

Third edition

Notes & Notes

For MRCP part 1 & 11

By

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Cardiology

Updated

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Coronary arteries: anatomy and clinical correlation

- Right coronary artery (RCA)

⇒ supplies:

- AV node, so heart block following inferior MI is common. However, heart block following anterior MI is a grave prognostic marker as this indicates a large anterior wall infarct. RCA supplies SA node in 60%, AV node in 90%
- Right ventricle, hence, problems relating to a right ventricular infarct are commonly associated with an inferior MI.
- Inferior myocardium and occlusion causes ST elevation in II, III and aVF.
- Posterior descending artery a branch of the right coronary artery in 85% of people (a branch of the circumflex in the remaining population).
 - supplies the **posterior left ventricular myocardium**
 - occlusion causes posterior MI (ST depression in V1-V4 with a dominant R wave in V1).
 - The concept of **coronary dominance** refers to which coronary artery supplies the posterior descending coronary artery (PDA).
 - 85% of patients having a dominant right coronary artery
 - 15% of patients having a dominant left circumflex.

- Left main stem left coronary artery (LCA)

⇒ **Supplies** most of the left ventricle.

⇒ **Complete left main stem occlusion** is invariably fatal. It would produce extensive ST elevation across all the chest leads, I and aVL and possibly aVR, too.

⇒ **LCA branches** into → Left Anterior Descending (LAD) + Left Circumflex artery (LCX)

- Left Anterior Descending (LAD) artery
 - supplies : anterior and septum
 - Occlusion → **ST segment elevation in leads V1-V4**
 - Right bundle branch block in acute anterior myocardial infarction suggests obstruction prior to the first septal branch of the left anterior descending coronary artery
- Left Circumflex artery (LCX)
 - Supplies : lateral
 - Occlusion produces ST elevation in V5, V6, I and aVL.

- ECG localization of STEMI

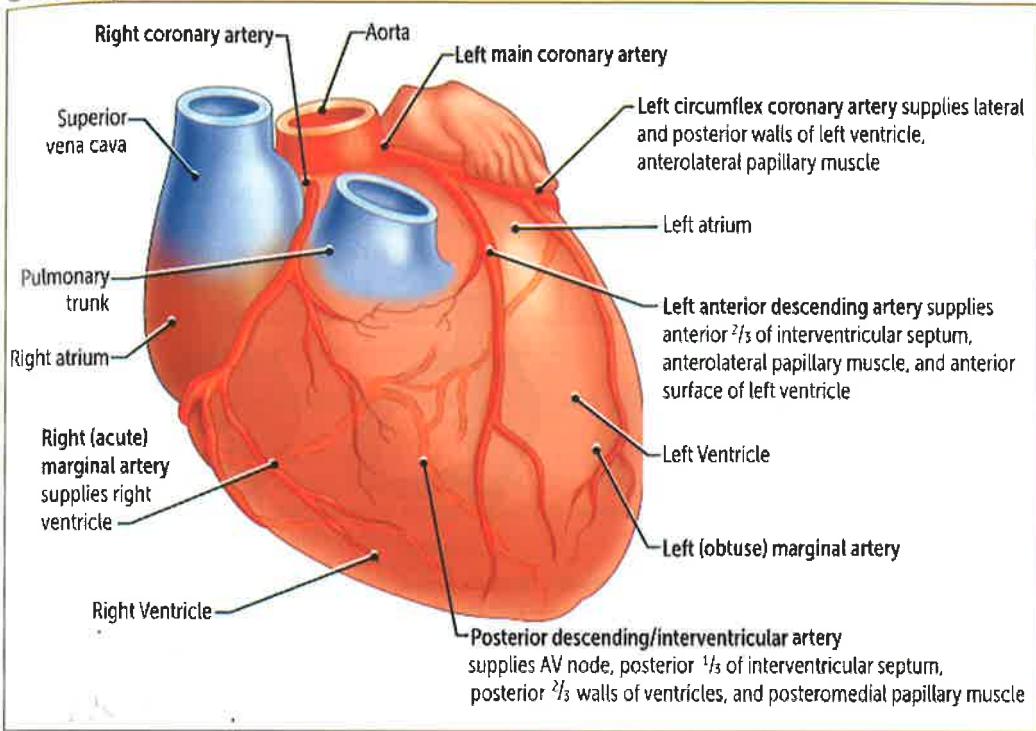
ECG leads with ST elevation	Infarction location
V1 – V2	Anteroseptal (LAD)
V3- V4	Antero-apical (distal LAD)
V5- V6	Antero-lateral (LAD or LCX)
I, aVL	Lateral (LCX)
II, III, aVF	Inferior (RCA)
V7 – V9, ST depression V1- V3 with tall R waves	Posterior (PDA)

ST-segment elevations or Q waves in leads II, III, and aVF on ECG signify a likely inferior MI, supplied by the right coronary artery.

Coronary arterial dominance

- **Right-dominant** (~ 85% of the population): PDA supplied by the RCA
- **Left-dominant** (~ 8% of the population): PDA supplied by the left circumflex artery (LCX)
- **Codominant** (balanced; ~ 7% of people): PDA supplied by both RCA and LCX

Coronary circulation



The left atrium is the posteriomost part of the heart, located directly in front of the esophagus. It can be visualized using TEE. The right ventricle is the anteriomost part of the heart and is at greatest risk of injury following chest trauma.

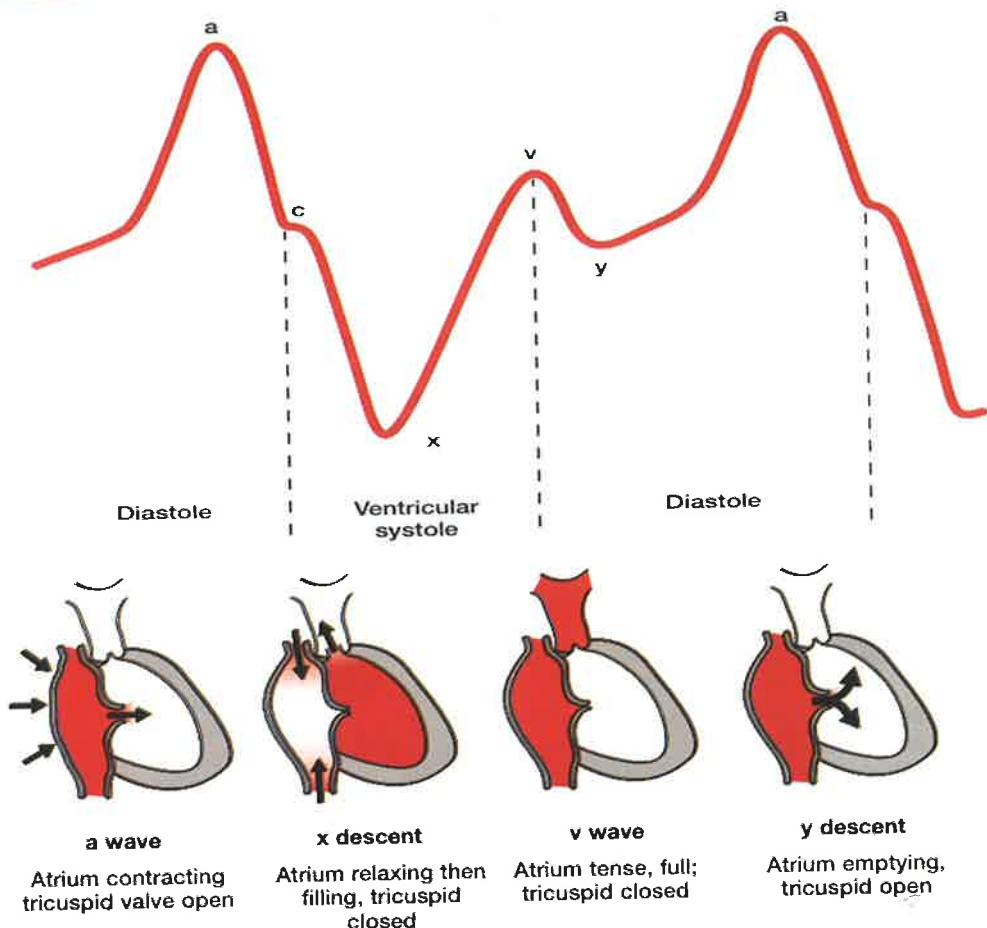
Jugular venous pulse (JVP)

JVP: C wave - closure of the tricuspid valve

JVP: x descent = fall in atrial pressure during ventricular systole

JVP: y descent = opening of tricuspid valve

JVP: giant v waves in tricuspid regurgitation



Clinical importance of JVP

- providing information on right atrial pressure,
- may provide clues to underlying valvular disease.
- A non-pulsatile JVP is seen in superior vena caval obstruction.
- Kussmaul's sign describes a paradoxical rise in JVP during inspiration seen in constrictive pericarditis.

JVP waves and abnormalities

Wave	Description	Abnormalities
a wave	<ul style="list-style-type: none"> The first peak caused by atrial contraction 	<ul style="list-style-type: none"> Absent in atrial fibrillation
c wave	<ul style="list-style-type: none"> The second peak caused by tricuspid valve closure, contraction of the right ventricle, and bulging of the tricuspid valve into the right atrium 	<ul style="list-style-type: none"> cv wave : severe tricuspid valve regurgitation
x descent	<ul style="list-style-type: none"> A drop in JVP caused by atrial relaxation during ventricular systole 	<ul style="list-style-type: none"> Absent in: <ul style="list-style-type: none"> Tricuspid valve regurgitation Right heart failure
v wave	<ul style="list-style-type: none"> The third peak caused by venous refilling of the right atrium against the closed tricuspid valve 	<ul style="list-style-type: none"> Prominent in: <ul style="list-style-type: none"> Tricuspid valve regurgitation Right heart failure
y descent	<ul style="list-style-type: none"> A drop in JVP caused by decreased right atrial pressure as blood flows into the right ventricle after opening of the tricuspid valve 	<ul style="list-style-type: none"> Prominent in: <ul style="list-style-type: none"> Tricuspid valve regurgitation Constrictive pericarditis Absent in: <ul style="list-style-type: none"> Cardiac tamponade Tricuspid valve stenosis

Cannon 'a' waves

- Caused by atrial contractions against a closed tricuspid valve
- Causes
 - Regular cannon waves
 - ventricular tachycardia (with 1:1 ventricular-atrial conduction)
 - atrio-ventricular nodal re-entry tachycardia (AVNRT)
 - Irregular cannon waves
 - complete heart block

A left sided internal jugular central venous catheter has been inserted and you are reviewing the chest radiograph to check the position of the tip of the catheter. What is the safest position to leave the catheter tip?

- ⇒ In the lower superior vena cava

Central venous access of the Subclavian Vein

Anatomy

- Each subclavian vein is a continuation of the axillary vein and runs from the outer border of the first rib.
- The subclavian and internal jugular vein unite to form the brachiocephalic vein, subsequently the left and right brachiocephalic veins unite to form the superior vena cava.

Procedure

- Left-sided subclavian access is associated with lower rates of catheter malposition and vessel trauma. It is preferred when immediate cardiac access is needed (eg, temporary transvenous pacer and pulmonary artery catheter insertion) since the guidewire and catheter are more easily directed into the superior vena cava and right heart.
- The optimal point of needle insertion :
 - ⇒ 1 cm inferior to the junction of the middle and medial third of the clavicle.

Advantages

- the cleanest site for central venous access (lower potential for infection).
- It also the most tolerated by patients.
- consistent landmarks (lower potential for arterial injury compared with other sites of access).

Complications

- Arrhythmias (e.g. premature atrial and ventricular contractions) caused by contact of the guidewire to the right atrium.
- Venous air embolism, pneumothorax, and pneumomediastinum are other common complications of central line placement.
- subclinical pneumothorax even in the hands of experienced clinicians.

Central venous access

- Ultrasound guidance improves initial cannulation success.
- Obtain a postprocedural chest x-ray to confirm catheter position and exclude pneumothorax in jugular and subclavian catheters. Femoral catheters do not require radiographic confirmation and can be used immediately following insertion.
- The internal jugular vein are a commonly used site for central venous access. The distal tip of jugular catheters should lie in the lower superior vena cava. Carotid artery puncture is a well-recognized complication.**
- Femoral site cannulation is often recommended as a secondary site due to higher rates of delayed complications.

Subclavian steal syndrome

Brainstem features (vertigo, diplopia, dysarthria, and drop attacks) **with disparity in BP > 15 mm Hg and pain precipitated by exercise** → **Subclavian steal syndrome**

Pathophysiology

- Stenosis of the subclavian artery proximal to the origin of the vertebral artery → hypoperfusion distal to the stenosis → reversal of blood flow in ipsilateral vertebral artery → compensation through collateral arteries → reduced blood flow in the basilar artery → reduced cerebral perfusion upon exertion involving the affected arm
- characterized by **retrograde flow** into the vertebral or internal thoracic arteries, **due to stenosis and/ or occlusion of the subclavian artery.**

What is the most likely mechanism that maintains blood flow to the affected extremity?

- Blood from the **contralateral vertebral artery** is shunted away from the basilar artery (away from the brainstem) and **retrograde into the ipsilateral vertebral artery** to supply the affected arm .

Causes

- Atherosclerosis
- Takayasu's arteritis

Symptoms

- The most common symptoms** are those related to **upper limb ischemia (arm pain and numbness, especially during exertion)** and exercise with the arm above the head, such as **painting a wall.**)

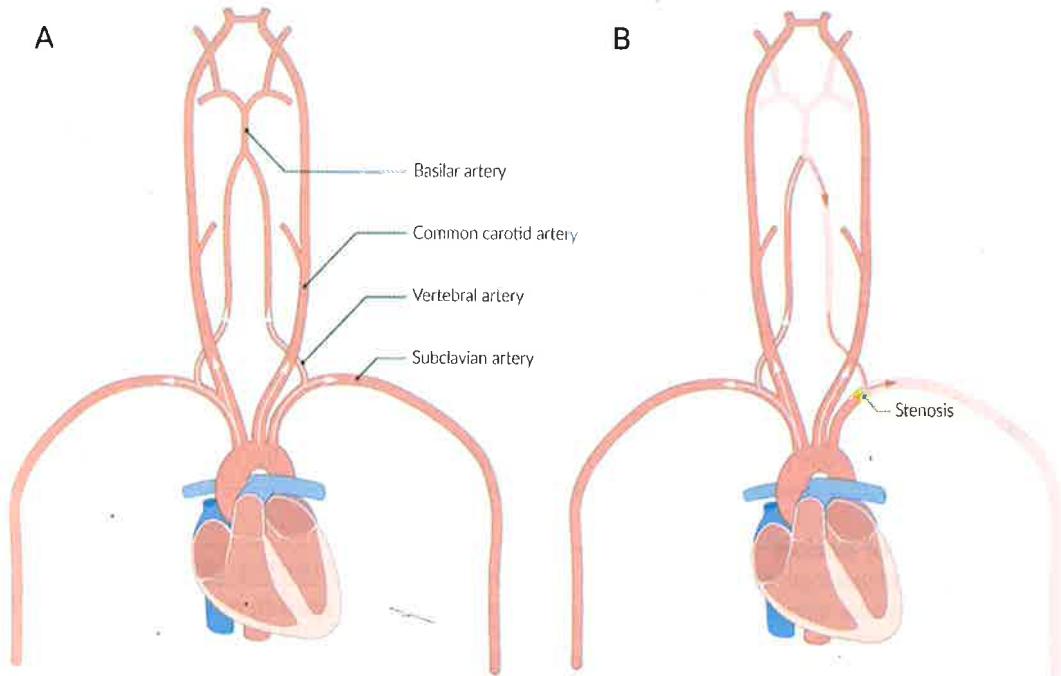
- brainstem features due to vertebrobasilar insufficiency such as: Vertigo, Diplopia, Dysarthria, and Drop attacks.
- **blood pressure is different between the upper limbs by at least 15 mmHg.**

Diagnosis

- **Duplex ultrasound** is **the best initial** radiological test \Rightarrow shows reversal of blood flow (retrograde flow in the ipsilateral vertebral artery)
- **Angiography** of the subclavian vessels (MRA) is the **most accurate** test.

Management

- Most patients require no intervention
- Symptomatic patients: angioplasty and stenting or surgical revascularization



Pathophysiology of subclavian steal syndrome

Under normal conditions, the subclavian artery distributes blood equally to the arteries in the brain and arms (A). If there is stenosis of the subclavian artery proximal to the origin of the vertebral artery, this leads to hypoperfusion of the upper extremities on the affected side (B). This is compensated by a contralateral circulation, in which there is increased blood flow from the unaffected side to the affected side via the vertebral arteries. As a result, there is hypoperfusion of the vertebrobasilar territory and corresponding central nervous system symptoms.

Atrial natriuretic peptide (ANP)

Secretion

- Released from atrial myocytes (**right > left**) in response to blood volume and atrial pressure.
- Acts via cGMP

ANP secretion pathway and actions

- ↑ Volume → ↑ atrial stretch receptors stimulation → release of ANP from atrial cardiomyocytes which results in:
 - ⇒ ↑ Excretion of NaCl and water by the kidneys (via afferent arterioles dilations and efferent arterioles constriction)
 - ⇒ ↓ Na⁺ reabsorption at the renal collecting tubule (via ↑ cGMP)
 - ⇒ Inhibition of renin
 - ⇒ Vasodilation of veins and arteries (↓ preload and ↓ afterload)
- ↓ Volume → ↓ atrial stretch receptors stimulation → ↓ Release of ANP → ↓ excretion of NaCl and water by the kidneys

How is the "aldosterone escape" mechanism mediated by atrial natriuretic peptide (ANP)?

- ANP causes cGMP-mediated dilation of the afferent arteriole and constriction of the efferent arteriole, promoting diuresis and counteracting the effects of aldosterone

B-type (Brain) Natriuretic Peptide (BNP)

BNP - actions:

- vasodilator
- diuretic and natriuretic
- suppresses both sympathetic tone and the renin-angiotensin-aldosterone system

Definition

- B-type natriuretic peptide (BNP) is a hormone produced mainly by the **left ventricular** myocardium in response to strain (myocyte stretch).

Mechanism of action

- Similar physiologic action to ANP with longer half-life.
- ↑ intracellular smooth muscle cGMP → **arterial and venous smooth muscle vasodilatation** → ↓ pre-load → ↓BP
- ↓ sodium reabsorption, leading to natriuresis and diuresis.
- suppresses both sympathetic tone and the renin-angiotensin-aldosterone system

Causes of raised BNP levels

- **heart failure is the most obvious cause**
- age over 70 years,
- ventricular hypertrophy, ischaemia, tachycardia, hypoxaemia [including pulmonary embolism], chronic obstructive pulmonary disease,
- renal dysfunction [eGFR less than 60 ml/minute/1.73 m²]
- sepsis,
- diabetes
- cirrhosis of the liver
- BNP synthesis is increased by thyroid hormones as well as glucocorticoids, endothelin-1, angiotensin-II, and tachycardia, independent of the haemodynamic effects of these factors.

Factors which reduce BNP levels

- Obesity
- African or African-Caribbean family origin
- treatment with diuretics, angiotensin-converting enzyme (ACE) inhibitors, beta-blockers, angiotensin II receptor blockers (ARBs) or mineralocorticoid receptor antagonists (MRAs)

Clinical uses of BNP

- Diagnosing patients with acute dyspnoea (very good negative predictive value for heart failure). (NICE guidelines - 2018).

NT-proBNP level	Note
> 2,000 ng/litre	Refer urgently to specialist and echocardiography within 2 weeks.
Between 400 and 2,000 ng/litre	Refer to specialist and echocardiography within 6 weeks.
< 400 ng/litre	makes a diagnosis of heart failure less likely

- Prognosis in patients with chronic heart failure: very high levels of NT-proBNP carry a poor prognosis
- Guiding treatment in patients with chronic heart failure: effective treatment lowers BNP levels

Brain natriuretic peptide (BNP)

- BNP a hormone **secreted from ventricular myocytes in response to ventricular volume overload**, as seen in congestive heart failure.
- BNP acts on the renal collecting duct to decrease sodium reabsorption and increases glomerular filtration rate, leading to urinary sodium loss.
- BNP has a good negative predictive value, so a patient with a normal BNP likely does not have heart failure.

Cardiovascular physiology

- The basic muscle unit of the myocardium \Rightarrow Sarcomere
- The normal resting cell membrane potential of a cardiac myocyte \Rightarrow - 90 mV
- Left ventricular ejection fraction (LV EF) = (Stroke volume/end diastolic volume) * 100%
- Cardiac output (CO) measure how much blood ejected by the heart in one minute. CO = Stroke volume (SV) x Heart rate (HR)
- Stroke Volume (SV) volume of blood ejected per heart beat = CO/HR = End-Diastolic Volume (EDV) - End-Systolic Volume (ESV)
- Stroke volume is decreased by hypovolaemia
- normal ejection fraction is more than 55% of the blood volume.
- In systolic dysfunction, EF is low. In diastolic dysfunction, EF is normal (called HF with preserved LV EF) e.g. hypertrophic heart failure
- Pulse pressure = Systolic Pressure - Diastolic Pressure
 - \Rightarrow Factors which increase pulse pressure
 - less compliant aorta (this tends to occur with advancing age)
 - increased stroke volume
 - \Rightarrow Factors which reduced pulse pressure
 - Reduced stroke volume,
 - high aortic compliance,

- reduced venous return, and
 - reduced peripheral resistance
- **Sinoatrial node**
 - ⇒ has the **fastest firing rate** of all potential pacemakers in the heart.
 - ⇒ Sinoatrial node impulses must occur at a rate **slower than 200 impulses per minute** to be considered in normal sinus rhythm.
- **Endothelin**
 - ⇒ preferentially **constricts renal afferent arterioles**.
 - **Efferent arteriole vasoconstriction is mediated by angiotensin-II**, to defend GFR in states of generalised vasoconstriction and reduced blood flow.
 - ❖ efferent arteriole vasodilation will occur when angiotensin-II levels fall.
 - ⇒ Stimulates the renin-angiotensin-aldosterone system
 - ⇒ Leads to release of atrial natriuretic peptide
 - ⇒ Inhibits the action of vasopressin
 - ⇒ Two types of endothelin receptor have been characterised, A and B.
 - Binding of endothelin to the A receptor induces vasoconstriction,
 - binding to the B receptor leads to nitric oxide release and hence vasodilatation.

Coronary circulation physiology

- The three most potent factors for vasodilation of the coronaries are:
 1. Increased adenosine
 2. Increased nitric oxide
 3. Opening of ATP-sensitive potassium (KATP) channels by low ATP concentrations, which hyperpolarizes the vascular smooth muscle

Physiological changes during pregnancy

- Heart rate: increases by 10-20 bpm
- Cardiac output and blood volume increase from the second month up to the thirtieth week to 30 - 50% above the normal levels.
- The increase in cardiac output is mediated via **increase in both stroke volume** and to a lesser extent **heart rate**, along with a dramatic **fall in total peripheral vascular resistance**.
- Venous pressure: remain the same due to a 25% reduction in systemic and pulmonary vascular resistance.
- Blood pressure: drop in the first and second trimester **due to vasodilatation** and then climb to pre-pregnancy levels by the third trimester.
- The increase in blood volume and increased cardiac output lead to **all stenotic murmurs becoming more prominent** (there is increased flow across the valve, with more turbulence and pressure gradient, leading to a louder sound).
- Increased metabolic workload
- Apex beat is displaced, because of cardiomegaly and a raised diaphragm
- The increased blood flow may produce a **pulmonary systolic murmur** and a **third heart sound**.

Which murmur is diminished during pregnancy?

- ⇒ **Aortic regurgitation**
 - The fall in diastolic blood pressure during pregnancy leads to a reduction in the murmur of aortic regurgitation.

Physiological changes during exercise

Increases during exercise

- cardiac output → Systemic arterial pressure
- ↑ venous return → ↑ stroke volume
- ↑ heart rate

Decreases during exercise

- Venous compliance
- Peripheral vascular resistance
- Diastolic pressure
- Pulmonary vascular resistance

- Dilatation of the blood vessels within the exercising muscles causes a **fall** in total **peripheral resistance**, resulting in a **decrease** in **diastolic blood pressure**.
- Decrease in venous compliance (dilatation), caused by sympathetic stimulation, helps to maintain ventricular filling during diastole.
- The **pulmonary vessels** undergo **passive dilatation** as more blood flows into the pulmonary circulation, **decreasing** **pulmonary vascular resistance**.

Physiological changes associated with age

- Decrease elasticity and compliance of the aorta → increased resistance to ejection of blood from the left ventricle → increased ventricular afterload.
- Diastolic dysfunction and reduced stroke volume
- ↓↓diastolic pressure (the pressure responsible for subendocardial perfusion) → subendocardial ischemia and interstitial fibrosis. (These changes are related to an increase in the magnitude of the L-type Ca⁺⁺)
- Higher systolic arterial pressure and increased impedance to left ventricular ejection
- ↑ **systolic** + ↓ **diastolic** → ↑ **pulse pressure**
- Increased sino-atrial conduction time
 - ⇒ Because of the delayed LV relaxation and the stiffer left ventricle, the force of left atrial contraction increases and the contribution of the atrial contraction to LV end-diastolic volume increases
- There is apoptosis of atrial pacemaker cells with a loss of 50%-75% of cells by age 50. The number of atrioventricular nodal cells is preserved and there is fibrosis and cellular loss in the His bundle
- Left ventricular hypertrophy
- **Which physiological change associated with age during exercise?**
 - ⇒ Reduced tachycardic response

Valsalva manoeuvre

Definition

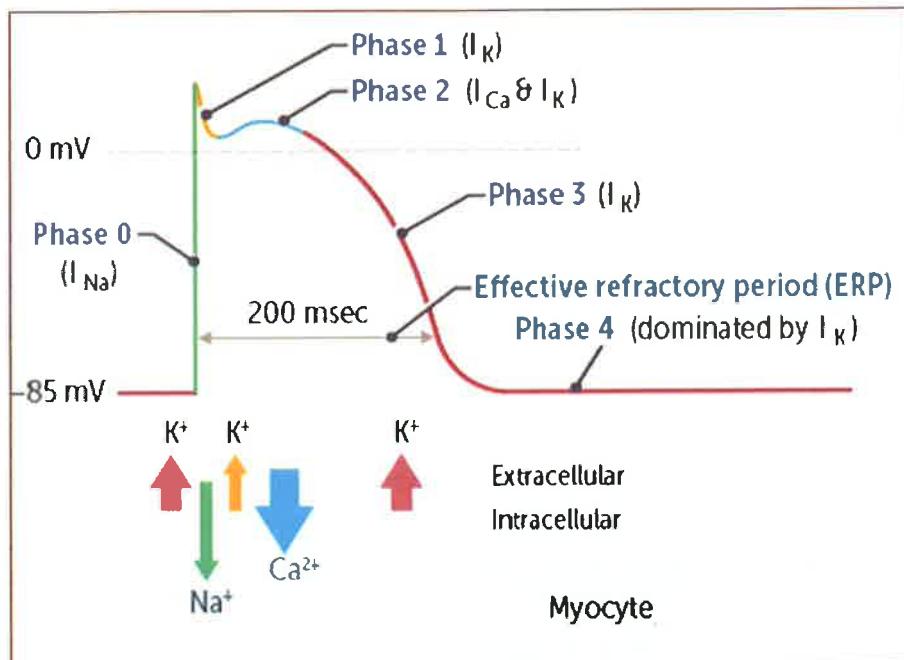
- The Valsalva manoeuvre describes a forced expiration against a closed glottis. This leads to increased intrathoracic pressure which in turn has a number of effects on the cardiovascular system.

Uses

- to terminate an episode of supraventricular tachycardia
- normalizing middle-ear pressures

Cardiac action potential

Cardiac action potential



Phase	Description	Mechanism
0	Rapid depolarisation	Rapid sodium influx These channels automatically deactivate after a few ms
1	Early repolarisation	Efflux of potassium
2	Plateau	Slow influx of calcium
3	Final repolarisation	Efflux of potassium
4	Restoration of ionic concentrations	Resting potential is restored by Na^+/K^+ ATPase There is slow entry of Na^+ into the cell decreasing the potential difference until the threshold potential is reached, triggering a new action potential

NB cardiac muscle remains contracted 10-15 times longer than skeletal muscle

Conduction velocity

Site	Speed
Atrial conduction	Spreads along ordinary atrial myocardial fibres at 1 m/sec
AV node conduction	0.05 m/sec
Ventricular conduction	Purkinje fibres are of large diameter and achieve velocities of 2-4 m/sec (this allows a rapid and coordinated contraction of the ventricles)

Pulses

Patent ductus arteriosus - large volume, bounding, collapsing pulse

Pulsus alternans - seen in left ventricular failure

Pulse	Causes
Pulsus paradoxus (>10 mmHg fall in systolic BP on inspiration)	cardiac tamponade (common)
Pulsus alternans (regular alternation of the force of the arterial pulse between strong and weak)	severe LVF
Bisferiens pulse ('double pulse' - two systolic peaks)	mixed aortic valve disease
Collapsing	aortic regurgitation, patent ductus arteriosus, hyperkinetic (anaemia, thyrotoxic, fever, exercise/pregnancy).
Slow-rising/plateau	aortic stenosis
Jerky pulse	hypertrophic obstructive cardiomyopathy

Pulsus paradoxus

- **Definition**

- ⇒ a greater than 10 mmHg fall in systolic BP on inspiration
- ⇒ → faint or absent pulse in inspiration

- **Mechanism**

- ⇒ Inhalation → ↑ venous return → expands right ventricle (RV) → compresses left ventricle (LV) → ↓ blood pressure.
- ⇒ **Inhale = Big RV = Smaller LV = BP drop > 10 mm Hg**

- **Causes**

- ⇒ **cardiac tamponade** (common)
- ⇒ constrictive pericarditis (less commonly than tamponade)
- ⇒ asthma,
- ⇒ obstructive sleep apnoea
- ⇒ croup.

Heart sounds

First heart sound (S1) : Closure of the mitral and tricuspid valves

Changes in first heart sound (S1)	Causes
Loud S1	<ul style="list-style-type: none"> mitral stenosis left to right shunts short PR interval (e.g. WPW type B). (shortened diastole) atrial premature beats hyperdynamic states
Quiet (soft) S1	<ul style="list-style-type: none"> mitral regurgitation immobile mitral stenosis if closure of the mitral valve is delayed e.g.: <ul style="list-style-type: none"> ⇒ LBBB, ⇒ long PR hypodynamic state
Split S1	<ul style="list-style-type: none"> right bundle branch block, left bundle branch block, ventricular tachycardia, Ebstein's anomaly
Variable intensity	<ul style="list-style-type: none"> Atrial fibrillation

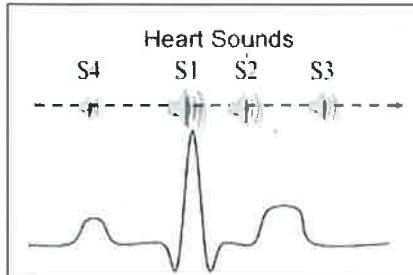
Third heart sound (S3)

Gallop rhythm (S3) is an early sign of LVF

Sound	Origin	Causes	Notes
Third heart sound (S3)	caused by rapid ventricular filling during diastole.	<ul style="list-style-type: none"> Physiological: <ul style="list-style-type: none"> ⇒ young individuals (< 40 years of age), athletes, or pregnant women Pathological <ul style="list-style-type: none"> ⇒ Chronic mitral regurgitation ⇒ Aortic regurgitation ⇒ Heart failure ⇒ Dilated cardiomyopathy • Thyrotoxicosis 	<ul style="list-style-type: none"> Early diastolic sound that is heard immediately after S2 Ventricular gallop: S1 is followed by S2 and S3 .
Fourth heart sound (S4)	caused by <u>atrial contraction</u> against a stiff ventricle	<ul style="list-style-type: none"> Physiological: advanced age Pathological if palpable <ul style="list-style-type: none"> ⇒ Ventricular hypertrophy (e.g., hypertension, aortic stenosis, cor pulmonale) ⇒ Ischemic cardiomyopathy Acute myocardial infarction 	<ul style="list-style-type: none"> Late diastolic (presystolic) sound heard immediately before S1 P wave on ECG

Gallops that originate from the left side of the heart (the most common) become softer with inspiration, while those that originate from the right side become louder.

Heart sound and ECG correlation



Second heart sound (S2)

Second heart sound (S2)

- loud: hypertension
- soft: AS
- fixed split: ASD
- reversed split: LBBB

Second heart sound (S2): Closure of the aortic valve (A2) (louder) and pulmonary valve (P2) (softer).

Changes in S2	Causes
Loud A2	arterial hypertension, coarctation of the aorta
Loud P2	pulmonary hypertension
Physiological split (A2 precedes P2).	during inspiration → ↓ intrathoracic pressure → ↑ venous return to the right side of the heart → prolonged right ventricular systole → delayed closure of P2. Especially pronounced among young individuals
Wide split	Mechanism <ul style="list-style-type: none"> • Increased right ventricular afterload → prolonged right ventricular systole • Decreased left ventricular preload → shortened left ventricular systole Causes <ul style="list-style-type: none"> • Pulmonary hypertension • Pulmonary valve stenosis • RBBB • Massive pulmonary embolism • Severe mitral regurgitation • Wolff-Parkinson-White syndrome • Constrictive pericarditis
Fixed split (Does not change with respiration and tends to be wide,, i.e., the split is also audible during expiration)	<ul style="list-style-type: none"> • Atrial septal defect (ASD)→ RV volume overload → delay in the closure of the pulmonary valve • Severe RV failure • Right bundle-branch block with heart failure (right bundle-branch block widens the split, and heart failure makes the split fixed).
reversed (paradoxical) split S2 (P2 occurs before A2)	<ul style="list-style-type: none"> • Due to delayed A2 <ul style="list-style-type: none"> ⇒ left bundle-branch block (LBBB) ⇒ aortic stenosis (the aortic leaflets are thickened and so close slowly) ⇒ hypertrophic obstructive cardiomyopathy • Due to early P2 <ul style="list-style-type: none"> ⇒ Early excitation of the right ventricle (e.g., RV pacing, Wolff-Parkinson-White syndrome type B) where the right-sided accessory pathway causes early RV depolarisation.
Absent split (No splitting of S2)	<ul style="list-style-type: none"> • Severe aortic stenosis (geriatric) • VSD with Eisenmenger syndrome (paediatric)

Murmurs

Most murmurs of stenosis or regurgitation are exaggerated during squatting and get softer with the Valsalva manoeuvre. **The exceptions are HOCM where the opposite occurs (↑ by Valsalva & ↓ by squatting)** and mitral valve prolapse where the murmur gets longer.

Relation between murmurs intensity and respiration:

- Murmurs that **increase** in intensity with **inspiration** originate from the **right side of the heart** (tricuspid or pulmonary)
- Murmurs that **increase** in intensity with **expiration** originate from the **left side of the heart** (mitral or aortic).

Mnemonic: RILE (Right Inspiration, Left Expiration)

Murmur	Causes
Ejection systolic	<ul style="list-style-type: none"> Aortic stenosis, HOCM Pulmonary stenosis ASD Fallot's
Holosystolic (pansystolic)	<ul style="list-style-type: none"> mitral/tricuspid regurgitation (high-pitched and 'blowing' in character) VSD ('harsh' in character)
Late systolic	<ul style="list-style-type: none"> Mitral valve prolapse Coarctation of aorta
Early diastolic	<ul style="list-style-type: none"> Aortic regurgitation (high-pitched and 'blowing' in character) Graham-Steel murmur (pulmonary regurgitation, again high-pitched and 'blowing' in character)
Mid-late diastolic	<ul style="list-style-type: none"> Mitral stenosis Austin-Flint murmur (severe aortic regurgitation, indistinguishable from that of mitral stenosis). It is due to partial closure of the anterior leaflet of the mitral valve by the regurgitant jet.
Continuous machine-like murmur	<ul style="list-style-type: none"> patent ductus arteriosus

Murmurs and the Effects of Maneuvers

Lesion	Squatting/ leg raising	Standing/ Valsalva
Mitral and aortic stenosis	Increases both	Decreases both
Mitral and aortic regurgitation	Increases both	Decreases both
Mitral valve prolapse	Decrease	Increase
HOCM	Decrease	Increase

More blood increases **all** murmurs except MVP and HOCM.

Standing and Valsalva **decrease** venous return to the heart.

Murmurs in pregnancy

- The intensity of Aortic regurgitation murmur diminishes during pregnancy.**
- Diastolic blood pressure is lower due to vasodilatation, and this is responsible for the fading of the aortic regurgitation murmur
- All stenotic murmurs become more prominent**

Mitral murmurs are heard best during expiration and while the patients lies on the left side.

All right-sided heart murmurs are intensified during deep inspiration.

Isometric **handgrip exercises** increase blood pressure and afterload significantly. Therefore, murmurs caused by the backward flow of blood will be **accentuated**:

- aortic regurgitation,
- mitral valve regurgitation,**
- mitral valve prolapse and
- ventricular septal defect.

Syncope

Definition

- Syncope is a transient loss of consciousness due to transient global cerebral hypoperfusion, characterised by rapid onset, short duration, and spontaneous complete recovery.

Cases

- Syncope can be classified as
 - non-cardiovascular causes:
 - neurally-mediated (reflex syncope)
 - vasovagal
 - situational syncope:** provoked by straining during micturition (usually while standing) or by coughing or swallowing.
 - secondary to orthostatic hypotension
 - cardiovascular causes (such as arrhythmias or ischaemia)
- In older patients, non-cardiovascular causes are twice as common as cardiovascular causes

Evaluation

- The initial evaluation after T-LOC consists of:
 - a careful history
 - orthostatic BP measurements
 - ECG**
 - ECG is the most useful test for classifying syncopal episodes into high risk and low risk categories:**
 - High risk: history of heart disease or abnormal ECG.
 - Low risk: no underlying diseases and a normal ECG.

- The initial evaluation can define the cause of syncope in 23-50% of patients and should answer three key questions:
 - ⇒ Is it a true syncopal episode or not?
 - ⇒ Has the aetiological diagnosis been determined?
 - ⇒ Are there findings suggestive of a high risk of cardiovascular events or death?
- **What you were doing during the episode of blackout?**
 - ⇒ **during exercise**: exercise-induced syncope occurred (cardiac arrhythmic cause is probable)
 - offer urgent (within 7 days) exercise testing, unless there is a possible contraindication (such as suspected aortic stenosis or hypertrophic cardiomyopathy requiring initial assessment by imaging).
 - Advise the person to refrain from exercise until informed otherwise following further assessment.
 - offer an ambulatory ECG and do not offer a tilt test as a first-line investigation.
 - ❖ **TLoC at least several times a week**, → offer Holter monitoring (up to 48 hours)
 - ❖ If no further TLoC occurs during the monitoring period, → offer external event recorder that provides continuous recording with the facility for the patient to indicate when a symptomatic event has occurred.
 - ❖ **TLoC every 1–2 weeks** → offer an external event recorder.
 - ❖ If the person experiences further TLoC outside the period of external event recording, → offer an implantable event recorder.
 - ❖ **TLoC infrequently (less than once every 2 weeks)** → offer an implantable event recorder. A Holter monitor should not usually be offered unless there is evidence of a conduction abnormality on the 12-lead ECG.
 - ⇒ **shortly after stopping exercise** (vasovagal cause is more likely).
 - Unexplained syncope → offer ambulatory ECG. Do not offer a tilt test before the ambulatory ECG.
 - For people with suspected **carotid sinus syncope** and for people with unexplained syncope who are aged 60 years or older, → offer **carotid sinus massage as a first-line investigation**.
 - ⇒ This should be conducted in a controlled environment, with ECG recording, and with resuscitation equipment available.
 - ⇒ Diagnose carotid sinus syncope if carotid sinus massage reproduces syncope due to marked bradycardia/asystole and/or marked hypotension.
 - ⇒ Do not diagnose carotid sinus syncope if carotid sinus massage causes asymptomatic transient bradycardia or hypotension
 - **Tilt test**
 - ⇒ Do not offer a tilt test to people who have a diagnosis of vasovagal syncope on initial assessment.
 - ⇒ For people with suspected vasovagal syncope with recurrent episodes of TLoC adversely affecting their quality of life, or representing a high risk of injury, → consider a tilt test only to assess whether the syncope is accompanied by a severe cardioinhibitory response (usually asystole).
 - If a person has persistent TLoC, consider psychogenic non-epileptic seizures (PNES) or psychogenic pseudosyncope if:
 - ⇒ the nature of the events changes over time
 - ⇒ there are multiple unexplained physical symptoms
 - ⇒ there are unusually prolonged events.
 - **Driving**
 - ⇒ must not drive while waiting for a specialist assessment.
 - ⇒ Following specialist assessment → report the TLoC event to (DVLA)

Implantable loop recorder (ILR)

- subcutaneous, single-lead, (ECG) monitoring device
- used for diagnosis in patients with recurrent unexplained episodes of palpitations or syncope,
- The device is typically implanted in the left parasternal region and is capable of storing ECG data automatically in response to a significant bradyarrhythmia or tachyarrhythmia or in response to patient activation.
- It is particularly useful either when symptoms are infrequent (and thus not amenable to diagnosis using short-term external ECG recording techniques) or when aggregate long-term data (eg, burden of AF) are required.

Vasovagal syncope (VVS)

- Vasovagal syncope (VVS) is the most common type of syncope.

Causes

- features suggestive of uncomplicated vasovagal syncope (**the 3 'P's**):
 - ⇒ Posture – prolonged standing, or similar episodes that have been prevented by lying down
 - ⇒ Provoking factors (such as pain or a medical procedure)
 - common during dental procedures, mainly induced by pain (as the dentist started drilling).
 - ⇒ Prodromal symptoms (such as sweating or feeling warm/hot before TLoC).

Feature

- VVS is usually preceded by a prodrome of symptoms such as dizziness, nausea, and diaphoresis.
 - ⇒ The syncope lasts briefly, but nausea, warmth and sweating may persist for some time.
- Twitching and jerking are often seen with vasovagal or cardiac syncope, which can be differentiated from rhythmic jerking of all the limbs in tonic-clonic seizures.
- **It is common to have jerking of limbs due to brain hypoxia.**
- **Incontinence of urine can occur, but not biting of the tongue.**

Diagnosis

- Recover very quickly supports the diagnosis of syncope.
- ECG is always normal.
- **Tilt table test is a useful test to support the diagnosis**
 - ⇒ If structural heart disease is excluded and syncope is reproduced on tilt table testing along with fall in blood pressure and heart rate, then this is diagnostic of vasovagal syncope.

Treatment

- Midodrine may be indicated in patient with VVS refractory to life style management
 - ⇒ Midodrine is a prodrug of Desglymidodrine
 - ⇒ a sympathomimetic (alpha receptor agonist) that acts on the blood vessels to raise blood pressure.

Postural hypotension

- Causes: mnemonic (HANDI)**

- ⇒ H = Hypovolemia, Hypopituitarism (dehydration, bleeding)
- ⇒ A = Addison's disease
- ⇒ N = Neuropathy (autonomic due to diabetics, amyloidosis)
- ⇒ D = Drugs (Vasodilators, TCA, antipsychotic, Diuretics etc.)
- ⇒ I = Idiopathic orthostatic hypotension

- Management of postural hypotension**

- ⇒ if the standing BP is clearly acceptable (110 systolic) , **the most obvious first step is stopping the causative drug (eg: indapamide)** and monitoring his blood pressure over the subsequent 2-4 weeks.
- ⇒ If he still has significant postural hypotension then **the next steps** would be to add elastic stockings, **then** fludrocortisone.
- ⇒ The history of pre-syncope is much more suggestive of changes in blood pressure rather than changes in blood glucose.

Vertigo & Dizziness

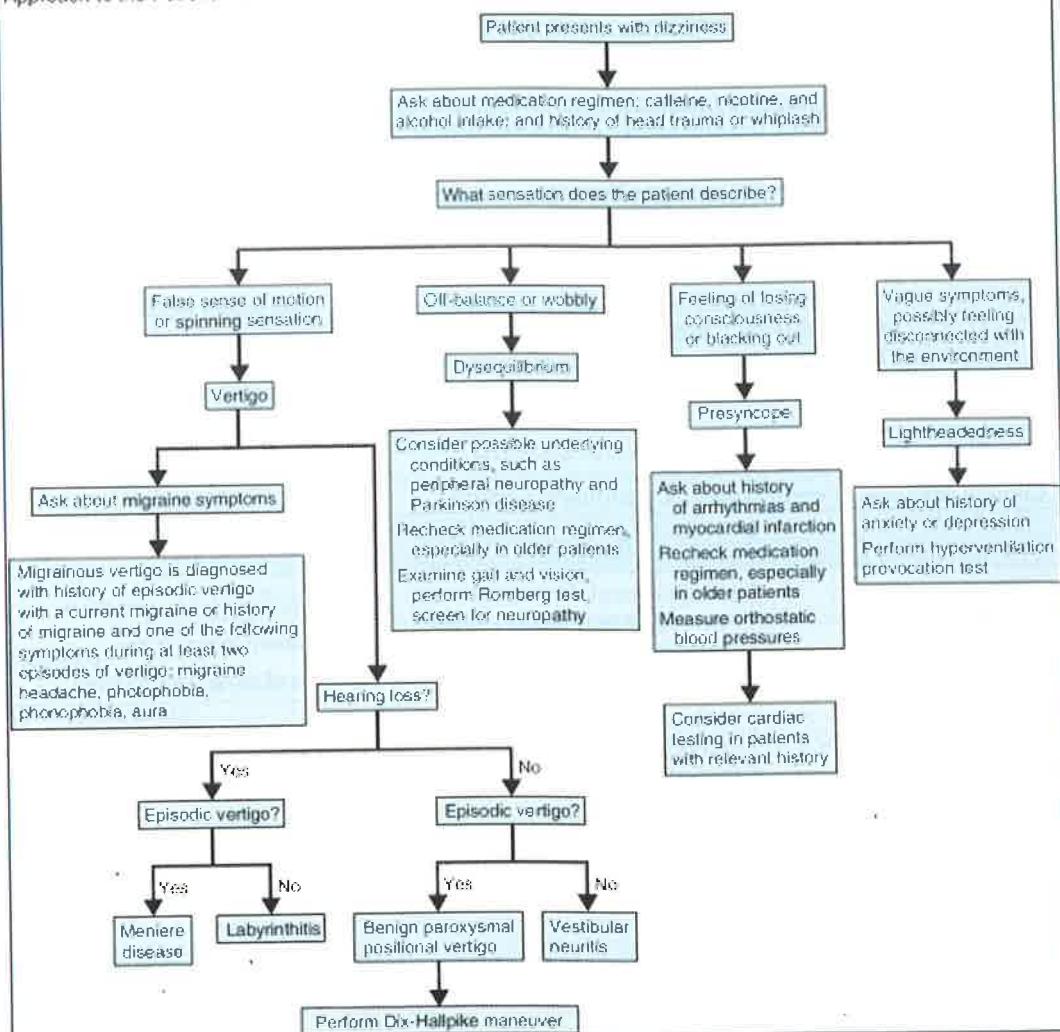
Clinical features of central versus peripheral vertigo

	Peripheral	Central
Nystagmus Direction	Unidirectional, fast component toward the normal ear; never reverses direction	Sometimes reverses direction when patient looks in the direction of slow component
Nystagmus Type	Horizontal with a torsional component, never purely torsional or vertical	Can be any direction
Nystagmus Effect of visual fixation	Suppressed	Not suppressed
Other neurologic signs	Absent	Often present
Postural instability	Unidirectional instability, walking preserved	Severe instability, patient often falls when walking
Deafness or tinnitus	May be present	Absent

Dix-Hallpike maneuver for positional nystagmus: Findings in central versus peripheral vertigo

	Peripheral disorder	Central disorder
Latent period before onset of positional nystagmus	2 to 20 seconds	None
Duration of nystagmus	Less than 1 minute	Greater than 1 minute
Fatigability	Fatiguing with repetition	Non-fatiguing
Direction of nystagmus	Only one type, may change direction with gaze	May change direction with a given head position
Intensity of vertigo	Severe	Less severe, sometimes none

Approach to the Patient with Dizziness



Algorithm for the initial evaluation of a patient with dizziness.

The HINTS exam: (Head Impulse, Nystagmus, Test for Skew)

- A three step physical exam testing oculomotor function (The HINTS exam) was able to differentiate between peripheral causes of vertigo and stroke with a sensitivity of 100%, and a specificity of 96%.
- Remember, the patient needs to be currently experiencing vertigo in order to perform the HINTS exam.

Head Impulse Test (HI)

- Method:
 - ⇒ Patient looks at your nose
 - ⇒ Hold skull (not jaw) firmly
 - ⇒ Slow movement to relax neck muscles
 - ⇒ Quick movement about 20 degree from lateral to midline
 - ⇒ Activate your biceps and forearm, not just wrists
 - ⇒ Random side tested
- Interpretation:
 - ⇒ In peripheral vertigo where the vestibulo-ocular reflex (VOR) reflex is impaired, rapid head rotation toward the affected side will cause the patients eyes to slowly move away from the target and force a corrective saccade (fast) back to the target.
 - ⇒ In central vertigo the VOR reflex remains intact.

