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PAE 503

CARDIOVASCULAR AND RESPIRATORY DISORDERS

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Course code: PAE 503

Course title: Cardiovascular and Respiratory Disorders

Course credit load: 2 Units

Series 1: Foundations of Cardiovascular Evaluation

- **Part 1.1: Overview of the Cardiovascular System and Classification of Disorders**
  - Sub Part 1.1.1: Anatomy and function of the cardiovascular system
  - Sub Part 1.1.2: Epidemiology and aetiology of cardiovascular disorders in children
  - Sub Part 1.1.3: Classification of cardiovascular disorders
- **Part 1.2: General Examination of the Child with Cardiovascular Disease**
  - Sub Part 1.2.1: General inspection and growth parameters
  - Sub Part 1.2.2: Examination of the peripheral pulses
  - Sub Part 1.2.3: Blood pressure measurement technique
- **Part 1.3: Jugular Venous Pressure and Precordial Examination**
  - Sub Part 1.3.1: Anatomy and physiology of the jugular venous pulse
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  - Sub Part 1.3.3: Systematic examination of the precordium
- **Part 1.4: Diagnostic Investigations in Cardiovascular Evaluation**
  - Sub Part 1.4.1: The chest radiograph in cardiovascular diagnosis
  - Sub Part 1.4.2: Electrocardiography in cardiovascular diagnosis
  - Sub Part 1.4.3: Echocardiography in cardiovascular diagnosis

Introduction

Picture a busy paediatric outpatient clinic. A mother brings in her six month old son because he tires easily during feeding and seems to breathe faster than her other children did at the same age. Findings such as these are common triggers for a **cardiovascular evaluation** in children, and how confidently you respond to them depends entirely on how well you understand the systematic approach to assessing the cardiovascular system. This Series opens the PAE 503 course by building that foundation, moving from the basic anatomy and classification of

cardiovascular disorders through to the physical examination skills and diagnostic tools that will support everything you study in the Series that follow.

The aim of this Series is to equip you with the knowledge and clinical skills required to conduct a thorough and systematic **cardiovascular evaluation** of a child, from history and classification through to physical examination and an appreciation of the key diagnostic investigations used to confirm a diagnosis.

This Series introduces the **anatomy, epidemiology, and classification** of cardiovascular disorders in children, the **general examination** of a child with suspected cardiovascular disease, including the peripheral pulses and blood pressure measurement, the **jugular venous pressure and precordial examination**, and the **diagnostic investigations**, namely the chest radiograph, electrocardiography, and echocardiography, that support the clinical diagnosis of cardiovascular disorders in children.

Series Learning Outcomes

At the end of this Series, you should be able to:

- **SLO 1.1:** describe the cardiovascular system and its clinical relevance in the evaluation of a child,
- **SLO 1.2:** enumerate the classification of cardiovascular system disorders encountered in paediatric practice,
- **SLO 1.3:** list the general examination findings suggestive of cardiovascular disease in a child,
- **SLO 1.4:** describe the technique of peripheral pulse examination and blood pressure measurement,
- **SLO 1.5:** explain the technique and clinical significance of jugular venous pressure measurement,
- **SLO 1.6:** describe the systematic examination of the precordium,
- **SLO 1.7:** discuss the role of the chest radiograph in the diagnosis of common cardiovascular disorders, and
- **SLO 1.8:** discuss the role of electrocardiography and echocardiography in cardiovascular diagnosis.

1.1 Overview of the Cardiovascular System and Classification of Disorders

1.1.1 anatomy and function of the cardiovascular system

How blood moves through the four chambers

The **cardiovascular system** comprises the heart, blood vessels, and the blood that circulates within them, working together to deliver oxygen and nutrients to every tissue of the body while removing waste products of metabolism. The heart itself is a four chambered muscular organ made up of the **right atrium**, **right ventricle**, **left atrium**, and **left ventricle**, separated by the **interatrial septum** and **interventricular septum**. Deoxygenated blood returns from the body through the **superior vena cava** and **inferior vena cava** into the right atrium, passes through the **tricuspid valve** into the right ventricle, and is pumped through the **pulmonary valve** into the **pulmonary artery** towards the lungs.

Oxygenated blood returns from the lungs through the **pulmonary veins** into the left atrium, crosses the **mitral valve** into the left ventricle, and is ejected through the **aortic valve** into the **aorta** to supply the rest of the body. In children, the heart is proportionally larger relative to body size than in adults, and the **heart rate** is correspondingly faster, particularly in infancy, a normal difference worth remembering before you begin interpreting vital signs in a young child.

Fill in the blank: Deoxygenated blood passes from the right atrium into the right ventricle through the _____ valve.

1.1.2 epidemiology and aetiology of cardiovascular disorders in children

Cardiovascular disorders in children arise from a variety of causes, broadly grouped into **congenital** causes, present from birth due to abnormal cardiac development in utero, and **acquired** causes, which develop after birth as a result of infection, inflammation, or other insults to an initially normal heart. Congenital heart disease affects approximately 8 in every 1,000 live births worldwide, and its aetiology is often multifactorial, involving genetic factors such as **chromosomal abnormalities**, maternal factors such as poorly controlled diabetes mellitus or rubella infection during pregnancy, and environmental exposures.

Acquired cardiovascular disorders are frequently linked to infection and inflammation. In Nigeria and much of sub-Saharan Africa, **acute rheumatic fever** following untreated streptococcal throat infection remains an important cause of acquired heart disease in children, alongside conditions such as **infective endocarditis** and **Kawasaki disease**. Recognising these epidemiological patterns helps you anticipate which cardiovascular disorders are most likely in a given clinical setting.

Fill in the blank: A child presenting with fever, joint pain, and a new heart murmur several weeks after an untreated sore throat should raise suspicion for _____.

