supravalvular membrane).
- Subvalvular: below the valve (e.g. subvalvular membrane).

Pathophysiology

Stenosis of the aortic valve leads to pressure overload and:

- Left ventricular hypertrophy.
- Left ventricular failure.
- Low output state.

Presentation

- Symptomatic patients may present with chest pain, syncope and shortness of breath; 50% of patients presenting with syncope and AS will die in 3 years without a valve replacement.

Assessment of severity

- Echocardiographic parameters for grading peak AS are:
  - Mild AS: 20–40 mmHg.
  - Moderate AS: 40–60 mmHg.
  - Severe AS: >60 mmHg.

Treatment

- Surgical intervention is recommended for patients with severe AS and symptoms.

Box G Secondary causes of hypertension

- Intrinsic renal disease (glomerulonephritis, polycystic kidneys, polyarteritis nodosa, etc.)
- Renovascular disease (renal artery stenosis).
- Endocrine causes (Cushing's syndrome, Conn's syndrome, phaeochromocytoma, acromegaly, hyperparathyroidism).
- Coarctation of the aorta.

KEY POINT

Percutaneous aortic valve replacement is currently available at some tertiary centres for elderly patients with critical AS who are unable to undergo open heart surgery. Further developments in this area are expected in the future.

Mitral stenosis (MS)

Epidemiology

Incidence of MS has significantly declined in developed countries due to reduction in the major cause – rheumatic fever.

Aetiology

MS may be caused by narrowing of the mitral valve orifice at the:

- Cusps (thicken and calcify).
- Commissures (fuse with the valve cusps but they are still mobile).

Pathophysiology

Narrowing of the mitral valve orifice leads to:

- A rise in left atrial pressures.
- Left atrial dilation.
- Pulmonary hypertension.
- Atrial fibrillation.

Presentation

- Breathlessness, fatigue, atrial fibrillation, peripheral embolism.

Specific medical treatments

- Anticoagulation is very important, the incidence of peripheral embolism is high.
- Rate control of atrial fibrillation.
- Diuretics for heart failure.

Indications for invasive intervention

- Severe MS.
- Poor symptom control.
- Progressive pulmonary hypertension.
- Recurrent peripheral embolism.

Interventions

- Percutaneous balloon valvotomy *(offered only to carefully selected patients)*.
- Surgical mitral valve replacement.

Pulmonary stenosis

- Rare.
- Often associated with congenital heart disease.
- Can be treated by balloon valvotomy.

Tricuspid stenosis (TS)

- Rare.
- Carcinoid is the most common causes.
- Tricuspid valve replacement rarely required.

Aortic regurgitation (AR)

Epidemiology

- Accounts for 10% of all valvular heart disease.

Aetiology

- Onset may be acute (e.g. infective endocarditis) or chronic (e.g. secondary to bicuspid aortic valve).
- May be caused by destruction of the valve leaflets (e.g. infective endocarditis, rheumatic fever, bicuspid aortic valves, trauma and degenerative calcific AS), *or by*
- Dilation of the aortic root, leading to failure of the valve leaflets to coapt (aortic dissection, Marfan's syndrome and aortitis).

Pathophysiology

AR leads to:

- Increased volume load as blood leaks through the aortic valve back into the left ventricle.
- Increased stroke volume in the short term.
- Dilatation and failure of the left ventricle in the long term.

Presentation

- Shortness of breath, palpitations, fatigue.
- Chronic AR can be tolerated well for many years and is associated with a good prognosis.

Treatment

- Medical treatment includes good blood pressure control (afterload reduction) with calcium channel blockers and vasodilators (ACE inhibitors).
- Acute severe AR is associated with a high mortality and requires immediate intervention.
- Aortic valve replacement surgery is considered when patients develop symptoms of heart failure, deterioration in left ventricular function, or significant dilation of the left ventricle (based on echocardiographic criteria).

Mitral regurgitation (MR)

Epidemiology

- Common valve lesion.

Aetiology

- Onset may be acute (e.g. papillary muscle rupture) or chronic (e.g. mitral valve prolapse).

• May occur due to destruction or malfunction the valve leaflets or chordae (e.g. mitral valve prolapse, infective endocarditis, rheumatic fever), *or*
• Damage to the papillary muscles (e.g. post-myocardial infarction), *or*
• Dilation of the left ventricle, causing mitral annular dilatation and failure of mitral valve leaflet coaptation.

Pathophysiology
MR leads to:
• Increase in left atrial volume and size.
• Increase in left atrial pressure and pulmonary oedema.
• Increase in left ventricular size and left ventricular failure (volume overload).

Presentation
• Shortness of breath, palpitations, atrial fibrillation, fatigue.
• Prognosis is poor in severe MR, with 33% survival at 8 years without surgical intervention.