Nystagmus (N)

- In peripheral vertigo:
 - ⇒ unidirectional horizontal nystagmus with the fast phase beating away from the affected side.
- In central vertigo:
 - ⇒ vertical or rotational nystagmus, or bidirectional horizontal nystagmus where the fast phase changes directions.

Test for skew (TS)

- Method
 - ⇒ alternating covering the patients eyes while the patient fixes their gaze on a fixed target.
- Interpretation
 - ⇒ In central vertigo:
 - the patients sometimes have vertical misalignment of their eyes due to impaired gravity sensing. As the cover moves back and forth between the two eyes, the uncovered eye will correct its gaze to refocus on the target. This correction should be observed repeatedly as the cover moves back and forth.
 - ⇒ In peripheral vertigo:
 - no skew deviation.

In summary: The HINTS exam:

- **Peripheral** = Positive head impulse test, unidirectional nystagmus, no skew
- **Central** = Negative head impulse test, bidirectional, vertical or rotational nystagmus,

Sudden cardiac death

- In those aged greater than 35 years:
 - ⇒ The most common cause of sudden cardiac death is ischemic heart disease.
 - ⇒ Up to 80% of individuals who suffer sudden cardiac death have coronary heart disease.
- In those under the age of 35 years of age:
 - ⇒ HOCM is the most common cause of sudden cardiac death, coronary artery disease being the second most common cause.
 - ⇒ In competitive athletes <35 years of age HOCM is by far the most common cause of sudden cardiac death (prevalence is 1 in 500).
- Arrhythmogenic right ventricular dysplasia (ARVD)
 - ⇒ the second most common cause of sudden cardiac death in the young after HOCM.
- Catecholaminergic polymorphic ventricular tachycardia (CPVT)
 - ⇒ an autosomal dominant inherited cardiac disease
 - ⇒ prevalence of around 1:10,000.
- Brugada syndrome
 - ⇒ an autosomal dominant inherited cardiovascular disease.
 - ⇒ prevalence of 1:5,000-10,000.
 - ⇒ more common in Asians.

Exercise tolerance tests

Indications: Exercise tolerance tests (ETT, also exercise ECG) are used for a variety of indications:

- assessing patients with suspected angina - however the 2010 NICE Chest pain of recent onset guidelines do not support the use of ETTs for all patients
- risk stratifying patients following a myocardial infarction
 - ⇒ the best predictor of mortality post-STEMI → exercise capacity
 - ⇒ **Above average exercise capacity → good prognosis after a STEMI**
- assessing exercise tolerance
- risk stratifying patients with hypertrophic cardiomyopathy

Sensitivity and specificity of ETT: (high number of false positives and false negatives)

- **ETT has a sensitivity of around 80% and a specificity of 70% for ischaemic heart disease.** Thus, a negative test may not necessarily be true and further testing may be advised.
 - ⇒ Exercise ECG testing has a **relatively high sensitivity but only moderate specificity for the diagnosis of CAD.**
- Diagnostic accuracy is poor in women and this may relate to smaller heart size.

Heart rate:

- maximum predicted heart rate = $220 - \text{patient's age}$
- the target heart rate is at least 85% of maximum predicted to allow reasonable interpretation of a test as low-risk or negative

Contraindications

- myocardial infarction less than 7 days ago
- unstable angina
- uncontrolled hypertension (systolic BP > 180 mmHg) or hypotension (systolic BP < 90 mmHg)
- Any condition where left ventricular output is reduced - eg, aortic stenosis or hypertrophic obstructive cardiomyopathy (HOCM).

- Abnormal baseline ECG (eg, bundle branch block patterns or left ventricular hypertrophy); these make interpretation of the ETT difficult.

Stop if:

- exhaustion / patient request
- 'severe', 'limiting' chest pain
- > 3mm ST depression
- > 2mm ST elevation. Stop if rapid ST elevation and pain
- systolic blood pressure > 230 mmHg
- systolic blood pressure falling > 20 mmHg
- attainment of maximum predicted heart rate
- heart rate falling > 20% of starting rate
- arrhythmia develops

Interpreting the exercise tolerance test

- The patient is normally considered to have been adequately 'stressed' if they achieve 85% or more of their maximum heart rate (calculated as 220 - age in years for men and 210 - age for women).
- If ECG criteria for inducible ischaemia (chest pain is not mandatory). The next step is → Coronary angiography
 - ⇒ this will define the coronary anatomy and give a better guide to prognosis.
- If an inadequate test was performed, further non-invasive investigations may be indicated, such as myocardial perfusion scanning, cardiac MRI, or stress echocardiogram.

Notes

- Beta-blockers and digoxin can interfere with the results so are usually stopped before the ETT.
 - ⇒ If ETT performed on beta blocker and there is an adequate rise in heart rate (85% of (220 - age)) → so there is no indication for stopping beta blocker and repeat the test

Cardiac enzymes and protein markers

Myoglobin rises first following a myocardial infarction

Key points for the exam

- myoglobin is the first to rise
- CK-MB is useful to look for reinfarction as it returns to normal after 2-3 days (troponin T remains elevated for up to 10 days)

	Begins to rise	Peak value	Returns to normal
Myoglobin	1-2 hours	6-8 hours	1-2 days
CK-MB	2-6 hours	16-20 hours	2-3 days
CK	4-8 hours	16-24 hours	3-4 days
Trop T	4-6 hours	12-24 hours	7-10 days
AST	12-24 hours	36-48 hours	3-4 days
LDH	24-48 hours	72 hours	8-10 days

Troponin

Troponin C: Binds to calcium to activate actin: myosin interaction

Troponin T: Binds to tropomyosin

Troponin I: Blocks or inhibits actin: myosin interaction

- Troponin is a **component of thin filaments**
- Cardiac-specific marker with high sensitivity for myocardial necrosis
- The degree of elevation correlates with the size of the infarct and risk of mortality. **Levels act as a prognostic factor following an acute coronary syndrome**
- **Other causes of an elevated troponin are:**
 - ⇒ Pulmonary embolism, Pulmonary hypertension
 - ⇒ Hypertension, Hypotension, especially with arrhythmias
 - ⇒ Hypertrophic obstructive cardiomyopathy, Myocarditis including Kawasaki's disease
 - ⇒ Sepsis, Burns, Trauma, Cardioversion, Rhabdomyolysis
 - ⇒ Subarachnoid haemorrhage and stroke
 - ⇒ Infiltrative/autoimmune disorders including sarcoidosis, amyloidosis, haemochromatosis and scleroderma.
 - ⇒ Drugs including: Adriamycin, **Herceptin** and 5-fluorouracil.

CK-MB

- No longer commonly used clinically; has been replaced by cardiac troponin in the diagnosis of ACS
- CK-MB is more specific to cardiac tissue than total CK (but may also be due to skeletal muscle injury).
- Can be helpful for evaluating reinfarction because of its short half-life but is no longer commonly used
- The degree of elevation often correlates with the size of the infarct.

Serum creatine kinase

- **Causes of high CK**
 - ⇒ Myocardial infarction
 - ⇒ **Racial variant : serum CK activity in Afro-Caribbean people is often up to three times the upper limit of normal for white populations**
 - ⇒ Hypothyroidism
 - ⇒ Heavy exercise
 - ⇒ Statins

Glycogen phosphorylase isoenzyme BB (GPBB)

- GPBB exists in heart and brain tissue.
- **Rise significantly by three hours post mi. As such it is an appropriate marker for early cardiac muscle injury.**
- **Rise earlier than myoglobin**
 - ⇒ GPBB levels increase 1–3 h after the event.
 - ⇒ Myoglobin levels increase significantly 2 h after ischaemia.

ECG: axis deviation

Normal axis

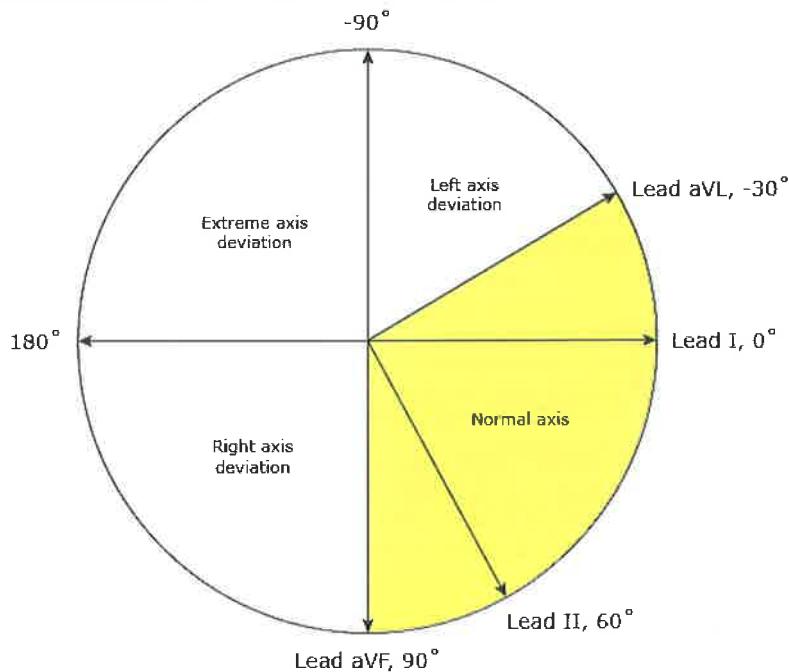
- between -30 and 90° (directed inferior and to the left)

Left axis deviation (LAD)

- **Definition**
 - ⇒ An axis between -30° and -90° (directed superior and to the left)
- **Prevalence**
 - ⇒ LAD (≥ -30 degrees) is the most common "abnormality" in adults occurring in over 8%.
- **Causes of LAD:**
 - ⇒ left ventricular hypertrophy
 - ⇒ left bundle branch block
 - ⇒ **left anterior hemiblock**
 - Marked LAD (≥ -45 degrees) is called left anterior hemiblock or left anterior fascicular block
 - ⇒ Wolff-Parkinson-White syndrome* - right-sided accessory pathway
 - *in the majority of cases, or in a question without qualification, Wolff-Parkinson-White syndrome is associated with left axis deviation
 - ⇒ congenital: ostium **primum** ASD, tricuspid atresia, endocardial cushion defect
 - ⇒ Inferior wall myocardial infarction
 - ⇒ hyperkalaemia
 - ⇒ Normal variation (physiologic, often with age), minor LAD in obese people
 - ⇒ Mechanical shifts, such as expiration, high diaphragm (pregnancy, ascites, abdominal tumor)
 - ⇒ Emphysema
 - ⇒ Ventricular ectopic rhythms
- **Recommendations:** (If LAD is present):
 - ⇒ Exclude hypertension. (If borderline → ambulatory BP monitoring).
 - ⇒ check for borderline indicators of LVH (i.e., the voltage criteria and left atrial enlargement).
 - ⇒ Note whether diagnostic inferior Q waves are present since an inferior MI can cause LAD.

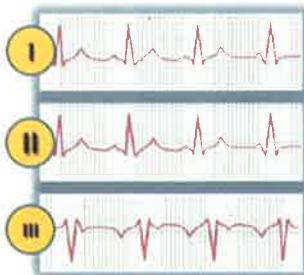
Right axis deviation (RAD)

- **Definition**
 - ⇒ An axis between 90° and 180° (directed inferior and to the right),
- **Causes of RAD:**
 - ⇒ right ventricular hypertrophy
 - ⇒ right bundle branch block
 - ⇒ **left posterior hemiblock**
 - ⇒ **Wolff-Parkinson-White syndrome** - left-sided accessory pathway
 - ⇒ ostium **secundum** ASD
 - ⇒ chronic lung disease → cor pulmonale
 - ⇒ pulmonary embolism
 - ⇒ Dextrocardia
 - ⇒ Ventricular ectopic rhythms
 - ⇒ Lateral wall myocardial infarction
 - ⇒ Normal variation (vertical heart with an axis of 90°).
 - normal in youngsters (less than 21 years of age), tall people, thin adults and athletes

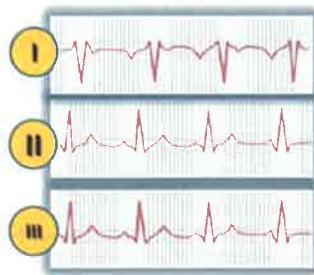


- If the QRS complex is **positive (upright) in leads I and II**, it falls between -30 and 90° and is **normal**, as indicated by the yellow area.
- If the QRS complex is **negative in I and positive in aVF**, there is **right axis deviation**.
- If the QRS complex is **positive in I and negative in II**, there is **left axis deviation**.
- If the QRS complex is **negative in I and aVF**, there is **extreme axis deviation**.

Left axis deviation



Right axis deviation



ECG: coronary territories

The table below shows the correlation between ECG changes and coronary territories:

Localization of myocardial infarct on ECG

	ECG changes	Coronary artery
Anteroseptal	V1-V4	Left anterior descending (LAD)
Inferior	II, III, aVF	Right coronary
Anterolateral	V4-6, I, aVL	Left anterior descending (LAD) or left circumflex
Lateral	I, aVL +/- V5-6	Left circumflex
Posterior	Tall R waves V1-2	Usually left circumflex, also right coronary

High lateral wall MI

- **ST segment elevation in leads I and aVL → High lateral wall MI**
- usually due to occlusion of the first diagonal branch of the left anterior descending artery, though occlusion of other arteries like branches of the left circumflex or a short left anterior descending artery may cause the same picture.

Postero-lateral MI → prominent R wave in lead V1 and ST depression in V1-V3 + ST elevation in leads V5 and V6.

Posterior MI (ESC guidelines 2017)

- posterior wall (now termed inferobasilar), usually supplied by the posterior descending artery — a branch of the **right coronary artery** in 80% of individuals.
- isolated ST-segment depression ≥ 0.5 mm in leads V₁-V₃ represents the dominant finding. These should be managed as a STEMI.
- The use of additional posterior chest wall leads [elevation V₇-V₉ ≥ 0.5 mm (≥ 1 mm in men, 40 years old)] is recommended.

Left main stem (LMS)

- LMS occlusion typically presents dramatically with cardiogenic shock.
- ECG findings include ST elevation in aVR with diffuse ST depression in other leads.
- The presence of ST depression ≥ 1 mm in six or more surface leads, coupled with ST-segment elevation in aVR and/or V₁, suggests **multivessel ischemia** or **left main coronary artery obstruction**, particularly if the patient presents with haemodynamic compromise. (ESC guidelines 2017)

Which ECG changes may be seen earlier in ischaemia ?

↪ **hyper-acute T-waves**, which may precede ST-segment elevation.

ECG criteria for STEMI (ESC guidelines 2017)

- ST-segment elevation (measured at the J-point) is considered suggestive of ongoing coronary artery acute occlusion in the following cases:
 - ⇒ **Numbers of leads:** at least two contiguous leads with ST-segment elevation
 - ⇒ ST-segment elevation:
 - ≥ 2.5 mm in men < 40 years,
 - ≥ 2 mm in men ≥ 40 years, or
 - ≥ 1.5 mm in women in leads V₂–V₃ and/or
 - ≥ 1 mm in the other leads
 - ⇒ In patients with **inferior MI**, it is recommended to record right precordial leads (V_{3R} and V_{4R}) seeking ST-segment elevation, to identify **concomitant right ventricular (RV) infarction**.
 - ⇒ Likewise, ST-segment depression in leads V₁–V₃ suggests myocardial ischaemia, especially when the terminal T-wave is positive (ST-segment elevation equivalent), and confirmation by concomitant ST-segment elevation ≥ 0.5 mm recorded in leads V₇–V₉ should be considered as a means to identify **posterior MI** (circumflex occlusion).

ECG: digoxin

ECG features

- down-sloping ST depression ('reverse tick')
- flattened/inverted T waves
- short QT interval
- arrhythmias e.g. AV block, bradycardia

ECG: hypothermia

The following ECG changes may be seen in hypothermia

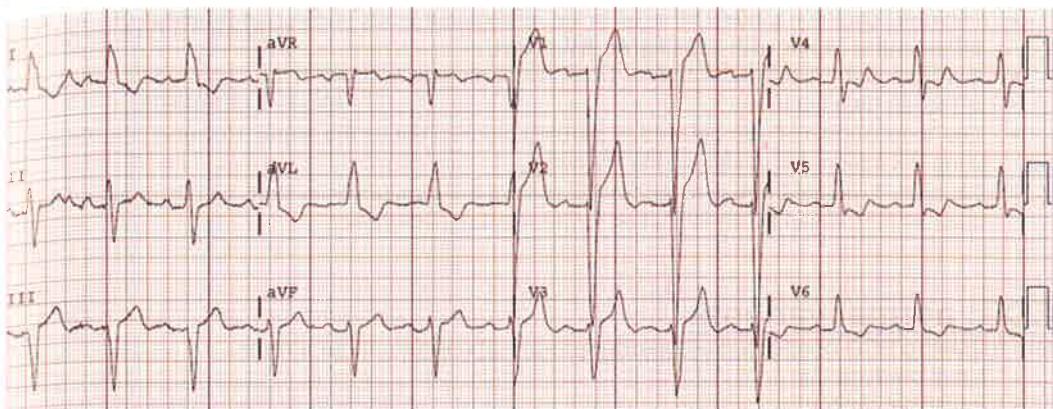
- bradycardia
- 'J' wave - small hump at the end of the QRS complex
- first degree heart block
- long QT interval
- atrial and ventricular arrhythmias

ECG: left bundle branch block

- The diagram below shows the typical features of left bundle branch block (LBBB):



- The ECG would show:
 - ⇒ broad QRS complex (>120ms),
 - ⇒ tall R waves in the lateral leads (I, V₅-V₆) and deep S waves in the right precordial leads (V₁-V₃)
 - ⇒ usually leads to left axis deviation.
- One of the most common ways to remember the difference between LBBB and RBBB is WILLiaM MaRRoW
 - WILLiaM : in LBBB there is a 'W' in V1 and a 'M' in V6
 - MaRRoW: in RBBB there is a 'M' in V1 and a 'W' in V6

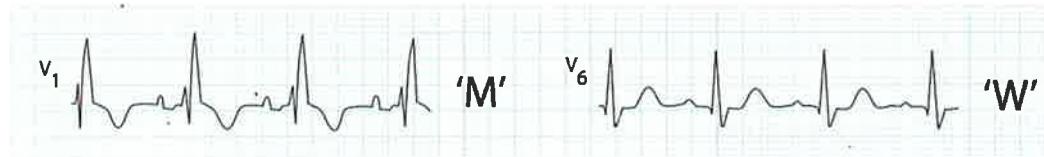


ECG showing typical features of LBBB

- Causes of LBBB
 - ⇒ ischaemic heart disease
 - ⇒ hypertension
 - ⇒ aortic stenosis
 - ⇒ cardiomyopathy
 - ⇒ rare: idiopathic fibrosis, digoxin toxicity, hyperkalaemia

Right bundle branch block (RBBB)

- Patients with MI and right bundle branch block (RBBB) have a poor prognosis. (ESC guidelines 2017)
 - ⇒ It may be difficult to detect transmural ischaemia in patients with chest pain and RBBB.
 - ⇒ Therefore, persistent **ischaemic symptoms** occur in the presence of **RBBB** → primary **PCI** strategy (emergent coronary angiography and PCI if indicated) **should be considered**



Trifascicular block

The evidence of trifascicular block (RBBB, LAD and prolongation of the PR interval) in the context of dizziness and collapses. This is an indication for dual chamber (DDDR) pacing for likely complete heart block.

- Trifascicular block is not strictly an ECG diagnosis but is a term used for the combination of:
 1. right bundle branch block,
 2. left hemiblock (typically left anterior hemiblock (LAHB)) (LAHB is diagnosed because the net QRS deflection in lead II is negative).
 3. long PR interval.
- **the site of the lesion → AV node and Purkinje fibres**

- The most common pattern referred to as "trifascicular block" is the **combination of bifascicular block with 1st degree AV block**.
- It implies that the bundle branches (Purkinje fibres) are blocked in the right bundle and one of the left hemibundles.
- The 'third' bundle is also delayed or partially blocked hence the name. However, the delay (long PR interval) is usually at the AV node.
- Clinically it means there is extensive disease of the conduction system and, in a patient such as this, would be an indication for permanent pacemaker.

ECG: normal variants

The following ECG changes are considered normal variants in an athlete:

- sinus bradycardia
- junctional rhythm
- first degree heart block
- Wenckebach phenomenon

ECG: PR interval

Causes of a prolonged PR interval

- idiopathic
- ischaemic heart disease
- digoxin toxicity
- hypokalaemia: hyperkalaemia can rarely cause a prolonged PR interval, but this is a much less common association than hypokalaemia
- rheumatic fever
- aortic root pathology e.g. abscess secondary to endocarditis
- Lyme disease
- sarcoidosis
- myotonic dystrophy
- A prolonged PR interval may also be seen in athletes

short PR interval is seen in Wolff-Parkinson-White syndrome

ECG: ST depression

Causes of ST depression

- secondary to abnormal QRS (LVH, LBBB, RBBB)
- ischaemia
- digoxin
- hypokalaemia
- syndrome X

T wave

- The T wave should be analyzed for:
 - orientation: upgoing, downgoing (inverted) or biphasic
 - concordance with QRS
 - Concordant:** (normal) both QRS and T wave are on the same direction (upgoing or downgoing) (downgoing is common in aVR for normal ECG's)
 - Discordant:** (abnormal) QRS is upgoing, T wave is downgoing or vice versa
 - morphology (size and shape)

Biphasic T wave

BIPHASIC T-WAVE



- Biphasic T waves can be "up then down", or "down, then up".
- There are 2 causes of biphasic T waves:
 - ⇒ Ischemia
 - Wellens' syndrome (type II):
 - ❖ Two types of Wellens' syndrome are identified:
 1. Type I: The most common (75% of cases), characterised by deep negative T waves in V2–V3 and often in V4.
 2. Type II: less common (one third of patients) ,present with **biphasic T waves in V2–V3**
 - ❖ pathognomonic of critical stenosis of the proximal left anterior descending coronary artery (LAD)
 - ❖ It is also known as the "widow maker" sign because of the high risk of an acute coronary syndrome within days/weeks if it is untreated
 - ⇒ Hypokalaemia

Q waves

- A Q wave is any negative deflection that precedes an R wave on the ECG.
- The evolution of Q waves is the most suggestive of an infarct. (**more specific than ST elevation and cardiac enzyme for MI**)
 - ⇒ **the most specific for a diagnosis of myocardial infarction**
- Small Q-waves are normal in most leads, and they can be prominent in leads III and aVR as a normal variant but should not be seen in leads V1-V3.
- They are **considered pathological if** they are:
 - ⇒ more than 1mm wide,
 - ⇒ more than 2mm deep,
 - ⇒ more than 25% of the depth of the QRS complex, or
 - ⇒ seen in leads V1-V3.
- Such pathological Q-waves usually indicate prior full thickness myocardial infarct.

ECG: ST elevation (STE)

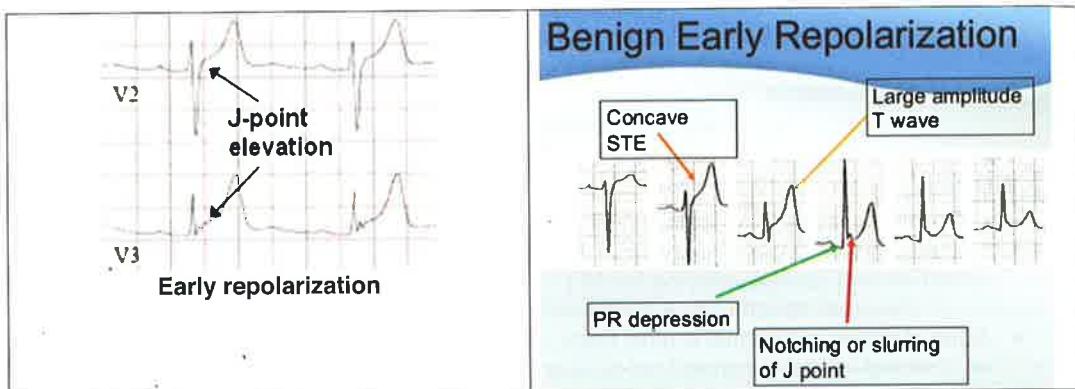
Causes of ST elevation

- myocardial infarction
- pericarditis
- normal variant - 'high take-off'
- left ventricular aneurysm
- Prinzmetal's angina (coronary artery spasm)
- rare: subarachnoid haemorrhage, part of spectrum of changes in hyperkalaemia

Early repolarization

- Definition

- ⇒ It appears as mild ST segment elevation (appears like an elevated "J point.") that can be diffuse; however, it is more prominent in the precordial leads.
- Causes
 - ⇒ common finding in young, healthy individuals.
 - ⇒ Prevalence: occurs in up to 13% of the general population
- Differential diagnosis
 - ⇒ Early repolarization (benign finding)
 - ⇒ acute myocardial infarction (convex and not diffuse)
 - ⇒ pericarditis
 - The ST elevation seen in early repolarization is very similar: diffuse and concave upward.
 - Three things may help to distinguish pericarditis from early repolarization:
 - 1) The ratio of the T wave amplitude to the ST elevation should be > 4 if early repolarization is present. In other words, the T wave in early repolarization is usually 4 times the amplitude of the ST elevation. Another way to describe this would be that the ST elevation is less than 25% of the T wave amplitude in early repolarization.
 - 2) The ST elevation in early repolarization resolves when the person exercises.
 - 3) Early repolarization, unlike pericarditis, is a benign ECG finding that should not be associated with any symptoms.



QT Interval

Definition

- The QT interval is the time between the onset of the **QRS** complex and the end of the **T** wave.

Physiology

- It represents the ventricular diastole
- QRS corresponds with ventricular depolarization (when it contracts) and T wave corresponds with ventricular repolarization (when contraction stops).

Which phase of the cardiac cycle shortens the most with increasing heart rate?

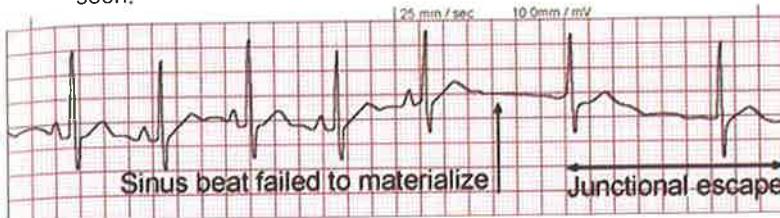
- it is diastole.
 - ⇒ Diastole is usually the longest portion of the cardiac cycle, and its duration diminishes the most (more than the reduction seen in the duration of systole) with increasing heart rate.

Which ECG interval will show the greatest reduction during ECG stress test?

- **QT interval**

ECG: Junctional escape rhythm

- Junctional escape rhythm describes an abnormal heart rhythm that arises within the AV node or from an adjacent area.
- There is a slow, regular pulse rate.
- Common after a pause in the underline rhythm
- ECG shows absent P waves, narrow QRS complexes, and a heart rate of 40 to 60 bpm.
- Retrograde P waves, which appear *immediately* before or after the QRS complex may be seen.



Cardiac amyloidosis

Amyloid

- Low-voltage ECG**
- Speckled pattern on echo**

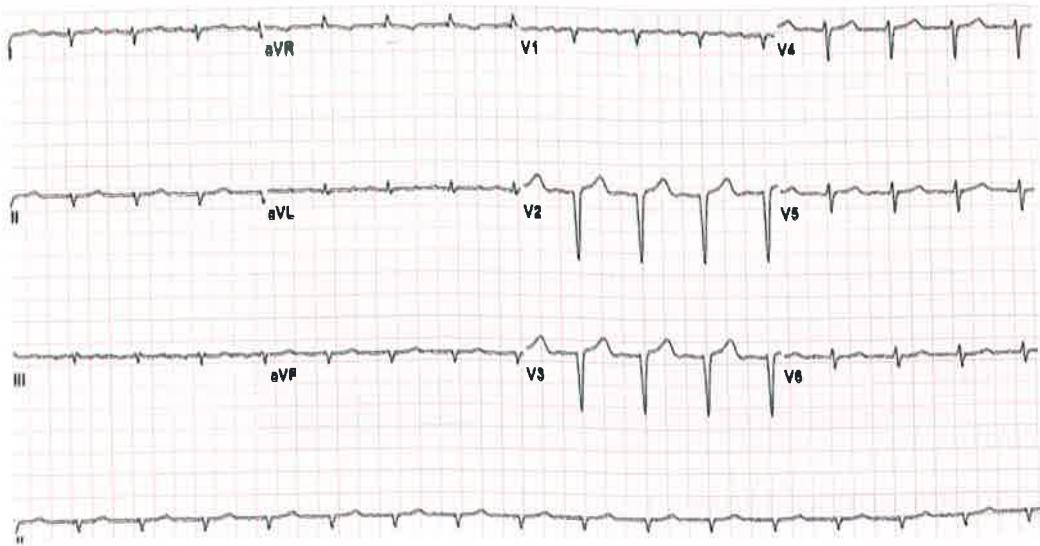
Features

- most commonly presents as restrictive cardiomyopathy.
- clinical findings are those of right heart failure, i.e. jugular venous distension and peripheral oedema,
- orthopnoea and paroxysmal nocturnal dyspnoea are **typically absent**.
- systolic dysfunction (In more advanced stages,)
- Postural hypotension can occur as a result of poor ventricular filling or associated autonomic neuropathy.

Investigations

- ECG**
 - ⇒ The combination of low-voltage ECG and thickened ventricular walls is one of the characteristic features of cardiac amyloidosis.
- Echo**
 - ⇒ echocardiographic abnormalities include atrial dilatation, thickened interatrial septum, diastolic dysfunction and small-volume ventricles.
 - ⇒ The most distinctive feature of cardiac amyloidosis is a sparkling, granular appearance of the myocardium, but this is a relatively insensitive feature occurring only in about 25% of cases.
 - ⇒ 'global speckled' pattern on echo.
- The history of rheumatoid arthritis and the echocardiographic finding of bi-atrial dilatation, ventricular hypertrophy and a speckled appearance to the myocardium make amyloidosis the most likely underlying cause.
- Digoxin is contraindicated in amyloid patients as the digoxin binds irreversibly to the amyloid fibrils.**

The ECG typically shows low-voltage complexes with poor R wave progression in the chest leads (a pseudo-infarction pattern).



ECG: Wrong leads

- They are normally labelled red (right arm) and yellow (left arm). The other leads are green (left leg) and black (right leg).
- If the wires to the right and left arms have been accidentally swapped over → It gives the appearance of **abnormal T wave inversion in the lateral leads I and aVL**.
- The clue to recognising it is the **inverted P waves in lead I** and the **upright aVR** which are both highly unusual for a 12-lead ECG.
⇒ **The correct course of action → Repeat the ECG again**

Early repolarization variant

Mechanism

- It expresses as an early uptake of the ST segment before the descending limb of the R wave has reached the baseline.

Features

- benign but often alarming ST segment elevation
⇒ **Classically the ST segment elevation during early exercise returns to normal as heart rate increases further**
- It is common in black males
- Clinical evaluation is entirely normal
- ST elevation is usually seen in the precordial leads

ECG: U wave

Causes of prominent U waves are:

- Hypokalaemia
- Cardiovascular drugs, e.g. digitalis, quinidine, amiodarone
- Psychotropic drugs, e.g. phenothiazines, tricyclic antidepressants.

Cardiac catheterisation and oxygen saturation levels

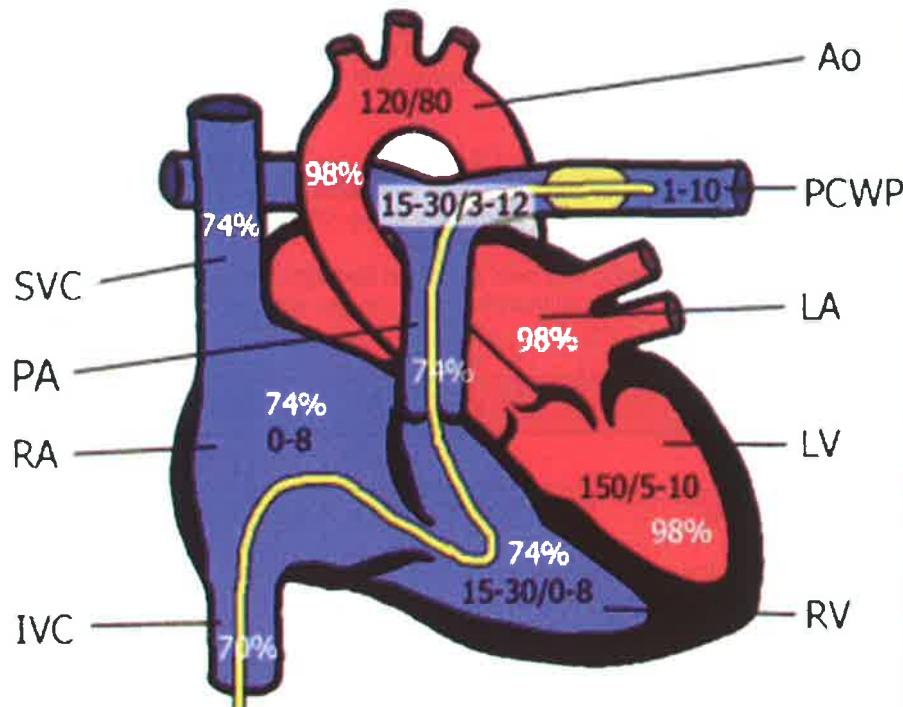
- Questions regarding cardiac catheterisation and oxygen saturation levels can seem daunting at first but a few simple rules combined with logical deduction can usually produce the answer.

Let's start with the basics:

- deoxygenated blood returns to the right side of the heart via the superior vena cava (SVC) and inferior vena cava (IVC). It has an oxygen saturation level of around **70%**. The right atrium (RA), right ventricle (RV) and pulmonary artery (PA) normally have oxygen saturation levels of around 70%
- the lungs oxygenate the blood to a level of around **98-100%**. The left atrium (LA), left ventricle (LV) and aorta should all therefore have oxygen saturation levels of 98-100%

Some examples:

Diagnosis & notes	RA	RV	PA	LA	LV	Aorta
Normal	70%	70%	70%	100%	100%	100%
Atrial septal defect (ASD) The oxygenated blood in the LA mixes with the deoxygenated blood in the RA, resulting in intermediate levels of oxygenation from the RA onwards	85%	85%	85%	100%	100%	100%
Ventricular septal defect (VSD) The oxygenated blood in the LV mixes with the deoxygenated blood in the RV, resulting in intermediate levels of oxygenation from the RV onwards. The RA blood remains deoxygenated	70%	85%	85%	100%	100%	100%
Patent ductus arteriosus (PDA) Remember, a PDA connects the higher pressure aorta with the lower pressure PA. This results in only the PDA having intermediate oxygenation levels	70%	70%	85%	100%	100%	100%
VSD with Eisenmenger's	70%	70%	70%	100%	85%	85%
PDA with Eisenmenger's	70%	70%	70%	100%	100%	85%
ASD with Eisenmenger's	70%	70%	70%	85%	85%	85%



Cardiac catheter data

Normal cardiac pressures & oxygen saturation

Guidelines for the Interpretation of Cardiac Catheter Data

- Right-heart saturations do not exceed 75%. Saturations more than this are suggestive of a left-to-right shunt.
- **Atrial septal defect (ASD) :** The oxygen saturation in the RA and SVC should be the same. But in ASD there is a step-up in oxygen saturation at the level of the RA. This can only result from the addition of oxygenated blood to the deoxygenated blood in the right heart circulation, that is, an abnormal connection between the right and left sides of the heart.
 - ⇒ **Primum ASD:**
 - ❖ The location of the step-up is suggestive of a primum defect since these lesions occur low down in the A-V septum, lying immediately above the atrioventricular valves.
 - ❖ These lesions can affect the function of the anterior leaflet of the mitral valve, causing mitral regurgitation.
 - ❖ **high pressures of Right ventricular are more likely to occur with primum ASDs.**
- **Patent ductus arteriosus (PDA)**
 - ⇒ unexpected step-up in oxygen saturation between the RV and PA.
 - ⇒ **high pulmonary artery pressures**
 - ⇒ high wedge pressure.

- ⇒ The change in O₂ saturation between the ascending and descending aorta strongly suggests the presence of a patent ductus.
- ⇒ Unfortunately the extremely elevated right sided pressures are indicative of advanced disease, not amenable to surgical correction. In late disease the machinery murmur said to be characteristic of the disease may well not be audible.
- Left-heart saturations vary from 96–98%. Saturations less than this are suggestive of a right-to-left shunt.
- In right-to-left shunts, the arterial saturations do not change with inspired high-concentration oxygen.
- **Ventricular septal defect (VSD)**
 - ⇒ There is a step-up in the oxygen saturation between the RA and RV. This can only occur when there is an abnormal connection between these two chambers, that is, via a VSD.
 - ⇒ This is confirmed by the raised right ventricular pressures.
 - ⇒ **VSD with Eisenmenger's syndrome**
 - the pressures in the RV and PA are markedly elevated, but RA pressure is normal.
 - The left ventricular oxygen saturation is low, which raises the possibility of a right to left cardiac shunt mixing desaturated RV blood with LV saturated blood (due to right ventricular pressures exceeding left ventricular pressure).
 - ⇒ post-MI VSD and papillary rupture are difficult to distinguish clinically.
 - The diagnosis is established by demonstration of a left to right shunt.
 - ❖ if there is a step-up in the oxygen saturation between the RA and PA → VSD
 - ❖ if there is no step-up, → papillary muscle rupture.
- **Fallot's tetralogy**
 1. **VSD:** step-down in oxygen saturation between LA and LV, indicating right to left shunt at the level of the ventricles.
 2. **Pulmonary stenosis:** there is ↑mmHg gradient across the pulmonary valve (RV systolic - PA systolic).
 3. **RVH:** Right ventricular pressures are high and there is a right to left shunt, which indicated by the oxygen saturations.
 4. **Over-riding aorta:**
 - ❖ there is a further step-down in oxygen saturation between the LV and aorta.
 - ❖ This could occur in either Fallot's or with a patent ductus arteriosus with right to left shunting.
 - ❖ However, given the other features of Fallot's, this is most likely to be caused by an over-riding aorta with reduced saturations due to a mixture of deoxygenated blood from the RV entering the left heart circulation.
 - ❖ The over-riding aorta receives a mixture of blood from the left and right ventricles as is formed above a VSD.
- Pulmonary hypertension does not occur in Fallot's tetralogy due to narrowing of the right ventricular outflow tract/ subpulmonary valve stenosis.
- A VSD with a right-to-left shunt and pulmonary stenosis can be differentiated from Fallot's tetralogy by examining the oxygen saturation in the left ventricle and the ascending aorta.
 - ⇒ In the case of a VSD, the saturations in the left ventricle and the aorta will both be low and very similar.

- ⇒ In the case of **Fallot's tetralogy**, the aortic oxygen saturation will be much lower than the oxygen saturation in the left ventricle because the right ventricle pumps most of the deoxygenated blood into the overriding aorta.
- A pulmonary artery pressure exceeding 35 mmHg is suggestive of **pulmonary hypertension**.
- A pressure drop of more than 10 mmHg across the aortic or pulmonary valve is suggestive of **aortic or pulmonary stenosis**, respectively.
- The diagnosis of **mitral regurgitation** cannot be made unless you are given the PCWP 'v-wave'. A v-wave higher than 20 mmHg is highly suggestive of mitral regurgitation.
- The right and LVEDP and the left and right atrial pressures are roughly equal in pericardial constriction
- When interpreting right heart catheter data, remember the saturation should decrease gradually as the venous blood reaches the pulmonary capillary wedge saturation, which should be equal to arterial blood.
- In Ebsteins anomaly there should be elevated RA pressure due to significant tricuspid regurgitation.
- **Hypertrophic cardiomyopathy**
 - ⇒ Left ventricular pressures are high with a steep drop-off between the LV and aortic systolic pressures.
- Anomalous pulmonary venous drainage to SVC
 - ⇒ normally oxygenation in the superior vena cava should always be lower than the inferior vena cava, due to the high oxygen demands from the brain.
 - ⇒ If SVC sats is markedly higher than the IVC, suggest a diagnosis of **anomalous pulmonary venous drainage** of more highly oxygenated blood into the SVC (left to right shunt).

What is meaning of "valve gradient"?

- The valve's gradient describes the severity of the narrowing of the valve by the increase in pressure behind it.
- It helps to measure the amount of blood that is able to pass through the valve.
- It also indicates whether the "velocity" (or speed of movement) of the blood flow is increased because of the increased pressure behind the narrowed valve.

Diagnosis of tricuspid stenosis

- mean gradient by echocardiogram or cardiac catheterisation of 2 mmHg or greater, but is usually found to be >7 to 10 mmHg in severe TS

Diagnosis of pulmonary hypertension

- If the pulmonary arterial pressure is greater than the normal one-fifth of systolic measurements → pulmonary hypertension is present.

Diagnosis of right ventricular failure

- The right atrial pressure is grossly elevated, with a normal wedge pressure.
 - ⇒ Normal right atrial pressure = (4–8) mmHg.
 - ⇒ Normal indirect left atrial mean pressure (wedge) = (5–10) mmHg.
 - normal wedge pressure excludes acute left ventricular failure or acute mitral regurgitation.

Diagnosis of aortic stenosis

- a greater than 25mmHg gradient across the aorta valve, demonstrating moderate aortic stenosis.
- systolic gradient of ↑ mmHg across the aortic valve (LV systolic pressure - aortic systolic pressure), indicating critical aortic stenosis.
- Hypertrophic cardiomyopathy may result in similar pressure differences, but given the clinical information, aortic stenosis is far more likely than hypertrophic obstructive cardiomyopathy (HOCM) in an old patient.
- A guide to determining the severity of aortic stenosis is given below:

Severity of aortic stenosis	Severity Valve area (cm ²)	Mean gradient (mmHg)
Mild	>1.5	<25
Moderate	1.0-1.5	25-50
Severe	<1.0	>50
Critical	<0.7	>80

Diagnosis of mitral stenosis

- A normal mitral valve expects less than 5mmHg pressure difference.
- Using these inferences, the mitral valve gradient is calculated by the capillary wedge pressure of mmHg (same as the left atrial pressure) minus the diastolic left ventricular pressure of mmHg: **the mmHg difference more than 5 demonstrates mitral stenosis.**
- The PCWP is equal to the LVEDP. When the PCWP exceeds the LVEDP, the diagnosis of **mitral stenosis** should be considered.
- The gradient across the mitral valve (LA pressure - LV end diastolic pressure); it is usual to use the PCWP as a surrogate for LA pressure.**
- There is also evidence of right ventricular hypertrophy, with **markedly elevated RV pressures due to secondary pulmonary hypertension.**
- The severity of mitral stenosis can be graded:

Severity of mitral stenosis	Severity Valve area (cm ²)	Gradient (mmHg)
Mild	1.6-2.0	<5
Moderate	1.0-1.5	5-10
Severe	<1.0	>10

Aortic incompetence

- wide pulse pressure in the aorta
- high left ventricular end-diastolic pressure (LVEDP).
 - ⇒ LVEDP greater than 20 mmHg is suggestive of irreversible LV dysfunction.
- All left heart valve diseases can ultimately cause elevated right heart pressures

Coarctation of the aorta

- There is a steep systolic gradient between the left ventricle and the femoral artery**

Pulmonary artery floatation catheter findings:

- if the pulmonary artery occlusion pressure is low with a relatively low cardiac index, suggesting the patient is hypovolaemic**, even in spite of high right atrial pressure.
 - ⇒ A fluid challenge should be performed, and values re-measured to assess response.
 - ⇒ In a fluid replete patient, the occlusion pressure would be higher (usually >13 mmHg)
- if the Pulmonary artery occlusion pressure is high and cardiac index low (i.e. <2.5 L/min/m²) this would be more suggestive of cardiogenic shock.

Pulmonary artery floatation catheter findings:

- Low pulmonary artery occlusion pressure + low cardiac index** → hypovolaemia
- High pulmonary artery occlusion pressure + low cardiac index** → cardiogenic shock

Hyperthyroidism and cardiac catheterisation:

- Cardiac catheterisation requires the use of an iodine-containing contrast.
- This may worsen hyperthyroidism caused by toxic multinodular goitre, whereas it may improve the symptoms in patients with Grave's disease (Wolff-Chaikoff effect).
- The most reliable diagnostic method is a radionuclide (**99Tcm, 123I or 131I**) scan of the thyroid, which will distinguish the diffuse, high uptake of Grave's disease from nodular thyroid disease.**
- If a toxic multinodular goitre or toxic adenoma is detected, the patient should receive an antithyroid drug before undergoing catheterisation.
- The antithyroid medication must be continued for at least 2 weeks after the procedure.

Pulmonary capillary wedge pressure

- Pulmonary capillary wedge pressure (PCWP) is measured using a balloon tipped Swan-Ganz catheter which is inserted into the pulmonary artery.
- The pressure measured is similar to that of the left atrium (normally 6-12 mmHg).
- The PCWP provides an indirect measurement of the left atrial pressure, and since the left atrial pressure is increased, the PCWP will also be increased.
- One of the main uses of measuring the PCWP is determining whether pulmonary oedema is caused by either heart failure or acute respiratory distress syndrome.
- In many modern ITU departments PCWP measurement has been replaced by non-invasive techniques.

Which method is an appropriate of measuring adequate intravascular filling?

- PiCCO (pulse contour cardiac output)**
 - ⇒ PiCCO gives indications of cardiac output, extravascular lung water, intravascular filling and only requires a central line and a PiCCO femoral arterial line and as such is relatively simple to use.

Cardiac imaging: non-invasive techniques excluding echocardiography**Nuclear imaging**

- These techniques use radiotracers which are extracted by normal myocardium.
- Examples include:
 - ⇒ Thallium
 - Nuclear isotopes are picked up by the Na/K ATPase of normal myocardium.
 - If cardiac tissue is alive and perfused, it will pick up the nuclear isotope.
 - To the myocardium, thallium looks like potassium.
 - Decreased uptake = Damage
 - ⇒ technetium (99mTc) sestamibi:
 - a coordination complex of the radioisotope technetium-99m with the ligand methoxy-iso-butyl isonitrile (MIBI), used in 'MIBI' or cardiac Single Photon Emission Computed Tomography (SPECT) scans
 - ⇒ **fluorodeoxyglucose (FDG):**
 - used in Positron Emission Tomography (PET) scans
 - Cardiac PET is predominately a research tool at the current time

SPECT

- The primary role of SPECT is to assess myocardial perfusion and myocardial viability.
- Two sets of images are usually acquired. First the myocardium at rest followed by images of the myocardium during stress (either exercise or following adenosine / dipyridamole).
- By comparing the rest with stress images any areas of ischaemia can be classified as reversible or fixed (e.g. Following a myocardial infarction).

MUGA

- Multi Gated Acquisition Scan, also known as radionuclide angiography
- radionuclide (technetium-99m) is injected intravenously
- the patient is placed under a gamma camera
- may be performed as a stress test
- can **accurately measure left ventricular ejection fraction**.
- Typically used before and after cardiotoxic drugs are used

Cardiac Computed Tomography (CT)

- Cardiac CT is useful for **assessing suspected ischaemic heart disease**, using two main methods:
 - ⇒ **calcium score**:
 - there is known to be a correlation between the amount of atherosclerotic plaque calcium and the risk of future ischaemic events.
 - Cardiac CT can quantify the amount of calcium producing a 'calcium score'
 - ⇒ **contrast enhanced CT**:
 - allows visualisation of the coronary artery lumen
- If these two techniques are combined cardiac CT has a **very high negative predictive value** for **ischaemic heart disease**.
- The updated NICE guidelines recommends that **cardiac CT** is the **first-line investigation** for patients presenting with **new-onset chest pain due to suspected CAD**.

Cardiac MRI

- Cardiac MRI (commonly termed CMR) has become the gold standard for providing **structural images of the heart**.
- It is particularly **useful in**:
 - ⇒ assessing congenital heart disease,
 - ⇒ determining right and left ventricular mass and
 - ⇒ differentiating forms of cardiomyopathy.
 - ⇒ Myocardial perfusion can also be assessed following the administration of gadolinium.
- Currently CMR provides limited data on the extent of coronary artery disease.