1.1.3 classification of cardiovascular disorders

Cardiovascular disorders in children are commonly classified into two broad categories, **congenital heart disease** and **acquired heart disease**. Congenital heart disease is further subdivided into **acyanotic lesions**, in which oxygenated and deoxygenated blood do not mix significantly, such as **ventricular septal defect**, **atrial septal defect**, and **persistent ductus arteriosus**, and **cyanotic lesions**, in which mixing of blood produces visible bluish discolouration of the skin, such as **tetralogy of Fallot**.

Acquired heart disease includes conditions such as **rheumatic valvular heart disease**, **cardiomyopathy**, **myocarditis**, **pericarditis**, and **infective endocarditis**, each with distinct clinical presentations but a shared potential to impair cardiac function if not identified early. A clear grasp of this classification will help you organise your clinical reasoning as you move into Series 2 and 3, which explore congenital and acquired heart diseases in much greater depth.

Fill in the blank: Congenital heart disease is subdivided into acyanotic lesions and _____ lesions.

Category	Mechanism	Examples
Congenital Heart Disease	Structural malformations present at birth	Ventricular septal defect, atrial septal defect, tetralogy of Fallot
Acquired Heart Disease	Secondary to infection, inflammation, or systemic illness	Rheumatic heart disease, infective endocarditis, myocarditis
Arrhythmias	Abnormal impulse generation or conduction	Supraventricular tachycardia, congenital long QT syndrome, Wolff-Parkinson-White syndrome
Cardiomyopathies	Primary myocardial dysfunction	Dilated cardiomyopathy, hypertrophic cardiomyopathy, restrictive cardiomyopathy
Pericardial Disorders	Inflammation or fluid accumulation around the heart	Pericarditis, pericardial effusion, cardiac tamponade
Vascular Disorders	Abnormalities of blood vessels	Kawasaki disease, coarctation of the aorta, vasculitis syndromes

Table 1.1: Classification of cardiovascular disorders in children.

Pilot questions 1.1

1. Which four chambers make up the normal human heart? (Linked to SLO 1.1)
2. State the approximate incidence of congenital heart disease among live births. (Linked to SLO 1.1)

3. Distinguish between acyanotic and cyanotic congenital heart disease. (Linked to SLO 1.2)
4. List three examples of acquired heart disease in children. (Linked to SLO 1.2)
5. Explain why acute rheumatic fever remains an important cause of acquired heart disease in Nigeria. (Linked to SLO 1.2)

1.2 General Examination of the Child with Cardiovascular Disease

1.2.1 general inspection and growth parameters

What to look for before you touch the child

A cardiovascular examination begins the moment you see the child, well before you touch a stethoscope to the chest. General inspection should note the child's **respiratory rate**, the presence or absence of **cyanosis**, **clubbing** of the fingers and toes, and any visible **chest deformity** such as a precordial bulge, which may suggest chronic cardiac enlargement. You should also assess the child's overall nutritional state, since children with significant cardiovascular disease frequently present with **failure to thrive** because increased metabolic demand from cardiac work outpaces caloric intake.

Plotting the child's weight, length or height, and head circumference on an appropriate **growth chart** is therefore essential, as a faltering growth trajectory is often one of the earliest indicators of an underlying cardiac problem, sometimes preceding the detection of a murmur. Signs of respiratory distress such as **nasal flaring**, **intercostal recession**, and **grunting** should also be documented, as these frequently accompany cardiac failure in infancy.

Fill in the blank: A faltering growth trajectory is often one of the earliest indicators of underlying _____ disease.

1.2.2 examination of the peripheral pulses

Peripheral pulse examination provides valuable clues to underlying cardiovascular pathology and should be performed systematically, comparing pulses on both the upper and lower limbs. The **radial pulse**, **brachial pulse**, and **femoral pulse** should each be palpated, noting the **rate**, **rhythm**, **volume**, and **character** of the pulse. A classic and clinically important finding is **radiofemoral delay**, in which the femoral pulse is felt distinctly later than the radial pulse or is weak or absent altogether, a sign strongly suggestive of **coarctation of the aorta**.

Pulse character can also point towards specific diagnoses: a **bounding pulse** with a wide pulse pressure may indicate a **persistent ductus arteriosus** or **aortic regurgitation**, while a **weak, thready pulse** may reflect poor cardiac output in **heart failure** or **shock**. Counting the pulse for a full minute is recommended in infants and young children because of natural beat to beat variability.

Fill in the blank: A pulse that is easily felt in the arm but weak or delayed in the femoral artery is described as _____ and should raise suspicion for coarctation of the aorta.

1.2.3 blood pressure measurement technique

Accurate **blood pressure measurement** in children depends heavily on selecting the correct **cuff size**, since a cuff that is too small will give a falsely elevated reading while one that is too large will underestimate the true blood pressure. The recommended cuff width should cover approximately 40 percent of the circumference of the upper arm, and the bladder length should encircle at least 80 to 100 percent of the arm. Blood pressure should ideally be measured in a calm, seated or supine child after several minutes of rest, as crying or agitation can significantly raise readings.

Blood pressure should be measured in all four limbs when coarctation of the aorta is suspected, since a normal finding is a slightly higher pressure in the upper limbs compared with the lower limbs; a reversal of this pattern is an important diagnostic clue. Readings should always be compared against normative values for the child's age, sex, and height.

Fill in the blank: Blood pressure cuff width should cover approximately _____ percent of the circumference of the upper arm.