Treatment
• Medical treatment includes diuretic therapy. Patients in atrial fibrillation are treated with anticoagulation and rate-controlling medications.
• Surgical treatment is indicated in those with severe MR and symptoms or those with deteriorating left ventricular function. Surgical options include: mitral valve repair (preferred if technically feasible) or mitral valve replacement.

Tricuspid regurgitation (TR)

Epidemiology
TR is a common valve lesion.

Aetiology
TR may occur due to:
• Destruction of valve cusps (e.g. rheumatic fever, endocarditis, carcinoid).
• Dilatation of the right ventricle leading to tricuspid annular dilatation (e.g. right heart failure, pulmonary hypertension).

Presentation
• May present with symptoms of right heart failure (see p. 21).

Treatment
• Right heart failure symptoms are treated with diuretics.
• Surgical valve replacement is accompanied by a high operative mortality and is rarely indicated. Valve repair may be undertaken with tricuspid annuloplasty in conjunction with surgery for left-sided valvular disease.

Pulmonary regurgitation

• Mild regurgitation is often seen in normal individuals and is of no clinical consequence.
• May be secondary to pulmonary hypertension.
• Rarely requires surgical intervention.

General approach to treatment of valve pathology

Valve disease treatments include:
• Valvotomy:
  ○ Fused valve leaflets are divided surgically.
• Balloon valvuloplasty:
  ○ Fused valve leaflets are divided by inflating a balloon, which is passed percutaneously.
• Valve repair:
  ○ Preferable to valve replacement if possible, as native valve tissue is preserved (commonly carried out for isolated posterior mitral valve leaflet prolapse).
• Valve replacement with a:
  ○ Bioprosthesis (types: porcine or allograft). No need for anticoagulation, but shorter life span than mechanical valves (10 years).
  ○ Mechanical (types: ball and cage, tilting disc or bileaflet), require anticoagulation (target international normalised ratio [INR] 2.5–4.5, depending on valve prosthesis and position).

Infective endocarditis

• Incidence of 1500 cases/annum in the UK.
• Infection may settle on:
  ○ Native valves – diseased valves (e.g. AS, mitral valve prolapse) or the sites of vascular or myocardial abnormalities (e.g. coarctation, VSD or PDA).
  ○ Right-sided valves – commonly occurs in intravenous drug users.
  ○ Prosthetic valves – common in the immediate post operative period (1%) but declines thereafter.
• Patients may present with:
  ○ Fever, new murmur, malaise, weight loss and non-specific symptoms.
  ○ Diagnosis can be made using the Dukes classification (see Box H).
• Causative organisms include:
  ○ Streptococci (*S. viridens, S. pneumoniae*, Lancefield groups B, C and G, *S. bovis*), staphylococci (90% *Staph aureus*), enterococci, Gram-negative organisms and fungal infections (see Box 14.1 p. 133).

Box H Duke criteria for diagnosis of endocarditis

Major criteria
- Positive blood cultures
  - Typical microorganisms consistent with infective endocarditis from two separate blood cultures
  - Persistent positive blood cultures of blood samples taken >12 hours apart
  - Three or more positive cultures taken over more than 1 hour apart
- Evidence of endocardial involvement noted on echocardiography
  - New valvular regurgitation
  - Abscess
  - Vegetations

Minor criteria
- Predisposing valvular or cardiac abnormality
- Fever: temperature >38°C
- Vasculitic phenomena
- Embolic phenomena
- Microbiological evidence: positive blood culture but does not meet major criteria
- Suggestive echocardiographic findings

Clinical diagnosis for infective endocarditis requires:
- Two major criteria, or
- One major and three minor criteria, or
- Five minor criteria

- There are multiple systemic complications.
- The associated mortality is high 10–20%.
- Treatment includes long-term intravenous antibiotics and surgery in complex cases (see Table 14.3, p. 134).

Heart failure

Heart failure is defined as a state in which the cardiac output is unable to match metabolic needs of the tissues. Prevalence of heart failure is around 3% of the general population, 20% in 70-to-80-year-olds.

Classification

- Anatomical – left, right and biventricular failure:
  - *Left-sided heart failure*: occurs when the left ventricle is predominantly affected. Patients present with left-sided heart failure symptoms (pulmonary oedema).
  - *Right-sided heart failure*: occurs when the right ventricle is predominantly affected. Patients present with right-sided heart failure symptoms (oedema, ascites, hepatomegaly).
  - *Biventricular failure*: both ventricles are affected. Patients present with a mixture of right- and left-sided heart failure symptoms and signs.
- Systolic and diastolic heart failure:
  - *Systolic*. Symptoms and signs of left heart failure with reduced left ventricular ejection fraction.
  - *Diastolic*. There are symptoms and signs of left heart failure with a preserved left ventricular ejection fraction. Also known as HFNEF (heart failure with normal ejection fraction).
- Pathology:
  - Cardiomyopathy:
    - Restrictive e.g. amyloid.
    - Dilated e.g. alcohol, post-viral, ischaemic.
    - Hypertrophic e.g. HOCM.