Mitral stenosis (MS)

Pathophysiology

- MS → mechanical obstruction of blood flow into the left ventricle (LV) → limited diastolic filling of the LV (\downarrow end-diastolic LV volume) → decreased stroke volume → decreased cardiac output (forward heart failure)
- MS → \uparrow left atrial pressure → backup of blood into lungs → \uparrow pulmonary capillary pressure → cardiogenic pulmonary edema → pulmonary hypertension → backward heart failure and right ventricular hypertrophy

Causes

- **Common → Rheumatic fever**
 - ⇒ Rheumatic valve disease is increasing uncommon in the UK, but can still be seen in other parts of the world.
 - ⇒ The physiological stress of pregnancy can exacerbate the features of rheumatic mitral stenosis.
- **Rare**
 - ⇒ Calcification of the mitral valve annulus
 - ⇒ Autoimmune diseases: systemic lupus erythematosus, rheumatoid arthritis
 - ⇒ Congenital
 - ⇒ Mucopolysaccharidoses
 - ⇒ carcinoid

Features

- Malar flush: Mauve discoloration of the cheeks due to low cardiac output and systemic vasoconstriction
- Dyspnea
- Low volume pulse
- **Tapping apex beat**
- **Auscultation**
 - ⇒ Loud first heart sound (S1)
 - ⇒ Mid-late diastolic murmur (with pre-systolic accentuation)
 - heard best at the 5th left intercostal space at the midclavicular line (the apex) in expiration.
 - ⇒ **Opening snap**
 - **A high frequency, early to mid-diastolic sound, heard after S2**
 - **suggests that the mitral valve is mobile**
 - **opening snap is not heard when the mitral valve is heavily calcified**
 - the high left atrial pressure → **rapid reversal of anterior mitral valve leaflet towards the left ventricle in early diastole** lead to early diastole sound.

Complications

- Compression by the enlarged left atrium
 - ⇒ Compression of the esophagus → Dysphagia
 - ⇒ Compression of the recurrent laryngeal nerve → Hoarseness (known as Ortner syndrome.)
- Atrial fibrillation
 - ⇒ Embolic disease (e.g., stroke, mesenteric ischemia)
 - ⇒ Patients with mitral stenosis often develop acute heart failure following the onset of atrial fibrillation.
- Leads to left atrial enlargement, but the **left ventricle is usually small**.
- Right heart failure (paroxysmal nocturnal dyspnea, orthopnea, lower limb pitting edema , bibasilar rales)
- Hemoptysis

Mechanism of opening snap earlier in worsening MS

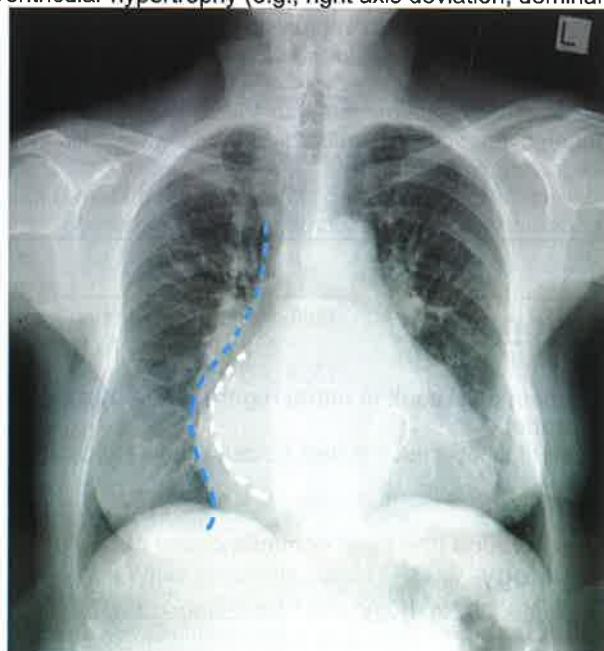
- The mitral valve opens when LA pressure > LV pressure. Worse MS = Higher LA pressure. Higher LA pressure pushes the mitral valve open earlier.

Features of severe MS

- length of murmur increases
- opening snap becomes closer to S2. (shorter interval between S2 and opening snap)
 - ⇒ **opening snap is characteristically lost with heavy valvular calcification**
- high transvalvular pressure gradient and high blood flow velocity.

Investigations

- **Transthoracic echocardiography (TTE)**
 - ⇒ TTE is the most important test for diagnosing and guiding the treatment of mitral stenosis.
 - ⇒ Characteristic findings include:
 - Reduced mitral valve area (MVA): $\leq 1.5 \text{ cm}^2$ is considered to be severe MS
 - Thickened, calcified leaflets with commissural fusion
 - RV dilation
 - LA enlargement
 - Evidence of pulmonary hypertension
- **Chest x-ray**
 - ⇒ Left atrial enlargement may be seen
 - The main bronchi appear elevated and have $> 90\%$ angulation (splayed).
 - Straightening or convexity of the left cardiac border
 - Double density sign (the silhouette of the enlarged left atrium appears near that of the right atrium.)
 - ⇒ Cardiomegaly
 - ⇒ Pulmonary congestion
- **ECG**
 - ⇒ Often normal
 - ⇒ Characteristic findings include:
 - ⇒ Left atrial enlargement/P mitrale
 - ⇒ Atrial fibrillation
 - ⇒ Right ventricular hypertrophy (e.g., right axis deviation, dominant R wave in lead V1)



Chest x-ray from a patient with mitral stenosis. This patient has had a sternotomy and a prosthetic mitral valve. There is splaying of the carina with elevation of the left main bronchus, a double right heart border and cardiomegaly. The features are those of left atrial enlargement. Although the entire heart is enlarged, a double contour is seen through the right side of the heart. The more medial line is the enlarged left atrium (white dotted line) and the heart border is more lateral (blue dotted line).

Management

- **Asymptomatic:** echocardiography follow-up
 - ⇒ every 3 to 5 years if the mitral valve area (MVA) is $>1.5 \text{ cm}^2$
 - ⇒ every 1 to 2 years if the MVA is 1.0 to 1.5 cm^2
 - ⇒ once per year if the MVA is $<1.0 \text{ cm}^2$.
- **Symptomatic with severe MS**
 - ⇒ 1st line: transcatheter valvotomy : in patients with favorable valve morphology
 - ⇒ 2nd line: surgical mitral valve replacement: if transcatheter valvotomy is unsuitable.

Indications for surgical mitral valve replacement

- Unfavorable anatomy for transcatheter valvotomy (Percutaneous mitral valve balloon commissurotomy)
- Presence of thrombus in the left atrium
- Mixed valvular disease (e.g., severe MR, tricuspid disease)

Mitral stenosis in pregnancy

- **Overview**
 - ⇒ **MS is poorly tolerated in pregnancy due to volume overload.**
 - ⇒ Pregnancy can unmask previously undiagnosed obstructive valvular heart disease. The symptoms may develop in the second trimester, when the demand for cardiac output increases by around 70%.
- **Treatment**
 - ⇒ **Medical therapy** for mild symptoms (beta blockers and/or diuretics)
 - ⇒ **Percutaneous mitral balloon valvuloplasty (PMBV) should be carried out for severe mitral stenosis in patients who remain symptomatic despite medical therapy.**
 - ⇒ Symptomatic patients with moderate to severe MS (mitral valve area $\leq 1.5 \text{ cm}^2$) should undergo intervention, preferably percutaneous balloon mitral valvotomy, before pregnancy.
 - ⇒ Vaginal delivery with assisted second stage is the preferred mode of delivery with caesarian delivery generally reserved for obstetric reasons.

Mitral regurgitation (MR)

Valvular anatomy

- left atrial enlargement can result in mitral regurgitation by affecting which leaflet?
 - ⇒ posterior leaflet
 - anterior leaflet is not affected, because of its attachment to the root of the aorta.

Pathology

- **Myxomatous degeneration (the most common cause of MR in UK).**

Risk factors and aetiology

- MR associated with Marfan syndrome and Ehlers-Danlos syndrome.
- cardiac complication seen 3-14 days post-myocardial infarction that occurs due to papillary muscle rupture.

Features

- Symptoms
 - ⇒ dyspnoea, usually on exertion, → decreased exercise tolerance.
 - ⇒ palpitations,

- Signs
 - soft S1, split S2
 - pan-systolic murmur
 - Typically presents as a holosystolic blowing murmur at the apex, radiating to axilla.
 - **intensified by** isometric exercise and thus helps to differentiate it from other systolic murmurs.
 - Sudden standing and amyl nitrite **decrease** the murmur.

Diagnosis

- Transthoracic echo is the diagnostic test of choice

Which feature suggests more severe mitral regurgitation?

- As mitral regurgitation becomes more severe, the left ventricle enlarges, and the **apex beat displaces**, and a **systolic thrill** can develop.

Management

- asymptomatic chronic MR:
 - ⇒ left ventricular ejection fraction >60% and/or left ventricular end-systolic diameter <45 mm → (ACE) inhibitors + beta-blockers
 - ⇒ left ventricular ejection fraction 60% or less and/or left ventricular end-systolic diameter 45 mm or more → surgery
- symptomatic chronic MR
 - ⇒ left ventricular ejection fraction 30% or more → surgery + medical treatment (ACE inhibitors, beta-blockers, and diuretics.)
 - ⇒ left ventricular ejection fraction <30% → medical treatment
 - intra-aortic balloon counterpulsation in severe acute cases

Mitral valve prolapse (MVP)

Epidemiology

- common, occurring in around 5-10 % of the population.
- the most common valvular defect in the United States
- more common in females.

Causes

- usually idiopathic
- inherited in an autosomal dominant fashion.
- may be associated with:
 - ⇒ congenital heart disease: PDA, ASD
 - ⇒ cardiomyopathy
 - ⇒ Turner's syndrome
 - ⇒ Marfan's syndrome,
 - ⇒ Fragile X
 - ⇒ osteogenesis imperfecta
 - ⇒ pseudoxanthoma elasticum
 - ⇒ Wolff-Parkinson White syndrome
 - ⇒ long-QT syndrome
 - ⇒ Ehlers-Danlos Syndrome
 - ⇒ polycystic kidney disease
 - ⇒ 15-40% of people with **panic disorder** have associated mitral valve prolapse.

Features

The late systolic murmur with mid systolic click is indicative of mitral valve prolapse where the posterior leaflets bulge during systole.

- atypical chest pain (the most common symptom)
- palpitations
- dyspnea, exercise intolerance,
- dizziness or syncope,
- panic and anxiety disorders.
- mid-systolic click (occurs later if patient squatting)
- late systolic murmur (longer if patient standing) heard best at the apex

Complications

- mitral regurgitation,
- arrhythmias (including long QT),
- emboli,
- sudden death

Treatment

- Mild to moderate mitral regurgitation
 - ⇒ follow-up in clinic with repeat echocardiograms to monitor progression.
- Mitral valve replacement is only indicated in:
 - ⇒ severe mitral regurgitation or
 - ⇒ if there are signs of concomitant LV compromise (reduced ejection fraction or new dilatation of the LV).
- If a surgical mitral valve replacement are indicated, coronary angiogram should be part of the pre-op work-up for potential concomitant coronary artery bypass grafting.

Aortic dissection

Aortic dissection

- type A - ascending aorta - control BP(IV labetalol) + surgery .
- type B - descending aorta - control BP(IV labetalol)
- It is most common between the ages of 50-70, being rare below the age of 40.

Stanford classification

- type **A** - Ascending aorta, (immediately above of the aortic valve) → **2/3 of cases**
- type B - descending aorta, (after the aorta arch) distal to left subclavian origin, 1/3 of cases

DeBakey classification

- type I - originates in ascending aorta, propagates to at least the aortic arch and possibly beyond it distally
- type II - originates in and is confined to the ascending aorta
- type III - originates in descending aorta, rarely extends proximally but will extend distally

Associations

- | | |
|--|---|
| <ul style="list-style-type: none"> • hypertension (The most common risk factor) • trauma (direct blunt chest trauma) • collagens: Marfan's syndrome, Ehlers-Danlos syndrome | <ul style="list-style-type: none"> • bicuspid aortic valve • Turner's and Noonan's syndrome • pregnancy • syphilis • Drugs (such as cocaine) |
|--|---|

Complications of backward tear

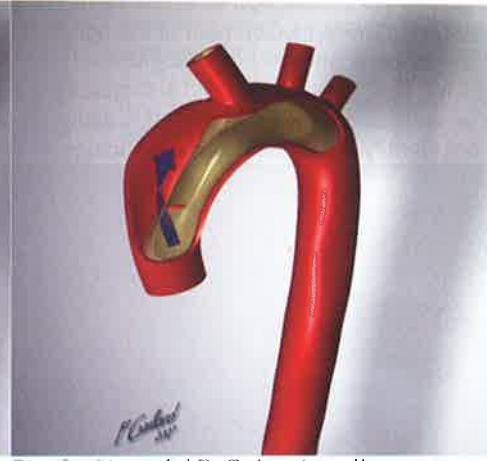
- aortic incompetence/regurgitation
- MI: inferior pattern often seen due to right coronary involvement

Complications of forward tear

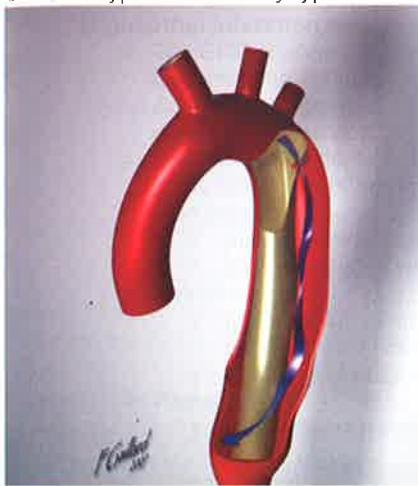
- unequal arm pulses and BP
- stroke
- renal failure



Stanford type A / DeBakey type I

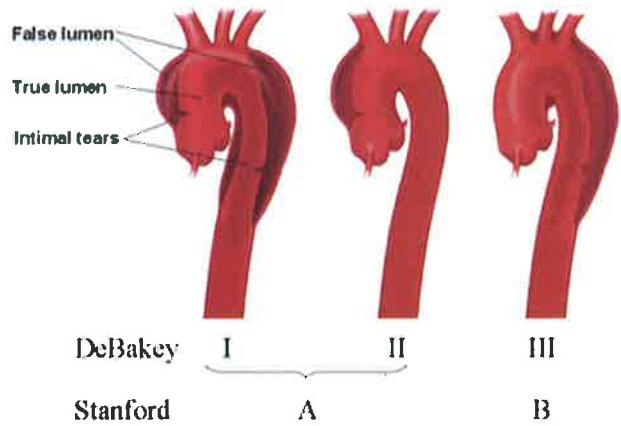


Stanford type A / DeBakey type II



Stanford type B / DeBakey type III

Anatomy and Classification of Aortic Dissection

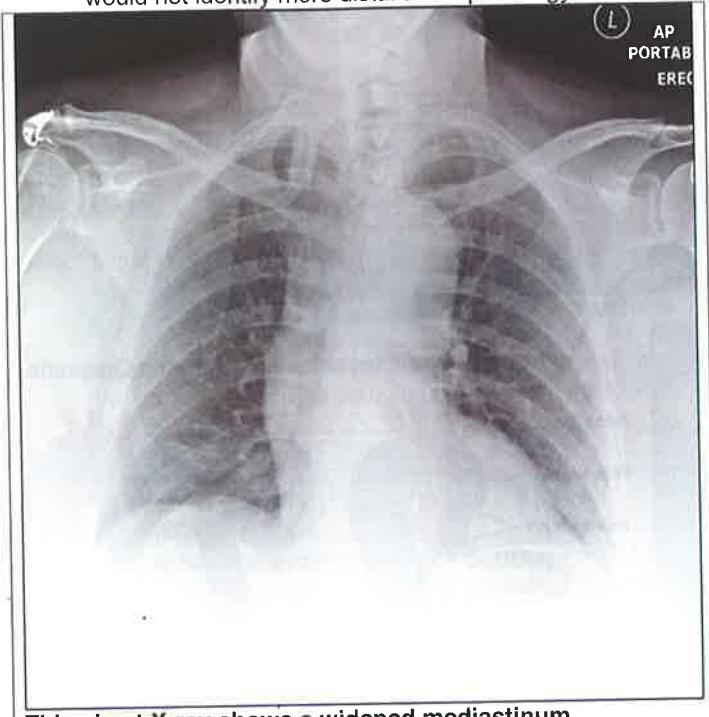


Investigations

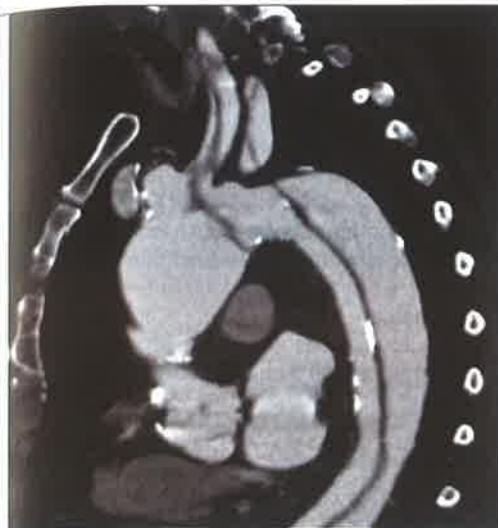
- **The best investigation is a CT chest with IV contrast (CT aortogram)** because the IV contrast will be able to best demonstrate the size and extent of the false lumen.
- Chest X-ray:
 - ⇒ is a useful **first line** investigation because its readily available it is, and useful for ruling out many other conditions.
 - ⇒ The chest X-ray may show a **widened mediastinum** (greater than 8 cm)
 - ⇒ but unfortunately, it is not a sensitive or specific investigation as 20% of patients present with normal chest X-ray and there are many causes of a widened mediastinum.

⇒ Looking for a separation of the intimal calcification from the outer aortic soft tissue border by 10 mm is an indication of the presence of a dissection.

- In a man with low blood pressure and vague abdominal pain, always be mindful of the possibility of dissection or aneurysmal rupture.
- Occasionally, there is involvement of the right coronary artery in the dissection process giving rise to the acute electrocardiographic changes.
- MRI has the best sensitivity (98%) and specificity (98%) for aortic dissection.
- Whilst an echocardiogram might identify disruption of the aortic root in a backwards tear, it would not identify more distal aortic pathology.



This chest X-ray shows a widened mediastinum



This computerised tomography (CT) scan demonstrates an obvious **flap in the thoracic aorta indicating aortic dissection**. The flap is in the middle of the descending aorta (the dark line) which separates the true lumen anteriorly from the intimal flap posteriorly. The aortic regurgitant murmur would alert the examiner to this and mediastinal widening may be seen on x ray.

Differential diagnosis

- **Myocardial infarction and aortic dissection: an important differential diagnosis**
 - ⇒ The ECG changes of inferior myocardial infarct suggest that the aneurysm has dissected the right coronary artery at its ascending aortic ostium.
 - ⇒ An inferior myocardial infarct is high in the differential; however thrombolysis will kill a patient with an aortic dissection. (delayed diagnosis and surgical treatment)
 - ⇒ up to 85% of patients with dissections may not receive appropriate medical treatment in the first hours of treatment due to an incorrect diagnosis
 - ⇒ pain onset
 - pain in aortic dissection is abrupt in onset and maximal at the time of onset.
 - pain associated with **MI** starts slowly and gains in intensity with time.
 - ⇒ Pain character
 - In dissection although tearing is the classical description, the pain is described as sharp more often than tearing, ripping, or stabbing.
 - In MI it is usually more oppressive and dull.
 - ⇒ Pain site
 - with distal dissections the pain location is between the scapulae and in the back.
- **Oesophageal rupture**
 - ⇒ **Features that favor oesophageal rupture over aortic dissection include:**
 - The history of onset while eating
 - Blood pressure equal in both arms
 - No diastolic murmur
 - Good peripheral pulses, and
 - Presence of a pleural effusion.

the history of chest pain **radiating to the back** is concerning., **early diastolic murmur** suggesting aortic valve regurgitation, **ECG changes in the inferior territory** and indicating occlusion of the right coronary artery. These features combined suggest that the **aortic dissection has tracked back to the heart** itself. The enlarged heart on chest X-ray may suggest a **haemopericardium**, and the patient should be assessed for **cardiac tamponade** given his **low blood pressure**. This patient is highly unstable and requires urgent cardiothoracic involvement . **the most appropriate next step in the management → Bedside echocardiogram and urgent cardiothoracic review**

Management

- **Type A**
 - ⇒ surgical management, but blood pressure should be controlled to a target systolic of 100–120 mmHg whilst awaiting intervention
 - ⇒ The most appropriate management strategy is to provide adequate analgesia and urgently reduce the blood pressure with intravenous antihypertensives: beta-blockers first line, and then nitroprusside. Then the cardiothoracic surgeons should be contacted.
 - ⇒ perioperative management of patients undergoing high risk vascular surgery
 - prophylactic beta blockers for high risk vascular surgery (including those **patients with COPD**).
 - ❖ Bisoprolol is the best clinical choice
 - ❖ **Atenolol is next best choice**; it is cardioselective and long acting, reducing risk of postoperative myocardial ischaemia and tachycardia.
- **Type B**
 - ⇒ conservative management
 - bed rest
 - reduce blood pressure IV labetalol to prevent progression
 - ⇒ endovascular repair of type B aortic dissection may have a role in the future

Complication

- **haemopericardium and cardiac tamponade**
 - ⇒ If the dissection (involving the ascending aorta (Stanford type A) results in a tear of the tunica externa, aortic blood can leak into the pericardium.
 - ⇒ **Management of aortic dissection complicated by haemopericardium and cardiac tamponade**
 - acute type A aortic dissection complicated by haemopericardium and cardiac tamponade:
 - ❖ **Relatively stable patient → immediate surgical repair and surgical evacuation of haemopericardium.**
 - ❖ Pericardiocentesis in these patients can increase the intra-aortic pressure and reopen the closed communication between false lumen and pericardium. This can lead to recurrent cardiac tamponade that may be lethal.
 - marked hypotension or electromechanical dissociation → pericardiocentesis

Prevention

- The management of patients with predisposing inherited diseases such as Marfan's syndrome and Ehlers-Danlos syndrome should include:
 - ⇒ Periodic aortic diameter screening.
 - ⇒ Lifelong beta-blockade.
 - ⇒ Consideration of prophylactic replacement of the aortic root if dilated.
 - ⇒ Moderate restriction of physical activity.

Prognosis

- Mortality for untreated aortic dissection is 25–30% at 24 h and 65–70% at 2 weeks
- dissections confined to the descending aorta are associated with better survival (80%).

Aortic aneurysms

Aortic aneurysms

- **Most common cause of aneurysms → atherosclerosis**
- The nice guidelines state that an aortic aneurysm of greater than 5.5 cm in diameter should be treated. Below this size, the risk of dissection is outweighed by the risk of surgery.

Definition

- Localized dilation of all three layers of the abdominal aortic wall (intima, media, and adventitia) to ≥ 3 cm

Epidemiology

- Sex: ♂ > ♀: ~ 2:1

Risk factors

- Advanced age
- Smoking (most important risk factor)
- Atherosclerosis
- Hypercholesterolemia and arterial hypertension
- Positive family history

Localization

- **Infrarenal: below the renal arteries : Most common location**
- Suprarenal: above the renal arteries

Features

- Aortic aneurysms are usually asymptomatic or have nonspecific symptoms.
- Lower back pain
- Pulsatile abdominal mass
- Bruit on auscultation

Abdominal vs. thoracic aortic aneurysm

Characteristics	Abdominal aortic aneurysm	Thoracic aortic aneurysm
Location	<ul style="list-style-type: none"> Below the renal arteries (most common) 	<ul style="list-style-type: none"> Ascending aorta (most common)
Epidemiology	<ul style="list-style-type: none"> Advanced age Predominantly men More common than TAA 	<ul style="list-style-type: none"> Advanced age Predominantly men
Etiology	<ul style="list-style-type: none"> Smoking (most important risk factor) Atherosclerosis Hypercholesterolemia and arterial hypertension 	<ul style="list-style-type: none"> Arterial hypertension Bicuspid aortic valve Tertiary syphilis [10] Connective tissue diseases (e.g., Marfan syndrome, Ehlers-Danlos syndrome) Trauma Smoking
Clinical features	<ul style="list-style-type: none"> Pulsatile abdominal mass Bruit on auscultation Lower back pain 	<ul style="list-style-type: none"> Feeling of pressure in the chest Thoracic back pain
Diagnostics	<ul style="list-style-type: none"> Abdominal ultrasound (best initial and confirmatory test) 	<ul style="list-style-type: none"> Chest x-ray and CTA of chest
Therapy	<ul style="list-style-type: none"> Indications for repair <ul style="list-style-type: none"> ⇒ Diameter: ≥ 5.5 cm ⇒ Expansion rate: ≥ 1 cm/year ⇒ Symptomatic aneurysm ⇒ Complications (e.g., rupture) 	<ul style="list-style-type: none"> Indications for repair <ul style="list-style-type: none"> ⇒ Diameter: ascending aneurysm ≥ 5.5 cm; descending aneurysm ≥ 6.5 cm ⇒ Expansion rate: ≥ 1 cm/year ⇒ Symptomatic aneurysm ⇒ Complications (e.g., rupture)

Aortic regurgitation (AR)

Turner's syndrome - most common cardiac defect is bicuspid aortic valve

Causes

- due to valve disease
 - ⇒ bicuspid aortic valve
 - the most common cause of chronic AR in a young patient is a congenital bicuspid valve.
 - Bicuspid valve is also a common cause of early-onset aortic stenosis.
 - ⇒ infective endocarditis
 - the vegetations prevent the valve from creating a proper seal to prevent backflow during diastole.
 - ⇒ rheumatic fever
 - ⇒ connective tissue diseases e.g. RA/SLE
- due to aortic root disease

- ⇒ aortic dissection
- ⇒ Spondyloarthropathies (e.g. **ankylosing spondylitis**)
 - Ankylosing spondylitis is strongly associated with aortic regurgitation (occurs in 4% of cases).
 - An aortitis leads to aortic root dilatation with subsequent failure of leaflet coaptation.
- ⇒ hypertension
- ⇒ syphilis
- ⇒ Marfan's,
- ⇒ Ehler-Danlos syndrome

Causes of acute aortic regurgitation:

- ascending aortic dissection,
- infective endocarditis,
- **collagen vascular disorders such as Marfan's**
- trauma,
- dehiscence of a prosthetic valve.

Features

- early diastolic murmur
 - ⇒ heard along the left sternal border
 - ⇒ heard best while the patient is leaning forward on deep expiration.
- collapsing pulse
- wide pulse pressure
- mid-diastolic Austin-Flint murmur
 - ⇒ **It is a low frequency mid/late diastolic murmur**
 - ⇒ due to partial closure of the anterior mitral valve cusps caused by the regurgitation streams.
 - ⇒ There is no correlation between the presence of murmur and severity of AR, or aetiology.
- Note that there is often an aortic systolic flow murmur because there is an increased volume of blood in the LV due to the regurgitation.
- **Isolated LV dilatation (other chambers are normal) on ECHO due to volume overload**
 - ⇒ (AS, HOCM & ↑ BP → hypertrophy and a smaller LV cavity)
- Pulsus bisferiens; increased pulse pressure; visible, forceful, and bounding peripheral pulses (water hammer)
- Corrigan's pulse - visible and vigorous arterial pulsations in neck
- **Musset's sign - Bobbing of the head**, due to the arterial pulsations in the neck
- Quincke's sign - Capillary pulsations of the nail bed
- Muller's sign - Pulsations of the uvula
- Traube's sign - Loud systolic sound over femoral arteries ('pistol-shot' femorals)
- Duroziez sign - diastolic murmur proximal to femoral artery compression (due to flow reversal).
- Hill's sign (Higher systolic in leg than arm)

Signs of severity of AR

- Soft S2
- S3
- Austin Flint murmur (functional mdm at the apex due to regurgitant jet striking the anterior leaflet of the MV, therefore obstructing flow from the LA into the LV)
- characteristic of the murmur. (Duration and loudness) (cf with AS)
 - ⇒ As the lesion becomes **more severe**, the murmur shortens.
- Apex beat displaced and thrusting

- CCF (pulmonary oedema)
- Wide pulse pressure
- collapsing pulse,
- Hill's sign (Higher systolic in leg than arm)

Investigations

- **Echocardiogram** (the most important test)
 - ⇒ **Echocardiographic markers of severe AR**
 - Width of AR jet on colour flow > 65 % of LVOT
 - regurgitant fraction (RF) > 50 %
 - left ventricular end-diastolic diameter (LVEDD) > 70mm
 - left ventricular end-systolic diameter (LVESD) > 50mm
- Cardiac catheterisation
 - ⇒ may be performed if there is doubt over the severity of the regurgitation;
 - ⇒ severity is estimated by the degree of contrast that fills the ventricles after injection into the aortic root.

Treatment

- **Asymptomatic:**
 - ⇒ Asymptomatic without signs of sever AR:
 - **ACEI improve the prognosis in asymptomatic left ventricular dysfunction.**
 - Beta blockers **should be avoided** as these prolong diastole and therefore would increase the regurgitant fraction.
 - ⇒ Asymptomatic with signs of sever AR: surgery (**Indications for surgery in asymptomatic**):
 - signs of sever AR (echo criteria):
 - ❖ **LV ejection fraction under 50%**
 - ❖ LV end diastolic diameter greater than 7 cm
 - ❖ LV end systolic diameter greater than 5 cm.
 - ⇒ Patient has moderate AR and is undergoing coronary artery bypass surgery or other surgery involving the ascending aorta = surgery
- **Symptomatic:** Surgical
 - ⇒ Symptomatic (CCF, angina)
 - ⇒ deteriorating exercise tolerance, or
 - ⇒ abnormal hemodynamic responses to exercise, such as inability to augment blood pressure during a treadmill study

Aortic stenosis (AS)

Aortic stenosis - most common cause:

- younger patients < 65 years: bicuspid aortic valve
- older patients > 65 years: calcification

Aortic stenosis - S4 is a marker of severity

Angiodysplasia is associated with aortic stenosis

Epidemiology

- Aortic stenosis (AS) is the **most common valve problem** in the United Kingdom.

Risk factors

- age >60 years
- congenitally bicuspid aortic valve
- rheumatic heart disease
- **chronic kidney disease**

Causes

- degenerative calcification (tricuspid aortic valve calcification)
 - ⇒ most common cause in older patients > 65 years
- **congenital bicuspid aortic valve (BAV)**
 - ⇒ most common cause in younger patients < 65 years
 - ⇒ BAV is the most common form of congenital heart disease in adults (1-2% of population).
 - ⇒ The European Society of Cardiology states that there is an estimated 10% chance of a first degree relative being affected, which increases to 20-30% if you consider aortopathy. NOTCH1 gene mutations may be responsible.
 - It is possible that up to a third of relatives of patients with a bicuspid valve have valve or aortic abnormalities (often a dilated aorta).
 - NOTCH1 gene mutations may be responsible.
 - ⇒ **most helpful in establishing a diagnosis of congenital bicuspid valve as the aetiology is → Systolic ejection click** (best heard at the apex)
 - ⇒ **aortic valve replacement is eventually likely to be required**
 - Only 15% of patients with a bicuspid aortic valve will have a normally functioning valve in the fifth decade, and this often continues to deteriorate with age.
- William's syndrome (supravalvular aortic stenosis)
- post-rheumatic disease → fibrosis → Commissural fusion on ECHO
- subvalvular: HOCM

Pathophysiology

- **Pathophysiological response in aortic stenosis**
 - ⇒ **The LV hypertrophies increase (in the size of myocytes) in a concentric - rather than an eccentric (asymmetric) - manner in response to the increase in afterload.**

⇒ There is also an increase in interstitial collagen and little fibrosis

The triad of angina, left ventricular failure and syncope is classical to aortic stenosis.

Features

Narrow pulse pressure and new murmur → aortic stenosis

- **Symptoms**
 - ⇒ heart failure
 - ⇒ **SAD**
 - **Syncope** (40%)
 - **Angina or chest pain** (50%)
 - **Dyspnea** (60%)
 - ❖ Exertional dyspnea is the most common initial complaint
- **Physical exam**
 - ⇒ pulse
 - narrow pulse pressure
 - slow rising pulse
 - pulsus parvus et tardus
 - ❖ weak pulses with a delayed peak
 - ⇒ **Displaced apex beat**
 - ⇒ thrill
 - ⇒ ejection systolic murmur (ESM)
 - crescendo-decrescendo murmur
 - typically, a mid-systolic ejection murmur
 - heard best with the diaphragm of the stethoscope in the 2nd intercostal space in a patient who is sitting upright leaning forward.
 - ❖ in the elderly the more high frequency components of aortic stenosis may be heard best at the apex, the so called (Gallavardin phenomenon)
 - may have ejection click
 - **radiates to carotid arteries** (left often louder than right). radiate to the right neck
 - decreases with standing, Valsalva, or handgrip
 - increases with amyl nitrate, squat, or leg raise
 - The **intensity of the systolic murmur does not correspond to the severity of aortic stenosis**;
 - ❖ As LV contractility decreases in critical AS, the murmur becomes softer and shorter. The intensity of the murmur may therefore be misleading in these circumstances.
 - **the timing of the peak and the duration of the murmur correspond to the severity of aortic stenosis**.
 - ❖ The more severe the stenosis, the longer the duration of the murmur and the more likely it peaks at late systole.
 - ⇒ **S4** heart sound
 - from stiff or hypertrophic ventricle
 - ⇒ **S2 (Character of S2)**
 - soft/absent S2
 - paradoxical splitting of S2
 - ❖ heard on expiration rather than inspiration

Associated conditions

- hemolytic anemia
- predisposes to bleeding due to an acquired von Willebrand deficiency caused by turbulent flow across the stenotic valve.
- chronic gastrointestinal bleeding that is associated with angiodyplasia.

Severity of aortic stenosis

Features of severe aortic stenosis

1. narrow pulse pressure
2. slow rising pulse
3. delayed ESM
4. soft/absent S2
5. S4
6. thrill
7. duration of murmur
8. left ventricular hypertrophy or failure

- The severity of (AS) can be accurately assessed with echocardiography.
- the severity of AS is difficult to assess with echocardiography when cardiac output is low.
- Catheterization to determine the severity of AS is reserved for patients in whom echocardiography is nondiagnostic
- The volume of the murmur has **NO** relationship to the severity of the stenosis

In a patient with aortic stenosis, what **will lead to an overestimation of the severity of the problem** when assessed by echocardiography?

⌚ Aortic regurgitation

- due to large volumes of blood passing over the valve at high velocities

Which condition is most associated with quietening of the aortic stenotic murmur?

- ⌚ Left ventricular systolic dysfunction → decreased flow-rate across the aortic valve and hence a quieter murmur.
- ⌚ Atrial fibrillation
 - Where the R-R interval is particularly short, such as in atrial fibrillation, flow across the valve is reduced, as such the intensity of the murmur is variable and may be significantly reduced.
 - Aortic regurgitation has no effect on the intensity of the murmur, such that in patients with mixed aortic valve disease, the stenotic murmur is still clearly audible.

Conditions which leads to accentuation of the murmur → increased flow across the murmur.

- High output cardiac failure
- severe thyrotoxicosis

The predominant component of mixed aortic valve disease is determined by the murmur that is louder (ejection systolic murmur in aortic stenosis and mid diastolic murmur for aortic regurgitation).

Evaluation

- Severe AS is defined by a valve area of less than 1.0 cm^2 .
- **distinguish patients with true severe (AS) with secondary LV dysfunction from those who have a falsely low calculated aortic valve area because of low cardiac output.**

- ⇒ calculated valve area in patients with severe left ventricular (LV) dysfunction can be falsely low because low cardiac output reduces the valve opening forces.
- ⇒ It is important to distinguish patients with true severe (AS) with secondary LV dysfunction from those who have a falsely low calculated aortic valve area because of low cardiac output.
- ⇒ **An important method of distinguishing between the two conditions is to assess the haemodynamics after increasing the cardiac output by dobutamine infusion during echocardiography or cardiac catheterisation.**
 - Patients with truly severe AS manifest an increase in trans-aortic pressure gradient while the valve surface area remains the same during dobutamine infusion;
 - those with falsely low calculated valve area manifest an increase in calculated valve surface area.
- ⇒ Dobutamine echocardiography is also important to assess LV contractile reserve.
 - Patients who have 20% or more increase in stroke volume after dobutamine infusion have a much better prognosis after surgery compared to those who do not have LV contractile reserve.

What is the difference between aortic stenosis and aortic sclerosis?

- Both aortic stenosis and aortic **sclerosis** are :
 - ⇒ senile degeneration of the valve
 - ⇒ **there is an ejection systolic murmur,**
- Unlike aortic stenosis, **aortic sclerosis** have:
 - ⇒ Occur in > 25% of > 65 year of age
 - Aortic stenosis occur in > 2% of > 65 year of age
 - ⇒ **Absence of stenosis**
 - **no carotid radiation,**
 - **normal pulse** (character and volume)
 - **normal S2.**

Investigations

- Echocardiography
 - ⇒ transthoracic echocardiogram (TTE) initially
 - ⇒ transesophageal echocardiogram (TEE) is more accurate
 - ⇒ Although echocardiography will aid in diagnosis, **gradient across the aortic valve may be underestimated because of the possibility of multiple echo signals and co-existent left ventricular dysfunction.**
- **Left heart catheterization**
 - ⇒ most accurate diagnostic test (**the definitive investigation of choice**)
 - ⇒ to assess pressure gradient across the valve
 - ⇒ only indicated to confirm the diagnosis if echocardiography is unclear
 - ⇒ findings
 - elevated pressure gradient (> 30 mmHg)
 - ❖ In the context of poor LV function, the aortic valve gradient may be normal or only mildly raised in the presence of a severely narrowed aortic valve area.
- **The next step in management after diagnosis → Coronary angiography**
 - ⇒ Coronary artery disease (CAD) is common in patients with AS
 - ⇒ Progressing straight to aortic valve replacement is not advised; significant coronary artery disease should be ruled out first, as CABG may be required at the same time as valve replacement.

Patients undergoing open surgical valve replacement should first undergo **coronary angiography to exclude any coronary stenosis that could simultaneously be treated with bypass grafting.**

Management

Aortic stenosis management: AVR if symptomatic, otherwise cut-off is gradient of 50 mmHg

- if asymptomatic then observe the patient is general rule
- if symptomatic then valve replacement
 - ⇒ **The patient's symptomatology is the most important determinant in terms of the decision to operate**

There are three important factors to consider regarding management of aortic stenosis:

- 1. Presence of symptoms
- 2. The gradient across the valve on echocardiogram
- 3. Evidence of left ventricular dysfunction.

- **Symptomatic patient**
 - ⇒ Fit for surgery → aortic valve replacement
 - **the best treatment option in an older person who can undergo the surgery.**
 - ⇒ Not fit for aortic valve replacement
 - **Transcatheter aortic valve implantation (TAVI)**
 - ❖ The catheter-delivered device produces similar one-year survival as aortic valve replacement but a higher risk of stroke, TIAs and vascular complications.
 - Balloon valvuloplasty
 - ❖ Balloon aortic valvuloplasty is a palliative procedure prone to restenosis for patients unsuitable for other interventions.
- **Asymptomatic patient**
 - ⇒ **with severe stenosis (transvalvular gradient > 50 mmHg, valve area < 1 cm²)**
 - **but has an ejection fraction of less than 50%.**
 - **should be referred for aortic valve replacement or TAVI if unsuitable.**
 - ⇒ with severe stenosis but has an ejection fraction greater than 50%.
 - Exercise testing would be recommended
 - ❖ If pass exercise testing, then → reviewed in six months.
 - ⇒ echo follow-up
 - asymptomatic with mild stenosis → every 3 to 5 years
 - asymptomatic with moderate stenosis → every 1 to 2 years
 - asymptomatic with severe stenosis → every 6 to 12 months.

Indicator of poor prognosis

- **Clinical features of left ventricular failure**
 - ⇒ deteriorating LV function (ejection fraction less than 40%)
- Symptomatology
 - ⇒ exertional breathlessness or presyncope/syncope
- Increasing gradient across the valve (above 70 mmHg)
- Age of patient

Heyde's syndrome

- association between microcytic anaemia and calcific aortic stenosis.
- Heyde syndrome refers to a triad of
 1. aortic stenosis,
 2. acquired coagulopathy (von Willebrand syndrome type 2A) and
 3. anaemia due to bleeding from intestinal angiodyplasia or from an idiopathic site.
 - Angiodysplasia most commonly occur in the ascending colon, particularly the caecum.
- **Pathophysiology**
 - ⇒ destruction of von Willebrand's factor as the platelets traverse the stenosed valve resulting in bleeding per rectum.
- **Investigation**
 - ⇒ The investigation of choice after valve replacement is mesenteric angiography as the bleeding vessels are poorly visualised on colonoscopy.
 - This would look for the presence of angiodysplasia, which may be associated with aortic stenosis.
 - ⇒ All patients with aortic stenosis should be screened for iron deficiency anaemia.
- **Treatment**
 - ⇒ replace the valve
 - ⇒ Resection of the diseased bowel has also been described as a treatment.
- There is an association with jaundice and aortic stenosis; this is thought to be due to microangiopathic haemolysis.

Williams syndrome

Supravalvular aortic stenosis is the congenital cardiovascular deformity most often associated with Williams syndrome.

- **Supra-valvar AS** is one of the characteristic findings of Williams syndrome along with:
 - ⇒ unusual elfin facies,
 - ⇒ excellent verbal skills contrasted with intellectual disability and lack of social inhibition,
 - ⇒ hypercalcemia (due to increased sensitivity to vitamin D.)
- caused by a microdeletion of the **elastin** gene on long arm of chromosome 7

Coarctation of the aorta

Definition:

- congenital narrowing of the descending aorta, most commonly at the site of insertion of the ductus arteriosus

Overview

- more common in males (despite association with Turner's syndrome)
- a bicuspid valve is found in approximately 50% of patients with coarctation of the aorta.
- **site of coarctation:**
 - ⇒ distal to the origin of the left subclavian artery
 - **The commonest site**
 - The systolic BP in the arms exceeds that in the leg.
 - ⇒ proximal to the origin of the left subclavian artery

- occurs in 15% of cases of coarctation
- if the systolic BP in the right arm is higher than that of the left arm by more than 30 mmHg, the left subclavian is involved in the coarctation (ie coarctation is proximal to the origin of the subclavian)

Features

- Most patients are asymptomatic
- infancy: heart failure
- claudication of the calf muscles.
 ⇒ pain in calves is almost certainly due to poor distal blood supply.
- Hypertension
 ⇒ the **most common** presenting feature in adults
- headache and nose bleeds occur due to hypertension proximal to the coarctation,
- differential blood pressures between the right and left arms
- radio-femoral delay
- mid systolic murmur, and thrill
 ⇒ maximal over back.
 ⇒ **continuous** murmur over the thoracic spine usually originates **from small, tight coarctation (< 2 mm)**.
- apical click from the aortic valve

Complications

- Secondary hypertension
- development of cerebral aneurysms
 ⇒ may present with intracranial haemorrhage from a ruptured berry aneurysm
- Left ventricular failure,
- Bacterial endocarditis.

Associations

- **Bicuspid aortic valve**
 ⇒ the **commonest associated congenital abnormality**
 ⇒ occurs in 50% of the coarctations.
- patent ductus arteriosus (PDA)
- Turner's syndrome
 ⇒ Female patients diagnosed with coarctation of the aorta should have a **karyotype analysis** to rule out Turner syndrome.
- berry aneurysms
- neurofibromatosis

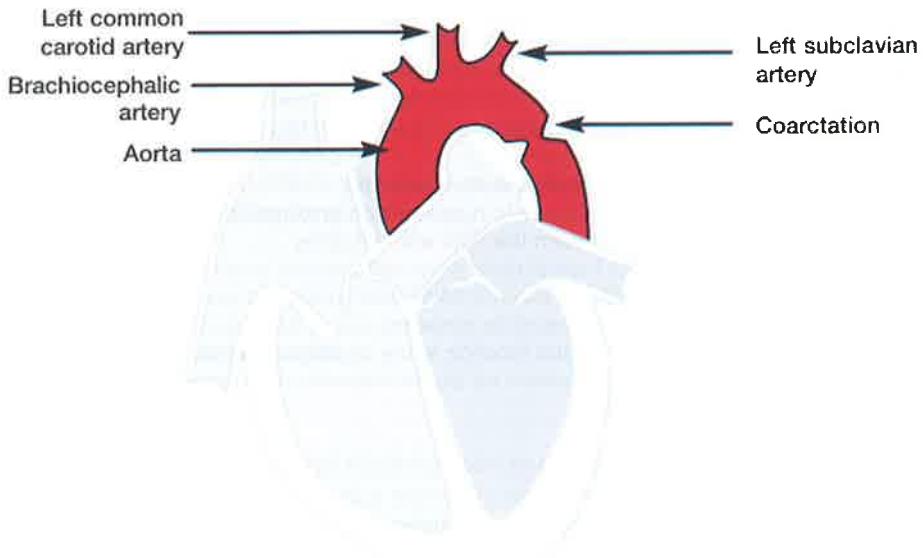
Investigations

- Radiograph
 - ⇒ Cardiomegaly
 - ⇒ ↑ pulmonary vascular markings
 - ⇒ rib notching
 - notching of the **inferior border of the ribs (due to collateral vessels)**
 - usually manifests in adults and older children, as it takes time to develop.
 - ⇒ may demonstrate an indentation of the aortic shadow at the site of the coarctation.
 - ⇒ rib notching is not seen in young children
- **Echocardiography with doppler (confirmatory test):**
 - ⇒ location and extent of stenosis;
 - ⇒ concurrent anomalies

Treatment

- Balloon angioplasty and stenting is
 - ⇒ the preferred intervention in adults.
 - ⇒ surgical correction is indicated if the **pressure gradient across the coarctation is above 20 mmHg**, even without associated hypertension.

- Prostaglandin E1 should be administered to neonates with aortic coarctation to keep the ductus arteriosus open.



Coarctation of the aorta.

Differences in blood pressure between arms:

- up to 10 mmHg difference → Normal variant (physiological)
- difference of greater than 10 mmHg: → abnormal:
 - ⇒ + radio-radial or **radio-femoral delay** (NO Leg claudication) → proximal coarctation of the aorta (involves the left subclavian artery origin)
 - ⇒ + **arm claudication**, intermittent vertigo, ataxia or diplopia, or facial sensory symptoms (NO Leg claudication) → Subclavian steal syndrome
 - ⇒ + **Leg claudication** (chronic intermittent leg pain, exacerbated by exercise and relieved by rest) → Peripheral vascular disease

Bicuspid aortic valve

Overview

- occurs in 1-2% of the population
- Bicuspid aortic valve tends to be sporadic although there is a reported familial incidence of approximately 9%.
- usually asymptomatic in childhood
- the majority eventually develop aortic stenosis or regurgitation

associated with:

- left dominant coronary circulation (the posterior descending artery arises from the circumflex instead of the right coronary artery)
- Turner's syndrome
- coarctation of the aorta (around 5% of patients)

Complications

- aortic stenosis/regurgitation as above
- higher risk for aortic dissection and aneurysm formation of the ascending aorta

Tricuspid regurgitation

Signs

- pan-systolic murmur
- giant V waves in JVP
- pulsatile hepatomegaly
- left parasternal heave

Causes

- pulmonary hypertension e.g. COPD (The most common cause)
- right ventricular dilation
- rheumatic heart disease
- infective endocarditis (especially intravenous drug users)
- **Ebstein's anomaly**
- carcinoid syndrome

Prosthetic valves

Prosthetic heart valves - mechanical valves last longer and tend to be given to younger patients

Prosthetic heart valves - antithrombotic therapy:

- bioprosthetic: aspirin
- mechanical: warfarin + aspirin

Mechanical valves - target INR:

- aortic: 2.0-3.0
- mitral: 2.5-3.5

- The most common valves which need replacing are the aortic and mitral valve.
- There are two main options for replacement: biological (bioprosthetic) or mechanical.

Biological (bioprosthetic) valves	Mechanical valves
Usually bovine or porcine in origin	The most common type now implanted is the bileaflet valve. Ball-and-cage valves are rarely used nowadays
Advantages : not requiring Long-term anticoagulation Warfarin may be given for the first 3 months depending on patient factors. Low-dose aspirin is given long-term.	Advantages : have a low failure rate
Disadvantages calcification over time. must be replaced within 5 to 10 years. Most older patients (> 65 years for aortic valves and > 70 years for mitral valves) receive a bioprosthetic valve	Disadvantages ↑ risk of thrombosis meaning long-term anticoagulation is needed. Aspirin is normally given in addition unless there is a contraindication. Target INR <ul style="list-style-type: none"> • aortic: 2.0-3.0 • mitral: 2.5-3.5

Following the 2008 NICE guidelines for prophylaxis of endocarditis → **antibiotics are no longer recommended for common procedures such as dental work.**

Which pathological findings in the bioprostheses has most likely led to the need for replacement?

⇒ **Calcification with stenosis**

Supraventricular tachycardia (SVT)

Definition

- The term 'SVT' literally indicates tachycardia [atrial rates >100 beats per minute at rest, the mechanism of which involves tissue from the His bundle or above. Traditionally, SVT has been used to describe all kinds of tachycardias apart from ventricular tachycardias (VTs) and AF.