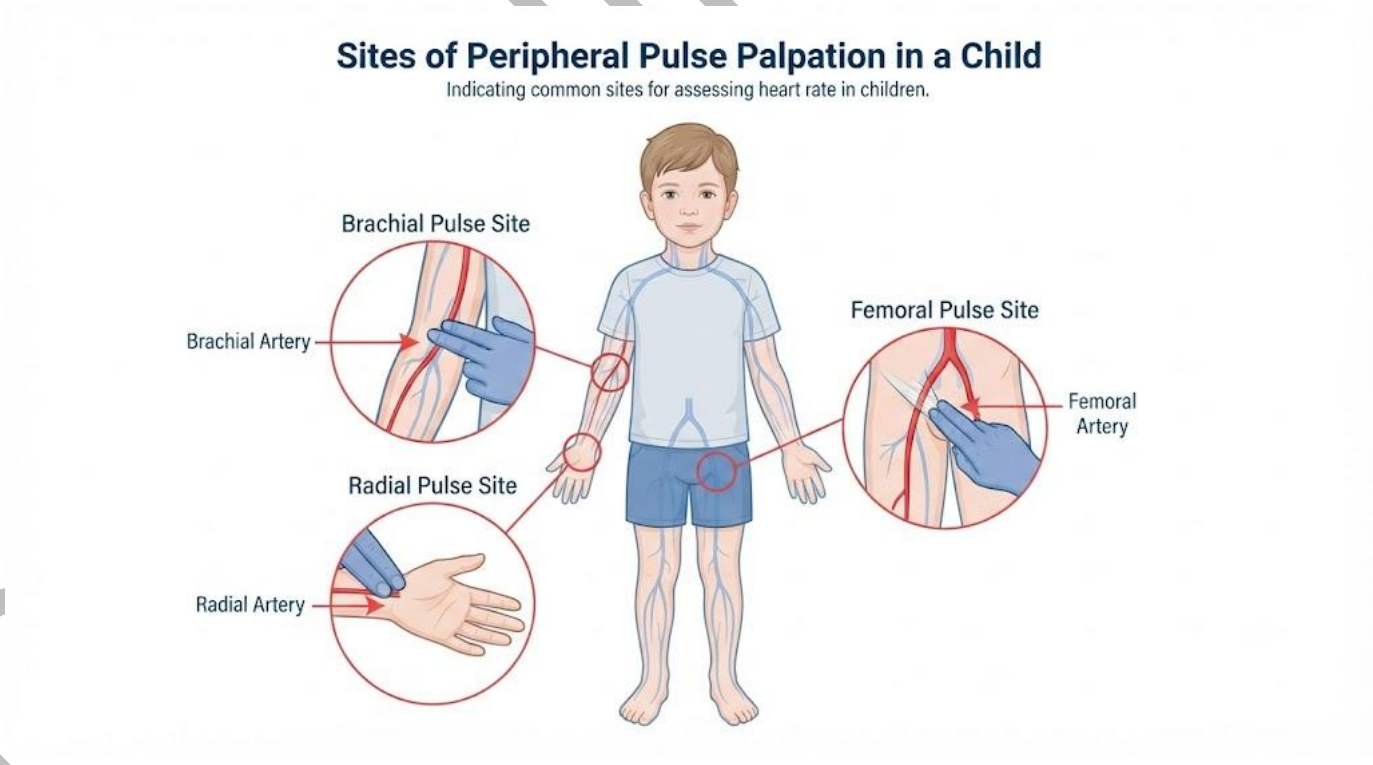

Figure 1.2: Sites of peripheral pulse palpation used in cardiovascular examination.

Pilot questions 1.2

1. List three features to note when performing general inspection of a child for cardiovascular disease. (Linked to SLO 1.3)
2. Why is plotting growth parameters important in the cardiovascular assessment of a child? (Linked to SLO 1.3)
3. Define radiofemoral delay and state its clinical significance. (Linked to SLO 1.4)
4. What determines the correct blood pressure cuff size for a child? (Linked to SLO 1.4)
5. Why should blood pressure be measured in all four limbs when coarctation of the aorta is suspected? (Linked to SLO 1.4)

1.3 Jugular Venous Pressure and Precordial Examination

1.3.1 anatomy and physiology of the jugular venous pulse

Reading the waveform in the neck

The **jugular venous pulse** reflects pressure changes within the right atrium as it fills and empties during the cardiac cycle, and is best assessed by observing the **internal jugular vein**, which lacks valves and therefore transmits atrial pressure waves directly. The normal waveform consists of three positive waves, the **a wave**, produced by atrial contraction, the **c wave**, produced by bulging of the tricuspid valve during early ventricular systole, and the **v wave**, produced by atrial filling against a closed tricuspid valve, along with the **x descent** and **y descent**.

In children, examination of the jugular venous pulse can be technically challenging because of a short neck and limited cooperation, but it remains a valuable, non-invasive window into **right atrial pressure** and, indirectly, overall cardiac function.

Fill in the blank: The a wave of the jugular venous pulse is produced by _____ contraction.

1.3.2 technique of jugular venous pressure measurement

To measure the **jugular venous pressure**, the child should be positioned reclining at approximately 45 degrees, with the head turned slightly away from the side being examined and adequate lighting directed tangentially across the neck. The examiner identifies the highest point of visible pulsation in the internal jugular vein and measures the vertical distance from this point to the **sternal angle**, a fixed anatomical reference point corresponding to approximately 5 centimetres above the centre of the right atrium.

A jugular venous pressure of more than 3 to 4 centimetres above the sternal angle is generally considered elevated and suggests **right heart failure**, **fluid overload**, or **pericardial disease**. The jugular pulse is characteristically **non-palpable**, has a **double waveform** per cardiac cycle, and its height falls with inspiration, features that help confirm you are examining the correct vessel.

Fill in the blank: A jugular venous pressure exceeding _____ centimetres above the sternal angle is considered elevated.