Presentation

May present as acute, chronic and acute-on chronic heart failure:
- *Acute heart failure*: sudden-onset of heart failure symptoms (shortness of breath and orthopnoea). May be due to 'de novo' causes (e.g. acute myocardial infarction, acute myocarditis, valvular rupture) or acute decompensation on the background of chronic heart failure (acute-on chronic)
- *Chronic heart failure*: gradual onset of symptoms of heart failure (e.g. due to cardiomyopathy).

Pathophysiology (Figure Z)

Heart failure pathophysiology is a vicious cycle which is initiated when there is a reduction in cardiac output leading to decreased blood pressure and reduced tissue perfusion or an increased demand (see Box E, p. 9).

Treatment

- Preload and afterload reduction with diuretics and nitrates.
- Disease-modifying drugs: prognostic benefit is gained from ACE inhibitors, aldosterone antagonists and beta blockers.
- Devices: cardiac resynchronization devices and intracardiac defibrillators can reduce mortality and improve symptoms in carefully selected patient groups.
- Transplantation: considered in patients with severe symptoms despite optimal medical treatments but is limited by shortage of donor organs.

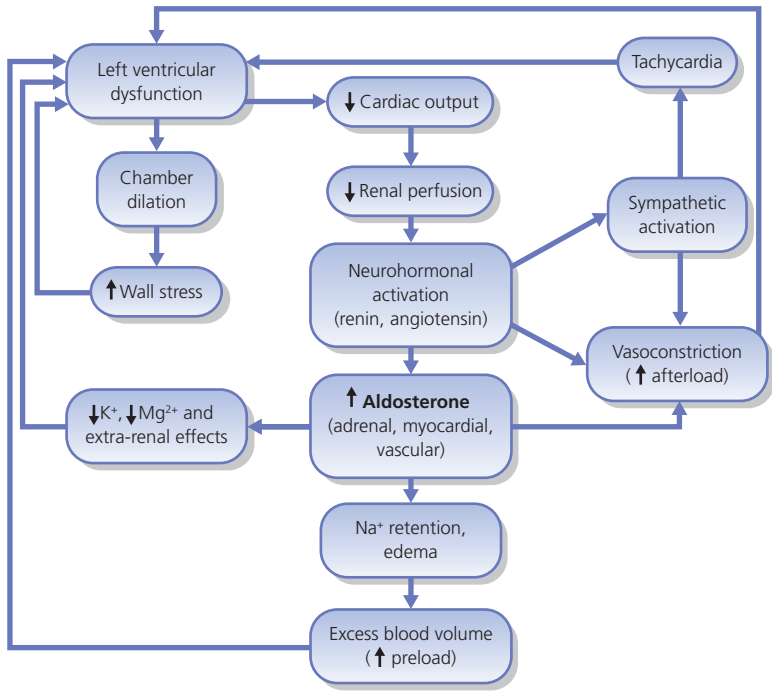


Figure Z The vicious cycle of heart failure.

• Prognosis: poor with median survival 3–5 years from time of diagnosis.

Pericardial diseases

Pericarditis

• Results from primary and secondary inflammation of the parietal and visceral pericardium.
• The majority of cases are viral or idiopathic.
• Patients present with sharp pleuritic chest pain, which is relieved on sitting forward.
• Outcome is largely benign. However, a minority of patients develop chronic relapsing pericarditis.

Pericardial effusion and tamponade

• There is normally a small amount of fluid (<50 ml) in the pericardial sac, which comes from the visceral pericardium.
• Large effusions can become life threatening and cause haemodynamic compromise because the heart is in a confined space and cannot fill properly.
• The most common causes of large pericardial effusions are malignancy, uraemia or tuberculosis (TB).
• Patients most commonly present with shortness of breath.
• Clinical signs of tamponade are tachycardia, pulsus paradoxus, muffled heart sounds and hypotension.
• Small pericardial effusions are harmless and can be managed conservatively.
• Treatment is based on the degree of haemodynamic compromise.

Constrictive pericarditis

• Pericardium becomes thickened and fibrotic and attaches to the myocardium; diastolic filling of the heart becomes restricted.
• Most cases are seen following acute viral or bacterial pericarditis, cardiac surgery or radiotherapy. TB is a likely cause in developing countries.
• Patients present with symptoms and signs of right heart failure.
• Diagnosis can be difficult and is aided by Doppler echocardiography, right- and left-heart catheterisation, cardiac CT and cardiac MR.
• Pericardectomy (stripping) of the pericardium can be carried out for relief of symptoms but carries a high mortality.

References

Durack DT, Lukes AS, Bright DK. New criteria for diagnosis of infective endocarditis: utilization of specific echocardiographic findings. Duke Endocarditis Service. *American Journal of Medicine* 1994: **96**(3);200–9.