Causes

- Atrioventricular nodal re-entry tachycardia (AVNRT).
 - ⇒ **the most common supraventricular tachycardia,**
 - ⇒ twice as **common in females** as in males
 - ⇒ the incidence is 1–3 per 1000
 - ⇒ Small elevations in troponin are occasionally seen in this situation, but there are no ECG changes to suggest a myocardial infarction.
- Atrioventricular re-entry tachycardias (AVRT)
- Junctional tachycardias.

Differential diagnosis

Paroxysmal SVT ⇒ would start and stop suddenly, **not gradually**.

Panic attacks ⇒ breathlessness and **palpitations** start and stop **gradually**.

Management

Vagal manoeuvres and adenosine are the treatments of choice for the acute therapy of SVT, and may also provide important diagnostic information.

- **Acute management**

⇒ haemodynamically **stable** patient:

- **1st line :** vagal manoeuvres : e.g. Valsalva manoeuvre
 - ❖ **Carotid sinus massage is contraindicated in patients with carotid vascular disease**
- **2nd line:** intravenous adenosine 6mg → 12mg → 12mg
 - ❖ Adenosine can cause flushing, chest pain, and dizziness.
 - ❖ contraindicated in asthmatics - **verapamil** is a preferable option
- **3rd line:** Verapamil or diltiazem i.v. or Beta-blockers (i.v. esmolol or metoprolol) should be considered if vagal manoeuvres and adenosine fail.
- **4th line:** Synchronized DC cardioversion

⇒ haemodynamically **unstable** patient:

- Synchronized DC cardioversion: start with **70-120 J biphasic** (100 J monophasic).

- **Prevention of episodes**

⇒ **1st line: beta-blockers or**
⇒ 2nd line: radio-frequency ablation

- Do not use flecainide or propafenone in patients with LBBB, or ischaemic or structural heart disease.
- Verapamil is not recommended in wide QRS-complex tachycardia of unknown aetiology.
- Flecainide and propafenone are not recommended as first-line antiarrhythmic drugs in patients with ventricular dysfunction and severe fibrosis.

SVT in pregnancy

- Tachyarrhythmias may increase during pregnancy although the causes are not entirely clear.

- **Termination of acute SVT:**

⇒ haemodynamic stable:

- Vagal manoeuvres and, if these fail, adenosine (**adenosine appears to be safe in pregnancy**).
- An i.v. beta-1 selective blocker (except atenolol) should be considered for acute conversion or rate control of SVT.

- **Prevention of recurrent SVT**

⇒ **in patients without WPW syndrome :**

- If possible, avoid all antiarrhythmic drugs during the first trimester of pregnancy.
- **1st line :** beta-1 selective agents (but not atenolol) beta-blockers.
 - ❖ The cardio-selective beta-1-blockers include atenolol, betaxolol, bisoprolol, esmolol, acebutolol, **metoprolol**, and nebivolol.
 - ❖ **Metoprolol** is the preferred and safest Beta-blocker in prophylaxis for SVT in pregnancy (it is a short acting β blocker and a TDS regimen is required).
- **2nd line:** verapamil
- **3rd line:** Fluoroless catheter ablation
- ⇒ **Prevention of recurrent SVT in patients with WPW syndrome :**
- **1st line : Flecainide or propafenone**
- **2nd line:** Fluoroless catheter ablation

Sinus arrhythmia

- The (ECG) shows normal P wave, PR interval, QRS complex and each P wave conducted to ventricles.
- **There is a gradual decrease in R-R interval and then an increase again. This slight beat-to-beat variation (rhythmic and cyclical variation) is termed as sinus arrhythmia.**
- the most common cause is respiration.
 - ⇒ Respiratory sinus arrhythmia is thus heart rate variability in synchrony with respiration, and is normal in children and young adults.
 - ⇒ The R-R interval decreases with inspiration and increases with expiration.
- Anxiety → reassured.



Premature ventricular ectopic (PVEs)

The first line management of supraventricular **ectopics** is generally **reassurance and lifestyle modifications** (eg: reduce alcohol and caffeine intake). If symptoms persisted, then a **beta blocker** would be **first line**.

- usually seen in normal hearts;
- palpitations are described as an early beat with a pause followed by an unusually strong or 'pounding' beat, or simply as a 'flip-flop';
 - ⇒ Symptoms are usually worse at rest and may disappear with exercise.
 - Symptoms which increase on exercise are more worrying and significant.
- may be associated with caffeine intake
- Investigations
 - ⇒ baseline ECG without symptoms: typically normal
 - ⇒ ambulatory ECG: isolated wide QRS complexes
 - If symptoms are short-lived but frequent (>2-3 times per week), use a 24-hour Holter monitor
 - If symptoms are short-lived and infrequent (<1 per week), use an event monitor or transtelephonic recorder
 - ⇒ Exercise stress testing
 - the relation of extrasystoles to exercise may have prognostic importance.
 - ⇒ Echocardiography - to assess LV function and heart structure.
- For PVE to be **significant** they have to meet the following criteria:
 - ⇒ Occurring frequently (6 or more beats/min)
 - ⇒ PVE in bigeminal rhythm
 - ⇒ PVE in short runs of ventricular tachycardia
 - ⇒ PVE exhibiting R-on-T phenomenon
 - ⇒ PVE associated with serious organic heart disease and left ventricular decompensation.
- Treatment
 - ⇒ **Not significant PVE → Reassurance**
 - ⇒ **Significant PVE**
 - **beta-blockers**
 - Radiofrequency catheter ablation of the ectopic focus
 - ❖ Curative with good outcome

Ventricular extrasystoles are the most common type of arrhythmia that occurs after myocardial infarction.

Management of symptomatic atrial extrasystoles

- beta-blockers (atenolol or metoprolol).
- Atrial extrasystoles arising from the pulmonary veins may be treatable by the procedure of pulmonary vein isolation.

Arrhythmogenic right ventricular cardiomyopathy(ARVC)

Overview

- Arrhythmogenic right ventricular cardiomyopathy (ARVC, also known as arrhythmogenic right ventricular dysplasia or ARVD) is a form of inherited cardiovascular disease which may present with syncope or sudden cardiac death.
- It is generally regarded as **the second most common cause of sudden cardiac death** in the young after hypertrophic cardiomyopathy.
- Although ARVC was initially described in the right ventricle, most patients have biventricular involvement.

Pathophysiology

- inherited in an autosomal dominant pattern with variable expression
- the right ventricular myocardium is replaced by fatty and fibrofatty tissue
- around 50% of patients have a mutation of one of the several genes which encode components of desmosome

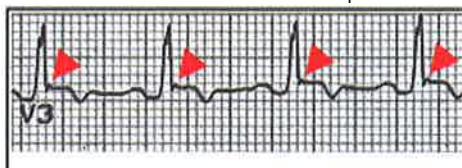
Presentation

- palpitations
- syncope
- sudden cardiac death

Investigation

epsilon potential is seen on the ECG of patients with → Right ventricular dysplasia

- ECG abnormalities in V1-3:
 - Typically, T wave inversion.
 - An **epsilon wave** is found in about 50% of those with ARV - this is best described as a terminal notch in the QRS complex



The epsilon wave

The epsilon wave (marked by red triangle), seen in ARVD.

- echo changes are often subtle in the early stages but may show an enlarged, hypokinetic right ventricle with a thin free wall
- magnetic resonance imaging is useful to show fibrofatty tissue

Management

- drugs: sotalol is the most widely used antiarrhythmic
- catheter ablation to prevent ventricular tachycardia
- implantable cardioverter-defibrillator

Naxos disease

- an autosomal recessive variant of ARVC
- a triad of ARVC, palmoplantar keratosis, and woolly hair

Atrial fibrillation (AF) (NICE guideline April 2021)

Overview

- AF is the most commonly encountered cardiac arrhythmia.
- Hypertension is the most common risk factor for AF.
- In 15% of cases, AF is idiopathic
- AF **most commonly originates from the roots of the pulmonary veins.** (longitudinal smooth muscle fibers in the pulmonary vein)

Classification

Definition	Duration of atrial fibrillation
Paroxysmal	Up to 7 days
Persistent	Longer than 7 days
Permanent	Cardioversion failed or not attempted

Classification of atrial fibrillation.

Classification of atrial fibrillation (AF): AF classified into 3 patterns:

1. **first detected episode** (irrespective of whether it is symptomatic or self-terminating)
2. **recurrent episodes**, when a patient has 2 or more episodes of AF:
 - **paroxysmal AF:**
 - ❖ episodes of AF terminate spontaneously.
 - ❖ episodes last less than 7 days (typically < 24 hours).
 - **persistent AF**
 - ❖ the arrhythmia is not self-terminating.
 - ❖ episodes usually last greater than 7 days
3. **permanent AF**
 - there is continuous atrial fibrillation which cannot be cardioverted or if attempts to do so are deemed inappropriate.
 - Treatment goals are therefore rate control and anticoagulation if appropriate

Symptoms and signs

- Symptoms
 - ⇒ Palpitations
 - ⇒ Dyspnea
 - ⇒ chest pain
- Signs
 - ⇒ irregularly irregular pulse

Complications

- AF is poorly tolerated in elderly and often leads to **pulmonary oedema** even in the presence of a relatively normal left ventricle (LV).

Diagnosis

- if an irregular pulse is detected ⇒ Perform a 12-lead electrocardiogram (ECG)
- In people with suspected paroxysmal AF undetected by ECG:
 - ⇒ if asymptomatic episodes are suspected or symptomatic episodes are less than 24 hours apart ⇒ use a 24- hour ambulatory ECG monitor
 - ⇒ if symptomatic episodes are more than 24 hours ⇒ use an ambulatory ECG monitor, event recorder or other ECG technology.

Assessment

- **Assessment of stroke and bleeding risks**
 - ⇒ Assess stroke risk by using the **CHA₂DS₂-VASc** score
 - ⇒ Assess the bleeding risk when considering starting anticoagulation by using the **ORBIT** bleeding risk score
 - ⇒ modify risk factors for bleeding:
 - uncontrolled hypertension
 - poor control of international normalised ratio (INR) in patients on vitamin K antagonists
 - concurrent medication, including antiplatelets, selective serotonin reuptake inhibitors (SSRIs) and non-steroidal anti-inflammatory drugs (NSAIDs)
 - harmful alcohol consumption
 - reversible causes of anaemia.
- **Assessment of cardiac function** by transthoracic echocardiography (TTE) as a baseline and to look for underlying structural or functional heart disease .

Management

- **Anticoagulation for stroke prevention**
 - ⇒ 1st line: direct-acting oral anticoagulant (e.g. Apixaban, dabigatran, edoxaban, rivaroxaban), if CHA₂DS₂-VASc score ≥ 1 for men or ≥ 2 for women.
 - ⇒ 2nd line: If DOAC are contraindicated or not tolerated ⇒ vitamin K antagonist.
 - ⇒ 3rd line: If anticoagulation is contraindicated or not tolerated ⇒ consider left atrial appendage occlusion (LAAO).
- **Rate and rhythm control**
 - ⇒ **Rate control:**
 - the first-line treatment for AF **except** in:
 - 1) AF due to reversible cause
 - 2) heart failure caused by AF
 - 3) new-onset AF
 - 4) atrial flutter which considered suitable for an ablation strategy to restore sinus rhythm
 - 5) if rhythm-control strategy would be more suitable based on clinical judgement.
 - Use beta-blocker (other than sotalol) or a rate-limiting calcium-channel blocker (diltiazem or verapamil)
 - Consider digoxin monotherapy for initial rate control if the person does no or very little physical exercise or other rate-limiting drug options are ruled out because of comorbidities.
 - ⇒ **Rhythm control:**
 - Do not offer class 1c antiarrhythmic drugs such as flecainide or propafenone to people with known ischaemic or structural heart disease.
 - If drug treatment for long-term rhythm control after successful cardioversion is needed:
 - ❖ 1st line: beta-blocker
 - ❖ 2nd line: dronedarone
 - Amiodarone for people with left ventricular impairment or heart failure.
 - In people with infrequent paroxysms and few symptoms, or if symptoms are induced by known precipitants (such as alcohol, caffeine), a '**no drug treatment**' strategy or a '**'pill-in-the-pocket'** strategy (in which antiarrhythmic drugs are taken only when an episode starts) should be considered
 - a '**'pill-in-the-pocket'** strategy: In people with paroxysmal AF if:

- ❖ infrequent symptomatic episodes + no left ventricular dysfunction, or valvular or ischaemic heart disease + systolic BP >100 mmHg and a resting heart rate > 70 bpm + able to understand how to, and when to take the medication.
- try to get the patient back into, and maintain, normal sinus rhythm. This is termed cardioversion.
- Drugs (pharmacological cardioversion) and synchronised DC electrical shocks (electrical cardioversion) may be used for this purpose
- indications of **Rhythm control** :
 - ❖ coexistent heart failure,
 - ❖ first onset AF or
 - ❖ where there is an obvious reversible cause.

Do not use rhythm control therapy in asymptomatic AF patients, nor in patients with permanent AF.

Rhythm control has no survival benefit over a rate control strategy

Reducing stroke risk → Anticoagulation

Young man with AF, no TIA or risk factors, no treatment is now preferred to aspirin (NO treatment)

Do not use antiplatelet therapy for stroke prevention in AF

- Some patients with AF are at a very low risk of stroke whilst others are at a very significant risk.
- NICE in 2014 suggest using the **CHA₂DS₂-VASc** score to determine the most appropriate anticoagulation strategy

	Risk factor	Points
C	Congestive heart failure	1
H	Hypertension (or treated hypertension)	1
A ₂	Age ≥ 75 years	2
	Age 65-74 years	1
D	Diabetes	1
S ₂	Prior Stroke or TIA	2
V	Vascular disease (including ischaemic heart disease and peripheral arterial disease)	1
S	Sex (female)	1

The table below shows a suggested anticoagulation strategy based on the score:

Score	Anticoagulation
0	No treatment
1	Males: Consider anticoagulation Females: No treatment (this is because their score of 1 is only reached due to their gender)
2 or more	Offer anticoagulation

Atrial fibrillation related to mitral stenosis

- **atrial fibrillation related to valvular heart disease → Warfarin**
 - ⇒ In patients with **non-valvular atrial fibrillation**, **novel oral anticoagulants** have the same efficacy as warfarin in preventing stroke.
- NICE guidelines suggest that valvular disease have high risk for thromboembolic events, and would benefit from anticoagulation.
- Mitral stenosis patients were excluded from the studies developing the CHADS-VASC score.
- None of the 'novel' anticoagulants currently available (rivaroxaban, apixaban, dabigatran) are indicated or licensed for atrial fibrillation related to valvular heart disease.

CHADS2-VASC scoring is generally used as a tool to assess need to anticoagulate a patient with AF. However, the following are **conditions that, if present, may trump the decision to anticoagulate**:

1. **valvular heart disease**
2. prior peripheral embolism, \leq nd
3. intracardiac thrombus.

Bleeding risk assessment (using the HASBLED scoring system)

- NICE recommend that we offer patients a choice of anticoagulation, including warfarin and the novel oral anticoagulants (NOACs).
- Aspirin is no longer recommended for reducing stroke risk in patients with AF
- Doctors have always thought carefully about the risk/benefit profile of starting someone on warfarin.
- A history of falls, old age, alcohol excess and a history of previous bleeding are common things that make us consider whether warfarinisation is in the best interests of the patient.
- NICE now recommend we formalise this risk assessment using the HASBLED scoring system.

	Risk factor	Points
H	Hypertension, uncontrolled, systolic BP > 160 mmHg	1
A	Abnormal renal function (dialysis or creatinine > 200) Or Abnormal liver function (cirrhosis, bilirubin > 2 times normal, ALT/AST/ALP > 3 times normal)	1 for any renal abnormalities 1 for any liver abnormalities
S	Stroke, history of	1
B	Bleeding, history of bleeding or tendency to bleed	1
L	Labile INRs (unstable/high INRs, time in therapeutic range < 60%)	1
E	Elderly (> 65 years)	1
D	Drugs Predisposing to Bleeding (Antiplatelet agents, NSAIDs) Or Alcohol Use (>8 drinks/week)	1 for drugs 1 for alcohol

- There are no formal rules on how we act on the HAS-BLED score although a **score of >= 3 indicates a 'high risk' of bleeding**, defined as intracranial haemorrhage, hospitalisation, haemoglobin decrease >2 g/L, and/or transfusion.

Atrial fibrillation: cardioversion

Atrial fibrillation - cardioversion:

- if no structural heart disease → flecainide
- With structural heart disease → amiodarone

offer rate or rhythm control if the onset of the arrhythmia is less than 48 hours, and start rate control if it is more than 48 hours or is uncertain

• Cardioversion indications

- ⇒ Haemodynamically unstable patient → electrical cardioversion (DC cardioversion 200J → 360J → 360J)
 - Adverse signs necessitating DC cardioversion are:
 - ❖ Blood pressure (BP) ≤90 mmHg

- ❖ Chest pain
- ❖ Heart failure
- ❖ Impaired consciousness, and
- ❖ Heart rate ≥ 200 bpm.
- ⇒ **Elective procedure** where a rhythm control strategy is preferred → electrical or pharmacological cardioversion
 - **Onset < 48 hours**
 - ❖ Anticoagulation
 - ⇒ patients should be heparinised.
 - ⇒ Patients who have risk factors for ischaemic stroke should be put on lifelong oral anticoagulation.
 - ❖ Cardioversion method:
 - ⇒ electrical - 'DC cardioversion'
 - ⇒ pharmacology:
 - ➡ amiodarone if structural heart disease,
 - ➡ flecainide or amiodarone in those without structural heart disease
 - ❖ Post-cardioversion:
 - ⇒ further anticoagulation is unnecessary
 - **Onset > 48 hours**
 - ❖ **prior to cardioversion:**
 - ⇒ anticoagulation
 - ➡ **for at least 3 weeks prior to cardioversion. OR**
 - ➡ exclude a left atrial appendage (LAA) thrombus by transoesophageal echo (TOE). If excluded patients may be heparinised and cardioverted immediately.
 - ⇒ If there is a high risk of cardioversion failure (e.g. Previous failure or AF recurrence) then it is recommended to have at least **4 weeks amiodarone or sotalol** prior to electrical cardioversion
 - ⇒ If the patient has a **slow ventricular response of AF** in the absence of anti-arrhythmic drugs, cardioversion should be performed after the **insertion of a temporary transvenous-pacing catheter**
 - ❖ Cardioversion method:
 - ⇒ NICE recommend electrical cardioversion, rather than pharmacological.
 - ⇒ The initial shock strength should be 100 J, followed by a second 200-J shock and a third 360-J shock
 - ⇒ If AF persists, a second 360-J shock with the paddles in the anteroposterior position can be attempted
 - ❖ Post-cardioversion:
 - ⇒ **Following electrical cardioversion patients should be anticoagulated for at least 4 weeks.** After this time decisions about anticoagulation should be taken on an individual basis depending on the risk of recurrence
 - **Catheter AF ablation**
 - ⇒ **Radiofrequency pulmonary vein isolation with ablation**
 - **the treatment of choice for patients who remain poorly controlled despite medical therapy,**
 - in selected patients as first-line therapy for symptomatic paroxysmal AF
 - Anticoagulation for stroke prevention should be continued indefinitely in patients at high risk of stroke, **even after apparently successful ablation of AF.**

- Surgical AF ablation**

- ⇒ Ablation can be performed in symptomatic patients during cardiac surgery for other reasons, or by stand-alone surgery either using open-chest techniques or by thoracoscopy.
- ⇒ Anticoagulation for stroke prevention should be continued indefinitely in patients at high risk of stroke, even after apparently successful ablation of AF.

The enlarged left atrial size suggests that a repeat DC cardioversion is unlikely to work for a sustained period.

H/O AF + enlarged left atrial size with previous DC cardioversions, the best long term treatment option → Refer for consideration of atrial fibrillation ablation → longer term good result.

AV node ablation:

- AV node ablation is reserved for those patients where pharmacological rate control is unsuccessful or not tolerated.
- The procedure is invasive and requires permanent pacemaker implantation.
- Patients who are candidates for this therapy include those with tachycardia induced cardiomyopathy despite pharmacologic efforts** at rate control and intolerable symptoms despite aggressive attempts at pharmacologic therapy (in some cases, much of the symptom burden is due to medications rather than AF itself).

Do not perform cardioversion or catheter ablation without anticoagulation, unless an atrial thrombus has been ruled out by transoesophageal echocardiogram.

Amiodarone or **vernakalant** have been efficient in converting post-operative AF to sinus rhythm.

Vernakalant

- A Novel agent for the Termination of Atrial Fibrillation
- blocks sodium channels
- more prominent in vernakalant's mechanism of action is its ability to block certain potassium channels.**
- Specifically, it blocks the atrial-selective potassium current, I_{Kur} , which is involved in atrial repolarization.**

Atrial fibrillation: pharmacological cardioversion

Atrial fibrillation - cardioversion: amiodarone + flecainide

- Agents with proven efficacy in the pharmacological cardioversion of atrial fibrillation**
 - ⇒ amiodarone
 - ⇒ **flecainide (if no structural heart disease)**
 - with large doses of **oral agents** or with intravenous agents.
 - Large single doses of flecainide (300 mg) or propafenone (450-600 mg) given orally have been shown to convert patients to sinus rhythm.
 - Flecainide and propafenone are not used in people with :
 - known or suspected ischaemic heart disease,
 - individuals who are already on antiarrhythmic therapy,

- ❖ those with a prolonged QT interval because these agents may have pro-arrhythmic effects (torsade de pointes).
- ⇒ others (less commonly used in UK): quinidine, dofetilide, ibutilide, propafenone
- **Less effective agents**
 - ⇒ beta-blockers (including sotalol)
 - ⇒ calcium channel blockers
 - ⇒ digoxin
 - ⇒ disopyramide
 - ⇒ procainamide

Atrial fibrillation: rate control and maintenance of sinus rhythm

Atrial fibrillation: rate control - beta blockers preferable to digoxin

The patient with very recent onset of atrial fibrillation is more likely to stay in sinus rhythm

- **Agents used to control rate** in patients with atrial fibrillation
 - ⇒ Beta-blockers
 - should be used **first line for rate control**.
 - cardioselective beta-blockers should be tried in patients with **left ventricular systolic dysfunction even if they have a diagnosis of:**
 - ❖ Chronic obstructive pulmonary disease (COPD)
 - ❖ Peripheral vascular disease
 - ❖ Diabetes
 - ❖ Erectile dysfunction, or
 - ❖ Interstitial pulmonary disease.
 - Beta-blockers should not be commenced in the setting of acute exacerbations of COPD or cardiac failure
 - If one drug does not control the rate adequately NICE recommend combination therapy with diltiazem or digoxin
 - ⇒ calcium channel blockers (diltiazem)
 - ⇒ digoxin:
 - not considered first-line anymore as they are less effective at controlling the heart rate during exercise.
 - they are the preferred choice if the patient has coexistent **heart failure**
 - **with borderline hypotension (eg: 95/70), COPD and AF, rate control without the possibility of worsening hypotension is the aim of intervention. Digoxin is therefore the optimal intervention.** It will both slow the ventricular rate and support the blood pressure.
 - ⇒ If the duration of AF is unknown caution should be used when considering the use of drugs which may cardiovert the patient - amiodarone and flecainide.
- **Agents used to maintain sinus rhythm** in patients with a history of atrial fibrillation
 - ⇒ sotalol
 - ⇒ amiodarone
 - ⇒ flecainide
 - ⇒ others (less commonly used in UK): disopyramide, dofetilide, procainamide, propafenone, quinidine
 - The table below indicates some of the factors which may be considered when considering either a rate control or rhythm control strategy

Factors favouring rate control	Factors favouring rhythm control
<ul style="list-style-type: none"> Older than 65 years History of ischaemic heart disease 	<ul style="list-style-type: none"> Younger than 65 years Symptomatic First presentation Lone AF or AF secondary to a corrected precipitant (e.g. Alcohol) Congestive heart failure

MRCPUK-part-2-march-2018: H/O borderline hypotension (BP: 95/70), COPD and AF. What is the most appropriate intervention?

⇒ **Digoxin 500 mg IV loading**

- B-blockers and verapamil are best avoided because of the potential for worsening hypotension here.

Atrial flutter

Tachycardia with a rate of 150/min ?atrial flutter

Overview

- Atrial flutter is a form of supraventricular tachycardia characterised by a succession of rapid atrial depolarisation waves.
- usually caused by a single macroreentrant rhythm within the atria.
- What is the differences between atrial flutter and focal atrial tachycardia?**
 - ⇒ Atrial flutter is caused mechanically by macro- reentry and has atrial rate (P wave/flutter morphology) usually >250 bpm.
 - ⇒ Focal atrial tachycardia is caused mechanically by micro-reentry or increased automaticity and has atrial rates of 100-250 bpm.

Epidemiology

- Sex: ♂ > ♀ (5:2)
- Peak incidence: risk of atrial flutter increases with age

Etiology:

- similar to atrial fibrillation

ECG findings

- Regular, narrow QRS complexes
- flutter waves, which are a **saw-tooth pattern** of atrial activation
 - ⇒ **most prominent in leads II, III, aVF, and V1.**
- as the underlying atrial rate is often around 300/min the ventricular or heart rate is dependent on the degree of AV block. For example if there is 2:1 block the ventricular rate will be 150/min
- flutter waves may be visible following carotid sinus massage or adenosine

Management

- is similar to that of atrial fibrillation although medication may be less effective
- atrial flutter is more sensitive to cardioversion however so lower energy levels may be used
- Anticoagulate patients with atrial flutter similar to AF.
- Catheter ablation** is the definitive treatment for atrial flutter.
 - ⇒ **radiofrequency ablation of the tricuspid valve isthmus** is curative for most patients

Multifocal atrial tachycardia (MAT)

Multifocal atrial tachycardia has ≥ 3 P-wave morphologies on ECG

Definition

- it is an **irregular** cardiac rhythm caused by at least three different sites in the atria, which may be demonstrated by morphologically distinctive **P waves**.
- It is more common in elderly patients with chronic lung disease, for example COPD

Management

- correction of hypoxia and electrolyte disturbances
- rate-limiting calcium channel blockers are often used first-line
- cardioversion and digoxin are not useful in the management of MAT

Atrial myxoma

Atrial myxoma - commonest site = left atrium

Overview

- Benign cardiac tumor
- the most common primary cardiac tumors in adults.
 - (**rhabdomyoma**) is the most common primary cardiac tumor in pediatric patients and strongly associated with tuberous sclerosis.
- 75% occur in **left atrium**, arising from a pedicle on the fossa ovalis.
- more common in females
 - Three-quarters of cases of atrial myxoma occur in females
- Although most cases of atrial myxoma are sporadic, an **autosomal dominant** variety may also exist within families.
- 10% are inherited

Features

- One third present with emboli
- One third with systemic inflammation (ESR $\uparrow\uparrow$ in 1/3)
- One third are **asymptomatic** when detected.
- There are 3 groups of manifestations:
 1. **Obstructive features:** like MS, signs vary with posture. Occasionally, there is a low-pitched sound called tumor plop.
 - Dyspnoea, (Exertional dyspnoea is present in three-quarters of patients).
 - Dizziness or syncope
 - results from the atrial myxoma obstructing the mitral valve.
 - ❖ **Mitral valve obstruction is the most likely complication**
 - ❖ Myxomas are more likely to have a stalk and be freely mobile.
 - atrial fibrillation
 - mid-**diastolic** murmur, 'tumour plop' (low-pitched sound)
 - ❖ murmur change with posture.
 - Elevated left atrial pressures cause dilatation.
 2. **Embolic features:** either systemic or pulmonary embolism.
 3. **Constitutional features:** such as fever, malaise, weakness, loss of weight, myalgia, arthralgia, clubbing, skin rash, Raynaud's phenomenon.

Investigations

- echo: pedunculated heterogeneous mass typically attached to the fossa ovalis region of the interatrial septum

- on histology
 - gelatinous appearance
 - abundant ground substance.

Treatment

- surgical removal by median sternotomy.

Prognosis

- sudden death may occur in 15% of patients.

Carney's complex is a familial multiple neoplasia and lentiginosis syndrome, associated with

1. Primary adrenal hypercortisolism
2. Lentigines and naevi of the skin
3. Various tumours including **myxoma**.

Heart block

Types of heart block

If the R is far from P,
then you have a **FIRST DEGREE**.



Longer, longer, longer, drop!
Then you have a **WENKEBACH**.



If some Ps don't get through,
then you have **MOBITZ II**.

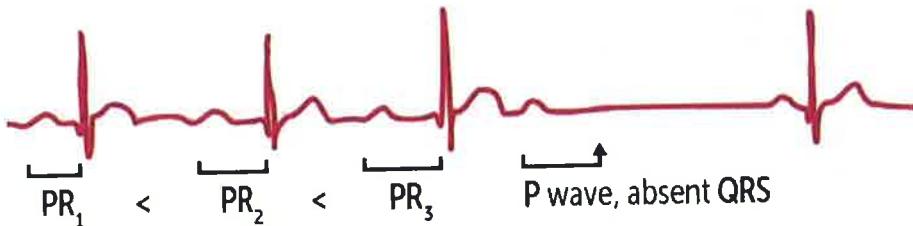


First-degree heart block

- PR interval > 0.2 seconds (> five small squares in the ECG.)
- Causes:
 - ⇒ Increased vagal tone (such as in trained athletes)
 - ⇒ Ischaemic heart disease
 - ⇒ Rheumatic fever
 - ⇒ Hyperkalaemia
 - ⇒ Hypokalaemia, and
 - ⇒ Drug therapy such as digoxin or beta-blockers.
- A long PR interval on the ECG may also be caused by structural abnormalities such as an atrial septal defect.
- No treatment is usually required.

Second-degree heart block

- Type 1 (Mobitz I, Wenckebach):
 - ⇒ progressive prolongation of the PR interval until a dropped beat occurs
 - ⇒ Mobitz Type I with symptoms is a **relative indication** for a permanent pacemaker
 - ⇒ **Asymptomatic → NO treatment → Discharge him from the clinic**
 - The risk of progression to complete heart block with Mobitz type I in an asymptomatic man is very low, unlike in Mobitz type II.



- Type 2 (Mobitz II):
 - ⇒ PR interval is constant, but the P wave is often not followed by a QRS complex
 - ⇒ **the most appropriate next management step → Transvenous cardiac pacing**
 - ⇒ Mobitz type II or complete heart block does not respond to atropine. Atropine may be useful for sinus or junctional bradycardia.
 - ⇒ In patients with Mobitz type II AV block, or complete heart block, a DDD or DDDR pacemaker is indicated.
 - DDD pacemaker will sense and pace both atria and ventricles.
 - DDD pacemakers ensure that the atrial and ventricles beat in synchrony thus preventing pacemaker syndrome.
- **Second-degree heart block with RBBB implies that this patient has a significantly increased risk of complete heart block.**
 - ⇒ prior to committing to pacemaker insertion, repeat tape is **the most likely next step**, with an **electronic patient diary** to see if the recorded arrhythmia corresponds to her symptoms.

Third degree (complete) heart block

Complete heart block causes a variable intensity of S1

Third degree (complete) heart block

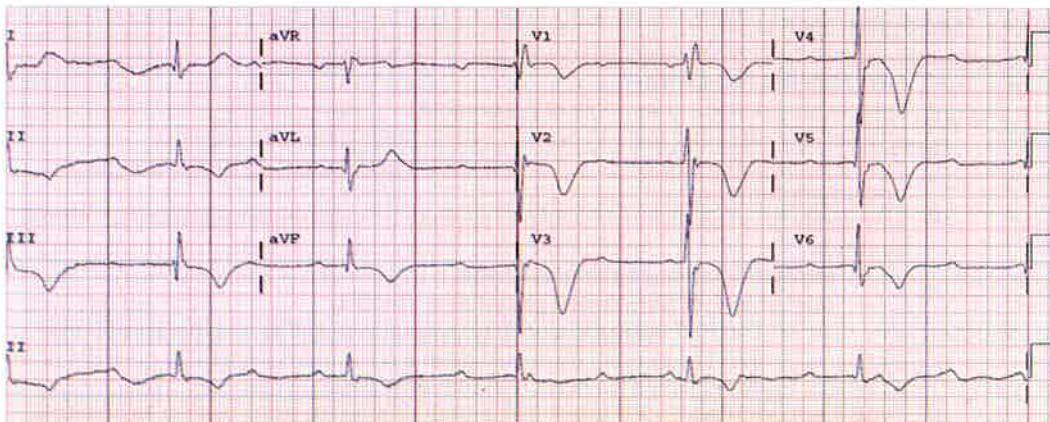
- there is no association between the P waves and QRS complexes
- **Complete heart block (whether symptomatic or not) is an absolute indication for a permanent pacemaker**

Causes

- myocardial ischemia
 - ⇒ The most common cause of third degree atrioventricular block is myocardial ischemia.
 - ⇒ Complete heart block is related most to **right coronary artery** occlusion because this commonly involves both the AV nodal artery and the right superior descending artery.
 - Prognosis is favourable, and revascularisation normally leads to restoration of sinus rhythm.
 - **As the AV nodal artery arises proximally from the right ventricular artery, distal right coronary artery occlusion is not commonly associated with complete heart block.**
 - **the artery most likely to be affected → Proximal right coronary**
 - ⇒ **Left coronary artery occlusion** leads to anterior myocardial infarction. As it is less commonly associated with complete heart block, when it does occur, the prognosis is very poor.
 - ⇒ Third degree atrioventricular block that is resulting from obstruction of the left anterior descending coronary artery is usually irreversible.
- Lyme disease
- Drugs eg: B. blockers
- Congenital third degree atrioventricular block might be due to maternal lupus.

Features

- Syncope
- heart failure
- regular bradycardia (30-50 bpm) that does not vary with exercise
- wide pulse pressure
- JVP: **irregular** cannon waves in neck
- variable intensity of S1
- compensatory increase in stroke volume with a large-volume pulse and **systolic flow**
- The escape rhythm of third-degree atrioventricular block resulting from obstruction of the right coronary artery is usually narrow-complex.
- the atrial rhythm is usually regular
- **The bizarre, wide, inverted T-waves can be seen in Stokes-Adams attacks** and do not necessarily imply new ischaemia.



ECG showing third degree (complete) heart block

Treatment

In patients with Mobitz type II AV block, or complete heart block, a DDD or DDDR pacemaker is indicated

- Asymptomatic or mild symptoms (stable)
 - ⇒ Condition-specific management
- Symptomatic (unstable): (syncope, ventricular rate is significantly low (<40 to 45 bpm) or low BP (mean arterial pressure <65 mmHg))
 - ⇒ Whilst arrangements are being made for temporary pacing, the options to be considered, prior to temporary transvenous pacing, in this context are:
 1. **Atropine** 0.5-1.0 mg intravenous bolus repeated as required up to 3mg .
 2. Isoprenaline, intravenous infusion at 2-10 microg/min.
 - ❖ it is a non-selective β agonist that is analog of epinephrine (adrenaline)
 3. External cardiac pacing.
 - ⇒ temporary (transcutaneous or transvenous) pacing
 - **Transvenous pacing is much more reliable than transcutaneous pacing**
- **Condition-specific management** includes:
 - ⇒ treating acute coronary syndrome (i.e., antiplatelet medications, urgent revascularisation)
 - ⇒ medication toxicity (e.g., **glucagon for beta-blocker toxicity**, calcium for calcium-channel toxicity, or digoxin antibody for digitalis toxicity).
- Intravenous aminophylline is useful in complete heart block, as the heart block is often mediated by adenosine which aminophylline inhibits
- Post- MI:
 - ⇒ Following anterior MI → pace-maker insertion
 - ⇒ **Following posterior MI and patient is haemodynamically stable → observation**
 - Often spontaneously resolved

Pacemakers

Definition

- A permanent pacemaker is an implanted device that provides electrical stimuli, thereby causing cardiac contraction when intrinsic myocardial electrical activity is inappropriately slow or absent.

Conditions definitely needs a permanent pacemaker

- Symptomatic bradycardia due to sinus node dysfunction (sick sinus syndrome)
- Third-degree heart block
- **second-degree (AV) block** associated with any of the following:
 - ⇒ symptomatic bradycardia
 - ⇒ documented periods of asystole of 3 s or more
 - ⇒ **any escape rate less than 40 bpm in awake, asymptomatic patients**
 - ⇒ **type II second-degree AV block and a ventricular rate of 45 bpm when awake and asymptomatic**
 - ⇒ asymptomatic sinus rhythm resulting in **periods of asystole longer than 3.0 seconds**
 - **asystolic pause causing syncope.**
 - **dual chamber permanent pacemaker (DDDR).**
 - ❖ The R in this code stands for responsive, and in an otherwise fit and well 76-year-old, he should have a responsive element to his PPM (that is, increases his heart rate with exercise).
 - ⇒ **Type II second-degree AV block has a high chance of progressing to asystole (35%) each year**
- Generally, permanent pacing can be justified for any degree of heart block associated with symptoms of bradycardia.

Indications for a temporary pacemaker

- symptomatic/haemodynamically unstable bradycardia, not responding to atropine
- post-ANTERIOR MI: type 2 or complete heart block
 - ⇒ post-INFERIOR MI complete heart block is common and can be managed conservatively if asymptomatic and haemodynamically stable
- trifascicular block prior to surgery
- Other indications for transvenous pacing in setting of acute MI are:
 - ⇒ asystole
 - ⇒ new bundle branch block (BBB) with first-degree heart block
 - ⇒ an old right BBB with first degree atrioventricular (AV) block and a new fascicular block

Notes

- All modern ICDs also function as pacemakers.
- Chest pain in **Ventricular pacing**
 - ⇒ Pacemaker rhythm may prevent interpretation of ST-segment changes and may require **urgent angiography** to confirm diagnosis.
 - ⇒ **Reprogramming the pacemaker**—allowing an evaluation of ECG changes during intrinsic heart rhythm—may be considered in patients who are not dependent on ventricular pacing, without delaying invasive investigation

Types of Pacemakers

- Pacemakers are classified by the nature of their pacing mode using a code of up to five letters.
- The NBG Pacemaker code was developed by the North American Society of Pacing and Electrophysiology (NASPE) and the British Pacing and Electrophysiology Group (BPEG):

I	II	III	IV	V
Chamber(s) Paced	Chamber(s) Sensed	Mode(s) of Response	Rate Modulation	Multisite Pacing
O = None	O = None	O = None	O = None	O = None
A = Atrium	A = Atrium	T = Triggered	R = Rate modulation	A = Atrium
V = Ventricle	V = Ventricle	I = Inhibited		V = Ventricle
D = Dual (A+V)	D = Dual (A+V)	D = Dual (T+I)		D = Dual (A+V)

Single-chamber pacemakers

- utilised **for patients in permanent atrial fibrillation**.
- VVI means there is **one lead in the ventricle** (pacing and sensing the ventricle, indicated by the 'VV').
- VVI pacemaker will pace and sense the right ventricle.
- VVI pacemaker** is useful when we are not too concerned about atrial activity (e.g. in patients with atrial fibrillation).
- In the presence or organised atrial activity, a VVI pacemaker may pace the ventricles out of sync with the atria resulting in pacemaker syndrome.
 - ⇒ Since organised atrial activity is present, a **DDI pacemaker** would be preferred, as this **senses and paces both atria and ventricle to preserve synchrony**.

Dual-chamber pacemakers

- Have pacing electrodes in both the right atrium and the right ventricle.
- They allow maintenance of the physiological relationship between atrial and ventricular contraction and also allow the paced heart to follow the increase in sinus rate that occurs during exercise.

Biventricular pacemakers

- Pacemaker leads are placed in the right atrium, right ventricle and left ventricle.
- Useful in the management of patients with heart failure who have evidence of abnormal intraventricular conduction (most often evident as left bundle branch block (LBBB) on ECG) which causes deranged ventricular contraction or dyssynchrony.
- **In a patient with severe ischaemic heart failure and is on optimal medical therapy.**

Despite this he is still symptomatic → ICD with biventricular pacing

- ⇒ very prolonged QRS duration is indicating left dyssynchrony which is an indication for biventricular pacing according to NICE guidance.
- ⇒ Documented VT in the context of ischaemic LV impairment necessitates the need for and a secondary prevention ICD.

Pacemaker complications

- Pacemaker complications are more common in the period following insertion.
- can be divided into early complications (<6 weeks) or late (>6 weeks).
- Most frequent complications are those related to implantation procedure, such as lead dislodgement and pneumothorax.
- **pneumothorax can occur up to forty-eight hours following pacemaker insertion.**
 - ⇒ It occurs in 1-2% of procedures and most patients will require chest drain insertion.
- The most common complication is **lead dislodgement** (higher rate atrial dislodgment than ventricular dislodgment).
- Lead dislodgement can occur following trauma or sporadically and can be either atrial or ventricular.
- Atrial dislodgment affects up to 3% of people whereas ventricular is less common affecting 1%.
- **If the ECG shows loss of sensing and capture around the QRS complex → ventricular lead displacement in a dual chamber pacemaker.**
 - What would be the likely ECG findings in ventricular lead displacement?
 - ⇒ **Loss of sensing and capture of the QRS complex**
- Atrial lead displacement would show an ECG with loss of atrial sensing and capture.
 - ⇒ **The ECG in atrial lead displacement would show an ECG with loss of atrial sensing and capture in a dual chamber or single chamber pacemaker.**
- On occasion lead displacement can be seen on chest X-Ray, however, **it may not be seen**, in this case → a lateral chest X-Ray may be of use in this scenario.
- **Pacemaker syndrome** would show AV dyssynchronisation.
- **Subclavian vein obstruction** is a fairly common complication over time but many patients may remain asymptomatic due to collateral vein formation. It can present with symptoms of superior vena cava (SVC) obstruction in severe cases.
- **Twiddler's syndrome** is when the patient intentionally or accidentally turns the pacemaker on its longitudinal axis which can cause lead dislodgement.
- **Reel's syndrome** is Twiddler's syndrome but on the horizontal axis.
- **Pacemaker lead fracture**
 - ⇒ occurs in 1-4% of pacemakers
 - ⇒ **usually following excessive exercise or direct trauma.**
 - ⇒ patient will require lead extraction and replacement.
- **myocardial rupture:**
 - ⇒ incidence is relatively small (<1%)

- ⇒ can be divided into early or late rupture with respect to the time it occurs following procedure.
- ⇒ Delayed perforations are less likely to cause such acute symptoms as well as a reduced incidence of tamponade and sudden cardiac death.
- ⇒ Risk factors for perforation include physician technique, patient independent factor (i.e obesity or difficult anatomy) and lead design.
- ⇒ presenting features :pericardial effusion, haemodynamically compromised following pacemaker insertion and is likely to develop cardiac tamponade and needs urgent intervention with pericardiocentesis.

Pacemaker syndrome

Pacemaker syndrome

- Loss of AV synchrony.
- Retrograde VA conduction.
- Absence of rate response to physiological need.

Pacemaker syndrome (breathlessness associated with ventricular pacing in the context of normal atrial activity).

VVI pacemaker will pace and sense the right ventricle. In the presence of organised atrial activity, a VVI pacemaker may pace the ventricles out of sync with the atria resulting in pacemaker syndrome.

Overview

- pacemaker syndrome is related to nonphysiologic timing of atrial and ventricular contractions, which may occur in a variety of pacing modes
- also named as "AV dyssynchrony syndrome,"
- **typically associated with a VVI pacemaker** that results in simultaneous atria and ventricle conduction.

Risk factors

- Sick sinus syndrome as have preserved AV conduction.
- Single-chamber ventricular pacing.

Features

- Hypotension, tachycardia, tachypnoea, dizziness, syncope
- Ventricular contraction against closed tricuspid and mitral valves can result in raised JVP (pulsation and fullness in the neck) cannon waves

Complications of AV dyssynchrony:

- Atrial fibrillation, thromboembolic events, and heart failure.

What are the characteristic ECG findings associated with this syndrome?

- ⇒ Small P waves with dissociation from QRS complex

Management

- In patients with other pacing modes, upgrading the pacemaker to a **dual-chamber pacing** or **reprogramming the pacemaker parameters** - eg, AV delay, post-ventricular atrial refractory period, sensing level, and pacing threshold voltage.

DC cardioversion in patients with pacemakers (eg : in AF)

- DC cardioversion is not contraindicated in patients with pacemakers
- **Pacemaker function should be checked after cardioversion and antiarrhythmic therapy added**

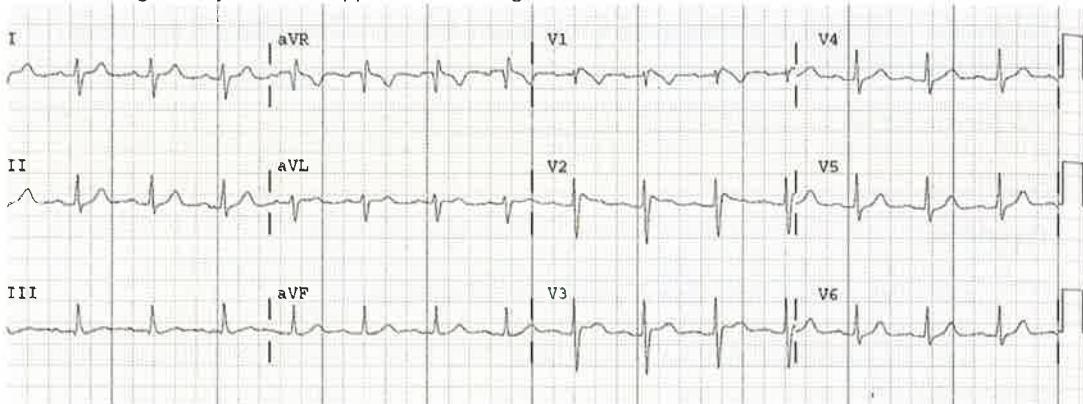
Brugada syndrome

Overview

- Inherited cardiovascular disease which may present with sudden cardiac death.
- Prevalence → 1:5,000-10,000.
- More common in Asians.
- Autosomal dominant
- A large number of variants exist
- Around 20-40% of cases are caused by a mutation in the SCN5A gene which encodes the myocardial sodium ion channel protein

ECG changes

- Convex ST segment elevation > 2mm in > 1 of V1-V3 followed by a negative T wave
- Partial right bundle branch block
- Changes may be more apparent following flecainide



ECG showing Brugada pattern, most marked in V1, which has an incomplete RBBB, a downslloping ST segment and an inverted T wave

Management

- implantable cardioverter-defibrillator

Catecholaminergic polymorphic ventricular tachycardia (CPVT)

Overview

- CPVT is a form of inherited cardiac disease associated with sudden cardiac death.
- Inherited in an autosomal dominant fashion
- Prevalence of around 1:10,000.

Pathophysiology

- the most common cause is a defect in the ryanodine receptor (RYR2) which is found in the myocardial sarcoplasmic reticulum
- uncontrolled calcium release from the sarcoplasmic reticulum
- induced by adrenergic stress.

Features

- exercise or emotion induced polymorphic ventricular tachycardia resulting in syncope
- sudden cardiac death
- symptoms generally develop before the age of 20 years

Management

- beta-blockers
- There is strong evidence that flecainide is effective when prescribed in addition to beta blockers
- implantable cardioverter-defibrillator
- Left cervical sympathetic denervation
- All first-degree relatives should be evaluated with ECG, Holter monitoring and exercise stress testing.

Ventricular tachycardia

Definition

- wide QRS complex (duration >120 milliseconds) at a rate greater than 100 bpm, originating from a ventricular ectopic focus.
 - ⇒ Whilst a broad complex tachycardia may result from a supraventricular rhythm with aberrant conduction, the European Resuscitation Council advise that in a peri-arrest situation it is assumed to be ventricular in origin.
- It has the potential to precipitate ventricular fibrillation and hence requires urgent treatment.

Pathophysiology

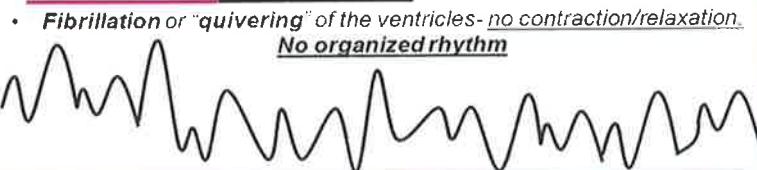
- Among patients with **prior MI** or non-ischaemic cardiomyopathy, VT is usually due to **re-entry involving regions of slowed conduction adjacent to scar**.
 - ⇒ **Post MI ventricular tachycardia (VT) is most commonly due to scar tissue.**
 - **The definitive investigation would be → Electrophysiological study (EPS)**
 - ❖ due to the fact that if this were scar related VT, the site could be localised and even possibly ablated.
 - ❖ If not, then an implantable cardiac defibrillator (ICD) implantation may be warranted if left ventricular (LV) dysfunction exists.
 - ✓ MADIT-2 trial showed a 5.6% 20-month absolute survival benefit in patients with LV dysfunction ($EF < 30\%$), post MI, treated prophylactically with an ICD.
 - (VT) may also arise from triggered activity due to **early after-depolarisations (EADs)** leading to torsades de pointes, a polymorphic ventricular tachycardia seen in the setting of a prolonged QT interval,
 - **delayed after-depolarisations (DADs)**, which are seen in:
 - ⇒ idiopathic right ventricular outflow tract VT or
 - ⇒ catecholaminergic polymorphic VT
 - cellular abnormalities of calcium handling → Increased intracellular calcium → predispose to VT. especially during periods of sympathetic stimulation.
 - EADs occur during phase 2 or 3 of the action potential, whereas DADs occur during phase 4.
 - When an EAD or DAD reaches a 'threshold' potential, it can result in triggering of another action potential.
 - Ventricular tachycardia originates below the bundle of His.