1.3.3 systematic examination of the precordium

Examination of the **precordium** follows the sequence of **inspection**, **palpation**, and **auscultation**. Inspection may reveal a visible **precordial bulge** in children with longstanding cardiac enlargement. Palpation begins with locating the **apex beat**, normally found in the fourth or fifth intercostal space in the midclavicular line depending on the child's age, noting whether it is displaced, diffuse, or unusually forceful. A **parasternal heave** and palpable **thrills** should also be sought, since a thrill corresponds to a loud murmur of grade 4 or greater.

Auscultation should proceed methodically across the **aortic area**, **pulmonary area**, **tricuspid area**, and **mitral area**, listening first for the **first and second heart sounds** before assessing for any **murmurs**, noting their timing, intensity, location, radiation, and character.

Fill in the blank: A palpable vibration on the chest wall corresponding to a loud murmur is called a _____.

Step
Inspection – Observe chest shape, symmetry, visible pulsations, scars, or deformities.
Palpation – Assess apex beat (location, character), thrills, heaves, and chest wall tenderness.
Auscultation – Listen systematically over valve areas for heart sounds, murmurs, added sounds, or pericardial rubs.

Box 1.3: A structured sequence for precordial examination.

Pilot questions 1.3

1. Which three waves make up the normal jugular venous pulse waveform? (Linked to SLO 1.5)
2. Describe the correct patient position for measuring jugular venous pressure. (Linked to SLO 1.5)
3. State two features that distinguish the jugular venous pulse from a carotid arterial pulse. (Linked to SLO 1.5)
4. List the four traditional auscultation areas of the precordium. (Linked to SLO 1.6)
5. Explain the clinical significance of a palpable thrill on precordial examination. (Linked to SLO 1.6)

1.4 Diagnostic Investigations in Cardiovascular Evaluation

1.4.1 the chest radiograph in cardiovascular diagnosis

Reading the cardiac silhouette

The **chest radiograph** remains a widely available and useful first-line investigation in the evaluation of a child with suspected cardiovascular disease. Key features to assess include the **cardiothoracic ratio**, considered abnormal when the cardiac shadow exceeds approximately 50 percent of the thoracic width, the overall **cardiac silhouette**, and the appearance of the **pulmonary vascular markings**, which may be increased in conditions with a left to right shunt or decreased in certain cyanotic lesions.

Specific silhouette shapes can be strongly suggestive of particular diagnoses; a **boot-shaped heart** classically suggests **tetralogy of Fallot**, while an **egg-on-a-string** appearance suggests **transposition of the great arteries**, patterns revisited in detail in Series 2. The chest radiograph also allows assessment of the lung fields for **pulmonary oedema** or **pleural effusion**.

Fill in the blank: A cardiac shadow exceeding approximately _____ percent of the thoracic width is considered an abnormal cardiothoracic ratio.

1.4.2 electrocardiography in cardiovascular diagnosis

The **electrocardiogram**, commonly abbreviated ECG, records the electrical activity of the heart and provides information on **heart rate**, **rhythm**, **chamber enlargement**, and **conduction abnormalities**. In children, normal ECG values differ considerably from those in adults and change further with age, so interpretation must always be made against age-appropriate reference ranges. Key features assessed include the **P wave**, reflecting atrial depolarisation, the **QRS complex**, reflecting ventricular depolarisation, and the **T wave**, reflecting ventricular repolarisation.

The ECG is particularly useful for detecting **arrhythmias**, **chamber hypertrophy**, and characteristic patterns associated with specific heart diseases. A prolonged **QT interval**, for example, may indicate an underlying risk of dangerous arrhythmia and warrants further specialist evaluation.

Fill in the blank: The P wave on an electrocardiogram represents _____ depolarisation.

1.4.3 echocardiography in cardiovascular diagnosis

Echocardiography uses ultrasound to produce real time images of the heart's structure and function, and it has become the single most important investigation for the definitive diagnosis of congenital and many acquired cardiovascular disorders in children. It allows direct visualisation of the cardiac chambers, valves, and great vessels, and, through **Doppler** techniques, allows assessment of blood flow direction and velocity across septal defects and valves.

Because echocardiography is non-invasive, does not involve ionising radiation, and can often be performed at the bedside, it is particularly well suited to the paediatric population and is typically the investigation that confirms a diagnosis suspected on clinical examination and supported by the chest radiograph and ECG.

Fill in the blank: Echocardiography uses _____ to produce real time images of the heart.

Investigation	Key Features	Clinical Use
Chest Radiograph	**Provides structural outline of heart and lungs; shows cardiac size, shape, pulmonary vasculature, and lung fields.**	**Detects cardiomegaly, pulmonary edema, congenital heart disease patterns, and associated lung pathology.**
ECG	**Records electrical activity of the heart; shows rhythm, conduction, and ischemic changes.**	**Diagnoses arrhythmias, conduction defects, myocardial ischemia, electrolyte disturbances.**
Echocardiography	**Ultrasound imaging of cardiac chambers, valves, and great vessels; can assess flow with Doppler.**	**Evaluates congenital and acquired heart disease, valve abnormalities, ventricular function, pericardial effusion.**

Table 1.4: Comparative role of chest radiograph, electrocardiography, and echocardiography.

Pilot questions 1.4

1. What cardiothoracic ratio on chest radiograph is generally considered abnormal in a child? (Linked to SLO 1.7)
2. Which congenital heart disease is classically associated with a boot-shaped heart on chest radiograph? (Linked to SLO 1.7)
3. What does the QRS complex on an ECG represent? (Linked to SLO 1.8)
4. Why must ECG findings in children be interpreted against age-specific reference ranges? (Linked to SLO 1.8)
5. Explain why echocardiography is considered the definitive investigation for most congenital heart diseases. (Linked to SLO 1.8)

Series Summary

This opening Series of the course introduced the **foundations of cardiovascular evaluation**, beginning with the anatomy, epidemiology, and classification of cardiovascular disorders, before

turning to the **general examination** of a child with suspected cardiovascular disease, including the peripheral pulses and blood pressure measurement.