- **Ventricular Tachycardia**

- *Tachycardia- firing from the ventricle*
- *WIDE "QRS", no "P"*



- **Ventricular Fibrillation**



Types: There are two main types of VT:

- **Monomorphic VT**
 - ⇒ organised, single-morphology QRS arising from one of the ventricles.
 - ⇒ most commonly caused by myocardial infarction
- **Polymorphic VT**
 - ⇒ multiple different wide QRS morphologies arising from one of the ventricles.
 - ⇒ results from abnormal myocardial repolarization.
 - ⇒ A subtype of polymorphic VT is **torsades de pointes** which is precipitated by prolongation of the QT interval. The causes of a long QT interval are listed below.

Other classifications of VT

- **Sustained VT**
 - ⇒ A ventricular rhythm faster than 100 bpm **lasting at least 30 seconds or requiring termination due to haemodynamic instability.**
 - ⇒ almost always symptomatic.
- **Non-sustained VT**
 - ⇒ A ventricular rhythm faster than 100 bpm lasting for at least 3 consecutive beats but **terminating spontaneously in less than 30 seconds**, and not resulting in significant haemodynamic instability.
 - ⇒ If these do not cause any haemodynamic compromise, **treatment is not needed.**
 - ⇒ **The most appropriate next step → Check potassium and magnesium levels**
 - During the GISSI-2 trial it was observed that a serum K⁺ level of <3.6 mmol/l was associated with a twofold increased risk of VF. Therefore serum K⁺ should be maintained >4 mmol/l by oral or intravenous (IV) supplementation in patients with acute MI.
 - Concomitant magnesium (Mg²⁺) deficiency is present in many patients with hypokalaemia and also makes correction of hypokalaemia difficult. Hence serum Mg²⁺ levels should also be checked and maintained >1 mmol/l.

Feature

- Patients may have a normal cardiac output or may be haemodynamically compromised
- Sustained VT is usually observed in ischaemic cardiomyopathy, but idiopathic VT may also be observed in patients without structural heart disease.
- jugular veins may show **cannon A waves** due to atrioventricular dissociation.

Differential diagnosis

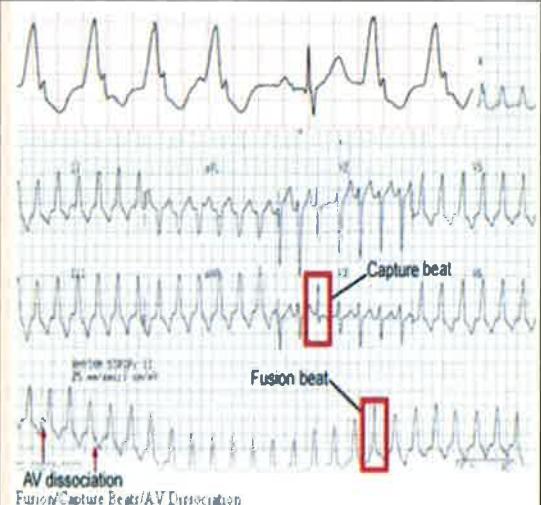
- Supraventricular tachycardia with bundle-branch block may resemble ventricular tachycardia on the ECG
 - ⇒ 80% of all broad complex tachycardias are secondary to VT and the proportion is even higher in patients with structural heart disease.
 - ⇒ In all cases of doubt, the rhythm should be treated as a VT.
 - ⇒ **the safest course of action is to consider a drug like adenosine**, which will cause short-lived AV block in SVT but not in VT. It is the presence of aberrant conduction which can lead to diagnostic confusion.
 - Amiodarone may be an appropriate next step for cardioversion, once the underlying rhythm has been elucidated.
- **Features suggesting VT rather than SVT with aberrant conduction**
 - ⇒ AV dissociation
 - ⇒ fusion or capture beats
 - ⇒ positive QRS concordance in chest leads ((same polarity QRS direction in all chest leads V1 -V6) (**Absence of an RS complex in all pre-cordial leads**, i.e., all the leads are concordant)
 - ⇒ marked left axis deviation
 - ⇒ history of IHD
 - ⇒ lack of response to adenosine or carotid sinus massage
 - ⇒ very broad QRS > 160 ms
 - ⇒ bifid upright QRS with a taller first peak in V1
 - ⇒ deep S wave in V6

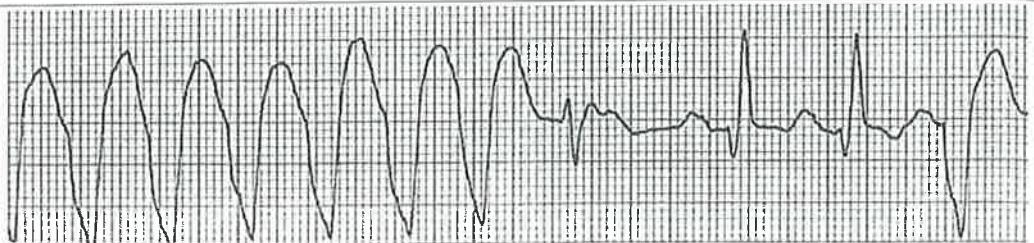
Capture beats

- intermittent narrow QRS complex owing to normal ventricular activation via the AV node
- occurs when a supraventricular and a ventricular impulse coincide to produce a hybrid complex.
- It indicates that there are two foci of pacemaker cells firing simultaneously: a supraventricular pacemaker (e.g. the sinus node) and a competing ventricular pacemaker (source of ventricular ectopics).
- Causes:
 - ⇒ Ventricular tachycardia
 - ⇒ Accelerated idioventricular rhythm (AIVR)

Fusion beats

(intermediate between ventricular tachycardia beat and capture beat) are seen





Fusion beats due to VT - the first of the narrower complexes is a fusion beat (the next two are capture beats)

Management

VT: cardioversion treatment

- ⌚ VT with pulse (not respond to medical treatment) → **LOW ENERGY synchronized cardioversion**
 - ☞ Synchronization avoids the delivery of a **LOW ENERGY** shock during cardiac repolarization (t-wave). If the shock occurs on the t-wave (during repolarization), there is a high likelihood that the shock can precipitate Ventricular Fibrillation (VF).
- ⌚ Pulseless VT or VF → **HIGH ENERGY asynchronized cardioversion**

- If the patient has adverse signs (systolic BP < 90 mmHg, chest pain, heart failure or **rate > 150 beats/min**) then **immediate cardioversion** is indicated.
 - ⇒ anaesthetist needs to be called to assist with direct current cardioversion (DCCV) which **should be 'synchronised' to limit the risk of conversion to VF.**
 - usually at a starting energy dose of 100 J (monophasic); comparable biphasic recommendations are not currently available).
 - ⇒ If deteriorate in the meantime and become pulseless, then a precordial thump should be given, followed immediately by DCCV if not successful.
 - ⇒ In cases of **pulseless VT**, the electrical cardioversion should be **unsynchronized**.
 - ⇒ **Amiodarone is the drug of choice for acute VT refractory to cardioversion shock.**
 - ⇒ Unstable polymorphic VT is treated with immediate defibrillation. The defibrillator may have difficulty recognizing the varying QRS complexes; therefore, synchronization of shocks may not occur.
- In stable patients (absence of adverse signs):
 - ⇒ **stable patients with monomorphic VT and normal LV function,**
 - If LV function is impaired, amiodarone (or lidocaine) is preferred to procainamide for pharmacologic conversion because of the latter drug's potential for exacerbating heart failure.
 - restoration of sinus rhythm is typically achieved with **IV procainamide, amiodarone, or sotalol.**
 - ⇒ **If LV function is impaired**, amiodarone (or lidocaine) is preferred to procainamide for pharmacologic conversion because of the latter drug's potential for exacerbating heart failure.
 - ⇒ In the absence of such signs antiarrhythmics may be used. If these fail, then electrical cardioversion may be needed with **synchronised DC shocks**
 - ⇒ If medical therapy is unsuccessful, synchronized cardioversion (50-200 J monophasic) following sedation is appropriate.
 - ⇒ prophylactic implantable cardioverter defibrillator implantation is recommended in high-risk patients.

- Polymorphic VT in stable patients
 - ⇒ typically terminates on its own.

	Unsynchronized	Synchronized
When to deliver electricity	At any point in cycle	Not during the T-wave
Indications	V-fib, pulseless VT	Everything except V-fib and pulseless VT

Drug therapy

Verapamil is contra-indicated in VT because it can cause a catastrophic fall in blood pressure.

- Amiodarone: ideally administered through a central line
☞ (i.e. given after the third shock). If amiodarone is not available lidocaine is a suitable alternative.
- Lidocaine: use with caution in severe left ventricular impairment
- Procainamide
- Adenosine is useful diagnostically when the diagnosis of regular wide complex tachycardia is in doubt.
- **Verapamil should NOT be used in VT**

Sotalol is recommended as the first-choice drug to prevent a recurrence of ventricular tachycardia (VT)

If drug therapy fails

- **electrophysiological study (EPS)**
- implantable cardioverter-defibrillator (ICD) - this is particularly indicated in patients with significantly impaired LV function

C.V Resuscitation:

- Guidelines from the Resuscitation Council (UK) state that if a patient has a monitored and witnessed VF/VT arrest in hospital, three quick successive (stacked) shocks should be given. Chest compressions should be started immediately after the third, with a compression to ventilation ratio of 30:2 for 2 minutes.
- **A precordial thump can be successful if given within seconds of the onset of a shockable rhythm.** Delivery should not delay calling for help, or accessing a defibrillator, but would be indicated here whilst awaiting the defibrillator. Chest compressions should start immediately if it is unsuccessful.
- Intravenous adrenaline would be given every 3-5 minutes once chest compressions had started.

QT interval

QT interval: Time between the start of the Q wave and the end of the T wave

- **Definition**

☞ **QT measured from the start of the QRS complex to the end of the T wave**

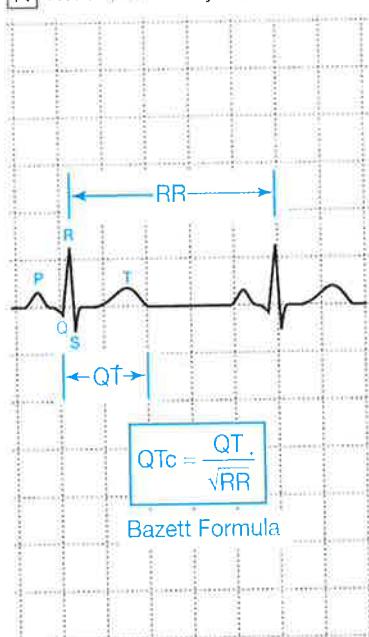
☞ represents the duration of activation and recovery of the ventricular myocardium

- **Normal duration** → should be between 0.33 and 0.44 seconds

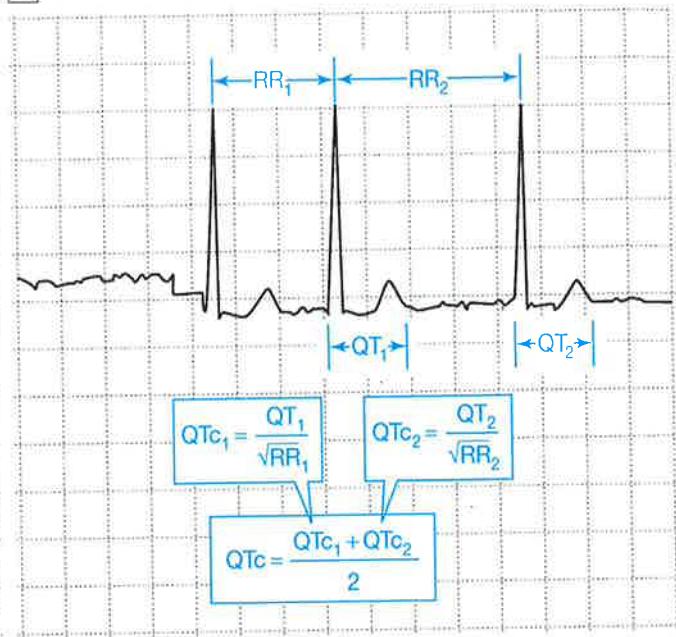
- **Corrected QT interval (QTc)** is calculated by dividing the QT interval by the square root of the preceding R - R interval. Normal = 0.42 s.

$$QTc_B = \frac{QT}{\sqrt{RR}}$$

A Normal Sinus Rhythm



B Atrial Fibrillation



Long QT syndrome

Definition

- Long QT syndrome (LQTS) is an inherited condition associated with delayed repolarization of the ventricles.
- A normal corrected QT interval is less than 430 ms in males and 450 ms (0.45 s) in females.
 - ☞ One large box represents 200 ms, one small box represents 40 ms

Mechanism

Long QT syndrome - usually due to loss-of-function/blockage of K⁺ channels

- the usual mechanism by which drugs prolong the QT interval is blockage of potassium channels → **delayed repolarization** of the ventricles.
- **Most drugs that prolong the QT_c interval act by blocking hERG-encoded potassium channels, although some drugs modify sodium channels.**
- The most common variants of LQTS (LQT1 & LQT2) are caused by defects in the alpha subunit of the slow delayed rectifier potassium channel.

Epidemiology

- more common in females.

Classification

	LQT1	LQT2	LQT3
Gene	KCNQ1	KCNQH2/ hERG	SCN5A
Irron	K _s (redifier potassium current, slow component)	K _r (redifier potassium current, rapid component)	Na
Pathophysiology	Decreased potassium outward current	Decreased potassium outward current	excessive sodium inward current
Triger of arrhythmia	Exercise stress	Emotional stress	Rest
Occurrence	> 50%	34 – 40%	10 – 15%

Causes of a prolonged QT interval

Methadone is a common cause of QT prolongation

Anti-arrhythmics	Antihistamines	Anti-infectives	Antimalarials
Amiodarone Disopyramide Dofetilide Ibutilide Procainamide Quinidine Sotalol	Astemizole Terfenadine	Clarithromycin Erythromycin Pentamidine Sparfloxacin	Chloroquine Halofantrine
Antipsychotics	Gastro-intestinal drugs	Opiate agonists	Other drugs
Chlorpromazine Haloperidol Mesoridazine Pimozide Thioridazone	Cisapride* Domperidone	Levomethadyl Methadone	tricyclic antidepressants, fluoxetine Arsenic trioxide Bepridil Droperidol Probuco
Congenital	Other conditions		
Jervell-Lange-Nielsen syndrome (includes deafness and is due to an abnormal potassium channel) Romano-Ward syndrome (no deafness)	<input type="checkbox"/> Electrolytes: <ul style="list-style-type: none"> ☞ hypocalcaemia ☞ hypokalaemia ☞ hypomagnesaemia <input type="checkbox"/> acute myocardial infarction <input type="checkbox"/> myocarditis <input type="checkbox"/> hypothermia <input type="checkbox"/> subarachnoid haemorrhage		

- *Cisapride have been withdrawn worldwide due to risk of QT prolongation
- **Jervell-Lange-Nielsen syndrome:**
 - ☞ **includes deafness** and is due to an abnormal potassium channel
 - ☞ **autosomal recessive**
 - ☞ **caused by Mutations in the KCNE1 and KCNQ1 genes**
 - Mutations in the KCNE1 and KCNQ1 genes → abnormal potassium channel → abnormal functions of **inner ear structures and cardiac muscle**.
- **Romano-Ward syndrome:**
 - ☞ congenital long QT syndrome
 - ☞ autosomal dominant
 - ☞ involves only cardiac (**no** deafness)
- The **human ether-à-go-go related gene (hERG)** is the gene affected by drugs that lengthen QT interval inadvertently; erythromycin, terfenadine, and ketoconazole.

- a non-sedating antihistamine are classic cause of prolonged QT in a patient, especially if also taking P450 enzyme inhibitor, e.g. Patient with a cold takes terfenadine and erythromycin at the same time

Features

A QT interval of greater than 0.44 seconds is associated with the development of ventricular arrhythmia, syncope and sudden cardiac death.

- asymptomatic
- may be picked up on routine ECG or following family screening
- Long QT1 - usually associated with exertional **syncope**, often swimming
- Long QT2 - often associated with syncope occurring following emotional stress, exercise or auditory stimuli
- Long QT3 - events often occur at night or at rest
- sudden cardiac death

Diagnosis

- corrected QT interval
 - ☞ Diagnosis is based upon the QTc (corrected QT interval),
 - ☞ QTc may be within the normal range at rest; hence **Holter ECG monitoring** is recommended.
- genetic testing of LQTS
 - ☞ Identification of an LQTS genetic mutation confirms the diagnosis.
 - ☞ However, a negative result on genetic testing is of limited diagnostic value because only approximately 50% of patients with LQTS have known mutations. The remaining half of patients with LQTS may have mutations of yet unknown gene. Therefore genetic testing of LQTS has high specificity but a low sensitivity.

Complications

- may lead to ventricular tachycardia → collapse/sudden death.

Management

Congenital long QT syndrome:

- Beta-blockers
 - ☞ **Beta-blockers are first-line initial treatment**
 - ☞ Beta blockers alone are enough to abate collapses in up to 70% of patients.
 - ☞ Beta blockers act by:
 1. decrease sympathetic activation from the left stellate ganglion,
 2. also decrease the maximal heart rate achieved during exertion and thereby prevent exercise-related arrhythmic events that occur in LQTS.
 - ☞ should be avoided in those congenital cases in which bradycardia is a prominent feature.
 - ☞ note sotalol may exacerbate long QT syndrome (due to blockage of K channel). This can be a particular risk in individuals with hypokalaemia. Therefore **Sotalol is better to be avoided in patients with thiazide diuretics.**
- patients who remain symptomatic despite receiving the maximally tolerated dose of beta-blockers → **Permanent pacing** and can be used in addition to beta-blockers.
- patients who remain refractory to beta-blockade and pacing → **High left thoracic sympathectomy**

- **Implantable cardioverter-defibrillators (ICDs)** are useful in rare instances when torsades still continues despite all of these treatments.
- Beta-blockers should be used along with ICDs because shock can further precipitate torsades by adrenergic stimulation.
- **Left stellate cardiac ganglionectomy** is an invasive procedure and results in Horner's syndrome. It is performed in patients who have symptoms despite βB and have frequent shocks with ICD.

Acquired long QT syndrome

- **avoid drugs which prolong the QT interval** and other precipitants if appropriate (e.g. Strenuous exercise)
- Long-term treatment in acquired cases is usually not required because the QT interval returns to normal once the predisposing factor has been corrected.
- Correction of any electrolyte disturbance
 - ☞ Due to the **pseudo-obstruction** it is very likely that the patient is hypokalaemic and as such this is the **first reversible aetiology for the non-sustained VT** that needs to be investigated
 - ❖ → **Check electrolytes**
 - ❖ Checking Magnesium would also be an appropriate step.
- **Beta-blockers are contra-indicated in acquired cases** because bradycardia produced by these agents can precipitate torsades. They should also be avoided in those congenital cases in which bradycardia is a prominent feature.
- Pacemaker implantation is effective in cases that are associated with heart block or bradycardia.
- ICDs are indicated in cases that cannot be managed by avoidance of any specific precipitating factor.

QT shortening: caused by:

- Hypercalcaemia
- Hypermagnesaemia
- Digoxin
- Thyrotoxicosis.

January 2019 exam: A patient develops torsades de pointes shortly after being started on sotalol. What effect does sotalol have on the cardiac cell membrane to make this more likely? **Blockage of potassium channels** → prolonged QT interval.

Torsades de pointes (TdP)

Overview

- Torsades de pointes ('twisting of the points') is a rare arrhythmia associated with a long QT interval.
- It may deteriorate into ventricular fibrillation and hence lead to sudden death
- In its most typical form, sudden slowing of heart rate (i.e., pauses) invariably precede each burst of TdP, and the recurrent arrhythmia is referred to as "pause-dependent TdP"

Risk factors

- **Female sex**
- causes of QT prolongation,
- **R-on-T phenomenon**
 - ☞ the R-wave, representing ventricular depolarization, occurs during the relative refractory period at the end of repolarization (represented by the latter half of the T-wave).

- Long QT intervals predispose the patient to an R-on-T phenomenon,
 - R-on-T can initiate torsades.
- bradycardia,
- congestive heart failure,
- digitalis therapy,
- severe alkalosis
- recent conversion from atrial fibrillation.

Management

- Stop all drugs which prolong QT
- Correct any electrolyte abnormalities
- IV magnesium sulphate (MgSO₄)
 - **the best initial drug**
 - **Mode of action: MgSO₄ → ↓ Ca influx → ↓ amplitude of the VT and helping terminate runs of torsade's.**
 - Dose : 2 gm as bolus over 10 minutes, followed by another bolus in 15 minutes if required, or continuous infusion at a rate of 5-20 mg/min.
 - It is effective even when serum magnesium level is normal.
- Temporary pacemaker/transvenous overdrive pacing (atrial or ventricular)
 - reserved for patients with long QT-related TdP who do not respond to intravenous magnesium.
- Isoproterenol
 - usually used as a temporizing measure prior to pacing in patients **who have failed to respond to magnesium and are awaiting placement of a temporary pacemaker.**
 - Action → Strong beta-1 & beta-2 stimulation

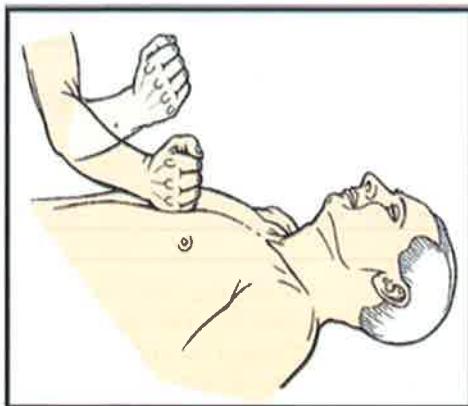
Adult advanced life support

Resuscitation Council (UK) 2021 guidelines

Major points include:

- **Point-of-care ultrasound (POCUS)**
 - ⇒ The guidelines recognise the increasing role of point-of-care ultrasound (POCUS) in peri-arrest care for diagnosis, but emphasises that it requires a skilled operator, and the need to minimise interruptions during chest compression.
 - ⇒ POCUS may be useful to diagnose treatable causes of cardiac arrest such as cardiac tamponade and pneumothorax.
 - ⇒ Right ventricular dilation in isolation during cardiac arrest should not be used to diagnose massive pulmonary embolism.
- **Immediately after the first shock (and each subsequent shock) chest compressions should be restarted immediately and pulse and rhythm reassessed after two minutes.**
- **Chest compression**
 - ⇒ Ratio of chest compressions to ventilation is 30:2
 - ⇒ Chest compressions are now continued while a defibrillator is charged
 - ⇒ After each shock chest compressions should be restarted immediately before anything else is done.
- **Adrenaline**
 - ⇒ should be used as soon as possible when the cardiac arrest rhythm is non-shockable
 - ⇒ after 3 defibrillation attempts for a shockable cardiac arrest rhythm.
 - ⇒ **during a VF/VT cardiac arrest, adrenaline 1 mg is given once chest compressions have restarted after the third shock** and then every 3-5 minutes (during alternate cycles of CPR).
 - ⇒ A 1 mg dose of adrenaline (epinephrine) would be administered with:
 - 0.1 ml of 1 in 100,
 - 1 ml of 1 in 1000 and
 - 10 ml of 1 in 10,000.
 - ⇒ **10 ml of 1 in 10,000 is the recommended dose and concentration** by the UK Resuscitation Council.
 - ⇒ **If not able to gain any venous access** within two minutes → **Obtain intraosseous access** (it provides adequate plasma levels of drugs and allows equivalent flow rates to IV access).
 - ⇒ Delivery of drugs via a tracheal tube is no longer recommended
- **Antiarrhythmic drugs (in VF/ pulseless VT)**
 - ⇒ Give Amiodarone 300 mg **after the third shock and 150 mg after the fifth shock.**
 - ⇒ If amiodarone is not available → use Lidocaine 100 mg after the third shock and 150 mg after the fifth shock.
- Atropine is no longer recommended for routine use in asystole or pulseless
- **Thrombolytic drugs**
 - ⇒ Consider thrombolytic drug therapy when pulmonary embolus is the suspected or confirmed as the cause of cardiac arrest.
 - ⇒ Consider CPR for 60-90 minutes after administration of thrombolytic drugs.
- **Waveform capnography during advanced life support**
 - ⇒ Use waveform capnography to confirm correct tracheal tube placement during CPR.
 - ⇒ Use waveform capnography to monitor the quality of CPR.
 - ⇒ An increase in ETCO₂ during CPR may indicate that ROSC has occurred. However, chest compression should not be interrupted based on this sign alone.

- **Recurrent or refractory VF**
 - ⇒ Consider escalating the shock energy, after a failed shock and for patients where fibrillation occurs.
 - ⇒ For refractory VF, consider using an alternative defibrillation pad position (e.g. anterior-posterior).
 - ⇒ **Magnesium sulphate IV is recommended for the treatment of refractory VF, if there is anything to suggest the patient may be hypomagnesaemic (such as on medications which might cause this, that is, thiazides).**
- **Precordial thump**
 - ⇒ Indicated only in witnessed or monitored cardiac arrest whilst awaiting the defibrillator within seconds of the onset of a shockable rhythm.
 - ⇒ It has a very low success rate for cardioversion. **There is more success with pulseless VT than with VF.**
 - ⇒ Chest compressions should start immediately if it is unsuccessful.
 - ⇒ The ulnar edge of a tightly clenched fist is used to deliver a sharp impact from a height of about 20 cm, then retract immediately (thereby creating an impulse-like stimulus). It delivers approximately 7-10 joules of energy.
 - ⇒ Only one thump should be delivered over the lower third of the sternum. Repeating a precordial thump is not recommended.



- **Electrical activity (PEA)**
 - ⇒ pulseless with no respiratory effort , ECG reveals small complexes with a normal morphology → CPR + Adrenalin 1mg repeated every 3-5 minutes
- **Defibrillation**
 - ⇒ Defibrillation is used to convert ventricular fibrillation to sinus rhythm
 - ⇒ Use single shocks where indicated, followed by a 2 minute cycle of chest compressions.
 - ⇒ The use of up to three-stacked shocks may be considered only if initial ventricular fibrillation/pulseless ventricular tachycardia (VF/pVT) occurs during a witnessed, monitored cardiac arrest with a defibrillator immediately available e.g. during cardiac catheterisation or in a high-dependency area.
 - ⇒ Antero-lateral pad position is the position of choice for initial pad placement. Ensure that the apical (lateral) pad is positioned correctly (mid-axillary line, level with the V6 ECG electrode position) i.e. below the armpit.
 - ⇒ In patients with an implantable device, place the pad > 8 cm away from the device
 - ⇒ **Range of the initial defibrillation energy**
 - **No clear evidence so , any level from 120-360 J is acceptable** followed by a fixed or escalating strategy up to maximum output of the defibrillator.

- **Cardiac arrest in profound hypothermia**
 - ⇒ **Prolonged cardiopulmonary resuscitation with re-warming is the management of choice.**
 - ⇒ Recovery with intact neurology has been reported even after very prolonged arrests, therefore resuscitation should be continued for far longer than would normally be considered.
 - ⇒ Hypothermic patients do not respond well to shocks or drugs and if there is no response to the first three shocks the patient should be rewarmed to at least 32°C before any drugs or shocks are administered.
- **Management of cold water drowning**
 - ⇒ **patients should be lifted out of the water in the prone position**
 - ⇒ **Re-warming** such patients should be undertaken in a hospital that has extracorporeal re-warming facilities
 - ⇒ **Defibrillation is ineffective if the myocardium is cold**
 - ⇒ **Hypothermia may render the carotid pulse impalpable** so it is important to commence chest compression with firm evidence of cardiac arrest.
 - ⇒ Continuous chest compression should be applied throughout transportation, which is as effective as chest compression with expired air resuscitation

Lance-Adams syndrome (Post-hypoxic myoclonus)

- Definition: a rare condition that can occur following a period of cerebral hypoxia (e.g. post cardiac arrest)
- Onset: **occurs within days to weeks of cardiac arrest**.
- Characterised by **intention myoclonus**.
- Treatment : antiepileptics (e.g. levetiracetam, valproate)

Peri-arrest arrhythmias

Resuscitation Council (UK) 2021 guidelines

Tachycardia

- To convert atrial or ventricular tachyarrhythmias, the shock must be **synchronised to occur with the R wave** of the ECG.
- **For atrial fibrillation:**
 - ⇒ An initial synchronised shock **at maximum defibrillator output** rather than an escalating approach is a reasonable strategy
- **For atrial flutter and paroxysmal supraventricular tachycardia:**
 - ⇒ Give an initial shock of 70 - 120 J **and stepwise increase energy for subsequent shocks**
- **For ventricular tachycardia with a pulse:**
 - ⇒ Give an initial shock of **120-150 J and stepwise increase energy for subsequent shocks**
- **If cardioversion fails to restore sinus rhythm and the patient remains unstable:**
 - ⇒ give IV amiodarone 300 mg over 10–20 minutes (or procainamide 10–15 mg kg⁻¹ and re-attempt electrical cardioversion.
 - ⇒ The loading dose of amiodarone can be followed by an infusion of 900 mg over 24 hours.

Bradycardia

- 1st line : give atropine 500 mcg IV (IO) and, if necessary, repeat every 3–5 minutes to a total of 3 mg.
- 2nd line (If atropine is ineffective): isoprenaline (5 mcg min^{-1} starting dose), and adrenaline ($2\text{--}10 \text{ mcg min}^{-1}$).
- For bradycardia caused by inferior myocardial infarction, cardiac transplant or spinal cord injury, consider giving **aminophylline** (100–200 mg slow intravenous injection).
- If bradycardia caused by beta-blockers or calcium channel blockers → give glucagon
- For bradycardia in patients with cardiac transplants
 - ⇒ Give aminophylline
 - ⇒ Do not give atropine, it can cause a high-degree AV block or even sinus arrest.
- For bradycardia refractory to drug therapies in patients who are unstable:
 - ⇒ transcutaneous pacing
 - ⇒ If transcutaneous pacing is ineffective, consider transvenous pacing.
- If atropine is ineffective and transcutaneous pacing is not immediately available, **fist pacing can be attempted while waiting for pacing equipment.**

Wolff-Parkinson White (WPW)

Pathophysiology

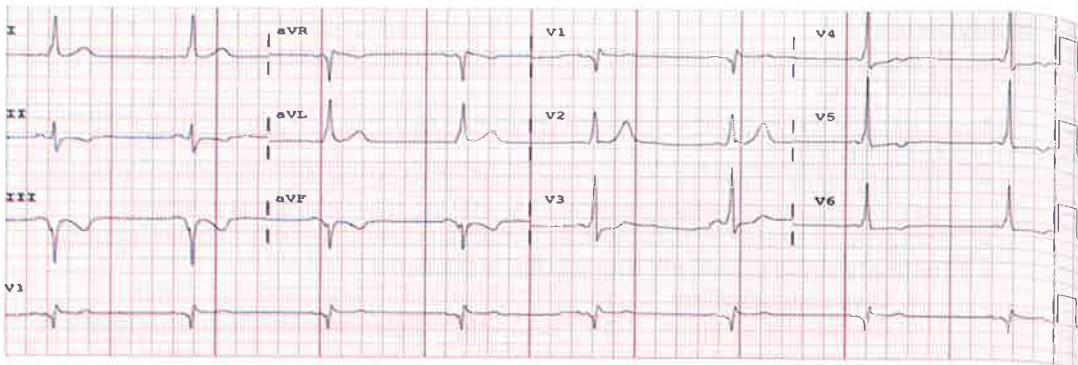
- Due to a congenital accessory conduction pathway, called the **bundle of Kent**, that connects the atria to the ventricles, bypassing the AV node and leading to **ventricular preexcitation**. As the accessory pathway does not slow conduction, AF can degenerate rapidly to VF

Presentation

- Most patients are asymptomatic.
- **WPW presents as SVT that can alternate with ventricular tachycardia (VT).**
- SVT is the most common type of tachycardia seen in a patient with WPW.
 - ⇒ often present with AV re-entrant tachycardia
- The other main clue to the diagnosis is **worsening of SVT after the use of calcium blockers or digoxin**

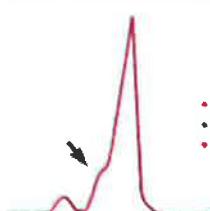
Possible ECG features

- short PR interval
- wide QRS complexes with a slurred upstroke - 'delta wave' (can be associated with negative delta waves in II, III and aVF)
- **ECG in sinus rhythm reveals right bundle-branch block**
- left axis deviation if right-sided accessory pathway*
 - ⇒ *in the **majority** of cases or in a question without qualification, Wolff-Parkinson-White syndrome is associated with **left axis deviation**
- right axis deviation if left-sided accessory pathway
- non-specific ST-T changes which are common in WPW and may be mistaken for ischaemia.

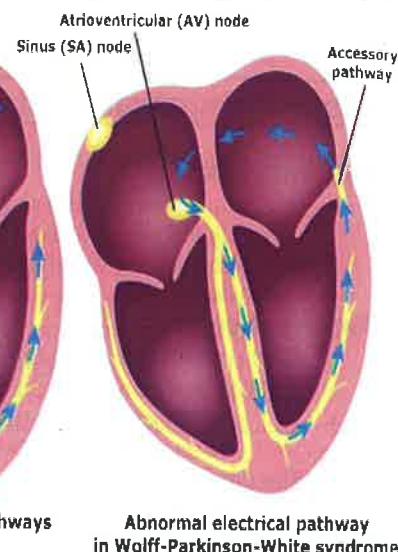


ECG showing short PR interval associated with a slurred upstroke (delta wave). Note the non-specific ST-T changes which are common in WPW and may be mistaken for ischaemia. The left axis deviation means that this is type B WPW, implying a right-sided pathway

Wolff-Parkinson-White Preexcitation



- Short PR
- Wide QRS
- Delta Wave (arrow)



Normal electrical pathways

Abnormal electrical pathway
in Wolff-Parkinson-White syndrome

Differentiating between type A and type B

- **type A** (left-sided pathway): dominant R wave in V1
- **type B** (right-sided pathway): no dominant R wave in V1
 - ⇒ In type B pre-excitation, the accessory pathway connects the right atrium to the right ventricle
- there is a rare type C WPW, WPW in which the delta waves are upright in leads V1-V4 but negative in leads V5-V6

Associations of WPW

- HOCM
- mitral valve prolapse
- **Ebstein's anomaly**
- thyrotoxicosis
- secundum ASD
- Leber's hereditary optic neuropathy (mitochondrial disease)

Investigations

- ECG
 - ⇒ Short PR interval
 - ⇒ ECG **delta wave**: a slurred upstroke at the start of the QRS complex, secondary to preexcitation
 - ⇒ Widened QRS
- **The most accurate test is electrophysiologic studies**

Management

Acute episodes

- **Hemodynamically unstable**: electrical cardioversion
- **Hemodynamically stable**: assess underlying rhythm
 - ⇒ **Narrow-complex tachycardia** (including Afib, atrial flutter)
 - Rhythm control measures (i.e. **IV procainamide** or cardioversion) are the safest treatment option.
 - Vagal maneuvers and AV nodal blocking agents (Adenosine, B-Blockers, Calcium Channel Blockers, Digoxin) are **contraindicated** (**may precipitate ventricular tachycardia or V-fib**)
 - ⇒ **Wide-complex regular or irregular tachycardia** → Determine whether the rhythm is more likely to be ventricular or supraventricular in origin (e.g., Brugada criteria)
 - **VT (~80%)**: pharmacological cardioversion or synchronized electrical cardioversion
 - **SVT (< 20%)**: Determine if an accessory pathway is present.
 - ⇒ Findings suggestive of an accessory pathway: **synchronized electrical cardioversion or IV procainamide**
 - HR > 200
 - Irregular rhythm
 - No bundle branch block on ECG
 - Signs of impending instability (e.g., clammy skin)
 - Baseline ECG findings that support the diagnosis
 - ⇒ No signs of an accessory pathway: manage as SVT
 - **Undifferentiated wide-complex tachycardia**: Treat as VT, with either electrical cardioversion or IV procainamide

Long-term management

- **High-risk patients** → **Catheter ablation**
 - ⇒ Syncpe
 - ⇒ Associated atrial fibrillation, atrial flutter, or atrial tachycardia
 - ⇒ Aborted sudden cardiac death
 - ⇒ Family history of sudden cardiac death
 - ⇒ High-risk occupations (e.g., pilots, athletes, school bus driver)
- **Low-risk patients**
 - ⇒ Asymptomatic patients: usually no treatment required
 - ⇒ symptomatic patients: First-line treatment → catheter ablation

Differentiating between VT and SVT

Brugada criteria

ECG finding	VT	SVT
Absence of RS in all precordial leads?	Yes	No
R:S interval > 100 ms in one precordial lead?	Yes	No
Signs of AV dissociation present?	Yes	No
QRS morphology consistent with VT in leads V ₁₋₂ and V ₆ ?	Yes	No

Interpretation

- If the answer to any is yes: most likely VT
- If none are present: most likely SVT

WPW management

- Asymptomatic : (incidentally found delta wave on ECG) → Reassurance
- Asymptomatic in high-risk professions (eg pilots, school bus driver) is best managed by catheter ablation of the accessory pathway
- Asymptomatic WPW in someone with a family history of sudden cardiac death is another indication for radiofrequency catheter ablation
- Chronic medical therapy: flecainide, amiodarone, procainamide
- Definitive treatment: radiofrequency ablation of the accessory pathway (first-line therapy)

Contraindications in WPW

A simple mnemonic to remember for drugs to avoid in WPW syndrome is ABCD (Adenosine, B-Blockers, Calcium Channel Blockers, Digoxin).

- **Digoxin**
- **Beta-blockers**
- **Diltiazem, verapamil**
- **Amiodarone**
- This is because blocking the AV node may enhance the rate of conduction through the accessory pathway, increasing the ventricular rate and potentially deteriorating into ventricular fibrillation.

If it is not possible to quickly identify the underlying rhythm as SVT or VT, it is safest to treat empirically as VT with synchronized electrical cardioversion (100 J) or with IV

If wide-complex tachycardia is present and the diagnosis of ventricular tachycardia (VT) cannot be excluded, the drugs of choice are IV procainamide or amiodarone.

Lown-Ganong-Levine (LGL) syndrome:

LGL syndrome is like WPW in the sense that it is a pre-excitation syndrome. However, the ECG changes present is only short PR interval without delta waves or abnormal QRS complex.

Implantable cardiac defibrillators (ICD)**Indications**

- Congenital long QT with family history of sudden cardiac death at young age.
- **hypertrophic obstructive cardiomyopathy (HOCM)**
- previous cardiac arrest due to VT/VF
- Sustained VT causing haemodynamic compromise
- previous myocardial infarction with non-sustained VT on 24 hr monitoring, inducible VT on electrophysiology testing and ejection fraction < 35%
- Brugada syndrome
- Arrhythmogenic right ventricular cardiomyopathy causing cardiac arrest.

Acute pericarditis

Overview

- Acute pericarditis: inflammation of the pericardium that either occurs as an isolated process or with concurrent myocarditis (myopericarditis).
- Pericarditis is one of the differentials of any patient presenting with chest pain.

Features

- Pleuritic chest pain
 - ⇒ Exacerbated by inspiration and lying flat, **relieved by sitting up and leaning forwards**
- Shoulder pain (referred pain): pericarditis is innervated by phrenic nerve
- Pericardial rub (present in 50% of cases.) → **pathognomonic feature**
- Other symptoms include non-productive cough, dyspnoea and flu-like symptoms

Types and causes

- **Fibrinous pericarditis (the most common type)**
 - ⇒ Causes:
 - **Viral infection is the most common cause of acute pericarditis: the most common viral cause is Coxsackie B virus**
 - Acute myocardial infarction (MI): more common than dressler syndrome
 - ⇒ friction rub is more common than pain
 - ⇒ Aspirin is the only NSAID that can be used in pericarditis complicating MI.
 - Post MI (Dressler syndrome): **rare**, autoimmune-mediated phenomenon to myocardial antigens, **occur 2 – 4 weeks post MI**
 - ⇒ Because of the risk of hemorrhagic pericarditis, anticoagulant therapy should be stopped in patients with dressler syndrome.
 - Radiation, trauma, severe infections
 - **Uremic pericarditis**
 - ⇒ blood urea nitrogen (BUN) level is usually greater than 60 mg/dL (22 mmol/L).
 - **Hemorrhagic effusions** are more common and result in part from **uremia-induced platelet dysfunction**.
 - does not present with the classic diffuse ST-elevations seen on ECG as in other types of pericarditis.
 - Uremic pericarditis is an indication for **urgent hemodialysis**.
- **Serous pericarditis**
 - ⇒ Usually caused by noninfectious inflammation such as: rheumatoid arthritis (RA) systemic lupus erythematosus (SLE).
 - ⇒ Fibrous adhesions rarely occur.
- **Purulent or suppurative pericarditis**
 - ⇒ Most commonly caused by staphylococcal and gram-negative species,
 - ⇒ high percentage of patients develop constrictive pericarditis.
- **Hemorrhagic pericarditis**
 - ⇒ Most commonly caused by:
 - **tuberculosis**, direct neoplastic invasion.
 - Severe bacterial infections
 - Bleeding diathesis, cardiac surgery or trauma (may cause tamponade).

- **Caseous pericarditis**
 - ⇒ caseation within the pericardial sac is **tuberculous in origin, until proven otherwise,**
 - ⇒ In tuberculous pericarditis, fever, night sweats, and weight loss are commonly noted (80%).
 - ⇒ Untreated, caseous pericarditis is **the most common antecedent to chronic constrictive pericarditis** of a fibrocalcific nature.
 - Approximately 50% of affected patients develop **constrictive pericarditis**.

ECG changes

- Stage 1 (initial)
 - ⇒ **Diffuse ST elevations**
 - ⇒ ST depression in aVR and V1
 - ⇒ PR segment depression (**most specific ECG marker for pericarditis**)
- Stage 2: **ST segment normalizes in ~ 1 week.**
- Stage 3: inverted T waves in all leads **~ 1 – 2 weeks**
- Stage 4: ECG returns to normal baseline after **weeks to months.**

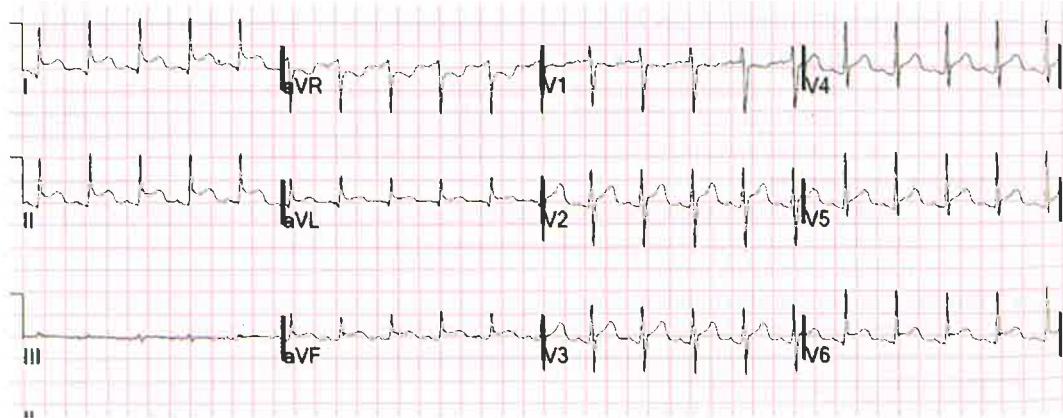
Which ECG changes would you expect to see in the next week or two?

⌚ **T-wave inversion in all leads**

Echocardiography is often normal in patients with pericarditis but is needed to rule out pericardial tamponade and pericardial constriction

Laboratory findings

- Elevation of inflammatory markers may support the diagnosis of pericarditis but are not considered to be a part of the diagnostic criteria.
- CBC: leukocytosis, ↑ **ESR, CRP**
- ↑ Troponin I : suggest some degree of myocarditis.
- ↑ Creatinine kinase



ECG showing pericarditis. Note the widespread nature of the ST elevation and the PR depression

Diagnosis

ESC guidelines defined the **diagnosis** of acute pericarditis as **2 out of 4** of the following:

- 1) pericarditic chest pain;
- 2) pericardial rub;
- 3) new widespread ST-elevation or PR depression; and
- 4) pericardial effusion (new or worsening).

- Rule out other causes of acute chest pain (e.g., myocardial infarction, myocarditis) before making a diagnosis of acute pericarditis.

Treatment

- **Pain management (analgesia, observation)**
 - ⇒ NSAID therapy (Aspirin, Ibuprofen)
 - ⇒ Post-myocardial infarction pericarditis: avoid NSAIDs other than aspirin.
 - ⇒ Colchicine (in combination with NSAIDs or as a monotherapy). **Useful both in acute episode and to prevent recurrence of pericarditis.**
- **Only consider prednisone in:**
 - ⇒ severe cases (not responded to NSAID and Colchicine)
 - ⇒ or in pericarditis caused by uremia, connective tissue disease, or autoreactivity.
- **Treat any known underlying causes**
- **Pericardectomy** is only indicated for recurrent pericarditis once medical interventions have failed.
- **Treatment duration:** until symptoms have resolved and CRP has normalized, but normally it is for 1-2 weeks duration.
- **Reduce physical activity**

Prognosis

- **Recurrence**
 - ⇒ Between 15 and 30% of patients with idiopathic acute pericarditis may have recurrent attacks, and this is considered to be an autoimmune phenomenon.
- **Poor prognostic factors** include:
 - ⇒ Temperature above 38°C
 - ⇒ Subacute disease course
 - ⇒ Presence of a large effusion or tamponade
 - ⇒ Unsuccessful therapy with nonsteroidal anti-inflammatory agents
- **Factors associated with complicated pericarditis** include:
 - ⇒ Early administration of high-dose corticosteroids
 - ⇒ Lack of colchicine treatment
 - ⇒ Elevated levels of high-sensitivity C-reactive protein

Acute pericarditis

- **Symptoms include** sharp, severe retrosternal chest pain worse with inspiration and a supine position.
- **The classic physical finding** is a pericardial friction rub. A low-grade fever is often present.
- **Diagnostic signs** include new widespread diffuse concave upwards ST elevation and/or PR depression on ECG and new or worsening pericardial effusion on echocardiography; blood tests generally suggest systemic inflammation.
- **Treatment:** All patients should be given a non-steroidal anti-inflammatory drug as first-line treatment. Colchicine should also be given unless the patient has tuberculous pericarditis.
- **Complications** include chronic recurrent pericarditis, cardiac tamponade, and constrictive pericarditis.

Pericardial effusion

Causes

- infectious pericarditis: viral, tuberculosis, pyogenic spread from septicaemia and pneumonia
- uraemia
- idiopathic
- post myocardial infarction (including Dressler's syndrome)
- malignancy
- heart failure
- nephrotic syndrome
- hypothyroidism
- **trauma**

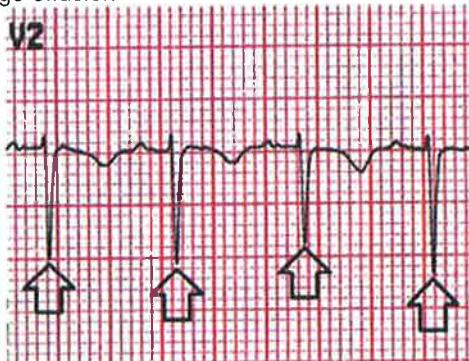
⇒ **CT is the most appropriate investigation**

- provide more information than Echo
- quicker to obtain than (MRI).

Investigations

- **ECG of pericardial effusion**

⇒ ECG reveals electrical alternans, which is caused by a "swinging" movement of the heart in a large effusion



Constrictive pericarditis

The right sided failure, ascites and pericardial calcification on x ray suggest a diagnosis of constrictive pericarditis.

Pathophysiology

- Inflammation of the pericardium → fibrosis and constriction

Risk factors

- **previous cardiac surgery**
- previous pericarditis,
- radiotherapy
- connective tissue disease

Causes

- **Mediastinal irradiation**
- TB : **Tuberculous pericarditis is the commonest cause of constrictive pericarditis worldwide.**
- any cause of purulent pericarditis

Features

- dyspnoea
- right heart failure: elevated JVP, ascites, oedema, hepatomegaly
- JVP shows prominent x and y descent
- pericardial knock - loud S3
- Kussmaul's sign is positive (**rise in JVP on inspiration**)

Investigations

- CXR
 - ⇒ pericardial calcification
 - ⇒ can detect effusions only if larger than 250 mL.
- Echocardiography
 - ⇒ Indication → to assess for pericardial effusion and cardiac tamponade
 - ⇒ the best diagnostic tool for diagnosing pericardial effusion.
 - ⇒ shows no increase in the venous return with inspiration.