We then examined the **jugular venous pressure and precordial examination**, before concluding with the **diagnostic investigations**, namely the chest radiograph, electrocardiogram, and echocardiogram, that support and confirm a clinical diagnosis. Having worked through this Series, you should now be able to conduct a systematic cardiovascular evaluation of a child. The next Series turns to **congenital heart diseases**.

Glossary of Terms

- **Cardiovascular system:** the heart, blood vessels, and blood, working together to circulate oxygen and nutrients throughout the body.
- **Congenital heart disease:** a structural abnormality of the heart or great vessels present from birth.
- **Acquired heart disease:** a cardiac condition that develops after birth in a heart that was initially structurally normal.
- **Cyanosis:** a bluish discolouration of the skin and mucous membranes due to low blood oxygen saturation.
- **Radiofemoral delay:** a palpable delay between the radial and femoral pulses, suggestive of coarctation of the aorta.
- **Jugular venous pressure:** the pressure within the internal jugular vein, used as a clinical estimate of right atrial pressure.
- **Sternal angle:** the fixed bony landmark used as the zero reference point for jugular venous pressure measurement.
- **Precordium:** the area of the chest wall overlying the heart.
- **Apex beat:** the lowest and most lateral point at which the cardiac impulse can be palpated.
- **Cardiothoracic ratio:** the ratio of the width of the cardiac silhouette to the width of the thoracic cage on a chest radiograph.
- **Echocardiography:** an ultrasound based investigation used to visualise cardiac structure and function.
- **Thrill:** a palpable vibration on the chest wall corresponding to a loud cardiac murmur.

Concept Check Questions [Series 1]

Objective questions

1. Deoxygenated blood passes from the right atrium to the right ventricle through the _____ valve.
2. A delay between the radial and femoral pulses is termed _____.
3. The fixed landmark used to measure jugular venous pressure is the _____.
4. A boot-shaped heart on chest radiograph is classically associated with _____.
5. The investigation that provides direct, real time visualisation of cardiac structure using ultrasound is _____.

Short-answer questions

6. Briefly distinguish between acyanotic and cyanotic congenital heart disease.
7. State two clinical features that should prompt plotting of growth parameters in a child with suspected cardiac disease.
8. Describe the correct patient position for jugular venous pressure measurement.
9. List the four traditional areas of precordial auscultation.
10. State one limitation each of the chest radiograph and the electrocardiogram in cardiovascular diagnosis.

Long-answer questions

11. Discuss the normal pathway of blood flow through the four chambers of the heart, from the venae cavae to the aorta. (Linked to SLO 1.1)
12. Describe the systematic approach to examination of the peripheral pulses in a child, including the clinical significance of an abnormal finding. (Linked to SLO 1.3, SLO 1.4)
13. Explain the technique of jugular venous pressure measurement and how an elevated reading is interpreted clinically. (Linked to SLO 1.5)
14. Discuss the role of the chest radiograph in the evaluation of a child with suspected cardiovascular disease. (Linked to SLO 1.7)
15. Compare the relative contributions of the chest radiograph, electrocardiogram, and echocardiogram to the diagnostic work-up of a child with suspected heart disease. (Linked to SLO 1.7, SLO 1.8)

Answers to Concept Check Questions [Series 1]

Objective questions

1. Tricuspid.
2. Radiofemoral delay.
3. Sternal angle.
4. Tetralogy of Fallot.
5. Echocardiography.

Short-answer questions

6. Acyanotic lesions do not cause significant mixing of blood and typically present with heart failure, whereas cyanotic lesions cause mixing of blood and produce visible cyanosis.
7. Failure to thrive or a faltering growth trajectory, and signs of respiratory distress such as nasal flaring or intercostal recession.
8. The child should be reclined at approximately 45 degrees with the head turned slightly away from the side being examined, with adequate tangential lighting on the neck.
9. The aortic area, pulmonary area, tricuspid area, and mitral area.
10. The chest radiograph cannot show intracardiac anatomy in detail, and ECG values change with age and must be interpreted against age-specific reference ranges.

Long-answer questions

11. **Basic response:** Deoxygenated blood enters the right atrium from the venae cavae, passes through the tricuspid valve into the right ventricle, and is pumped through the pulmonary valve into the pulmonary artery towards the lungs.

Value-added response: Oxygenated blood then returns via the pulmonary veins to the left atrium, crosses the mitral valve into the left ventricle, and is ejected through the aortic valve into the aorta, completing the circuit that delivers oxygenated blood to the body.

12. **Basic response:** Peripheral pulse examination involves palpating the radial, brachial, and femoral pulses, comparing their rate, rhythm, volume, and character, and checking specifically for radiofemoral delay.

Value-added response: Radiofemoral delay strongly suggests coarctation of the aorta, a bounding pulse suggests a persistent ductus arteriosus or aortic regurgitation, and a weak thready pulse suggests poor cardiac output as in heart failure.

13. **Basic response:** Jugular venous pressure is measured by observing the internal jugular vein with the child reclined at 45 degrees, identifying the highest point of pulsation, and measuring its vertical height above the sternal angle.

Value-added response: A jugular venous pressure exceeding 3 to 4 centimetres above the sternal angle is considered elevated and suggests right heart failure, fluid overload, or pericardial disease, making the measurement a rapid, non-invasive marker of right atrial pressure.

14. **Basic response:** The chest radiograph allows assessment of the cardiothoracic ratio, the shape of the cardiac silhouette, and the pulmonary vascular markings.

Value-added response: Specific silhouette shapes point towards specific diagnoses, such as a boot-shaped heart in tetralogy of Fallot, making the chest radiograph a useful first-line screening tool.