The key differences between constrictive pericarditis and cardiac tamponade are summarized in the table below:

	Cardiac tamponade	Constrictive pericarditis
JVP	Absent Y descent	X + Y present
Pulsus paradoxus	Present	Absent
Kussmaul's sign*	Rare	Present
Characteristic features		Pericardial calcification on CXR

- Kussmaul's sign* → a paradoxical rise in jugular venous pressure (JVP) on inspiration
- Kussmaul's sign (a rise in the JVP on inspiration) is more likely to be seen in constrictive pericarditis than cardiac tamponade.

Treatment

- The first line of treatment of symptomatic constrictive pericarditis is pericardiotomy.

Cardiac tamponade

Cardiac tamponade is characterised by **Beck's triad** of:

- **hypotension**
- raised JVP (with absent Y descent), and
- muffled heart sounds.

Definition

- an accumulation of pericardial fluid under pressure, leading to impaired cardiac filling and hemodynamic compromise

Features

- dyspnoea
- raised JVP, with an absent Y descent - this is due to the limited right ventricular filling
- tachycardia
- **Hypotension**
 - ⇒ **the best clinical features that distinguishes cardiac tamponade from constrictive pericarditis**

- hypotension is a **late** feature in constrictive pericarditis.
- muffled heart sounds
- **pulsus paradoxus**
 - ⇒ an exaggerated inspiratory decrease in systolic blood pressure
- Kussmaul's sign
 - ⇒ **Rare**
 - ⇒ **Most common in constrictive pericarditis**
- impalpable apex beat

Investigations

- ECG :
 - ⇒ tachycardia,
 - ⇒ **low voltage**,
 - ⇒ electrical alternans, (due to the swinging movement of the heart).
 - beat-to-beat variation in QRS-axis and amplitude.
- chest x-ray (enlarged cardiac silhouette with clear lung fields),
- echocardiogram (chamber collapses, abnormal venous flows, exaggerated respiratory variation of cardiac and venous flows).

Treatment

- pericardiocentesis.

Hypotension is the best clinical features that distinguishes cardiac tamponade from constrictive pericarditis

⇒ hypotension is a **late** feature in constrictive pericarditis.

Hypertension (NICE guidelines 2019)

Definition

- Essential hypertension is defined as blood pressure (BP) $\geq 140/90$ mmHg, with no secondary cause identified.

Causes

- Essential hypertension (95% of patients)
 - ⇒ No specific cause known. Multifactorial etiology including genetic and environmental factors
- Secondary hypertension (5% of patients)
 - ⇒ **RECENT:** Renal (e.g., renal artery stenosis, glomerulonephritis), **Endocrine** (e.g., Cushing syndrome, hyperthyroidism, Conn syndrome), **Coarctation of the aorta**, **Estrogen** (oral contraceptives), **Neurologic** (raised intracranial pressure, psychostimulants use), **Treatment** (e.g., glucocorticoids, NSAIDs) are the causes of secondary hypertension.

When a question says: 'What is the most likely diagnosis?' think about what is epidemiologically the most common cause of hypertension? Therefore the answer is essential hypertension. **The most likely cause of hypertension in an obese is still essential hypertension.**

Diagnosis

Hypertension - NICE now recommend ambulatory blood pressure monitoring to aid diagnosis

Confirm diagnosis of hypertension in people with a: clinic blood pressure of 140/90 mmHg or higher and ABPM daytime average or HBPM average of 135/85 mmHg or higher.

Measuring blood pressure

- **Palpate the radial or brachial pulse before measuring blood pressure with automated devices.** If pulse irregularity is present, measure BP **manually** using direct auscultation over the brachial artery, because **automated devices may not measure BP accurately if there is pulse irregularity** (for example, due to atrial fibrillation).
- **Measure BP in both arms**
 - ⇒ If the difference between arms > 15 mmHg ⇒ **repeat BP**. If the difference remains > 15 mmHg:
 - Subsequent BP **should be recorded from the arm with the higher reading**.
 - Look for cases of **unequal BP from the arms**, e.g. supravalvular aortic stenosis.
- **If BP in the clinic ≥ 140/90 mmHg:**
 - ⇒ Take a second measurement during the consultation.
 - ⇒ If the second measurement is substantially different from the first, take a third measurement.
 - ⇒ Record **the lower** of the last 2 measurements as the clinic blood pressure.
- **If clinic BP is between 140/90 mmHg and 180/120 mmHg, offer ambulatory blood pressure monitoring (ABPM) to confirm the diagnosis of hypertension.** If ABPM is unsuitable or not tolerated, offer home blood pressure monitoring (HBPM).
- **In people with symptoms of postural hypotension (falls or postural dizziness):**
 - ⇒ measure BP with the person either supine or seated and with the person standing for at least 1 minute before measurement.
 - ⇒ If the systolic BP falls by 20 mmHg or more when the person is standing:
 - review medication
 - measure subsequent BP with the person standing
 - consider referral to specialist care if symptoms of postural hypotension persist.
- **Ambulatory blood pressure monitoring (ABPM)**
 - ⇒ The use of ambulatory blood pressure monitoring (ABPM) aims to:
 - prevent diagnosing 'white coat hypertension' as having hypertension in patients whose blood pressure climbs 20 mmHg whenever they enter a clinical setting.
 - ABPM has been shown to be a more accurate predictor of cardiovascular events than clinic readings.
 - ⇒ at least 2 measurements per hour during the person's usual waking hours (for example, between 08:00 and 22:00)
 - ⇒ use the average value of at least 14 measurements
 - ⇒ If ABPM is not tolerated or declined HBPM should be offered.

- **Home blood pressure monitoring (HBPM)**

- ⇒ for each BP recording, two consecutive measurements need to be taken, at least 1 minute apart and with the person seated
- ⇒ BP should be recorded twice daily, ideally in the morning and evening
- ⇒ BP should be recorded for at least 4 days, ideally for 7 days
- ⇒ discard the measurements taken on the first day and use the average value of all the remaining measurements.

Hypertension terms used in NICE guidelines 2019

Term	Definition
Hypertension	clinic BP of $\geq 140/90$ mmHg or higher and ABPM daytime average or HBPM average of $\geq 135/85$ mmHg.
White-coat hypertension	A discrepancy of more than 20/10 mmHg between clinic and average daytime ABPM or average HBPM measurements at the time of diagnosis.
Masked hypertension	Clinic BP measurements are normal ($<140/90$ mmHg), but higher when taken outside the clinic using average daytime ABPM or average HBPM BP measurements.
Persistent hypertension	High blood pressure at repeated clinical encounters.
severe hypertension	Stage 3 hypertension: Clinic systolic BP ≥ 180 mmHg or clinic diastolic BP ≥ 120 mmHg.
Accelerated hypertension (malignant hypertension)	A severe increase in BP to $\geq 180/120$ mmHg (and often over 220/120 mmHg) with signs of retinal haemorrhage and/or papilloedema (swelling of the optic nerve). It is usually associated with new or progressive target organ damage and is also known as malignant hypertension.

Hypertension Stages (NICE guidelines 2019)

Stage	Criteria
Stage 1 hypertension	Clinic BP $\geq 140/90$ mmHg and subsequent ABPM daytime average or HBPM average BP $\geq 135/85$ mmHg
Stage 2 hypertension	Clinic BP $\geq 160/100$ mmHg and subsequent ABPM daytime average or HBPM average BP $\geq 150/95$ mmHg
Stage 3 or severe hypertension	Clinic systolic BP ≥ 180 mmHg, or clinic diastolic BP ≥ 120 mmHg

Management (NICE guidelines 2019)

Non-pharmacological management

- Lifestyle advice is the **first line** in hypertension management
 - ⇒ **weight reduction:** Of all the lifestyle modifications, weight reduction produces the greatest reduction in BP (A 10 kg weight loss is expected to decrease BP by 15–20 mmHg)
 - ⇒ **low salt diet**, aiming for less than 6g/day, ideally 3g/day. (reducing salt intake by 6g/day can lower systolic blood pressure by 10mmHg)
 - ⇒ low caffeine intake.

- ⇒ stop smoking, drink less alcohol
- If a patient on antihypertensive and drink alcohol → Reduction of alcohol intake is the next step in treatment.

Starting antihypertensive drug treatment

- any age with persistent stage 2 hypertension.
- age < 60 years with stage 1 hypertension and an estimated 10-year cardiovascular risk below 10%.
- age < 80 years with stage 1 hypertension who have 1 or more of the following:
 - ⇒ target organ damage
 - ⇒ established cardiovascular disease
 - ⇒ renal disease
 - ⇒ diabetes
 - ⇒ an estimated 10-year risk of cardiovascular disease of 10% or more.
- age > 80 years with stage 1 hypertension if their clinic BP >150/90 mmHg
- For patients < 40 years → consider specialist referral to exclude secondary causes.

Pharmacological management : Steps of hypertension treatment

- **Step 1 treatment**
 - ⇒ Age ≤ 55 OR any age, with T2DM with no black African origin → ACEi or ARB
 - ⇒ Age ≤ 55 OR any age, with T2DM with black African origin → ARB
 - ⇒ Age ≥ 55 without T2DM → CCB
 - ⇒ black African origin of any age without T2DM → CCB
 - ⇒ With heart failure → thiazide-like diuretic, such as indapamide in preference to a conventional thiazide diuretic such as bendroflumethiazide or hydrochlorothiazide.
- **Step 2 treatment**
 - ⇒ If BP not controlled on ACEi or ARB → Add CCB or thiazide-like diuretic.
 - ⇒ If BP not controlled on CCB → Add ACEi or ARB or thiazide-like diuretic.
 - ⇒ If BP not controlled on CCB in a black African → Add ARB (in preference to an ACEi)
- **Step 3 treatment**
 - ⇒ If BP not controlled with step 2 treatment → ACEi or ARB and CCB and thiazide-like diuretic.
- **Step 4 treatment**
 - ⇒ BP not controlled with the optimal tolerated doses of ACEi or ARB and CCB and thiazide-like diuretic → Resistant hypertension
 - ⇒ Before considering further treatment for a person with resistant hypertension:
 1. Confirm elevated clinic blood pressure measurements using ambulatory or home blood pressure recordings.
 2. Assess for postural hypotension.
 3. Discuss adherence
 - ⇒ Confirmed resistant hypertension:
 - blood potassium ≤ 4.5 mmol/l → further diuretic therapy with low-dose spironolactone
 - monitor blood sodium and potassium and renal function within 1 month of starting treatment and repeat as needed thereafter.
 - blood potassium > 4.5 mmol/l → alpha-blocker or beta-blocker

Calcium channel blockers are now preferred to thiazides in the treatment of hypertension

Hypertension in diabetics - ACE-inhibitors are first-line regardless of age

ACE inhibitors have reduced efficacy in black patients and are therefore not used first-line

Drug choice

- *hypertensive with benign prostatic hyperplasia → alpha-blockers*
- *hypertensive with heart failure or angina → beta-blockers*
- *hypertensive post myocardial infarction either a beta blocker or ACE inhibitor would be the agent of choice.*
- *calcium channel blockers are now considered superior to thiazides*
- *bendroflumethiazide is no longer the thiazide of choice*

Use of multiple anti-hypertensives at low doses is preferable to having fewer tablets at higher doses, in view of the synergistic effectiveness of targeting several mechanisms of hypertension.

Blood pressure targets

Blood pressure target (based on clinic readings) for patients < 80 years - 140/90 mmHg

	Clinic BP	ABPM / HBPM
Age < 80 years	140/90 mmHg	135/85 mmHg
Age > 80 years	150/90 mmHg	145/85 mmHg

Recommendations for BP target

- British Hypertension Society Guidelines for Hypertension Management (BHS-IV) recommend a goal BP of **less than 130/80 mmHg** for patients with **diabetes, renal impairment** and established **cardiovascular disease**;

Hypertensive emergency

Definition

Hypertensive emergency: systolic BP ≥ 180 or diastolic BP $\geq 110 +$ end organ damage

Presentation

- The most common clinical presentations of hypertensive emergencies are:
 - ⇒ cerebral infarction (24.5%)
 - ⇒ pulmonary edema (22.5%),
 - ⇒ hypertensive encephalopathy (16.3%),
 - ⇒ congestive heart failure (12%).
 - ⇒ Other presentations include intracranial hemorrhage, aortic dissection, and eclampsia as well as acute myocardial infarction.

Management

Labetalol has both alpha- and beta-adrenoreceptor antagonistic activity and is the first choice for hypertensive crises where the aetiology is initially unclear.

- Gradual blood pressure lowering over the first 24 hours
 - ⇒ in the first hour : reduce mean arterial pressure (MAP) by 10 – 20 %
 - ⇒ in the next 23 hours: 5% to 15%, so that the final BP is reduced by 25% compared with baseline.
- IV antihypertensive : e.g. Labetolol
 - ⇒ The major risk of any oral agent used for hypertensive emergencies is ischaemic symptoms (for example myocardial infarction, angina pectoris or stroke) due to an excessive and uncontrolled hypotensive response usually due to lowering of BP to below the autoregulatory threshold. Therefore the use of oral agents should generally be avoided in the treatment of hypertensive emergencies if parenteral drugs are available.
- The exceptions to gradual BP lowering over the first 24 hours are:
 - ⇒ Acute ischemic stroke – The BP should not lowered unless it is $\geq 185/110$ mmHg in patients who are candidates for reperfusion therapy or $\geq 220/120$ mmHg in patients who are not candidates for reperfusion therapy.
 - ⇒ Acute aortic dissection – The systolic BP should rapidly lowered to a target of 100 to 120 mmHg (to be attained in 20 minutes).
 - ⇒ Spontaneous hemorrhagic stroke – The systolic BP can be rapidly reduced if no contraindications exist.

MAP = diastolic blood pressure + [(systolic blood pressure - diastolic blood pressure)/3]
 Or MAP = (2x diastolic + systolic)/3

Parenteral drugs for treatment of hypertensive emergencies

	Side effects (SE)	Notes
Labetalol (Adrenergic inhibitor) Beta-Blocker With Alpha-Blocking Activity	Nausea/vomiting, paresthesias (eg, scalp tingling) , bronchospasm, dizziness, nausea, heart block	Avoid in acute decompensated heart failure. Use cautiously in obstructive or reactive airway. Beta-blocker should not be withdrawn abruptly, but gradually tapered to avoid acute tachycardia, hypertension, and/or ischemia.
Nitroglycerin (glyceryl trinitrate) (Vasodilators)	Hypoxemia, tachycardia (reflex sympathetic activation) , headache, vomiting, flushing, methemoglobinemia , tolerance with prolonged use	used as adjunctive therapy for patients with acute coronary syndrome or acute pulmonary edema. contraindicated in patients with increased intracranial pressure (eg, intracranial hemorrhage)
Nicardipine (Vasodilators) Calcium Channel Blocker, Dihydropyridine	Tachycardia, headache, dizziness, nausea, flushing, local phlebitis, edema	Avoid use in acute heart failure. Caution with coronary ischemia,
Clevidipine (Vasodilators) Calcium Channel Blocker, Dihydropyridine	Atrial fibrillation (most common SE), nausea, lipid formulation contains potential allergens (eg, soy, egg)	Avoid in patients with defective lipid metabolism (hypertriglyceridemia is an expected SE). Patients who develop hypertriglyceridemia (eg, >500 mg/dL) are at risk of developing pancreatitis. Dihydropyridine calcium channel blockers may cause negative inotropic effects and exacerbate HF.
Hydralazine (Vasodilators) Direct vasodilation of arterioles	Sudden precipitous drop in blood pressure , tachycardia, flushing, headache, vomiting, aggravation of angina	In general, hydralazine should be avoided due to its prolonged and unpredictable hypotensive effect. Contraindicated in coronary artery disease; mitral valve rheumatic heart disease and SLE.
Nitroprusside (Vasodilators)	Elevated intracranial pressure, decreased cerebral blood flow, reduced coronary blood flow in CAD, cyanide and thiocyanate toxicity , nausea, vomiting, muscle spasm, flushing, sweating	In general, nitroprusside should be avoided due to its toxicity. avoid in AMI, CAD, CVA, elevated intracranial pressure, renal or hepatic impairment.
Phentolamine (Adrenergic inhibitor) Alpha ₁ Blocker	Tachycardia, flushing, headache, nausea/vomiting	Alternative option for catecholamine excess (eg, adrenergic crisis secondary to pheochromocytoma or cocaine overdose).

Hypertensive urgency

Definition

Hypertensive urgency: systolic BP ≥ 180 or diastolic BP $\geq 110 +$ NO end organ damage

Presentation

- Asymptomatic patient with a BP in the "severe" range (ie, $\geq 180/\geq 120$ mmHg)
- Often a mild headache, but **no** signs or symptoms of acute end-organ damage.

Management

- All patients should be provided a quiet room in which to rest. This may produce a fall in blood pressure $\geq 20/10$ mmHg in approximately one-third of adults. If this is not effective, antihypertensive drugs may be given.
- Gradual lowering of the BP over a period of hours to days to $<160/<100$ mmHg or no more than 25 to 30% of baseline BP.
- The risk of adverse events (eg, stroke or myocardial infarction) that may occur if the BP is lowered too rapidly or to a level below the ability for autoregulation to maintain adequate tissue perfusion.
- Can often be safely managed in the clinician's office
- Add or modify oral antihypertensive

Malignant hypertension (Accelerated hypertension)

A patient with **malignant hypertension** always has retinal **papilledema**

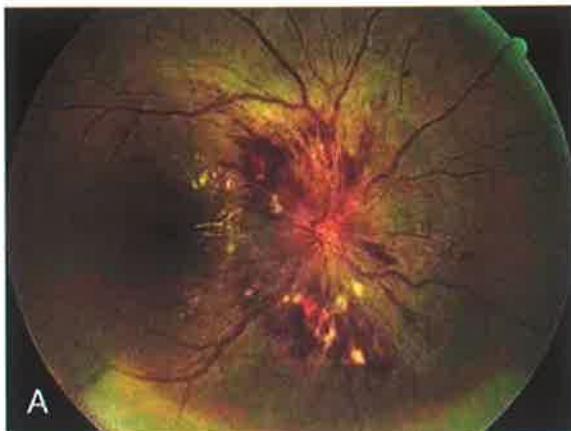
Definition

- BP $\geq 180/120$ mmHg (often over 220/120 mmHg) **with signs of retinal haemorrhage and/or papilloedema** (swelling of the optic nerve).
- It is usually associated with new or progressive **target organ damage**

Pathophysiology

- The pathologic hallmark of malignant hypertension is **fibrinoid necrosis of the arterioles** which occurs systemically, but specifically in the kidneys.

Management : as hypertensive emergency



Papilledema. Note the swelling of the optic disc, with blurred margins

Secondary hypertension

General indicators of secondary hypertension

- Young age (< 40 years) at onset of hypertension
- Onset of diastolic hypertension at an older age (> 55 years)
- Abrupt onset of hypertension
- End-organ damage that is disproportionate to the degree of hypertension
- Recurrent hypertensive crises
- Resistant hypertension: hypertension that is resistant to treatment with at least three antihypertensives of different classes including a diuretic

Causes

- Primary hyperaldosteronism, including Conn's syndrome (5-10% of hypertensive patients)
 - ⇒ the single most common cause of secondary hypertension
 - ⇒ ↑BP + ↓K⁺ + ↑Aldosterone
 - ⇒ CT or MRI of the abdomen identifies a secretory adrenal adenoma
- Renal diseases: include
 - ⇒ glomerulonephritis
 - ⇒ pyelonephritis
 - ⇒ Reflux-associated scarring is the commonest renal disease.
 - This will cause abnormalities on dimercaptosuccinic acid (DMSA) scan.
 - ⇒ adult polycystic kidney disease
 - ⇒ renal artery stenosis
- Coarctation of the aorta (the commonest non-renal cause)
- Endocrine disorders (other than primary hyperaldosteronism):
 - ⇒ phaeochromocytoma
 - ⇒ Cushing's syndrome
 - ⇒ Liddle's syndrome → (↑BP + ↓K⁺ + ↑Na⁺)
 - hypokalaemic hypertension
 - metabolic alkalosis
 - low plasma renin and aldosterone (called pseudo-hyperaldosteronism).

- ⇒ congenital adrenal hyperplasia (11-beta hydroxylase deficiency)
- ⇒ acromegaly
- **Fibromuscular dysplasia,**
 - ⇒ a rare cause of hypertension and hypokalaemia,
 - ⇒ more common in women.
 - ⇒ It causes hyperreninaemic hyperaldosteronism.
- **Pregnancy** (PIH, pre-eclampsia , eclampsia)
- **Drugs**
 - ⇒ **Liquorice ingestion**
 - causes a primary aldosterone type picture.
 - **It is caused by glycyrrhizic acid** contained in liquorice, blocking the enzyme 11b hydroxysteroid dehydrogenase. This prevents the inactivation of cortisol, which in turn activates mineralocorticoid receptors in the kidney. driving hypokalaemic metabolic alkalosis with hypertension.
 - ⇒ NSAIDs, combined oral contraceptive pill, steroids, MAOI

Different diagnostics for causes of secondary hypertension	
Diagnostic findings	Underlying condition
▪ Hypokalaemia	▪ Conn syndrome ▪ Renal artery stenosis
▪ Metabolic alkalosis and ↑ aldosterone-to-renin ratio	▪ Conn syndrome
▪ Difference in blood pressure in both arms	▪ Takayasu arteritis ▪ Aortic dissection ▪ Aortic arch syndrome ▪ Subclavian steal syndrome
▪ Of upper and lower limbs	▪ Coarctation of the aorta distal to the left subclavian artery
▪ Daytime sleepiness (Epworth scale, Berlin questionnaire)	▪ Obstructive sleep apnoea
▪ Nondipping in 24-hour blood pressure monitoring (the failure of BP to fall by ≥10% during sleep.)	
▪ Increased 24-hour urinary metanephhrines	▪ Pheochromocytoma
▪ ↑ Serum calcium, ↑ PTH level, ↓ serum phosphates	▪ Hyperparathyroidism
▪ ↑ Serum cortisol	▪ Excess of glucocorticoids (e.g., Cushing syndrome)
▪ ↓TSH, ↑ free T4	▪ Hyperthyroidism

MRCPUK- part 2- March 2017 : A 28-year-old woman of Afro-Caribbean ethnic origin c/o difficult-to-manage hypertension, despite taking maximal-dose amlodipine and indapamide. The GP trialled an ACE inhibitor, but this was discontinued due to a rise in serum creatinine. Renin and aldosterone are both Elevated. K is 3.1 mmol. Which of the following is the most likely diagnosis?

⇒ **Fibromuscular renal artery dysplasia**

- This patient's age and ethnicity suggest that her hypertension is related to fibromuscular dysplasia rather than to atherosclerotic renal artery stenosis.
- The renin and aldosterone elevation, coupled with hypokalaemia and deterioration in renal function on starting ACE inhibitors, are consistent with the diagnosis.

Differences in blood pressure between arms:

- Up to 10 mmHg difference → **Normal variant (physiological)**
- Difference > 10 mmHg: → **abnormal**:
 - ⇒ + radio-radial or radio-femoral delay (NO Leg claudication) → **proximal coarctation** of the aorta (Involves the left subclavian artery origin)
 - ⇒ + **arm claudication**, intermittent vertigo, ataxia or diplopia, or **facial sensory symptoms** (NO Leg claudication) → **Subclavian steal syndrome**
 - ⇒ + **Leg claudication** (chronic intermittent leg pain, exacerbated by exercise and relieved by rest) → **Peripheral vascular disease**

Hypokalaemia and hypertension

Liddle's syndrome: hypokalaemia + hypertension

Hypokalaemia with hypertension	Hypokalaemia without hypertension
<ul style="list-style-type: none"> ▪ Cushing's syndrome ▪ Conn's syndrome (primary hyperaldosteronism) ▪ Liddle's syndrome (autosomal dominant disorder that mimics hyperaldosteronism) • renal artery stenosis • 11-beta hydroxylase deficiency <ul style="list-style-type: none"> ⇒ 21-hydroxylase deficiency, which accounts for 90% of congenital adrenal hyperplasia cases, is not associated with hypertension • Carbenoxolone, an anti-ulcer drug, and liquorice excess 	<ul style="list-style-type: none"> ▪ Diuretics ▪ GI loss (e.g. Diarrhoea, vomiting) ▪ renal tubular acidosis (type 1 and 2) <ul style="list-style-type: none"> ⇒ type 4 renal tubular acidosis is associated with hyperkalaemia. ▪ Bartter's syndrome ▪ Gitelman syndrome

- The first step in case of (\uparrow BP + \downarrow K $^{+}$) should be further simple investigations → **Plasma renin and aldosterone levels**
 - ⇒ Cushing's & Conn's → high aldosterone and a low renin,
 - ⇒ Renal artery stenosis → high renin and aldosterone
 - ⇒ Liddle's syndrome → low renin and aldosterone.

Hypertension in pregnancy

Labetalol is first-line for pregnancy-induced hypertension

Physiology

- The blood pressure in normal pregnancy:
 - usually falls in the first trimester (particularly the diastolic), and continues to fall until 20-24 weeks
 - after this time the blood pressure usually increases to pre-pregnancy levels by term

Definition

- Hypertension in pregnancy is usually defined as:
 - systolic > 140 mmHg or diastolic > 90 mmHg
 - or an increase above booking readings of > 30 mmHg systolic or > 15 mmHg diastolic

Classification

Pre-existing hypertension	Pregnancy-induced hypertension (PIH, also known as gestational hypertension)	Pre-eclampsia
A history of hypertension before pregnancy or BP > 140/90 mmHg before 20 weeks gestation	Hypertension (as defined above) occurring in the second half of pregnancy (i.e. after 20 weeks)	Pregnancy-induced hypertension in association with proteinuria (> 0.3g / 24 hours)
No proteinuria, no oedema	No proteinuria, no oedema	Oedema may occur but is now less commonly used as a criterion
Occurs in 3-5% of pregnancies and is more common in older women	Occurs in around 5-7% of pregnancies	Occurs in around 5% of pregnancies
	Resolves following birth (typically after one month). Women with PIH are at increased risk of future pre-eclampsia or hypertension later in life	

Treatment of chronic hypertension with pregnancy

- **Pre-pregnancy advice:** If they are taking ACE inhibitors or ARBs, thiazide or thiazide-like diuretics and planning for pregnancy discuss an alternative antihypertensive treatment, stop it if they become pregnant. the limited evidence available has not shown an increased risk of congenital malformation with any other antihypertensive.
- **Best antihypertensive:**
 - ⇒ **1st line :** labetalol
 - ⇒ **2nd line:** nifedipine (if labetalol is not suitable)
 - ⇒ **3rd line:** methyldopa (if both labetalol and nifedipine are not suitable)
- **Target BP: 135/85 mmHg**
- **Aspirin** 75–150 mg once daily from 12 weeks.
- Offer placental growth factor (PIGF)-based testing to help rule out pre-eclampsia between 20 weeks and up to 35 weeks of pregnancy, if women with chronic hypertension or PIH are suspected of developing pre-eclampsia.

Treatment of hypertension in the postnatal period

- If women not planning to breastfeed → treat as hypertension in general
- If women planning to breastfeed:
 - ⇒ **1st line:**
 - non-black African or Caribbean women: enalapril
 - black African or Caribbean women: nifedipine **or** amlodipine if the woman has previously used this to successfully control her BP.
 - ⇒ **2nd line:** combination of nifedipine (or amlodipine) and enalapril
 - ⇒ **3rd line:** add atenolol or labetalol to the combination treatment **or** swapping 1 of the medicines already being used for atenolol or labetalol.
- avoid using diuretics or angiotensin receptor blockers for women who are breastfeeding.

Treatment of hypertension in the postnatal period

(NICE guidelines June 2019)

- | |
|---|
| <ul style="list-style-type: none"> • If women not planning to breastfeed → treat as hypertension in general |
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| <ul style="list-style-type: none"> • Avoid using diuretics or angiotensin receptor blockers for women who are breastfeeding. |

Pre-eclampsia/Eclampsia

Severe pre-eclampsia - restrict fluids

Eclampsia - give magnesium sulphate first-line

Definitions

- **Pre-eclampsia:** is a condition seen after 20 weeks gestation characterised by pregnancy-induced hypertension in association with proteinuria.
⇒ use albumin: creatinine ratio (8 mg/mmol) **or** protein: creatinine ratio (≥ 30 mg/mmol) to confirm significant proteinuria (Do not use 24-hour proteinuria or first morning urine void).
- **Eclampsia:** development of seizures in association pre-eclampsia.

Risk factors

- > 40 years old
- nulliparity (or new partner)
- multiple pregnancy
- body mass index $> 30 \text{ kg/m}^2$
- diabetes mellitus
- pregnancy interval of more than 10 years
- family history of pre-eclampsia
- previous history of pre-eclampsia
- **pre-existing vascular disease** such as **hypertension** or renal disease
- **There is some evidence to suggest that pre-eclampsia is actually less common in smokers**

Features of pre-eclampsia

- severe headache
- problems with vision, such as blurring or flashing before the eyes
- severe pain just below the ribs
- vomiting
- sudden swelling of the face, hands or feet.

Prevention of pre-eclampsia

- Aspirin 75–150 mg of aspirin daily from 12 weeks until the birth of the baby is indicated for pregnant with:
 - ⇒ **One** of the following **high risk factor** for pre-eclampsia:
 - hypertensive disease during previous pregnancies
 - chronic kidney disease
 - autoimmune disorders such as SLE or antiphospholipid syndrome
 - type 1 or 2 diabetes mellitus
 - chronic hypertension.
 - ⇒ **More than one** of the following **moderate risk factor** for pre-eclampsia:
 - first pregnancy
 - age 40 years or older

- pregnancy interval of more than 10 years
- body mass index (BMI) of 35 kg/m² or more at first visit
- family history of pre-eclampsia
- multi-fetal pregnancy.

Treatment

- **Pre-eclampsia**
 - ⇒ **Target blood pressure:** BP of 135/85 mmHg or less
 - ⇒ **Best antihypertensive:**
 - **1st line :** labetalol
 - **2nd line:** nifedipine (if labetalol is not suitable)
 - **3rd line:** methyldopa (if both labetalol and nifedipine are not suitable)
 - ⇒ Consider **magnesium sulfate** treatment, if 1 or more of the following **features of severe pre-eclampsia** is present:
 - ongoing or recurring severe headaches
 - visual scotomata
 - nausea or vomiting
 - epigastric pain
 - oliguria and severe hypertension
 - progressive deterioration in laboratory blood tests (such as rising creatinine or liver transaminases or falling platelet count).
- **Eclampsia**
 - ⇒ Magnesium sulphate is used to both prevent seizures in patients with severe pre-eclampsia and treat seizures once they develop.
 - IV bolus of 4g over 5-15 minutes followed by an infusion of 1g / hour for 24 hours.
 - Recurrent fits should be treated with a further dose of 2–4 g given intravenously over 5 to 15 minutes.
 - urine output, reflexes, respiratory rate and oxygen saturations should be monitored during treatment
 - treatment should continue for 24 hours after last seizure or delivery (around 40% of seizures occur post-partum)
 - Other important aspects of treating severe pre-eclampsia/eclampsia include **fluid restriction** to avoid the potentially serious **consequences** of fluid overload (limit maintenance fluids to 80 ml/hour)
 - delivery of the baby is the most important and definitive management step. The timing depends on the individual clinical scenario.

Pulmonary arterial hypertension (PAH)

Definition

- Sustained elevation in mean pulmonary arterial pressure of greater than 25 mmHg at rest or 30 mmHg after exercise.

Epidemiology

- More common in females
- Typically presents at 20-40 years old

WHO Classification & causes

1. Group 1: idiopathic pulmonary arterial hypertension (IPAH)

- Idiopathic (previously termed primary pulmonary hypertension (PPH))
 - 10% are familial (autosomal dominant)
 - Diagnosed when no underlying cause can be found
 - Endothelin thought to play a key role in pathogenesis

2. Group 2: Pulmonary hypertension with left heart disease

- Congenital heart disease with systemic to pulmonary shunts
- Left-sided atrial, ventricular or valvular disease such as left ventricular systolic and diastolic dysfunction, mitral stenosis and mitral regurgitation

3. Group 3: Pulmonary hypertension secondary to lung disease/hypoxia

- COPD
- Interstitial lung disease
- Sleep apnoea
- High altitude

4. Group 4: Pulmonary hypertension due to thromboembolic disease

5. Group 5: Miscellaneous conditions

- Lymphangiomatosis e.g. secondary to carcinomatosis or sarcoidosis
- Collagen vascular disease
- HIV (the mechanism by which HIV infection produces pulmonary hypertension remains unknown)
- Sickle cell disease
 - Haemoglobinopathies** (eg: sickle cell anaemia, thalassemia)
 - intravascular hemolysis** → ↓ nitric oxide (NO) → pulmonary vasoconstriction
- Drugs and toxins: cocaine and anorexigens (e.g. fenfluramine)

Increased pressure in pulmonary circuit → elevated right ventricular afterload → dilatation and/or hypertrophy of the right heart → right heart failure and arrhythmias

Lung disease can cause pulmonary hypertension by hypoxic vasoconstriction, whereas the heart can cause pulmonary hypertension by pump failure and subsequent fluid backup and stasis.

- Pulmonary arterial hypertension is caused by an intrinsic increase in the resistance of the pulmonary vasculature, while pulmonary hypertension can be caused by secondary aetiologies such as lung disease and heart failure.
- The most common cause of pulmonary arterial hypertension is idiopathic, while the most common overall cause of pulmonary hypertension is left-sided heart failure.

Features

Women with pulmonary hypertension should avoid becoming pregnant due to very high mortality levels

Bosentan - endothelin-1 receptor antagonist

- Symptoms
 - ⇒ exertional dyspnoea is the most frequent symptom
 - progressive SOB
 - ⇒ chest pain and syncope may also occur
- On examination:
 - ⇒ cyanosis
 - ⇒ Nail clubbing
 - ⇒ raised JVP with prominent 'a' waves,
 - ⇒ left parasternal heave (due to right ventricular hypertrophy)
 - ⇒ loud P2
 - ⇒ tricuspid regurgitation

Investigation

- **Doppler echocardiography**
 - ⇒ **the initial investigation of choice**
 - ⇒ the jet associated with tricuspid regurgitation can be visualised adequately (tricuspid regurgitant jet velocity)
- **Right heart catheterization**
 - ⇒ confirmatory test
 - ⇒ **the gold standard for the diagnosis**

World Health Organization (WHO) functional classification for pulmonary hypertension

Class	WHO functional classification for pulmonary hypertension
I	No limitations of physical activity. Ordinary physical activity does not cause undue fatigue or dyspnea, chest pain, or heart syncope.
II	Slight limitation of physical activity. Ordinary physical activity results in undue fatigue or dyspnea, chest pain, or heart syncope. Comfortable at rest.
III	Marked limitation of physical activity. Less than ordinary physical activity causes undue fatigue or dyspnea, chest pain, or heart syncope. Comfortable at rest
IV	Inability to carry on any physical activity without symptoms. Dyspnea and/or fatigue may be present even at rest.

Management

- Treatment of the underlying cause for example:
 - ⇒ Anticoagulants for PE
 - ⇒ Bronchodilators and inhalation corticosteroids for COPD,
 - ⇒ CPAP for patients with obstructive sleep apnea
- **Acute vasodilator testing** is central to deciding on the appropriate management strategy.

- ⇒ Acute vasodilator testing aims to decide which patients show a significant fall in pulmonary arterial pressure following the administration of vasodilators such as intravenous epoprostenol or inhaled nitric oxide
 - If there is a positive response → oral calcium channel blockers
 - If there is a negative response:
 - ❖ prostacyclin analogues: treprostinil, iloprost
 - ❖ endothelin receptor antagonists: bosentan
 - ❖ phosphodiesterase inhibitors: sildenafil
- Diuretics if right heart failure
- Heart-lung transplant

Whilst only 10-15% of patients appear to have a pulmonary vascular tree responsive to calcium antagonism, these agents still constitute the initial therapy of choice according to guidelines, but only in those patients who show a response to vasodilator testing.

Complication

- Cor pulmonale

Angina pectoris

Non-atherosclerotic angina would be associated with conditions such as

- Thyrotoxicosis
- **Aortic regurgitation**
- Aortic stenosis
- Hypertrophic cardiomyopathy
- Anaemia

Anginal pain is:

1. constricting discomfort in the front of the chest, or in the neck, shoulders, jaw, or arms
 2. precipitated by physical exertion
 3. relieved by rest or GTN within about 5 minutes.
- Three of the features above are defined as **typical angina**.
 - Two of the three features above are defined as **atypical angina**.
 - One or none of the features above are defined as **non-anginal chest pain**.

Features which make a diagnosis of stable angina unlikely are when the chest pain is:

- continuous or very prolonged **and/or**
- unrelated to activity **and/or**
- brought on by breathing in **and/or**
- associated with symptoms such as dizziness, palpitations, tingling or difficulty swallowing.
Consider causes of chest pain other than angina (such as gastrointestinal or musculoskeletal pain).

Investigations for stable chest pain

- **First-line:** 64-slice CT coronary angiography (CTCA)
- **Second-line:** non-invasive functional testing (if CTCA is non-diagnostic.)
 - ⇒ Myocardial perfusion scan (MPS) with single photon emission computed tomography (SPECT) (MPS with SPECT) or
 - ⇒ stress echocardiography or
 - ⇒ first-pass contrast-enhanced magnetic resonance perfusion or
 - ⇒ MRI for stress-induced wall motion abnormalities.
- **Third-line:** invasive coronary angiography (when the results of non-invasive functional imaging are inconclusive)

In the context of risk factors for ischaemic heart disease (hypertension, hypercholesterolaemia, smoking), the clinical diagnosis should be confirmed with non-invasive functional scanning such as myocardial perfusion scanning with SPECT.

- **High-risk patients** with **classic angina symptoms** should proceed directly to coronary angiography.
- Offer 64- slice (or above) CT coronary angiography if:
 1. clinical assessment indicates typical or atypical angina or
 2. clinical assessment indicates non-anginal chest pain but 12- lead resting ECG has been done and indicates ST- T changes or Q waves.
- **Low-risk patients** can be evaluated with **non-invasive stress imaging**.
- Offer non-invasive functional imaging for myocardial ischaemia if 64-slice (or above) CT coronary angiography has shown CAD of uncertain functional significance or is non-diagnostic.
 - ⇒ **non-invasive functional testing for myocardial ischaemia**
 1. **myocardial perfusion scintigraphy** with single photon emission computed tomography (MPS with SPECT)
 2. **stress echocardiography**
 3. first-pass contrast-enhanced magnetic resonance (MR) perfusion
 4. use adenosine or dipyridamole as stress agents
 - ⇒ Take account of locally available technology and any contraindications (for example, disabilities, frailty, limited ability to exercise) when deciding on the imaging method.
- Offer invasive coronary angiography as a third-line investigation when the results of non-invasive functional imaging are inconclusive.
- Treadmill exercise is no longer recommended in the work-up of new-onset chest pain.

Definition of significant coronary artery disease (CAD)

- CT coronary angiography is:
 - ⇒ $\geq 70\%$ diameter stenosis of at least one major epicardial artery segment **or**
 - ⇒ $\geq 50\%$ diameter stenosis in the left main coronary artery

Factors intensifying ischaemia

- Such factors allow less severe lesions (for example, $\geq 50\%$) to produce angina:
 - ⇒ reduced oxygen delivery: anaemia, coronary spasm
 - ⇒ increased oxygen demand: tachycardia, left ventricular hypertrophy
 - ⇒ large mass of ischaemic myocardium: proximally located lesions
 - ⇒ longer lesion length.

Factors reducing ischaemia which may render severe lesions ($\geq 70\%$) asymptomatic:

- Well-developed collateral supply.
- Small mass of ischaemic myocardium: distally located lesions, old infarction in the territory of coronary supply.

ESC guidelines 2017

- A reduction in chest pain after nitroglycerin (glyceryl trinitrate) administration can be misleading and is not recommended as a diagnostic manoeuvre
- In cases of symptom relief after nitroglycerin administration, another 12-lead ECG must be obtained.
- **A complete normalization of the ST-segment elevation after nitroglycerin administration, along with complete relief of symptoms, is suggestive of coronary spasm, with or without associated MI. In these cases, an early coronary angiography (within 24 h) is recommended.**
- In cases of recurrent episodes of ST-segment elevation or chest pain, **immediate angiography** is required.

Drug management

A beta-blocker or a calcium channel blocker is used first-line to prevent angina attacks

You should still use **bisoprolol** in patients with **COPD** and **IHD**, because it carries an important outcome benefit

Prinzmetal angina - treatment = dihydropyridine calcium channel blocker

Medication

- all patients should receive aspirin and a statin in the absence of any contraindication
- sublingual glyceryl trinitrate to abort angina attacks
- NICE recommend using either a beta-blocker or a calcium channel blocker first-line based on 'comorbidities, contraindications and the person's preference'
- if a calcium channel blocker is used as monotherapy a rate-limiting one such as verapamil or diltiazem should be used. If used in combination with a beta-blocker then use a long-acting dihydropyridine calcium-channel blocker (e.g. modified-release nifedipine). Remember that beta-blockers should not be prescribed concurrently with verapamil (risk of complete heart block)
- if there is a poor response to initial treatment then medication should be increased to the maximum tolerated dose (e.g. for atenolol 100mg od)
- if a patient is still symptomatic after monotherapy with a beta-blocker add a calcium channel blocker and vice versa
- if a patient is on monotherapy and **cannot tolerate the addition of a calcium channel blocker or a beta-blocker** then consider one of the following drugs:
 - ⇒ a long-acting nitrate,
 - ⇒ **ivabradine**,
 - ⇒ nicorandil or
 - ⇒ ranolazine

- if a patient is taking both a beta-blocker and a calcium-channel blocker then **only add a third drug whilst a patient is awaiting assessment for PCI or CABG**
- The FREEDOM trial demonstrated that **in diabetic patients CABG was superior to PCI** in that it significantly reduced rates of death and myocardial infarction.
- Cardioselective calcium antagonists such as verapamil and diltiazem do not affect prognosis in angina although they may impact on symptoms by reducing heart rate.
- If a patients don't tolerate beta-blockade, ivabradine may be a more appropriate intervention.

Nitrate tolerance

- many patients who take nitrates develop tolerance and experience reduced efficacy
- the BNF advises that patients who develop tolerance should take the second dose of isosorbide mononitrate after 8 hours, rather than after 12 hours. This allows blood-nitrate levels to fall for 4 hours and maintains effectiveness
- this effect is not seen in patients who take modified release isosorbide mononitrate
- the explanation for nitrate tolerance → generation of reactive oxygen species**
 - chronic nitrate therapy → ↑vascular oxidative stress → ↑ degradation of nitric oxide (NO) → reduced bioavailability

Ivabradine

- action
 - ⇒ **(I_f ('funny' ion) channel inhibitor** which is highly expressed in the sinoatrial node)
→ **reducing the heart rate**
- Indications
 - ⇒ a new class of anti-anginal drug
 - there is no evidence currently of superiority over existing treatments of stable angina
 - ⇒ **heart failure:**
 - with (NYHA) class II–IV stable chronic heart failure with systolic dysfunction and who are in sinus rhythm with a heart rate of 75 bpm or more and who are given ivabradine in combination with standard therapy including β-blocker therapy, angiotensin-converting enzyme (ACE) inhibitors and aldosterone antagonists, or when β-blocker therapy is contraindicated or not tolerated and with a left ventricular ejection fraction of 35% or less.
- adverse effects:
 - ⇒ visual effects, particular **luminous phenomena**, are common.
 - sensations of enhanced brightness in a fully maintained visual field
 - due to blockage of I_f ion channels in the retina, which are very similar to cardiac I_f.
 - mild, transient, and fully reversible.
 - ⇒ Bradycardia, due to the mechanism of action,

Ulceration of an atheromatous plaque of the abdominal aorta is the most common source of emboli in old man presented with acute pain, pallor and absent pulses in his leg.

MRCPUK-part-1-January 2018 exam: Which cell type is most implicated in the development of coronary artery plaques?

⇒ **Macrophages**

Coronary artery bypass graft (CABG)

- There are two main approaches.
 1. In one, the left internal thoracic artery (internal mammary artery) is diverted to the left anterior descending branch of the left coronary artery.
 2. In the other, a great saphenous vein is removed from a leg; one end is attached to the aorta or one of its major branches, and the other end is attached to the obstructed artery.
- CABG is superior to PCI in multivessel coronary disease.
- indicated when coronary arteries have a 50% to 99% obstruction.
- CABG guidelines state CABG is the preferred treatment for:
 - ⇒ Disease of the left main coronary artery (LMCA).
 - ⇒ Disease of all three coronary arteries (LAD, LCX and RCA).
 - ⇒ Diffuse disease not amenable to treatment with a PCI.
 - ⇒ high-risk patients such as those with severe ventricular dysfunction (i.e. low ejection fraction), or diabetes mellitus.
- **Benefits**
 - ⇒ relief of angina
 - ⇒ no survival benefit with bypass surgery vs. medical therapy in stable angina
 - ⇒ Bypass surgery does not prevent future myocardial infarctions.
- **Complications**
 - ⇒ **The incidence of acute coronary syndrome within 30 days of CABG is high, at around 17.5%.**
 - ⇒ Aneurysms are a rare and late complication of CABG.

Cardiac syndrome X

- consist of:
 - ⇒ angina-like chest pain during exertion
 - ⇒ characteristic ECG changes during exercise testing
 - ⇒ normal coronary arteries on cardiac catheterisation
 - ⇒ no inducible coronary artery spasm during catheterisation

Acute coronary syndrome

Poor prognostic factors

- age
- development (or history) of heart failure
- peripheral vascular disease
- reduced systolic blood pressure
- Killip class*
- initial serum creatinine concentration
- elevated initial cardiac markers
- cardiac arrest on admission
- ST segment deviation

Clinical factors which are good indicators of ACS:

- **typical pain lasting at least 15 minutes, associated nausea, and sweating.**
- Response to GTN **should not** be used as indicator of ACS

ACS referral**Chest pain Referral guidelines:**

- current chest pain or chest pain in the last 12 hours with an abnormal ECG: emergency admission
- chest pain 12-72 hours ago: refer to hospital the same-day for assessment
- chest pain > 72 hours ago: perform full assessment with ECG and troponin measurement before deciding upon further action

Myocardial infarction

DVLA advice post MI - cannot drive for 4 weeks

Inferior MI - right coronary artery lesion

- The most specific feature, which suggests that the pain is myocardial ischaemia, is the radiation to the jaw, which is relatively specific for pain of myocardial ischaemia.
- The clinical classification of MI includes: (NICE 2010)
 - ⇒ Type 1: ischaemia due to a primary coronary event such as plaque, fissuring or dissection.
 - ⇒ Type 2: ischaemia due to either increased oxygen demand or decreased supply, such as coronary spasm, coronary embolism, anaemia, arrhythmias, hypertension, or hypotension.

The diurnal variation of myocardial ischaemia

- There is a diurnal variation in presentation of myocardial ischaemia.
- Which physiological process is responsible for this? → Vasospasm
- The peak incidence of STEMI and the peak incidence of death due to ischaemic heart disease both coincide at around 8-9 am.
 - ⇒ The early morning is associated with several physiological and haematological factors which predispose to vasospasm, infarction and death.
 - ⇒ There is
 - ↑ adrenergic activity
 - ↑ plasma fibrinogen levels
 - ↑ inhibition of fibrinolysis and
 - ↑ platelet adhesiveness.
- Interestingly, NSTEMIs are not associated with this degree of diurnal rhythm.
- Precipitating factors for an infarct include:
 - ⇒ physical exertion
 - ⇒ Rest , Sleep
 - ⇒ Surgical procedure
 - ⇒ Emotional stressors.

Risk factors

The **worst** risk factor for CAD is diabetes mellitus, but the most **common** risk is hypertension.

The highest prevalence of myocardial infarction is 72 hours post operation. Patients with diabetes may not have chest pain due to autonomic dysfunction.

Investigations

- ECG (**best initial test**)
- Cardiac troponin levels: Measure as soon as possible and repeat after 1–6 hours.
- Transthoracic echocardiography: if the diagnosis is unclear. Findings:
 - ⇒ Wall motion abnormalities
 - ⇒ Decreased LV function

Typical Electrocardiographic Evolution of a STEMI

EKG Abnormality	Onset	Disappearance
Hyperacute T waves (tall, peaked T waves in leads facing infarction)	Immediately	6–24 hours
ST-segment elevation	Immediately	1–6 weeks
Q waves longer than 0.04 seconds	One to several days	Years to never
T wave inversion	6–24 hours	Months to years

Myocardial infarction: management

Primary percutaneous coronary intervention is the gold-standard treatment for ST-elevation myocardial infarction

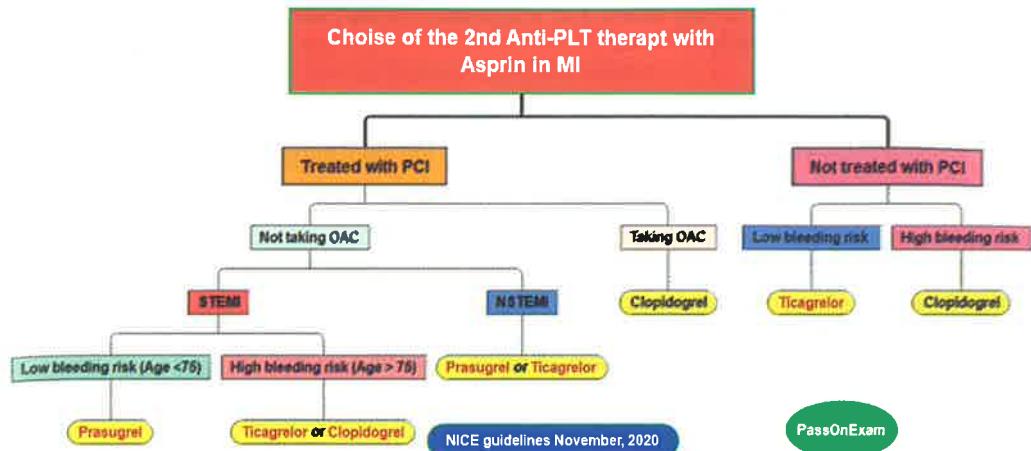
PCI: stent thrombosis - withdrawal of antiplatelets biggest risk factor

Clopidogrel inhibits ADP binding to platelet receptors

Ticagrelor has a similar mechanism of action to clopidogrel - inhibits ADP binding to platelet receptors

PCI - patients with drug-eluting stents require a longer duration of clopidogrel therapy

- Glyceryl trinitrate
 - ⇒ Sublingual glyceryl trinitrate and intravenous morphine + metoclopramide should be given to help relieve the symptoms.
 - ⇒ **ongoing pain despite the use of sublingual GTN is suggestive of continuing myocardial ischaemia/infarction → IV GTN**
- Aspirin 300mg.
 - ⇒ **the initial drug therapy**
 - ⇒ Aspirin 300mg should be given to all patients (unless contraindicated).
 - ⇒ **It is safe in the post-surgical patient with no signs of bleeding** at three days post operation.
 - ⇒ A second antiplatelet is normally given, usually ticagrelor, clopidogrel or prasugrel (all are antagonists of the P2Y₁₂ adenosine diphosphate receptor).
 - **(Aspirin + ticagrelor) is better than (aspirin + clopidogrel)**
 - ❖ ticagrelor was associated with a 13% relative reduction in cardiovascular events versus a conventional clopidogrel based regimen.
 - ❖ This has driven use of ticagrelor in place of clopidogrel in major guidelines on anti-platelet therapy post STEMI.
 - ❖ A loading dose of 180 mg stat is recommended at the time of diagnosis of STEMI.
 - ❖ This was also associated with increased risk of bleeding events when compared to aspirin and clopidogrel.
 - ⇒ NICE do not recommend giving other antiplatelet agents (i.e. Clopidogrel) outside of hospital. The dose of clopidogrel is 300 mg in ACS.
- Other treatments that may be given include bivalirudin (a direct thrombin inhibitor, usually given alongside aspirin + clopidogrel) and a form of heparin (either low-molecular weight or unfractionated).
 - ⇒ Heparin in Non-STEMI (has no benefit in ST elevation MI).
- do not routinely give oxygen, only give if sats < 94%*
- *NICE suggest the following in terms of **oxygen therapy**:
 - ⇒ do not routinely administer oxygen, but monitor oxygen saturation using pulse oximetry as soon as possible, ideally before hospital admission. Only offer supplemental oxygen to:
 - people with oxygen saturation (SpO₂) of less than 94% who are not at risk of hypercapnic respiratory failure, aiming for SpO₂ of 94-98%
 - people with chronic obstructive pulmonary disease who are at risk of hypercapnic respiratory failure, to achieve a target SpO₂ of 88-92% until blood gas analysis is available.
 - ⇒ ESC guidelines 2017 state that: routine oxygen is not recommended when **SaO₂** is ≥ 90%.
 - perform an ECG as soon as possible but do not delay transfer to hospital. A normal ECG does not exclude ACS
 - percutaneous coronary intervention (PCI)
 - ⇒ is the first-line and **the gold-standard treatment** management to revascularise the myocardium.
 - ⇒ but is not available in all centres. Thrombolysis should be performed in patients without access to primary PCI
 - ⇒ offer primary PCI to patients who present within 12 hours of onset of symptoms, if it can be delivered within 120 minutes of the time when fibrinolysis could have been given.
 - ⇒ A practical example may be a patient who presents with a STEMI to a small district general hospital (DGH) which does not have facilities for PCI. If they cannot be transferred to a larger hospital for PCI within 120 minutes then fibrinolysis should be given. If the patient's ECG taken 90 minutes after fibrinolysis failed to show resolution of the ST elevation then they would then require transfer for PCI.



Percutaneous coronary intervention (PCI)

- PCI is a technique used to restore myocardial perfusion in patients with ischaemic heart disease, both in patients with stable angina and acute coronary syndromes.
- Stents are implanted in around 95% of patients - it is now rare for just balloon angioplasty to be performed
- Following stent insertion migration and proliferation of smooth muscle cells and fibroblasts occur to the treated segment. The stent struts eventually become covered by endothelium. Until this happens there is an increased risk of platelet aggregation leading to thrombosis.
- Following insertion, the most important factor in preventing stent thrombosis is antiplatelet therapy. Aspirin should be continued indefinitely. The length of clopidogrel treatment depends on the type of stent, reason for insertion and consultant preference
- **How long should he continue dual antiplatelet therapy following stent insertion?**
 - **12 months**
 - When dual therapy is maintained for less than 12 months, early cessation of clopidogrel is associated with an increased risk of further ischaemic events.
 - Thrombosis of a drug-eluting stent is associated with high morbidity (42%) and mortality (71%). For this reason, dual antiplatelet therapy (usually aspirin and clopidogrel) is continued for at least twelve months following the insertion of this type of stent.
- **Elective surgery should be postponed for twelve months when it is considered safe to stop clopidogrel and continue with aspirin.**

Complications: Two main complications may occur

1. **Stent thrombosis:**
 - ⇒ due to platelet aggregation as above.
 - ⇒ Occurs in 1-2% of patients, most commonly in the first month.
 - ⇒ Usually presents with acute myocardial infarction
 - ⇒ Treated by **primary angioplasty**.
2. **Restenosis:**
 - ⇒ due to excessive tissue proliferation around stent.
 - ⇒ Occurs in around 5-20% of patients, most commonly in the first 3-6 months.
 - ⇒ Usually presents with the recurrence of angina symptoms.
 - ⇒ Risk factors include diabetes, renal impairment and stents in venous bypass grafts
 - **In patients with type-2 diabetes, uncoated coronary stents are liable to re-stenosis at a rate of 40-50% by the end of a 6-month**

- ⇒ Drug eluting stents have been shown to reduce the relative risk of re-stenosis by around 80%, but only where dual anti-platelet therapy with clopidogrel and aspirin is continued for at least 1 year.

Types of stent

- bare-metal stent (BMS)
- drug-eluting stents (DES): stent coated with paclitaxel or rapamycin which inhibit local tissue growth. Whilst this reduces restenosis rates the stent thrombosis rates are increased as the process of stent endothelialisation is slowed

Thrombolysis

Thrombolysis is no longer indicated except in the context of **STEMI** where **PCI** is not available within 90 minutes of first medical contact.

- **ECG criteria for thrombolysis** within 24 hours of typical pain include:
 - ⇒ ST elevation of more than 1 mm in two adjacent limb leads.
 - ⇒ ST elevation more than 2 mm in two adjacent anterior chest leads.
 - ⇒ new left bundle branch block.
- **Pre-hospital thrombolysis** is indicated if the time from the initial call to arrival at hospital is likely to be over 30 minutes.
 - ⇒ When primary percutaneous coronary intervention cannot be provided within 120 minutes of ECG diagnosis, patients with **STEMI** should receive immediate (pre-hospital or admission) thrombolytic therapy
 - ⇒ (NICE) recommends using **intravenous bolus** (**reteplase** or **tenecteplase**) rather than an infusion for pre-hospital thrombolysis
- **Thrombolitics**
 - ⇒ **tissue plasminogen activator (tPA)** has been shown to offer clear mortality benefits over streptokinase
 - ⇒ **streptokinase**
 - mechanism of action → Combining with plasminogen to form a complex
 - Streptokinase forms a 1:1 complex with plasminogen that induces structural changes in the protein that activates it without direct cleavage of the Arg-Val bond. It is not specific for fibrin-bound plasminogen.
 - ⇒ **alteplase**
 - Unlike streptokinase, alteplase activates plasminogen bound to fibrin without activating unbound plasminogen proteins.
 - It is not associated with hypotension or allergic reactions like streptokinase.
 - It has a much shorter half-life of only 3-4 minutes compared to 18 minutes for streptokinase.
 - ⇒ **tenecteplase**
 - easier to administer
 - has been shown to have non-inferior efficacy to alteplase with a similar adverse effect profile
- ECG should be performed 90 minutes following thrombolysis to assess whether there has been a greater than 50% resolution in the ST elevation
 - ⇒ if there has not been adequate resolution then rescue **PCI** is superior to repeat thrombolysis
 - ⇒ for patients successfully treated with thrombolysis PCI has been shown to be beneficial. The optimal timing of this is still under investigation
- **Contraindications to thrombolysis** include:
 - ⇒ Gastrointestinal (GI) bleeding in the preceding three weeks.
 - ⇒ Heavy vaginal bleeding
 - ⇒ Ischaemic stroke in last six months
 - ⇒ **Previous history of hemorrhagic stroke**
 - ⇒ Uncontrolled severe hypertension

- ⇒ Prolonged cardiopulmonary resuscitation (CPR) (more than half an hour).
- ⇒ Known or suspected aortic dissection
- ⇒ Known bleeding disorder
- ⇒ Major surgery or serious trauma within two weeks.
- ⇒ Lumbar puncture in the preceding week.

- **Relative contraindications**

- ⇒ Proliferative diabetic retinopathy,
- ⇒ allergy and
- ⇒ oral anticoagulants

- **Risk factors for bleeding**

- ⇒ Advancing age
- ⇒ Renal impairment
- ⇒ **Low body weight** and
- ⇒ Known bleeding problems.

Management of hyperglycaemia in acute coronary syndromes

- **the most appropriate treatment for his glycaemic control → Commence intravenous insulin infusion and stop metformin**
 - ⇒ metformin → increased risk of lactic acidosis.
- Nice in 2011 recommends using a dose-adjusted **insulin infusion** with regular monitoring of blood glucose levels to glucose below 11.0 mmol/l
- The Diabetes and Insulin-Glucose Infusion in Acute Myocardial Infarction (DIGAMI) study demonstrated **significant reductions in mortality** in subjects with diabetes and myocardial infarction (MI) treated with IV insulin infusion (followed by three months of sc insulin) compared with conventional therapy with their oral hypoglycaemic agents.
 - ⇒ intensive insulin therapy (an intravenous infusion of insulin and glucose with or without potassium, sometimes referred to as 'DIGAMI') regimes are not recommended routinely

MRCPUK-part-1-january-2018: What is the mode of action of alteplase?

⇒ **Plasminogen activator**

- Induce conversion of plasminogen to plasmin leading to the dissolution of a fibrin clot.

Myocardial infarction: complications

Complete heart block following a MI? - right coronary artery lesion

Complete heart block following an inferior MI is NOT an indication for pacing, unlike with an anterior MI

Myocardial infarction complications

Complication	Notes
Cardiac arrhythmia	Occurs within the first few days after MI. Important cause of death before reaching the hospital and within the first 24 hours post-MI. Ventricular fibrillation is the most common cause of death following a MI. Atrioventricular block is more common following inferior myocardial infarctions.
LV failure and pulmonary oedema	Can occur 2° to LV infarction, VSD, free wall rupture, papillary muscle rupture with mitral regurgitation.
Post infarction fibrinous pericarditis	1–3 days: common around 10% of patients. friction rub
Papillary muscle rupture (leads to acute mitral regurgitation).	2–7 days: posteromedial papillary muscle rupture. ↑ risk due to single blood supply from posterior descending artery. More common with infero-posterior infarction. Suddenly develops pulmonary oedema and a loud systolic murmur at the apex which radiated into the axilla with associated pulmonary oedema. often require emergency surgical repair.
Interventricular septal rupture	3–5 days: macrophage-mediated degradation →VSD →↑ O ₂ saturation and pressure in RV. acute heart failure associated with a pan-systolic murmur. An echocardiogram is diagnostic and will exclude acute mitral regurgitation which presents in a similar fashion. Urgent surgical correction is needed.
Ventricular pseudoaneurysm formation	3–14 days: free wall rupture contained by adherent pericardium or scar tissue; low cardiac output, risk of arrhythmia, embolus from mural thrombus.
True ventricular aneurysm	2 weeks to several months: outward bulge with contraction ("dyskinesia"), associated with fibrosis. typically associated with persistent ST elevation and left ventricular failure. Thrombus may form within the aneurysm increasing the risk of stroke. Patients are therefore anticoagulated .
Ventricular free wall rupture	5–14 days: present with acute heart failure secondary to cardiac tamponade (raised JVP, pulsus paradoxus, diminished heart sounds). LV hypertrophy and previous MI protect against free wall rupture. Urgent pericardiocentesis and thoracotomy are required.
Dressler syndrome	Several weeks: autoimmune phenomenon resulting in fibrinous pericarditis. characterised by a combination of fever, pleuritic pain, pericardial effusion, friction rub on auscultation and a raised ESR. Treated with NSAIDs.
Chronic heart failure	The most important factor predicting outcomes post-STEMI is the presence of new systolic heart failure.

Primary prevention

drugs which have evidence for the reduction of risk of developing a cardiac event?

- Angiotensin converting enzyme inhibitor
 - ⇒ **The most appropriate treatment to reduce cardiovascular risk should focus on adequate blood pressure control**
 - ⇒ ↓ BP is most important than control of DM and lipids in CV risk reduction
- Aspirin
- Metformin
 - ⇒ treatment of overweight, diabetic patients with metformin, lowers the relative risk of (MI) by 40%, as opposed to treatment with sulphonylureas or insulin.
- Statins

Myocardial infarction: secondary prevention

Patients with established CVD should take atorvastatin 80mg on

Flash pulmonary oedema, U&Es worse on ACE inhibitor,
asymmetrical kidneys → renal artery stenosis - do MR angiography

All patients should be offered the following drugs:

- dual antiplatelet therapy (aspirin plus a second antiplatelet agent)
- ACE inhibitor
- beta-blocker
- statin

Some selected lifestyle points:

- Diet:
 - ⇒ advise a Mediterranean style diet, switch butter and cheese for plant oil-based products.
 - ⇒ Do not recommend omega-3 supplements or eating oily fish
- Exercise:
 - ⇒ advise 20-30 mins a day until patients are 'slightly breathless'
- Sexual activity
 - ⇒ may resume 4 weeks after an uncomplicated MI.
 - ⇒ Reassure patients that sex does not increase their likelihood of a further MI.
 - ⇒ PDE5 inhibitors (e.g. sildenafil) may be used 6 months after a MI.
 - They should however be avoided in patient prescribed either nitrates or nicorandil

Clopidogrel

- STEMI:
 - ⇒ the European Society of Cardiology recommend dual antiplatelets for **12 months**. In the UK this means aspirin + clopidogrel
- Non-ST segment elevation myocardial infarction (NSTEMI):
 - ⇒ following the NICE 2013 guidelines, clopidogrel should be given for the first **12 months**

Aldosterone antagonists

- patients who have had an acute MI and who have symptoms and/or signs of heart failure and left ventricular systolic dysfunction, treatment with an aldosterone antagonist licensed for post-MI treatment (e.g. eplerenone) should be initiated within 3-14 days of the MI, preferably after ACE inhibitor therapy

Hyperlipidaemia: management

See endocrinology

Heart failure

Definition

- structural or functional impairment of ventricular filling and/or ejection of blood.

Types

- **Heart failure with reduced ejection fraction (HFrEF)**
 - ⇒ Reduced contractility → **systolic** ventricular **dysfunction** → decreased left ventricular ejection fraction (LVEF) → decreased cardiac output
 - ⇒ Causes include:
 - Damage and loss of myocytes (e.g., following myocardial infarction, coronary artery disease, dilated cardiomyopathy)
 - Cardiac arrhythmias
 - High-output cardiac failure
 - ❖ A state of heart failure characterized by increased cardiac output and lowered systemic vascular resistance. May be caused by arteriovenous fistulas, renal disease, anemia, beriberi, or Graves' disease.
- **Heart failure with preserved ejection fraction (HFpEF)**
 - ⇒ Decreased ventricular compliance → **diastolic** ventricular **dysfunction** → reduced ventricular filling and increased diastolic pressure → decreased cardiac output (while the left ventricular ejection fraction remains normal)
 - ⇒ Causes include:
 - Increased stiffness of the ventricle (e.g., long-standing arterial hypertension with ventricular wall hypertrophy, restrictive cardiomyopathy)
 - Impaired relaxation of the ventricle (e.g., constrictive pericarditis, pericardial tamponade)

NYHA classification

- The **New York Heart Association (NYHA)** classification is widely used to classify the severity of heart failure:
 - ⇒ **NYHA Class I**
 - no symptoms
 - no limitation: ordinary physical exercise does not cause undue fatigue, dyspnoea or palpitations
 - ⇒ **NYHA Class II**
 - mild symptoms
 - slight limitation of physical activity: comfortable at rest but **ordinary activity results in fatigue, palpitations or dyspnoea**
 - ⇒ **NYHA Class III**
 - moderate symptoms
 - marked limitation of physical activity: comfortable at rest but **less than ordinary activity results in symptoms**
 - ⇒ **NYHA Class IV**
 - severe symptoms
 - unable to carry out any physical activity without discomfort: **symptoms of heart failure are present even at rest with increased discomfort with any physical activity**

Diagnosis (NICE 2010)

- Patient with previous myocardial infarction
 - ⇒ arrange **echocardiogram** within 2 weeks
 - if transthoracic Doppler 2D echocardiography imaging is poor (eg: in obese)
 → consider other imaging methods, such as:
 - ❖ **radionuclide angiography**,
 - ❖ cardiac magnetic resonance imaging or
 - ❖ trans-oesophageal Doppler 2D echocardiography.
- No previous myocardial infarction
 - ⇒ measure serum natriuretic peptides (BNP)
 - if levels are '**high**' (> 400) arrange echocardiogram within 2 weeks
 - if levels are '**raised**' (100-400) arrange echocardiogram within 6 weeks → 40% of patients with raised BNP will have left ventricular systolic dysfunction on echo. the remaining will have other cardiac abnormalities.
 - if levels are '**normal**' (< 100) heart failure is unlikely (investigate for other causes)

B-type natriuretic peptide (BNP)

- Source
 - ⇒ produced mainly by the left ventricular myocardium in response to strain.
- Effect
 - ⇒ The net effect of these peptides is:
 - ↓BP (due to the decrease in systemic vascular resistance) and, thus, afterload on the heart.
 - ↓cardiac output (due to an overall decrease in central venous pressure) and preload as a result of the reduction in blood volume that follows natriuresis and diuresis.
- Uses
 - ⇒ normal level rules out acute heart failure in the emergency setting .
 - ⇒ Very high levels are associated with a poor prognosis.
- Excretion
 - ⇒ Less than 5% of BNP is cleared renally whereas NT-proBNP is reliant solely on the kidney for excretion and hence it is unreliable in patients with coexistent renal dysfunction.

	BNP	NTproBNP
High levels	> 400 pg/ml (116 pmol/litre)	> 2000 pg/ml (236 pmol/litre)
Raised levels	100-400 pg/ml (29-116 pmol/litre)	400-2000 pg/ml (47-236 pmol/litre)
Normal levels	< 100 pg/ml (29 pmol/litre)	< 400 pg/ml (47 pmol/litre)

Diagnosis of acute heart failure (Nice guidelines 2014):

- In people presenting with new suspected acute heart failure:
 - ⇒ rule out the diagnosis of heart failure if :
 - BNP less than 100 ng/litre
 - NT-proBNP less than 300 ng/litre.
 - ⇒ new suspected acute heart failure with raised natriuretic peptide levels → perform transthoracic Doppler 2D echocardiography (within 48 hours of admission)

Factors, which alter the BNP level:

Increase BNP levels	Decrease BNP levels
<ul style="list-style-type: none"> Left ventricular hypertrophy Aortic stenosis, Hypertension Ischaemia Tachycardia Right ventricular overload Hypoxaemia (including pulmonary embolism) GFR < 60 ml/min Sepsis COPD, Cor pulmonale Diabetes Age > 70 Liver cirrhosis Hyperaldosteronism Cushing's syndrome Stable angina, Acute coronary syndromes Atrial fibrillation (AF) 	<ul style="list-style-type: none"> Obesity Diuretics ACE inhibitors Beta-blockers Angiotensin 2 receptor blockers Aldosterone antagonists

Mechanism of central sleep apnea (CSA) in HF:

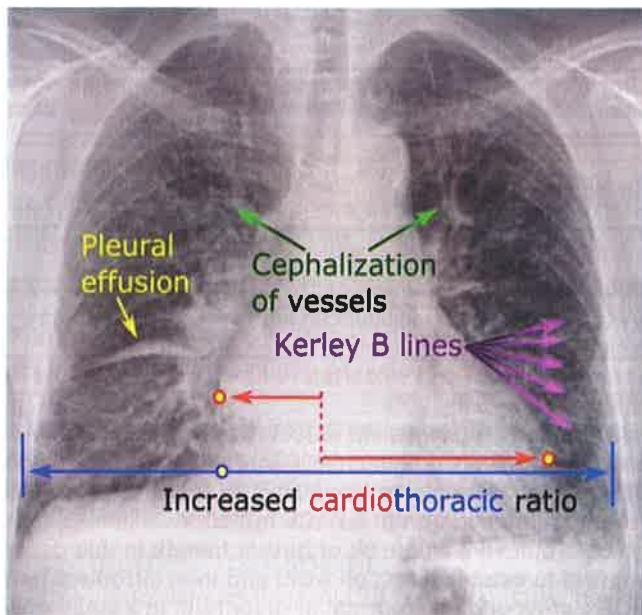
- Which mechanism is responsible for the patient's polysomnography findings in heart failure?
 - ⇒ Increased sensitivity to carbon dioxide and stimulation of the vagal receptors.
 - ⇒ increased sensitivity to PaCO_2 is a protective mechanism from hypercapnia due to heart failure.
 - HF → ↑duration of circulation of blood gases from the lungs to the brain.
 - When these blood gases reach the brain, the increased sensitivity to PaCO_2 → higher-than-normal response of hyperventilation → ↓ PaCO_2 lower than the apneic threshold.
 - As soon as the brain detects low PaCO_2 it will cease ventilation with apnea (central) so PaCO_2 can rise again.
 - As soon as the PaCO_2 rises again and reaches the brain (longer than normal due to heart failure), it will cause another episode of hyperventilation.
 - ⇒ supine position → ↑ venous return → pulmonary congestion → activate vagal receptors → hyperventilation.

Hyponatraemia in patients with CHF

- Water restriction is the first-line and mainstay of therapy
- Stopping furosemide will not be possible for a patient who has decompensated heart failure.
- Similarly, administration of hypertonic saline is only indicated if there is neurological manifestation of hyponatremia.
- Moreover hypertonic or isotonic saline administration will be poorly tolerated in a volume-overloaded patient.
- associated with the worst prognosis

Investigations

- Chest x-ray: Features of pulmonary oedema on a chest x-ray may include:
 - ⇒ interstitial oedema
 - ⇒ bat's wing appearance
 - ⇒ upper lobe diversion (increased blood flow to the superior parts of the lung)
 - ⇒ Kerley B lines
 - ⇒ pleural effusion
 - ⇒ cardiomegaly may be seen if there is cardiogenic cause



Typical CXR signs associated with heart failure

The most common cause of flash pulmonary oedema is myocardial ischaemia.
Bilateral renal artery stenosis is a less common cause of flash pulmonary oedema.

Pharmacological management

Acute heart failure management

- Initial pharmacological treatment
 - ⇒ intravenous diuretics
- Initial non-pharmacological treatment
 - ⇒ cardiogenic pulmonary oedema with severe dyspnoea and acidaemia consider starting non-invasive ventilation
 - in this case it is the most useful next step , before diuretics . The effect of the diuresis comes much later and has a modest overall contribution in managing the symptoms of shortness of breath.
 - ⇒ Consider invasive ventilation in acute heart failure that, despite treatment, is leading to or is complicated by: respiratory failure or reduced consciousness or physical exhaustion.

In a person presenting with acute heart failure who is already taking beta-blockers:

- continue the beta-blocker treatment unless they have a heart rate less than 50 beats per minute, second or third degree atrioventricular block, or shock.
- restart beta-blockers once their condition has been stabilised – for example, when intravenous diuretics are no longer needed.

In a person presenting with acute heart failure who is already taking frusemide 80 mg:

- in a patient with evidence of decompensated heart failure and fluid overload. The most appropriate initial management is to **Increase furosemide** and relieve the symptoms of fluid overload – pulmonary and peripheral oedema.

Chronic management

- **4 drugs have been shown to improve mortality in patients with chronic heart failure:**
 1. ACE inhibitors
 2. spironolactone
 3. beta-blockers
 4. hydralazine with nitrates
- No long-term reduction in mortality has been demonstrated for loop diuretics such as furosemide.
- In patients with symptoms of heart failure not controlled on ACE inhibitors alone, switching to the combination of ARB and neprilysin inhibitor can further improve symptoms and quality of life.
 - ⇒ e.g: **combination of sacubitril and valsartan reduced cardiovascular death and heart failure hospitalisations by 20%.**
- NICE issued updated guidelines on management in 2010, **key points** include:
 - ⇒ first-line treatment for all patients is both an ACE-inhibitor and a beta-blocker
 - With the persisting symptoms despite 80 mg of furosemide, guidelines would **initially suggest the addition of an ACE inhibitor.**
 - **Although beta-blockers would be of further benefit in this patient, it is important first to establish him on ACEi and then introduce beta-blockers like carvedilol, metoprolol or bisoprolol in a small dose and gradually increase.**
 - ⇒ second-line treatment is now either an aldosterone antagonist, angiotensin II receptor blocker or a hydralazine in combination with a nitrate
 - ⇒ if symptoms persist cardiac resynchronisation therapy or digoxin should be considered
 - digoxin has also not been proven to reduce mortality in patients with heart failure.
 - It may however improve symptoms due to its inotropic properties.
 - Digoxin is strongly indicated if there is coexistent atrial fibrillation
 - There is no evidence that increasing a dose of digoxin above 62.5 µg in a patient in sinus rhythm would have any added benefit.
 - ⇒ diuretics should be given for fluid overload
 - ⇒ offer annual influenza vaccine
 - ⇒ offer one-off pneumococcal vaccine
 - adults usually require just one dose but those with asplenia, splenic dysfunction or chronic kidney disease need a booster every 5 years

Drugs that improve prognosis are beta blockers, ACE inhibitors, ARNIs, aldosterone antagonists, and hydralazine with nitrate.

MRCPUK-part-1-jan-2018: In a patient with significant **heart failure on maximum medical therapy** (ramipril 10 mg OD, furosemide 80 mg OD, bisoprolol 10 mg OD and spironolactone 25 mg OD). Despite this, they have continued to deteriorate but **criteria for cardiac resynchronisation therapy (CRT) are not achieved. What is the most appropriate next step to improve mortality?**

⌚ **Ivabradine**

- acts as an inhibitor of the I_f current within the myocardium. This current, particularly present in the sino-atrial and atrio-ventricular nodes, acts as the cardiac pacemaker.
- By inhibiting this current, ivabradine reduces the heart rate without impacting the force of cardiac contraction.
- This has been shown to reduce heart failure hospitalisation and mortality in patients already on maximum medical therapy.
- Due to its mechanism, ivabradine is **only effective in patients in sinus rhythm.**

What is the management of a patient with severe CHF who develops gynecomastia? Switch spironolactone to eplerenone.

If known case of heart failure – on β -blocker – presented with acute pulmonary oedema → Increase diuretics, **stop β -blockers and restart β -blockers when his lungs are dry.**

A significant benefit from using IV iron in patients with heart failure and iron deficiency was demonstrated in a study

history of heart failure + iron deficiency. the first step → correcting iron deficiency

Non-pharmacological management

- **Cardiac resynchronisation therapy (CRT) (biventricular pacing): criteria for resynchronisation therapy recommended by NICE guidance**
 1. They are in sinus rhythm +
 - either with a QRS duration of ≥ 150 ms estimated by ECG (**LBBB**)
 - or with a QRS duration of 120-149 ms estimated by ECG and mechanical dyssynchrony that is confirmed by echocardiography
 2. They have a left ventricular ejection fraction of $\leq 35\%$.
 3. They are receiving optimal pharmacological therapy.
- **Benefit :** Improved symptoms and reduced hospitalisation in NYHA class III patients
- **Investigations :** **the most useful investigation in predicting symptomatic response to cardiac resynchronisation therapy is → transthoracic echocardiogram and ECG** (The echo will show asynchronous contraction of the LV and RV and subsequently reduced ejection fraction).
- **Complications:** When a CRT device is implanted the left ventricular lead is inserted in the coronary sinus. To obtain access to the coronary sinus a catheter with an aggressive tip is used. **There is a 1% risk of causing dissection/perforation to the coronary sinus which can lead to cardiac tamponade.**

Implantable cardioverter defibrillator (ICD)

- **Where there is no LBBB and QRS is between 120-149 ms, ICD is the recommended option according to NICE guidelines. This is because of the risk of VT on account of the low ejection fraction, (<35%), and symptomatic heart failure.**

Exercise training

- improves symptoms but not hospitalisation/mortality

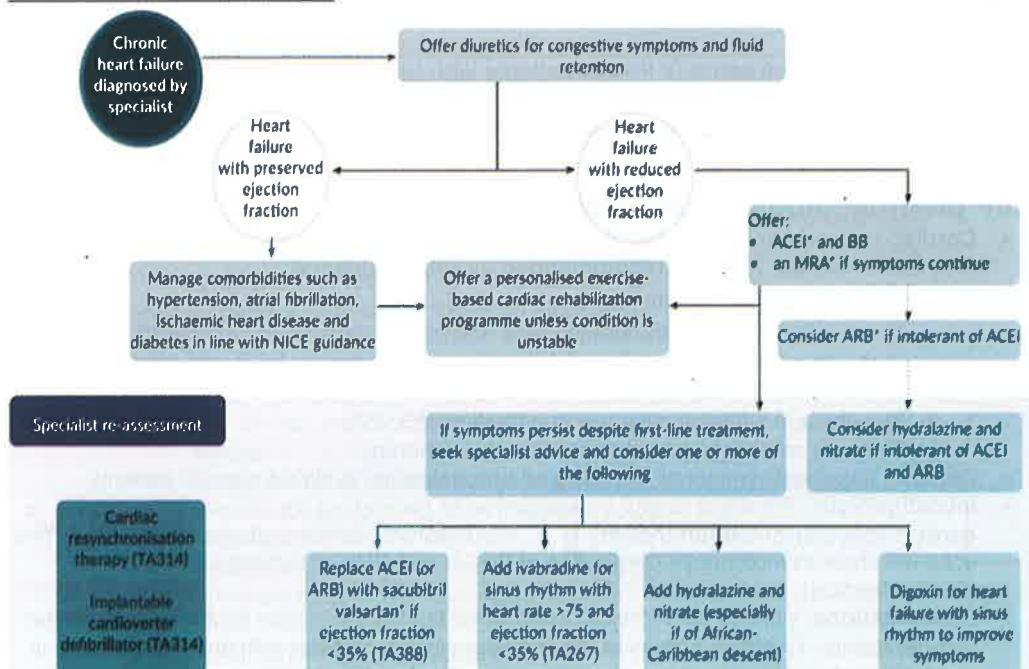
Tocolysis-associated pulmonary oedema

- Tocolytics are medications administered for the suppression of premature contractions.
- Acute pulmonary oedema can occur with administration of β_2 agonists for tocolysis in up to 5–15% of cases.**
- It usually occurs after 24 h of administration of these agents.
- The chest X-ray reveals pulmonary infiltrates and **normal heart size**.
- Concomitant use of corticosteroids that are often administered for lung maturation have also been implicated as risk factor for development of tocolysis-associated pulmonary oedema.
- Treatment involves stopping the tocolytics, oxygen and careful volume control.
- Deferential:
 - ⇒ Peripartum cardiomyopathy:
 - typically presents in the last month of pregnancy and up-to 6 months postpartum.
 - cardiomegaly on chest X-ray.

NICE management summary

Chronic heart failure: management

NICE National Institute for Health and Care Excellence



*Measure serum sodium, potassium and assess renal function before and after starting and after each dose increment.
If eGFR is 30 to 45 ml/min/1.73 m², consider lower doses or slower titration of ACEI or ARBs, MRAs, sacubitril valsartan and digoxin.

Other management options

If heart failure is caused or worsened by **other conditions**, these should be managed appropriately:

- **Revascularisation** (e.g. coronary artery bypass grafting)
- **Valve surgery** (e.g. aortic valve replacement)
- Implantable cardiac defibrillator (ICD): inserted if EF <30% for prevention of fatal arrhythmias
- **Cardiac resynchronisation therapy + defibrillator (CRT-D):** a biventricular pacemaker for EF <30% + QRS >130 m/sec to re-synchronise left and right ventricular contraction to improve EF
- **Cardiac transplantation** is rare and strict criteria must be met for consideration.
 - ⇒ By five years following cardiac transplantation, **nearly all patients have some degree of small coronary vascular narrowing (Coronary arteriopathy).**

Potentially harmful drugs to avoid in heart failure

Drug to avoid	Notes
Non-steroidal anti-inflammatory drugs (NSAIDs)	<ul style="list-style-type: none"> • May cause sodium and water retention, peripheral vasoconstriction, worsen heart failure, and decrease renal function. • Acute renal failure may be more likely when these agents are used in combination with an ACE inhibitor (ACEI) / angiotensin receptor blocker (ARB) and/or diuretic.
Non-dihydropyridine calcium channel blockers –verapamil and diltiazem1	<ul style="list-style-type: none"> • Negative inotropic effect may further depress cardiac function. Risk is greatest with verapamil, then diltiazem and least risk with dihydropyridines, but use with caution • Non-dihydropyridine calcium channel blockers are contraindicated in systolic heart failure , but may be useful in heart failure with preserved ejection fraction where slowing heart rate can increase filling time
Tricyclic antidepressants	May prolong QT interval and cause arrhythmias as well as hypotension from alpha-blocking effects
Thiazolidinediones (e.g. pioglitazone)	May cause fluid retention and heart failure by increasing renal sodium reabsorption
Corticosteroids	May worsen heart failure due to sodium and water retention (mineralocorticoid effect)
Clozapine	May cause cardiomyopathy and myocarditis
Oncology treatments such as anthracyclines (doxorubicin, daunorubicin), trastuzumab	may cause heart failure
Tumour necrosis factor antagonists (e.g. infliximab, etanercept)	May cause heart failure
Moxonidine (centrally acting antihypertensive)	Contraindicated in heart failure. Associated with increased mortality in heart failure

Prognosis

- Prognosis is poor overall, with approximately 50% of people with heart failure dying within five years of diagnosis
- **Factors indicating worse prognosis in heart failure**
 - ⇒ High BNP/NT-pro-BNP
 - ⇒ Anaemia
 - ⇒ **Hyponatraemia**
 - ⇒ Increased uric acid.

Mechanical support with the insertion of an intra-aortic balloon pump (IABP) in patient with Cardiogenic shock

- In case with hypotension and cardiogenic shock, what is the most appropriate intervention after failure of an inotropic support treatment?
 - ⇒ Intra-aortic balloon counter pulsation (IABCP) to support cardiac output.
- An intra-aortic balloon pump is inserted under echocardiographic guidance. At which point of the ECG should balloon inflation be timed?
 - ⇒ Middle of the T wave
 - Balloon inflation is timed with diastole once closure of the aortic valve has occurred; this corresponds to the middle of the T wave.
- What is the contraindication to placement of an intra-aortic balloon pump?
 - ⇒ For blood to be ejected antegrade to perfuse the tissues and retrograde to perfuse the coronaries, the aortic valve must be closed and competent. **Aortic regurgitation is therefore a contraindication to placement of an intra-aortic balloon pump.**

Hypertrophic obstructive cardiomyopathy (HOCM)

HOCM is the most common cause of sudden cardiac death in the young

- (HOCM) is an autosomal dominant disorder of muscle tissue caused by defects in the genes encoding contractile proteins.
- The most common defects involve a mutation in the gene encoding **β-myosin heavy chain** protein or myosin binding protein C.
- Mutations to various proteins including beta-myosin, alpha-tropomyosin and troponin T have been identified.
- **type of mutation → Frame-shift mutation**
- The estimated prevalence is 1 in 500.
- Septal hypertrophy causes left ventricular outflow obstruction.
- It is an important cause of sudden death in apparently healthy individuals.

Protein	Percentage
Beta-myosin heavy chain	35
Myosin-binding protein C	15
Troponin T	15
Alpha-tropomyosin	1
Myosin light chain	1

Mutations known to cause hypertrophic cardiomyopathy.

Features

Sudden death, unusual collapse in young person - ? HOCM

Symptoms and signs are similar to those of aortic stenosis, except that the character of the pulse in HOCM is jerky

- often asymptomatic
- dyspnea (the most common presenting symptom)
- angina,
- syncope
- sudden death (most commonly due to ventricular arrhythmias), arrhythmias, heart failure
- jerky pulse,
- large 'a' waves,
- double apex beat
- ejection systolic murmur: increases with Valsalva manoeuvre and decreases on squatting
 ⇒ Diastolic decrescendo murmur of aortic regurgitation (10% of patients)

Associations

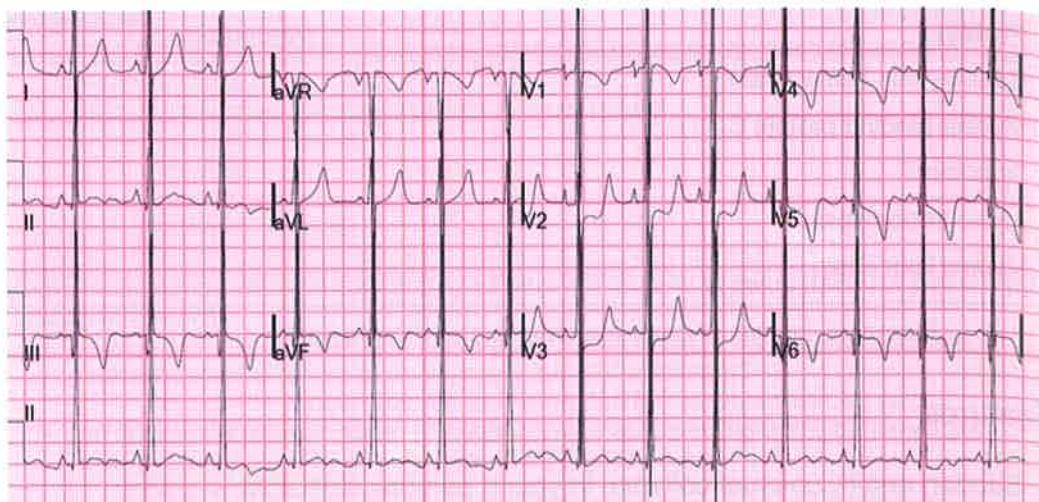
- Friedreich's ataxia
- Wolff-Parkinson White

Echo - mnemonic - MR SAM ASH

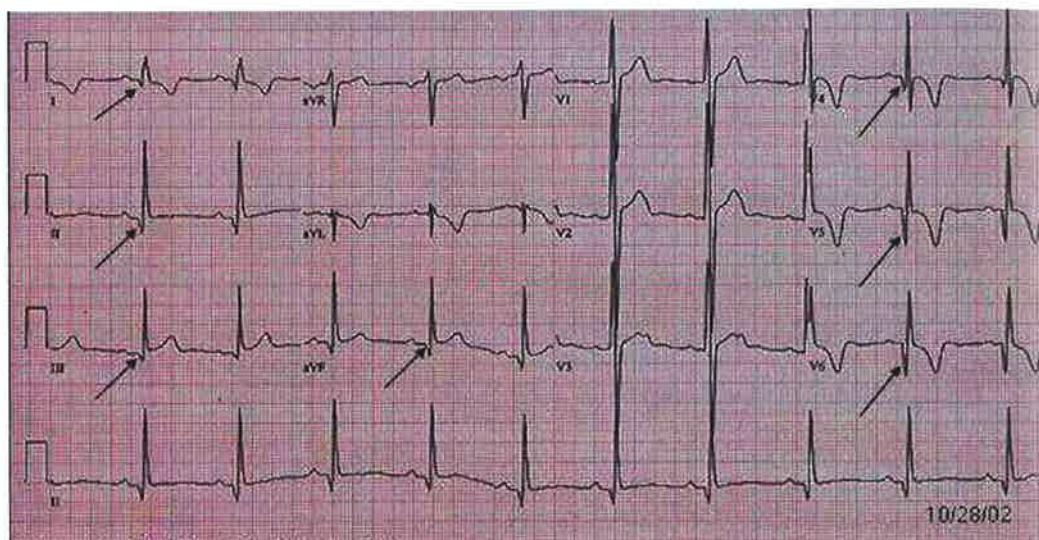
- mitral regurgitation (MR)
- systolic anterior motion (SAM) of the anterior mitral valve leaflet
- asymmetric hypertrophy (ASH)

ECG

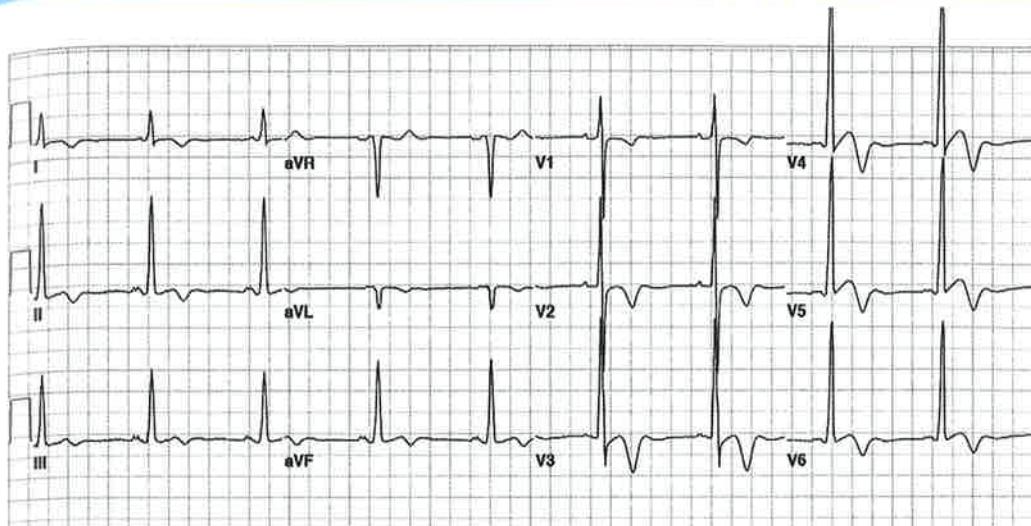
- left ventricular hypertrophy
- progressive T wave inversion
- deep Q waves
- right or left axis deviation
- PR prolongation
- atrial fibrillation may occasionally be seen
- **Right bundle branch block**
 - ⇒ **the most ECG FINDING which support a diagnosis of HOCM**
 - ⇒ RBBB is correlated with anterior, anteroseptal and mid-septal myocardial fibrosis in HOCM.



ECG showing typical changes of HOCM including LVH and T wave inversion



Dagger-like Q waves



This ECG shows the typical pattern of **apical HCM**:

- Large precordial voltages.
- Giant T wave inversions in the precordial leads
- Inverted T waves are also seen in the inferior and lateral leads.

Type of cardiomyopathy	Selected points
Hypertrophic obstructive cardiomyopathy	<ul style="list-style-type: none"> • Leading cause of sudden cardiac death in young athletes • Usually due to a mutation in the gene encoding β-myosin heavy chain protein • Common cause of sudden death • Echo findings include: <ul style="list-style-type: none"> ⇒ MR, ⇒ systolic anterior motion (SAM) of the anterior mitral valve ⇒ asymmetric septal hypertrophy
Arrhythmogenic right ventricular dysplasia	<ul style="list-style-type: none"> • Right ventricular myocardium is replaced by fatty and fibrofatty tissue • Around 50% of patients have a mutation of one of the several genes which encode components of desmosome • ECG abnormalities in V1-3, <ul style="list-style-type: none"> ⇒ typically T wave inversion. ⇒ An epsilon wave is found in about 50% of those with ARV - this is best described as a terminal notch in the QRS complex

Management

- Amiodarone
- Beta-blockers or verapamil for symptoms
- Cardioverter defibrillator
- Dual chamber pacemaker
- Endocarditis prophylaxis

Beta-blockers

- Generally first-line agents
 - ⇒ increase diastolic filling and decrease contractility
 - ⇒ Reduces provable gradient

disopyramide

- If β -blockers alone are ineffective, disopyramide, may be added (Class IA anti-arrhythmic drug)
- anticholinergic side-effects include dry eyes and mouth, urinary hesitancy or retention, and constipation.
- QTc interval should be monitored during dose up-titration and the dose reduced if it exceeds 480 ms.
- Disopyramide should be avoided in patients with glaucoma, prostatism, and in patients taking other drugs that prolong the QT interval, such as amiodarone and sotalol.

Verapamil

- Verapamil (starting dose 40 mg three times daily to maximum 480 mg daily) can be used when β -blockers are contraindicated or ineffective,
- close monitoring is required in patients with severe obstruction (≥ 100 mm Hg) or elevated pulmonary artery systolic pressures, as it can provoke pulmonary oedema.
- Verapamil should however be avoided in HOCM patients with coexistent Wolff Parkinson White as it may precipitate VT or VF.

Implantable cardioverter defibrillators (ICD) implantation → prevention of sudden cardiac death

- recommended in patients who have survived a cardiac arrest due to VT or VF or who have spontaneous sustained VT causing syncope or haemodynamic compromise

Invasive treatment (myectomy or alcohol septal ablation) (ESC Guidelines 2014)

- Left ventricular outflow tract obstruction (LVOTO) is defined as a peak instantaneous Doppler LV outflow tract gradient of ≥ 30 mm Hg, but the threshold for invasive treatment is usually considered to be ≥ 50 mm Hg.
- **Septal reduction therapy** is recommended in patients with LVOT gradient of ≥ 50 mm Hg, who are in NYHA functional Class III–IV, despite maximum tolerated medical therapy.
- The most commonly performed surgical procedure used to treat LVOTO is ventricular septal myectomy (Morrow procedure).
- Pre-operative determinants of a good long-term outcome are age < 50 years, left atrial size < 46 mm, absence of atrial fibrillation and male gender.
- surgery VS septal alcohol ablation (SAA)
 - ⇒ both procedures improve functional status with a similar procedural mortality.
 - ⇒ Septal alcohol ablation is associated with a higher risk of AV block, requiring permanent pacemaker implantation and larger residual LV outflow tract gradients.
 - ⇒ In contrast to myectomy, most patients develop right-, rather than left bundle branch block after SAA.

Drugs to avoid

- nitrates
- ACE-inhibitors
- Inotropes : Digoxin

Poor prognostic factors, which are predictive of sudden cardiac death

HOCM - poor prognostic factor on echo = septal wall thickness of > 3 cm

- syncope
- family history of sudden death
- **Maximum left ventricular wall thickness greater than 3 cm**
- young age at presentation
- non-sustained ventricular tachycardia on 24 or 48-hour Holter monitoring
- Abnormal blood pressure changes on exercise (**Blood pressure drop during peak exercise on stress testing**).

Screening of HOCM

- Current guidelines suggest that a resting ECG and TTE (transthoracic ECHO) are the most effective screening strategies for relatives of patients with HOCM.
- Genetic testing is not recommended as a first line screening tool given varying rates of penetrance.