15. **Basic response:** The chest radiograph shows cardiac silhouette and pulmonary vascular markings, the electrocardiogram shows heart rate, rhythm, and chamber enlargement, and echocardiography provides direct visualisation of cardiac structure and blood flow.

Value-added response: These investigations are complementary, with the chest radiograph and ECG typically used as first-line screening tools, while echocardiography most often provides the definitive structural diagnosis that guides management.

Answers to Pilot Questions [Series 1]

Part 1.1

1. The four chambers are the right atrium, right ventricle, left atrium, and left ventricle.
2. Congenital heart disease affects approximately 8 in every 1,000 live births.
3. Acyanotic lesions do not cause significant mixing of blood and present with heart failure signs, whereas cyanotic lesions cause mixing of blood and produce visible cyanosis.
4. Examples include rheumatic valvular heart disease, cardiomyopathy, and infective endocarditis.
5. Untreated streptococcal throat infection remains common in the region, and acute rheumatic fever is a well recognised complication.

Part 1.2

1. General inspection should note respiratory rate, cyanosis, and clubbing, among other features.
2. A faltering growth trajectory is often one of the earliest indicators of underlying cardiac disease.
3. Radiofemoral delay is a palpable delay between the radial and femoral pulses, suggestive of coarctation of the aorta.
4. Cuff size is determined by the circumference and length of the child’s upper arm.
5. Blood pressure is measured in all four limbs because a reversal of the normal upper-to-lower limb pressure gradient suggests coarctation of the aorta.

Part 1.3

1. The jugular venous pulse consists of the a, c, and v waves.
2. The child should be reclined at approximately 45 degrees with the neck turned slightly away and adequate tangential lighting.
3. The jugular pulse is non-palpable and shows a double waveform per cardiac cycle, unlike the arterial pulse.

4. The four traditional auscultation areas are the aortic, pulmonary, tricuspid, and mitral areas.
5. A palpable thrill corresponds to a loud murmur of grade 4 or greater and warrants careful further evaluation.

Part 1.4

1. A cardiothoracic ratio exceeding approximately 50 percent is generally considered abnormal.
2. A boot-shaped heart is classically associated with tetralogy of Fallot.
3. The QRS complex represents ventricular depolarisation.
4. ECG values change with age, so interpretation must always use age-appropriate reference ranges.
5. Echocardiography is definitive because it directly visualises cardiac structure, valve function, and blood flow using ultrasound and Doppler techniques.

References and Attributions [Series 1]

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Figures, Plates, Tables, and Boxes

- Table 1.1: Classification of cardiovascular disorders in children. AI-generated using the prompt: "Create a clean reference table titled Classification of Cardiovascular Disorders in Children, listing category, mechanism, and examples. Simple clinical reference table style with outside border."
- Figure 1.2: Sites of peripheral pulse palpation used in cardiovascular examination. AI-generated using the prompt: "Create a professional medical diagram titled Sites of Peripheral Pulse Palpation, showing the radial, brachial, and femoral pulse points on a child. Clean clinical educational diagram style."
- Box 1.3: A structured sequence for precordial examination. AI-generated using the prompt: "Create a simple single-column reference box titled A Structured Sequence for Precordial Examination, listing inspection, palpation, and auscultation with their key findings. Clean clinical reference box style."
- Table 1.4: Comparative role of chest radiograph, electrocardiography, and echocardiography. AI-generated using the prompt: "Create a simple three column

educational table titled Chest Radiograph, ECG, and Echocardiography Compared. Textbook style, plain labelling, no watermark."

Course code: PAE 503

Course title: Cardiovascular and Respiratory Disorders

Course credit load: 2 Units

Series 2: Congenital Heart Diseases

- **Part 2.1: Acyanotic Congenital Heart Disease: Septal Defects**
  - Sub Part 2.1.1: Classification and pathophysiology of acyanotic congenital heart disease
  - Sub Part 2.1.2: Ventricular septal defect
  - Sub Part 2.1.3: Atrial septal defect
- **Part 2.2: Persistent Ductus Arteriosus and Management of Acyanotic Heart Disease**
  - Sub Part 2.2.1: Pathophysiology and clinical features of persistent ductus arteriosus
  - Sub Part 2.2.2: Investigations in acyanotic congenital heart disease
  - Sub Part 2.2.3: Medical and surgical management of acyanotic heart disease
- **Part 2.3: Cyanotic Congenital Heart Disease and Tetralogy of Fallot**
  - Sub Part 2.3.1: Classification of cyanotic congenital heart disease
  - Sub Part 2.3.2: Pathophysiology and clinical features of tetralogy of Fallot
  - Sub Part 2.3.3: Diagnostic findings in tetralogy of Fallot
- **Part 2.4: Hypercyanotic Spells and Heart Failure in Infancy and Childhood**
  - Sub Part 2.4.1: Clinical features and management of hypercyanotic spells
  - Sub Part 2.4.2: Definition and pathophysiology of heart failure in children
  - Sub Part 2.4.3: Causes by age and management of heart failure

Introduction

In the last Series, we explored the **foundations of cardiovascular evaluation**, building the examination and diagnostic skills you will now put to direct clinical use. Imagine a two month old infant brought to clinic because his mother noticed he breathes fast and sweats heavily during feeds, and on examination you hear a loud, harsh murmur at the left sternal edge. Recognising and understanding conditions such as this is the focus of this Series, which turns from general

evaluation to the **specific congenital heart diseases** most commonly encountered in paediatric practice.

The aim of this Series is to equip you with the knowledge required to classify, recognise, and understand the management of the common **acyanotic and cyanotic congenital heart diseases**, together with the clinical presentation and management of **heart failure** in infancy and childhood.

This Series introduces the **acyanotic congenital heart diseases**, ventricular septal defect and atrial septal defect, followed by **persistent ductus arteriosus** and the general principles of managing acyanotic heart disease. Next, we turn to the **cyanotic congenital heart diseases**, with particular attention to **tetralogy of Fallot**, before concluding with **hypercyanotic spells** and **heart failure** in infancy and childhood.

Series Learning Outcomes

At the end of this Series, you should be able to:

- **SLO 2.1:** classify the acyanotic congenital heart diseases,
- **SLO 2.2:** describe the pathophysiology and clinical features of ventricular septal defect and atrial septal defect,
- **SLO 2.3:** describe the pathophysiology and clinical features of persistent ductus arteriosus,
- **SLO 2.4:** discuss the medical and surgical management options for acyanotic congenital heart disease,
- **SLO 2.5:** classify the cyanotic congenital heart diseases and describe the clinical features of tetralogy of Fallot,
- **SLO 2.6:** describe the chest radiograph, ECG, and echocardiographic findings in tetralogy of Fallot,
- **SLO 2.7:** describe the clinical features and emergency management of hypercyanotic spells, and
- **SLO 2.8:** define heart failure in infancy and childhood and discuss its causes by age and management.

2.1 Acyanotic Congenital Heart Disease: Septal Defects

2.1.1 classification and pathophysiology of acyanotic congenital heart disease

Why acyanotic lesions do not cause cyanosis

Acyanotic congenital heart disease refers to structural cardiac defects in which blood shunts from the higher pressure left side of the heart to the lower pressure right side, a **left to right**

shunt, without significant mixing of deoxygenated blood into the systemic circulation, so the child does not appear cyanosed. The three most common acyanotic lesions are **ventricular septal defect**, **atrial septal defect**, and **persistent ductus arteriosus**, each differing in the anatomical location of the shunt but sharing the common physiological consequence of increased **pulmonary blood flow**.

Over time, this increased pulmonary blood flow can lead to **pulmonary hypertension** and, if left uncorrected for many years, a reversal of the shunt direction known as **Eisenmenger syndrome**, in which the child eventually becomes cyanosed because pulmonary vascular resistance exceeds systemic resistance. Early recognition and appropriate timing of intervention is therefore essential to prevent this late, largely irreversible complication.

Fill in the blank: In acyanotic congenital heart disease, blood typically shunts from the _____ side of the heart to the right side.

2.1.2 ventricular septal defect

Ventricular septal defect, commonly abbreviated VSD, is the most common congenital heart defect, resulting from a failure of complete closure of the **interventricular septum** during fetal development. The clinical presentation depends on the size of the defect: small defects are often asymptomatic and detected incidentally as a **loud pansystolic murmur** at the left lower sternal edge, while large defects present in infancy with **failure to thrive**, **tachypnoea**, **excessive sweating during feeds**, and recurrent chest infections due to significant left to right shunting and pulmonary overcirculation.

On examination, a large VSD may also produce a palpable **thrill** and signs of heart failure, including hepatomegaly. Chest radiograph typically shows **cardiomegaly** and increased pulmonary vascular markings, while echocardiography confirms the size, location, and haemodynamic significance of the defect, guiding the decision on whether surgical or catheter-based closure is required.

Fill in the blank: A _____ VSD often presents in infancy with failure to thrive and tachypnoea, unlike a small VSD which is often asymptomatic.

2.1.3 atrial septal defect

Atrial septal defect, abbreviated ASD, results from a defect in the **interatrial septum**, most commonly at the site of the **ostium secundum**. Unlike VSD, most children with ASD are **asymptomatic** in childhood because the pressure difference between the atria is relatively small, and the defect is often first detected as an incidental finding of a **soft ejection systolic murmur** at the upper left sternal edge with a **fixed, widely split second heart sound**, a classic examination finding that helps distinguish ASD from other acyanotic lesions.

If left uncorrected into adulthood, a significant ASD can eventually cause **right heart enlargement**, arrhythmias, and pulmonary hypertension, so even asymptomatic defects are

usually closed electively, either surgically or via a catheter-delivered device, typically before school age to prevent these long-term complications.

Fill in the blank: A fixed, widely split second heart sound is a classic examination finding in _____.

Parameter	Ventricular Septal Defect (VSD)	Atrial Septal Defect (ASD)
Location	Defect in interventricular septum, usually membranous or muscular portion	Defect in interatrial septum, commonly secundum type
Murmur	Harsh pansystolic murmur best heard at left lower sternal border	Soft systolic ejection murmur at left upper sternal border; wide, fixed splitting of S2
Presentation	Symptoms of heart failure in infancy (tachypnea, poor feeding, failure to thrive); may present with recurrent chest infections	Often asymptomatic in childhood; may present later with exercise intolerance, palpitations, or recurrent respiratory infections

Table 2.1: Comparison of ventricular septal defect and atrial septal defect.

Pilot questions 2.1

- Define acyanotic congenital heart disease and name its three most common examples. (Linked to SLO 2.1)
- Explain what is meant by Eisenmenger syndrome. (Linked to SLO 2.1)
- Describe the clinical presentation of a large ventricular septal defect. (Linked to SLO 2.2)
- Describe the classic auscultatory finding in atrial septal defect. (Linked to SLO 2.2)
- Explain why an asymptomatic atrial septal defect is usually still closed electively. (Linked to SLO 2.2)

2.2 Persistent Ductus Arteriosus and Management of Acyanotic Heart Disease

2.2.1 pathophysiology and clinical features of persistent ductus arteriosus

Why the ductus normally closes after birth

The **ductus arteriosus** is a normal fetal vessel connecting the **pulmonary artery** to the **aorta**, allowing blood to bypass the fluid filled fetal lungs. It normally closes functionally within the first day or two of life in response to rising oxygen tension, and anatomically over the following weeks. When it fails to close, the condition is termed **persistent ductus arteriosus**, abbreviated

PDA, allowing continuous left to right shunting of blood from the aorta into the pulmonary artery throughout the cardiac cycle.