Dilated cardiomyopathy (DCM)

Overview

- Most common cardiomyopathy
- Sex: ♂ > ♀ (approx. 3:1)
- dilated heart leading to systolic (+/- diastolic) dysfunction
- all 4 chambers affected but LV more so than RV

Features

- arrhythmias,
- **emboli → cardio-embolic stroke,**
- mitral regurgitation
- absence of congenital, valvular or ischaemic heart disease

Causes

- Common causes
 - ⇒ Idiopathic (approx. 50%)
 - ⇒ alcohol: may improve with thiamine
 - ⇒ postpartum
 - ⇒ hypertension
- **Other causes**
 - ⇒ genetic inherited dilated cardiomyopathy:
 - around third of DCM patients
 - **the majority** of defects are inherited in an **autosomal dominant** fashion although other patterns of inheritance are seen
 - ⇒ infections e.g. Coxsackie B, HIV, diphtheria, parasitic
 - ⇒ endocrine e.g. Hyperthyroidism
 - ⇒ neuromuscular e.g. Duchenne muscular dystrophy
 - ⇒ nutritional e.g. Kwashiorkor, pellagra, thiamine/selenium deficiency
 - **Selenium deficiency is one of the reversible causes of dilated cardiomyopathy.**
 - ⇒ drugs e.g. Doxorubicin
 - ⇒ Infiltrative (may also lead to **restrictive cardiomyopathy**) e.g. Haemochromatosis, Sarcoidosis

Diagnosis → Echocardiogram

- The echo may show:
 - ⇒ Reduced left ventricular ejection fraction,
 - ⇒ myocardial dyssynchrony (myocardial segments contract at different points in time),
 - ⇒ thinning of the left ventricular wall
 - ⇒ dilated left ventricle.

Type of cardiomyopathy	Selected causes/points
Dilated cardiomyopathy	Classic causes include <ul style="list-style-type: none"> • alcohol • Coxsackie B virus • wet beri beri • doxorubicin
Restrictive cardiomyopathy	Classic causes include <ul style="list-style-type: none"> • amyloidosis • post-radiotherapy • Loeffler's endocarditis

Becker's muscular dystrophy

- **X-linked recessive** disorder resulting from a mutation in the **dystrophin gene**.
- The clinical picture is similar to that of Duchenne's muscular dystrophy but it is much **milder**.
- Patients usually present between the ages of 5 and 15 years, though presentation may not be until the fourth or fifth decade.
- **Patients may present with heart failure secondary to dilated cardiomyopathy rather than the classic proximal muscle weakness.**

Restrictive cardiomyopathy

Restrictive cardiomyopathy: amyloid (most common), haemochromatosis, Loffler's syndrome, sarcoidosis, scleroderma

Causes

- amyloidosis (e.g. secondary to myeloma) - **most common cause in UK**
 - ⇒ Cardiac involvement is the most common cause of death in patients with amyloidosis associated with an immunocyte dyscrasia - typically as restrictive cardiomyopathy
 - ⇒ **Transthyretin gene** mutations can lead to restrictive cardiomyopathy from amyloid deposition in the heart.
 - ⇒ Diagnosis is confirmed by **myocardial biopsy**, which shows **amyloid infiltration** when stained with **Congo Red**.
 - myocardial biopsy, which when stained with Congo Red will show "**apple green birefringence**" amyloid under polarized light.
- haemochromatosis
- Loffler's syndrome
- sarcoidosis
- scleroderma
- Radiotherapy
- Systemic sclerosis
- Carcinoid syndrome.

Pathophysiology:

- Proliferation of connective tissue → ↓ elasticity of myocardium → ↓ ventricular compliance → ↓ diastolic filling → atrial congestion → atrial enlargement and severe diastolic dysfunction

Features

- Physical examination reveals right heart failure with a raised JVP, characteristically showing a prominent deep Y descent
- Heart size is often normal.
- S 4 heart sound, due to ventricular noncompliance.
- Pericardial effusion is common, but rarely causes tamponade
- The most characteristic ECG finding of restrictive cardiomyopathy is diffusely diminished voltages**
- Echocardiography findings**
 - ⇒ small thick ventricles and a thick interatrial septum due to amyloid deposits, which have a 'granular sparkling' appearance
 - Amyloid deposits in the heart produce generalized thickening of the myocardium (as opposed to asymmetrical septal hypertrophy commonly seen in hypertrophic cardiomyopathy) and diastolic dysfunction.
 - ⇒ impaired relaxation in the diastolic phase.
 - ⇒ bright speckled appearance.

Differential diagnosis

- constrictive pericarditis**
 - ⇒ Features are very similar in constrictive pericarditis, but in constrictive pericarditis:
 - the apex is frequently non-palpable due to the thick pericardium**
 - chest X-ray may show pericardial calcifications

Features suggesting restrictive cardiomyopathy rather than constrictive pericarditis

- prominent apical pulse
- absence of pericardial calcification on CXR
- heart may be enlarged
- ECG abnormalities e.g. bundle branch block, Q waves

Clinical Features of Constrictive Pericarditis and Restrictive Cardiomyopathy

Clinical Features	Constrictive Pericarditis	Restrictive Cardiomyopathy
History	Prior history of pericarditis or condition that causes pericardial disease	History of systemic disease (eg, amyloidosis, hemochromatosis)
Systemic examination - Heart sounds	Pericardial knock, high-frequency sound	Presence of loud diastolic filling sound S₃ , Low-frequency sound
Murmurs	No murmurs	Murmurs of mitral and tricuspid insufficiency
apical pulse	apex is frequently non-palpable due to the thick pericardium	prominent apical pulse
Prior chest radiograph	Pericardial calcification	Normal results of prior chest radiograph

Management

- Cardiac transplant

Peripartum cardiomyopathy (PCM)

- biventricular heart failure during the third trimester.
- the aetiology: unknown, although both myocarditis and low levels of dietary selenium have been postulated as causes.

Management

- similar to the management of heart failure in any other situation with vasodilators, diuretics and beta blockade. ACE inhibition is reserved for the post-partum period.
 - ⇒ sodium restriction,
 - ⇒ diuretics to optimise the volume status,
 - ⇒ digoxin and afterload-reducing agents.
 - ⇒ Hydralazine
- For patients presenting with PCM, defined as left ventricular systolic dysfunction 1 month prior to delivery or 5 months postpartum, volume status should first be managed with diuretics after liaison with obstetricians. **Beta-blockers should be added once the patient's volume status is optimised.**
- Anticoagulation
 - ⇒ Patients with PCM are at risk of thromboembolism due to both hypercoagulable state of pregnancy and stasis of blood in the left ventricle. Therefore, anticoagulation with **heparin** is recommended.

Type of cardiomyopathy	Selected points
Peripartum cardiomyopathy	<ul style="list-style-type: none"> • Typical develops between last month of pregnancy and 5 months post-partum • More common in older women, greater parity and multiple gestations
Takotsubo cardiomyopathy	<ul style="list-style-type: none"> • 'Stress'-induced cardiomyopathy e.g. patient just found out family member dies then develops chest pain and features of heart failure • Transient, apical ballooning of the myocardium • Treatment is supportive

Takotsubo cardiomyopathy

Definition:

- Takotsubo cardiomyopathy is a type of non-ischaemic cardiomyopathy associated with a transient, apical ballooning of the myocardium.
- acute, stress-induced, reversible dysfunction of the left ventricle

Epidemiology:

- especially postmenopausal women > 60 years

Pathophysiology:

- emotional/physical stress → massive catecholamine discharge → cardiotoxicity, multi-vessel spasms and dysfunction → myocardial stunning

Features

- chest pain
- features of heart failure
- ST elevation
- normal coronary angiogram

Treatment

- supportive

Prognosis:

- spontaneous recovery if stressors are avoided

Congenital heart disease: types

Paradoxical embolus - PFO most common cause - do TOE

Congenital heart disease

- cyanotic: TGA most common at birth, Fallot's most common overall
- acyanotic: VSD most common cause

Acyanotic - most common causes

- **ventricular septal defects (VSD) - most common**, accounts for 30%
- atrial septal defect (ASD) 10%.
- patent ductus arteriosus (PDA)
- coarctation of the aorta
- aortic valve stenosis

VSDs are more common than ASDs. However, in adult patients ASDs are the more common new diagnosis as they generally presents later

Cyanotic - most common causes

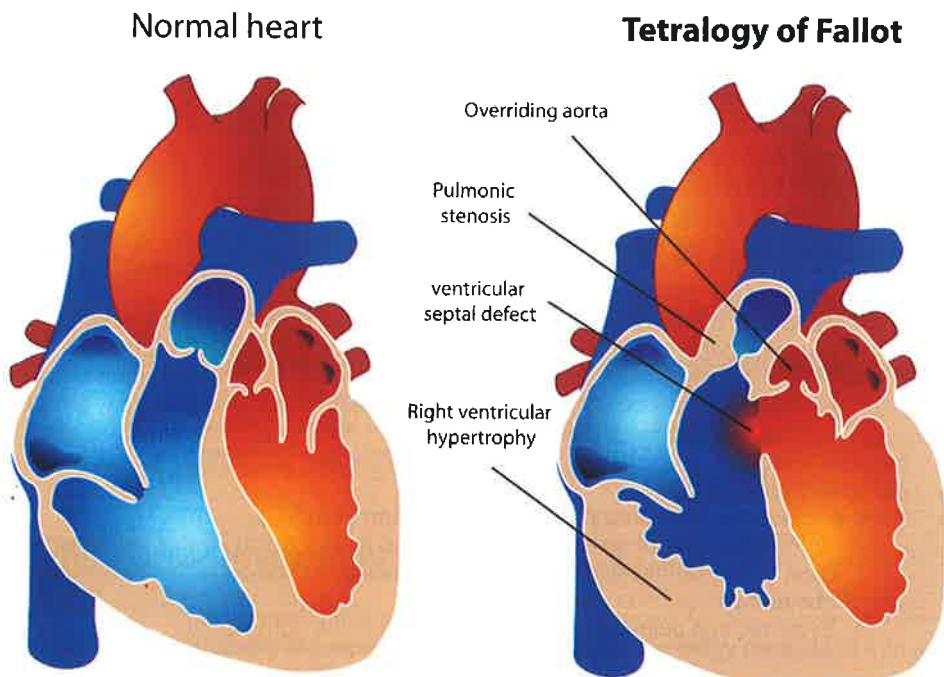
- tetralogy of Fallot
 - ⇒ There is a single sound in Fallot's because of an absent P2.
 - ⇒ A Blalock shunt (anastomosis of subclavian artery to pulmonary artery) used to be performed for Fallot's tetralogy and leads to a weak left radial pulse.
- transposition of the great arteries (TGA)
 - ⇒ **Fallot's** is more common than TGA. However, at birth TGA is the more common lesion as patients with Fallot's generally presenting at around 1-2 months
 - ⇒ TGA is usually treated by **prostaglandins in order to keep the ductus arteriosus patent** (from pulmonary artery to the descending aorta), so some oxygenated blood can reach systemic circulation.
- tricuspid atresia
- pulmonary valve stenosis
- **Total anomalous pulmonary venous connection (TAPVC)**
 - ⇒ TAPVC consists of an abnormality of blood flow in which all four pulmonary veins drain into systemic veins or the right atrium with or without pulmonary venous obstruction.
 - ⇒ Systemic and pulmonary venous blood mix in the right atrium.

Other notes

- Aortic regurgitation may be a feature of osteogenesis imperfecta.
- Ebstein's anomaly is associated with maternal LiCO₃ use if exposed in the first trimester.
- The majority of cases of neonates with complete heart block may be caused by autoimmune disease, particularly anti-ro antibodies in the mother.
- Left ventricle (LV) hypoplasia occurs when the left sided chambers fail to develop and blood enters the systemic circulation from the right ventricle via the pulmonary artery and a patent ductus arteriosus.

Tetralogy of Fallot (TOF)

- TOF is the most common cause of cyanotic congenital heart disease*.
 - ⇒ *however, at birth transposition of the great arteries is the more common lesion as patients with TOF generally present at around 1-2 months
- It typically presents at around 1-2 months, although may not be picked up until the baby is 6 months old
- TOF is a result of anterior malalignment of the aorticopulmonary septum. The four characteristic features are:
 1. ventricular septal defect (VSD)
 2. right ventricular hypertrophy
 3. right ventricular outflow tract obstruction, pulmonary stenosis
 - There is a single sound in Fallot's because of an absent P2.
 4. overriding aorta



- The severity of the right ventricular outflow tract obstruction determines the degree of cyanosis and clinical severity

Other features

- cyanosis
- causes a right-to-left shunt
- ejection systolic murmur due to pulmonary stenosis (the VSD doesn't usually cause a murmur)
- a right-sided aortic arch is seen in 25% of patients
- chest x-ray shows a 'boot-shaped' heart, ECG shows right ventricular hypertrophy

Management

- surgical repair is often undertaken in two parts
- cyanotic episodes may be helped by beta-blockers to reduce infundibular spasm

The most common residual lesion in repaired tetralogy of Fallot is pulmonary regurgitation.

Ventricular septal defects (VSD)**Overview**

- The second most common congenital heart defect
⇒ bicuspid aortic valve is the most common congenital heart defect
- They close spontaneously in around 50% of cases.
- **the most common site for a VSD → Perimembranous**
⇒ Perimembranous VSDs account for 70-80% of VSDs and are situated between the inlet and outlet portions of the septum.

Associations

- Congenital VSDs: associated with:
⇒ chromosomal disorders (e.g. Down's syndrome, Edward's syndrome, Patau syndrome)
- Non-congenital causes include:
⇒ Fetal alcohol syndrome
⇒ Intrauterine infection (e.g., TORCH)
⇒ post myocardial infarction

Features

- **Pan-systolic murmur** which is:
⇒ louder in smaller defects
⇒ **usually loudest at the left lower sternal edge (LSE)**
- Mid-diastolic murmur over cardiac apex
⇒ Due to increased flow through the mitral valve
- systolic thrill
- Loud pulmonic S2 (if pulmonary hypertension develops)

Investigations

- Chest x-ray
⇒ Enhanced pulmonary vascular markings
⇒ Left atrial and ventricular enlargement
- ECG
⇒ **The clue to diagnosis in the ECG finding → Biventricular hypertrophy**
 - Biventricular hypertrophy is classically described as having **biphasic QRS complexes in V2–5** – which is known as the **Katz Wachtel phenomenon** and is **classic for VSD**.
- Doppler echocardiography: confirms diagnosis

Complications

- Aortic regurgitation
⇒ due to a poorly supported right coronary cusp resulting in cusp prolapse
- Infective endocarditis
- Eisenmenger's complex
- Right heart failure
- Pulmonary hypertension
⇒ pregnancy is contraindicated in women with pulmonary hypertension as it carries a 30-50% risk of mortality.

Treatment

- small to moderate defects often heal spontaneously

- Symptomatic and large VSDs → Surgical (patch) repair
- Heart-lung transplant or lung transplant with concurrent VSD repair if Eisenmenger's reaction has occurred

Atrial septal defect (ASD)

- common congenital heart lesion
 - ⇒ VSD is more common

Types

- **Ostium secundum**
 - ⇒ **70% of ASDs**
 - ⇒ associated with Holt-Oram syndrome (tri-phalangeal thumbs)
 - ⇒ ECG: **RBBB with RAD**
- **Ostium primum**
 - ⇒ present earlier than ostium secundum defects
 - ⇒ associated with abnormal AV valves
 - ⇒ the AV node is displaced posteriorly and inferiorly and atrial and/or AV nodal conduction is often delayed.
 - ⇒ ECG: **RBBB with LAD, prolonged PR interval**

wide, fixed, split-second sound + right-axis deviation → Ostium secundum

wide, fixed, split-second sound + left-axis deviation → Ostium primum

Features

- Symptoms
 - ⇒ asymptomatic in youth
 - ⇒ often discovered on routine school health exams
 - ⇒ mild fatigue
 - ⇒ frequent respiratory infections
 - ⇒ Larger ones may lead to signs of right ventricular failure, such as shortness of breath and a parasternal heave.
- Physical exam
 - ⇒ **Mid-systolic ejection murmur** (over the left second ICS)
 - Due to → Relative pulmonary stenosis due to an increase in stroke volume
 - ⇒ **Soft mid-diastolic murmur** (over the lower left sternal border)
 - arises from increased flow across the **tricuspid valve**.
 - ⇒ **loud S1**
 - ⇒ **wide fixed-split S2**
 - The most frequently tested knowledge
 - splitting is fixed (does not vary with respiration)
 - ⇒ **heaving cardiac impulse (LLSB)**
- Other features
 - ⇒ The **grossly elevated D_Lco** is secondary to the left-right shunt and increased pulmonary blood flow. In contrast, chronic pulmonary emboli will cause a low D_Lco.

Predisposes patient to

- CHF
 - 2nd/3rd decades of life
- Eisenmenger's syndrome
 - pulmonary hypertension
 - right ventricular hypertrophy
 - reversal to a right-to-left shunt
- stroke
 - due to paroxysmal embolus

Associated condition

- Tricuspid atresia is the congenital cardiac disorder most commonly associated with an atrial septal defect.
- Down syndrome
- Fetal alcohol syndrome
- Holt-Oram syndrome
 - ⇒ Autosomal dominant disorder, which is also called hand-heart syndrome because affected children present with an ASD, a first degree heart block, and abnormalities of the upper limbs (e.g., absent radial bones). It affects approx. 1 in 100,000 children.

ECG

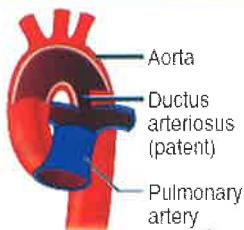
- **Right bundle branch block**

- ostium primum ASD → left axis deviation.
- ostium secundum ASD → right axis deviation.
- first degree heart block → **prolongation of the PR interval**
 - due to delayed conduction through the atria or through the AV node
- The QRS pattern typically is either an rSr' or an rsR' resulting from dilation and hypertrophy of the right ventricular outflow tract caused by volume overload of the right heart.

prominent left precordium in a young patient with an ejection murmur in the second left intercostal space indicate → **ASD with pulmonary hypertension**

- ⇒ A prominent left precordium suggests that:
 - the right ventricle was dilated during childhood
 - RV working against a high pressure

Patent ductus arteriosus



"**Indomethacin**" (indomethacin) **ends** patency of PDA; PGE **kEEps** it open (may be necessary to sustain life in conditions such as transposition of the great vessels). PDA is normal *in utero* and normally closes only after birth.

Overview

- acyanotic congenital heart defect
- connection between the pulmonary trunk and descending aorta
- more common in premature babies, born at high altitude or maternal rubella infection in the first trimester

Features

- left subclavicular thrill
- continuous 'machinery' murmur at the left upper sternal edge with late systolic accentuation
- large volume, bounding, collapsing pulse
- wide pulse pressure
- heaving apex beat

Management

- indomethacin closes the connection in the majority of cases
- if associated with another congenital heart defect amenable to surgery then prostaglandin E1 is useful to keep the duct open until after surgical repair

Patent foramen ovale (PFO)

- PFO is present in around 20% of the population.
- It may allow embolus (e.g. from DVT) to pass from right side of the heart to the left side leading to a stroke - 'a paradoxical embolus'
- There also appears to be an association between migraine and PFO.
 - ⇒ Some studies have reported improvement in migraine symptoms following closure of the PFO
- right heart catheter: left to right shunting of oxygenated blood at level of the atrium.
 - ⇒ oxygen saturation data show a step-up in the saturations between the vena cava and the right atrium.

Paradoxical embolisation

- For a right-sided thrombus (e.g. DVT) to cause a left-sided embolism (e.g. stroke) it must obviously pass from the right-to-left side of the heart.
- The following cardiac lesions may cause such events
 - patent foramen ovale - present in around 20% of the population
 - atrial septal defect - a much less common cause

Blue toe syndrome

- 80% of digital ischaemias have an emboli originating from the heart and so an urgent echocardiogram is crucial to prevent further and more severe events occurring.
- sudden onset of a cold, painful, and cyanotic big toe. the next steps → Therapeutic heparin and urgent echocardiogram

Eisenmenger's syndrome

Eisenmenger's syndrome - the reversal of a left-to-right shunt

Definition

- Eisenmenger's syndrome describes the reversal of a left-to-right shunt in a congenital heart defect due to pulmonary hypertension.
- This occurs when an uncorrected left-to-right leads to remodeling of the pulmonary microvasculature, eventually causing obstruction to pulmonary blood and pulmonary hypertension.

Associated with

- ventricular septal defect
 - Although patients with tetralogy of Fallot have, by definition, a ventricular septal defect they do not go on to develop Eisenmenger's syndrome
- atrial septal defect
- patent ductus arteriosus

Features

- original murmur may disappear
- cyanosis
- clubbing
- right ventricular failure
- polycythemia
- haemoptysis, embolism

Management

- heart-lung transplantation is required

Ebstein's anomaly

Definition

- Ebstein's anomaly is a congenital heart defect characterised by low insertion of the tricuspid valve resulting in a large atrium and small ventricle. It is sometimes referred to as 'atrialisation' of the right ventricle.

Causes

- Ebstein's anomaly may be caused by exposure to lithium in-utero

Features

- hypoplastic (atrialised) RV,
- apical displacement of the septal and posterior tricuspid valve leaflets,
- ASD,
- Right bundle branch block pattern on ECG.**

Associations

- tricuspid incompetence (pan-systolic murmur, giant V waves in JVP)
- Wolff-Parkinson White syndrome occurs in around 15% of the patients.

The presence of delta waves and short PR interval is indicative of WPW. When correlated with past surgical history (repair of atrial septal defect and tricuspid valve abnormalities as a child), Ebstein's anomaly is the most likely diagnosis.

Cardiac manifestations of genetic disorders

Genetic disorder	Associated cardiac manifestation
Marfan's syndrome	Aortic regurgitation (aortic dissection)
Down's syndrome	ASD, VSD
Turner's syndrome	Coarctation of the aorta
Spondyloarthritides, eg, ankylosing spondylitis	Aortic regurgitation

Peripheral vascular disease

- is a marker for increased risk of cardiovascular events even when it is asymptomatic.
- the femoropopliteal artery, the most common site of peripheral arterial disease.**
 - ⇒ paresthesia, intermittent claudication in calf and foot **and** palpable femoral pulses but absent pedal pulses

Risk factors

- age
 - ⇒ about 20% of people aged over 60 years have some degree of peripheral arterial disease.
- male gender
- Smoking
- Diabetes
- hypertension
- coronary artery disease.

Feature

- intermittent claudication (leg pain while walking) (The most common initial symptom).
- Critical limb ischaemia : ischaemic pain, ulceration, tissue loss and/or gangrene.

Investigations

- measuring the ankle brachial pressure index
 - ⇒ Calculate the index in each leg by dividing the highest ankle pressure by the highest arm pressure.
- Imaging before considering revascularization
 - ⇒ duplex ultrasound (first-line imaging)
 - ⇒ contrast-enhanced magnetic resonance angiography (after duplex ultrasound)
 - ⇒ computed tomography angiography (if contrast-enhanced magnetic resonance angiography is contraindicated or not tolerated.)

Treatment**Mild symptoms:**

- exercise programme
 - ⇒ 2 hours of supervised exercise a week for a 3- month period
 - ⇒ encouraging people to exercise to the point of maximal pain.
- management of cardiovascular risk factors (for example, with aspirin or statins)
- vasoactive drug treatment (for example, with naftidrofuryl oxalate).
- **Which drug might help improve pain-free walking distance?**
 - ⇒ **Naftidrofuryl**
 - ❖ Indicated only when exercise has not led to satisfactory improvement and the person refuse angioplasty or bypass surgery.
 - ❖ discontinue naftidrofuryl oxalate if there has been no symptomatic benefit after 3–6 months.
- Vasoactive drugs have limited benefit in treating intermittent claudication.
- There is modest evidence for the use of drugs such as naftidrofuryl and pentoxifylline, but little benefit from cinnarizine or inositol nicotinate.
- Simvastatin may be prescribed for patients with peripheral vascular disease who have elevated cholesterol levels, but there is no data on improvements in walking distance.

severe symptoms:

- endovascular treatment (such as angioplasty or stenting), bypass surgery, pain management and/or amputation.

Differential diagnosis of foot ulcers

	Venous ulcers	Arterial ulcer	Diabetic ulcer Neuropathic ulcer
Location	<ul style="list-style-type: none"> ▪ Gaiter region (above the ankle) 	<ul style="list-style-type: none"> ▪ Pressure points of the foot and shin (e.g., lateral malleolus, tips of the toes) 	<ul style="list-style-type: none"> ▪ Plantar pressure points of the foot (over the head of the metatarsal bones or the heel)
Mechanism	<ul style="list-style-type: none"> ▪ Chronic local venous hypertension → tissue ischemia 	<ul style="list-style-type: none"> ▪ Vessel occlusion → tissue ischemia 	<ul style="list-style-type: none"> ▪ Diabetic micro-vasculopathy and neuropathy → impaired tissue sustenance
Wound features	<ul style="list-style-type: none"> ▪ Irregular borders ▪ Exudative ▪ Superficial 	<ul style="list-style-type: none"> ▪ Punched-out appearance ▪ No exudation 	<ul style="list-style-type: none"> ▪ Hyperkeratotic borders ▪ Deep
Pain	<ul style="list-style-type: none"> ▪ Mild 	<ul style="list-style-type: none"> ▪ Severe 	<ul style="list-style-type: none"> ▪ Absent
Additional features	<ul style="list-style-type: none"> ▪ Varicose veins ▪ Oedema ▪ Stasis dermatitis 	<ul style="list-style-type: none"> ▪ Pale, shiny, cold, hairless surrounding skin ▪ Nail dystrophy ▪ Absent pulses 	<ul style="list-style-type: none"> ▪ Charcot joints ▪ Absent ankle reflex ▪ Impaired sensation (esp. vibration) ▪ Claw toes

- **Treatment of venous ulceration:**

- ⇒ control of oedema, treating any infection, and compression.
- ⇒ **Compressive dressings or devices should not be applied if the arterial circulation is impaired, and ankle-brachial pressure index is needed before application of compression**

Rheumatic fever

Definition

- an autoimmune process following infection with group A streptococci.

Overview

- Type II hypersensitivity is seen in rheumatic fever.
- Myocarditis is the most common cause of death during the acute phase of rheumatic fever.

Diagnosis: based on:

- **Evidence of recent streptococcal infection** accompanied by:
 - 2 major criteria
 - 1 major with 2 minor criteria
- Evidence of recent streptococcal infection
 - ⇒ ASOT > 200iu/mL
 - ⇒ history of scarlet fever
 - ⇒ positive throat swab
 - ⇒ increase in DNase B titre

Jones Criteria

Rheumatic fever major criteria: JONES

- Joints - polyarthritis;
- - carditis;
- Nodules (subcutaneous);
- Erythema marginatum;
- Sydenham's chorea.

• Major criteria

1. erythema marginatum
2. Sydenham's chorea
- 3. polyarthritis**
4. carditis (endo-, myo- or peri-)
5. subcutaneous nodules
 - Pea-sized, firm and non-tender.
 - characteristically seen on the extensor surfaces of joints such as knees and elbows and also over the spine.

• Minor criteria

1. raised ESR or CRP
2. pyrexia
3. arthralgia (not if arthritis a major criteria)
4. prolonged PR interval

Histology

- Aschoff bodies are foci of chronic inflammation seen histologically in the myocarditis of acute rheumatic fever.
 - ⇒ Anitschkow cells are reactive histiocytes with wavy, slender, caterpillar-like nuclei seen in Aschoff bodies of acute rheumatic fever.



*Erythematous patches
with central clearing*

Erythema marginatum

Erythema marginatum is seen in around 10% of children with rheumatic fever. It is rare in adults



Subcutaneous nodules
(*nodules of rheumatoid arthritis are larger*)

Infective endocarditis (IE)

The most common cause of endocarditis:

- ***Staphylococcus aureus* is now the most common cause of infective endocarditis**
- ***Staphylococcus epidermidis* if < 2 months post valve surgery.**

Definition

- an infection of the endocardium, the inner layer of the heart and valves.

Pathophysiology

- Damaged valvular endothelium → adherence of platelets and fibrin → sterile vegetation (microthrombus) → bacteremia → bacterial colonization of vegetation → valve destruction with loss of function

Risk factors

Infective endocarditis - strongest risk factor is previous episode of infective endocarditis

- **previous episode of endocarditis: The strongest risk factor for developing infective endocarditis.**
- previously normal valves (50%, typically acute presentation)
- rheumatic valve disease (30%)
- prosthetic valves
- congenital heart defects
- intravenous drug users (IVDUs, e.g. Typically causing tricuspid lesion)
- hemodialysis
- Hypertrophic cardiomyopathy.

Types

Acute Endocarditis	Subacute Endocarditis
Larger vegetations	Smaller vegetations
Attacks previously normal valves	Attacks damaged or abnormal valves
Destructive; 50% mortality rate despite treatment	Less destructive; most patients recover with treatment
High-virulence organisms, especially <i>S aureus</i>	Low-virulence organisms, especially the viridans streptococci <i>S mutans</i> and <i>S sanguinis</i>

The likelihood of infection

- The higher the valvular pressure, the greater the likelihood of infection. Thus, **mitral > aortic > tricuspid > pulmonary**. The exception to this rule is IV-related infective endocarditis; in this case, the tricuspid valve is the most commonly involved because it is the first valve encountered after venous injection.
- If the valve is already abnormal, then the likelihood of infection is greater and will be most likely on the aortic valve (*High-pressure systems create more blood turbulence and permit inoculation of the valve*).
- Diseases that affect the mitral valve, such as mitral valve prolapse and mitral regurgitation, are the most common valvular diseases. So **the mitral valve is the valve most frequently affected by endocarditis**. The exception is IV drug use. In these patients, the tricuspid valve is the most frequently involved valve

Causes

***Streptococcus bovis* endocarditis is associated with colorectal cancer**

- Staphylococcus aureus* (coagulase positive)** : the most common causative organism of IE (especially acute presentation, IVDUs).
 - ⇒ ***Staphylococcus aureus* endocarditis is an aggressive disease frequently associated with valve destruction and abscess formation.**
- Staphylococcus epidermidis*** (coagulase negative) most commonly associated with prosthetic valves < 2 months post operative.
- Streptococcus viridans***: commonly causing **subacute bacterial endocarditis**. The two most notable viridans streptococci are *Streptococcus mitis* and *Streptococcus sanguinis*. They are both commonly found in the mouth and in particular dental plaque so endocarditis caused by these organisms is linked with poor dental hygiene or following a dental procedure
- Streptococcus gallolyticus* (formerly *Streptococcus bovis*) is associated with colorectal cancer → colonoscopy should be done.**
- Bacteroides* is the most likely organism following bowel resection**, though *S. bovis* is also seen. Management is metronidazole.
- Candida endocarditis***: Risk factors: Intravenous drug abuse, immunodeficiency states and indwelling catheters. The aortic valve is the most common valve to be involved. **Treatment with Valve replacement followed by amphotericin B for 6 weeks**.
- Non-infective (sterile vegetations)**
 - ⇒ systemic lupus erythematosus (Libman-Sacks), commonly result in **mitral regurgitation**.
 - ⇒ malignancy: marantic endocarditis
- Culture negative causes**
 - ⇒ prior antibiotic therapy

- ⇒ *Coxiella burnetii* (Q fever agent), typically associated with exposure to animals (sheep and cattle).
- ⇒ *Bartonella* (from cats)
- ⇒ *Brucella*
- ⇒ *Chlamydia psittaci* (from birds).
- ⇒ **HACEK:** (*Haemophilus*, *Actinobacillus*, *Cardiobacterium*, *Eikenella*, *Kingella*)
 - slow-growing, **Gram negative bacteria**
 - These are normal flora of the upper respiratory tract
 - constitute 5-10% cases of endocarditis;
 - they require prolonged incubation in enriched media and increased carbon dioxide tension.
 - **The human bite injury and gram-negative culture make *Eikenella corrodens* the most likely causative organism.**
 - third-generation cephalosporin (**Ceftriaxone**) is effective against enteric gram-negative rods, including HACEK organisms

Associations	Most common cause
Generally	<i>Staphylococcus aureus</i>
prosthetic valves < 2 months post operative	<i>Staphylococcus epidermidis</i>
IV drug use	<i>Staphylococcus aureus</i>
Recent dental procedure	<i>Streptococcus viridans</i> : (<i>Streptococcus mitis</i> and <i>Streptococcus sanguinis</i>).
Colorectal cancer	<i>Streptococcus gallolyticus</i> (formerly <i>Streptococcus bovis</i>)

Diagnosis

Infective endocarditis: Modified Duke criteria

- **Infective endocarditis diagnosed if**
 - ⇒ pathological criteria positive, **or**
 - ⇒ 2 major criteria, **or**
 - ⇒ 1 major and 3 minor criteria, **or**
 - ⇒ 5 minor criteria
- **Pathological criteria**
 - ⇒ Positive histology or microbiology of pathological material obtained at autopsy or cardiac surgery (valve tissue, vegetations, embolic fragments or intracardiac abscess content)
- **Major criteria**
 - ⇒ **Positive blood cultures**
 - two positive blood cultures showing typical organisms consistent with infective endocarditis, such as *Streptococcus viridans* and the HACEK group, or
 - persistent bacteraemia from two blood cultures taken > 12 hours apart or three or more positive blood cultures where the pathogen is less specific such as *Staph aureus* and *Staph epidermidis*, or
 - positive serology for *Coxiella burnetii*, *Bartonella* species or *Chlamydia psittaci*, or
 - positive molecular assays for specific gene targets
 - ⇒ **Evidence of endocardial involvement**
 - positive echocardiogram (oscillating structures, abscess formation, new valvular regurgitation or dehiscence of prosthetic valves).

- **Minor criteria**

- 1) predisposing heart condition or intravenous drug use
- 2) microbiological evidence does not meet major criteria
- 3) fever > 38 C
- 4) vascular phenomena: major emboli, splenomegaly, clubbing, splinter haemorrhages, Janeway lesions, petechiae or purpura
- 5) immunological phenomena: **glomerulonephritis**, Osler's nodes, Roth spots

Classical symptoms of infective endocarditis can be remembered using the mnemonic FROM JANE: fever, Roth spots, Osler nodes, murmur, Janeway lesions, anaemia, nail-bed haemorrhage, emboli.

Ow for Owsler nodes: Osler nodes and Janeway lesions are similar in appearance, yet **Osler nodes are painful** and **Janeway lesions are painless**.

Peripheral signs associated with infective endocarditis

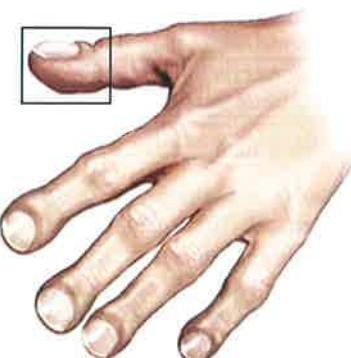


Clubbed fingers

Normal angle of nail bed



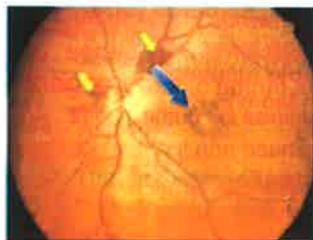
Distorted angle of nail bed



Splinter hemorrhage



Osler node



Roth's spot



Janeway lesion

Investigation

- Transthoracic echocardiography (TTE) is the initial test of choice for all patients with suspected IE.
- How should the blood samples be drawn to maximise the chances of obtaining positive cultures?
 - ⇒ Draw three samples of blood from different venepuncture sites with the first separated from the last by at least one hour over 24 hours

Aortic valve endocarditis can cause aortic root abscess which can cause damage to the AV node resulting in prolongation of the PR interval on ECG.

Management

Current antibiotic guidelines (source: British National Formulary)

Scenario	Suggested antibiotic therapy
Initial blind therapy	<ul style="list-style-type: none"> • Native valve: <ul style="list-style-type: none"> ⇒ Not allergic to penicillin , no MRSA , not sever sepsis: <ul style="list-style-type: none"> ▪ Amoxicillin + gentamicin ⇒ Allergic to penicillin , MRSA , or sever sepsis: <ul style="list-style-type: none"> ▪ vancomycin + gentamicin • prosthetic valve: <ul style="list-style-type: none"> ⇒ vancomycin + rifampicin + gentamicin
Staphylococci endocarditis	<ul style="list-style-type: none"> • Native valve: <ul style="list-style-type: none"> ⇒ Not allergic to penicillin , no MRSA , not sever sepsis: <ul style="list-style-type: none"> ▪ Flucloxacillin ⇒ Allergic to penicillin , MRSA , or sever sepsis: <ul style="list-style-type: none"> ▪ vancomycin + rifampicin • Prosthetic valve: <ul style="list-style-type: none"> ⇒ Not allergic to penicillin , no MRSA , not sever sepsis: <ul style="list-style-type: none"> ▪ Flucloxacillin + rifampicin + gentamicin ⇒ Allergic to penicillin , MRSA , or sever sepsis: <ul style="list-style-type: none"> ▪ vancomycin + rifampicin + gentamicin
Streptococci endocarditis	<ul style="list-style-type: none"> • Native valve and Prosthetic valve : <ul style="list-style-type: none"> ⇒ not allergic to penicillin <ul style="list-style-type: none"> ▪ Benzylpenicillin ± gentamicin ⇒ Allergic to penicillin: <ul style="list-style-type: none"> ▪ vancomycin + gentamicin

IV amoxicillin is the empirical treatment of choice in native valve endocarditis

The most useful laboratory test used to monitor the treatment of infective endocarditis is serial C reactive protein estimation.

- **length of treatment:**

- ⇒ 6 weeks of intravenous therapy is generally accepted as the length of treatment needed.

Indications for surgery

Infective endocarditis - indications for surgery:

- severe valvular incompetence
- aortic abscess (often indicated by a lengthening PR interval)
- infections resistant to antibiotics/fungal infections
- cardiac failure refractory to standard medical treatment
- recurrent emboli after antibiotic therapy
- organisms that are difficult to eradicate by medical therapy as such **fungi**, brucella, coxiella, pseudomonas aeruginosa, vancomycin-resistant enterococci
- persistent bacteraemia despite appropriate antibiotic therapy
- extension of infection to a extravalvular site
- early prosthetic valve endocarditis (within 2 months)
- dehiscence or obstruction of a prosthetic valve.
- large (more than 10 mm) vegetations

Prophylaxis

- NICE recommends the following procedures **do not require prophylaxis**:
 - ⇒ dental procedures
 - ⇒ upper and lower gastrointestinal tract procedures
 - ⇒ genitourinary tract; this includes urological, gynaecological and obstetric procedures and childbirth
 - ⇒ upper and lower respiratory tract; this includes ear, nose and throat procedures and bronchoscopy
- **Prophylaxis is only recommended** in those patients who are at highest risk of adverse outcomes on the development of endocarditis. These patient groups include:
 - ⇒ Prosthetic cardiac valve or prosthetic material used for cardiac valve repair
 - ⇒ Previous endocarditis
 - ⇒ **Unrepaired cyanotic congenital heart disease** including palliative shunts and conduits
 - ⇒ **Completely repaired congenital heart defect** with prosthetic material or device, whether placed by surgery or by catheter intervention, **during the first six months** after the procedure
 - ⇒ **Repaired congenital heart disease with residual defects** (persisting leaks or abnormal flow) at the site or adjacent to the site of a prosthetic patch or prosthetic device (which inhibit endothelialisation)
 - ⇒ Cardiac transplantation recipients who develop cardiac valve abnormalities.

Prognosis

Infective endocarditis - streptococcal infection carries a good prognosis

Poor prognostic factors

- Staph aureus infection (→ Acute endocarditis)
 - ⇒ (*Streptococcus viridans* → Subacute bacterial endocarditis has a better prognosis.)
- Prosthetic valve (especially 'early', acquired during surgery)
- Culture negative endocarditis
- Low complement levels
- Infection of the aortic rather than mitral valve
- Associated rhythm disturbance.
- Heart failure: **the most common cause of death from infective endocarditis**
- Intravenous drug abuse (often left and right sided disease)
- **Old age**
- Insulin dependent diabetes mellitus
- Severe co-morbidities.

Mortality according to organism

- Staphylococci - 30%
- Bowel organisms - 15%
- Streptococci - 5%

Myocarditis

The short prodromal illness coupled with the development of biventricular heart failure, tachycardia, T-wave inversion and elevated troponin is most consistent with viral myocarditis. The features, including the mild flu-like illness, are consistent with Coxsackie B.

Pathology

- Lymphocytic infiltrate with focal necrosis of myocardial tissue

Causes

- In 50% of cases, no cause can be identified; hence, myocarditis is commonly idiopathic.
- In patients with an identified cause:
 - ⇒ the most commonly implicated etiology is viral (similar to pericarditis), of which enteroviruses, notably **Coxsackie B**, are the most common.
- Viral:
 - ⇒ The **most common** in adults:
 - **Parvovirus B19**
 - **Human herpes virus 6**
 - ⇒ Other Viral Causes
 - **Coxsackie B virus**
 - ❖ most common in children
 - ❖ results in **dilated** cardiomyopathy.
 - Adenovirus, HIV, Hepatitis C, Influenza virus, Epstein-Barr virus
- Bacteria: diphtheria, clostridia
- Spirochaetes: Lyme disease (most commonly presents as heart block).
- protozoa
 - ⇒ Chagas' disease,
 - caused by *Trypanosoma cruzi*, a common pathogen in South America
 - Chagas disease myocarditis results in **dilated** cardiomyopathy.
 - ⇒ Toxoplasmosis
- Noninfectious

- ⇒ Autoimmune (e.g., systemic lupus erythematosus, sarcoidosis, dermatomyositis, polymyositis), Vasculitis (e.g., Kawasaki disease)
- ⇒ Toxins (e.g., carbon monoxide poisoning, black widow venom), Cocaine.
- ⇒ Medication (e.g., sulfonamides), chemotherapy (e.g., anthracycline, doxorubicin)
- ⇒ Radiation therapy

Presentation

- usually young patient with a history of viral prodrome 2 to 3 weeks prior to the onset (fever, arthralgia, myalgia, upper respiratory tract infections)
- typically present with symptoms of heart failure (dyspnea, orthopnea, and leg swelling).
- chest pain, due to involvement of the pericardium.
- Palpitations, typically **sinus tachycardia**.

Investigations

- **Markedly raised troponin.**
- ↑ ESR (and CRP)
- ECG:
 - ⇒ sinus tachycardia or ventricular arrhythmias
 - ⇒ nonspecific ST changes
 - ⇒ diffuse ST elevation in those with pericardial involvement (perimyocarditis).
- Echocardiography: **global systolic dysfunction**

Differential diagnosis

- **Acute coronary syndrome**
 - ⇒ differentiating factors: ECG changes (NSTEMI and STEMI) with increased troponins

Treatment

- Supportive, usually similar to heart failure.
- NSAIDs should be avoided in the acute phase of acute myocarditis as it may impair healing.

DVLA: cardiovascular disorders

	Group 1 car and motorcycle	Group 2 bus and lorry
Angina	<ul style="list-style-type: none"> ▪ Must not drive when symptoms occur at rest, with emotion or at the wheel. ▪ Need not notify the DVLA. 	<ul style="list-style-type: none"> ▪ Must not drive and must notify the DVLA when symptoms occur. ▪ Driving may be relicensed if no angina for at least 6 weeks.
Acute coronary syndromes (ACS)	<ul style="list-style-type: none"> ▪ After successful coronary angioplasty: can drive after 1 week. ▪ If no successful coronary angioplasty, drive after 4 weeks ▪ Need not notify the DVLA 	<ul style="list-style-type: none"> ▪ Can drive after 6 weeks ▪ Must notify the DVLA
Coronary artery bypass graft (CABG)	<ul style="list-style-type: none"> ▪ Can drive after 4 weeks ▪ Need not notify the DVLA 	<ul style="list-style-type: none"> ▪ Can drive after 3 months ▪ Must notify the DVLA
Arrhythmia	<ul style="list-style-type: none"> ▪ Can drive if arrhythmia is controlled for at least 4 weeks. ▪ may need to notify the DVLA. 	<ul style="list-style-type: none"> ▪ Can drive if arrhythmia is controlled for at least 3 months ▪ Must notify the DVLA
Successful catheter ablation	<ul style="list-style-type: none"> ▪ May drive after 2 days ▪ Need not notify the DVLA 	<ul style="list-style-type: none"> ▪ For arrhythmia causing incapacity: can drive after 6 weeks. ▪ For arrhythmia NOT causing incapacity: can drive after 2 weeks. ▪ Must notify the DVLA
Pacemaker implant	<ul style="list-style-type: none"> ▪ Can drive after 1 week ▪ Need not notify the DVLA 	<ul style="list-style-type: none"> ▪ Can drive after 6 weeks ▪ Must notify the DVLA
CRT pacemaker	<ul style="list-style-type: none"> ▪ Can drive after 4 weeks ▪ Must notify the DVLA 	<ul style="list-style-type: none"> ▪ Can drive after 6 weeks ▪ Must notify the DVLA
Implantable cardioverter defibrillator (ICD)	<ul style="list-style-type: none"> ▪ Can drive 6 months ▪ May need to notify the DVLA. 	<ul style="list-style-type: none"> ▪ Permanent bar ▪ Must notify the DVLA
Hypertension	<ul style="list-style-type: none"> ▪ May drive and need not notify the DVLA 	<ul style="list-style-type: none"> ▪ Must not drive and must notify the DVLA if resting BP is consistently: 180 mm Hg or higher systolic and/or 100 mm Hg or more diastolic.
Heart failure	<ul style="list-style-type: none"> ▪ Asymptomatic: May drive and need not notify the DVLA. ▪ Symptomatic: Must not drive but need not notify the DVLA. ▪ Left ventricular assist device implanted: Can drive after 3 months. Need not notify the DVLA. 	<ul style="list-style-type: none"> ▪ Asymptomatic: May drive and need not notify the DVLA. ▪ Symptomatic: Must not drive and must notify the DVLA. Relicensing would require LV ejection fraction at least 40% ▪ Left ventricular assist device implanted: Licence will be refused permanently. Must notify the DVLA.

ICD means:

- cannot drive a group 1 vehicle for 6 months
- Loss of a group 2 HGV license, regardless of the circumstances

DVLA advice post MI:

- if successfully treated by angioplasty → cannot drive for 1 week
- If does not undergo angioplasty → cannot drive for 4 weeks

DVLA: cardiovascular disorders**Acute coronary syndrome**

- if successfully treated by angioplasty → cannot drive for 1 week
- If does not undergo angioplasty → cannot drive for 4 weeks

Coronary artery bypass graft (CABG)

- Group 1 car: 4 weeks off driving
- Group 2 bus and lorry: Must not drive and must notify the DVLA.

pacemaker insertion: 1 week off driving**implantable cardioverter-defibrillator (ICD):**

- if implanted for sustained ventricular arrhythmia: cease driving for 6 months.
- If implanted prophylactically then cease driving for 1 month.
- for Group 2 drivers → permanent ban

Heart failure : LVEF of < 40% bars him from driving a lorry, even if he becomes asymptomatic with treatment

successful catheter ablation for an arrhythmia: 2 days off driving

Dextrocardia

Definition

- The heart is located on the right side of the chest.

Epidemiology

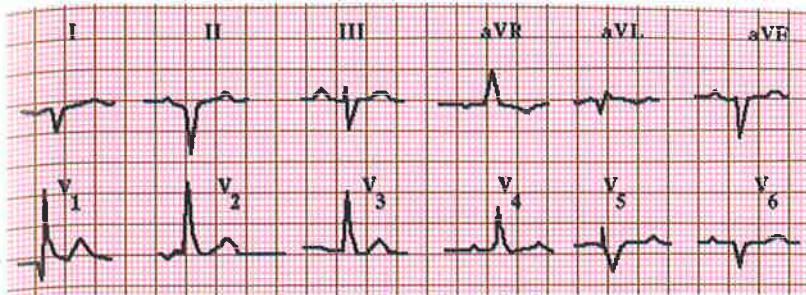
- No ethnic or gender-related predilection

Associations

- **Situs inversus totalis** (reversal in the position of other organs)
- **Kartagener syndrome:** classic triad of situs inversus (reversal in the position of the abdominal organs), recurrent sinusitis, and bronchiectasis
- When dextrocardia is associated with a normal position of other thoracoabdominal structures, it is called **situs solitus**.

ECG Features

- Right axis deviation
- Negative P wave and QRS complex in lead I.
- Upright p wave in AVL
- Reverse R wave progression across the precordium; the R wave is tallest in V1 and progressively decreases in amplitude in leads V2 to V6. The diagnosis may be confirmed by obtaining right-sided chest leads that demonstrate the normal progression of R wave amplitude.



Characteristic changes of dextrocardia include a negative P wave and QRS complex in lead 1, since atrial and ventricular depolarization begin on the left and spread to the right. There is also reverse R wave progression across the precordium; the R wave is tallest in V₁, and progressively decreases in amplitude in leads V₂ to V₆.

Differential diagnosis

- Cardiac dextroposition
 - ⇒ Dextrocardia also involves a change in the orientation of the heart with its base to the apex axis being directed to the right, in contrast to the normal heart orientation where the apex is directed to the left. This change in orientation differentiates it from cardiac dextroposition, where the heart is displaced to the right side as a result of extracardiac causes, such as a diaphragmatic hernia, right pneumonectomy, or right lung hypoplasia.

Prognosis

- Isolated dextrocardia is a benign condition often diagnosed incidentally.
- Typically, patients have a normal life expectancy if no cardiac anomalies are present.