Clinically, a PDA classically produces a **continuous, machinery-like murmur** heard best at the left infraclavicular area, and a large PDA may cause **bounding peripheral pulses** with a wide pulse pressure, together with signs of heart failure in more severe cases. PDA is particularly common in **premature infants**, in whom it can significantly complicate the course of respiratory illness in the neonatal period.

Fill in the blank: A persistent ductus arteriosus classically produces a continuous, _____-like murmur.

2.2.2 investigations in acyanotic congenital heart disease

Investigation of suspected acyanotic congenital heart disease follows the same three key modalities discussed in Series 1. The **chest radiograph** typically shows **cardiomegaly** and increased **pulmonary vascular markings** in significant shunts. The **electrocardiogram** may show evidence of **chamber enlargement** appropriate to the specific lesion, such as left ventricular hypertrophy in a large VSD or PDA, or right ventricular hypertrophy in a large ASD.

Echocardiography remains the definitive investigation, directly visualising the septal defect or the patent ductus, measuring its size, and using Doppler techniques to quantify the direction and velocity of shunt flow, information that is essential for planning the timing and type of intervention required.

Fill in the blank: In a large ventricular septal defect, the ECG may show evidence of _____ ventricular hypertrophy.

2.2.3 medical and surgical management of acyanotic heart disease

Management of acyanotic congenital heart disease depends on the size of the defect and the presence of symptoms. Small defects, particularly small VSDs and secundum ASDs, are frequently managed **conservatively** with regular follow up, since many small VSDs close spontaneously during early childhood. Symptomatic infants with heart failure may require medical treatment with **diuretics** and adequate nutritional support while awaiting definitive intervention.

Definitive management of significant defects involves either **surgical closure**, using cardiopulmonary bypass to directly repair the defect, or **catheter-based device closure**, an increasingly preferred, less invasive option for suitable ASDs, VSDs, and PDAs. A PDA can also be closed pharmacologically in preterm infants using **cyclo-oxygenase inhibitors** such as indomethacin or ibuprofen, which promote ductal closure.

Fill in the blank: Many small ventricular septal defects close _____ during early childhood without intervention.

Indication	Detailed Explanation
Heart Failure Symptoms	Persistent tachypnea, poor feeding, sweating during feeds, hepatomegaly, and failure to thrive despite optimal medical therapy.
Recurrent Respiratory Infections	Frequent chest infections due to pulmonary overcirculation from large left-to-right shunts (e.g., VSD, PDA).
Pulmonary Hypertension	Rising pulmonary artery pressures or early signs of Eisenmenger syndrome; intervention prevents irreversible vascular changes.
Growth Failure	Poor weight gain, delayed milestones, or developmental lag linked to chronic cardiac burden.
Significant Left-to-Right Shunt	Large shunt causing volume overload on ventricles and lungs, leading to cardiomegaly and pulmonary congestion.
Risk of Endocarditis	Structural lesions such as VSD or PDA predispose to bacterial endocarditis; closure reduces risk.
Progressive Chamber Enlargement	Enlargement of atria or ventricles on imaging/echo due to chronic volume overload, indicating need for correction.

Box 2.2: Indications for intervention in acyanotic congenital heart disease.

Pilot questions 2.2

1. Describe the normal function and postnatal closure of the ductus arteriosus. (Linked to SLO 2.3)
2. Describe the classic murmur of a persistent ductus arteriosus. (Linked to SLO 2.3)
3. What does echocardiography contribute to the assessment of acyanotic congenital heart disease? (Linked to SLO 2.4)
4. Under what circumstances might a small VSD be managed conservatively? (Linked to SLO 2.4)
5. Name a pharmacological option for closing a PDA in a preterm infant. (Linked to SLO 2.4)

2.3 Cyanotic Congenital Heart Disease and Tetralogy of Fallot

2.3.1 classification of cyanotic congenital heart disease

Why mixing of blood causes cyanosis

Cyanotic congenital heart disease occurs when deoxygenated blood bypasses the lungs or mixes with oxygenated blood before reaching the systemic circulation, lowering arterial oxygen saturation and producing visible **central cyanosis**. Common examples include **tetralogy of Fallot**, **transposition of the great arteries**, **tricuspid atresia**, and **total anomalous pulmonary**

venous connection, each with a distinct anatomical mechanism but the shared clinical hallmark of low oxygen saturation that does not improve significantly with supplemental oxygen.

This lack of significant improvement with oxygen, sometimes assessed formally with a **hyperoxia test**, helps distinguish cyanotic heart disease from a primary respiratory cause of cyanosis, in which oxygen saturation typically improves substantially once supplemental oxygen is given.

Fill in the blank: Central cyanosis in cyanotic congenital heart disease does not significantly improve with _____.

2.3.2 pathophysiology and clinical features of tetralogy of Fallot

Tetralogy of Fallot, the most common cyanotic congenital heart disease, comprises four anatomical features: a large **ventricular septal defect**, **pulmonary stenosis**, an **overriding aorta**, and **right ventricular hypertrophy**. The degree of pulmonary stenosis largely determines the severity of cyanosis, since a more severely obstructed right ventricular outflow tract forces a greater proportion of deoxygenated blood across the VSD into the aorta rather than towards the lungs.

Affected infants often present with cyanosis that may be mild at birth and progressively worsen over the following months as the pulmonary stenosis becomes relatively more severe with growth. On auscultation, a harsh **ejection systolic murmur** is heard at the left sternal edge, reflecting turbulent flow across the stenotic pulmonary outflow tract, and older, undiagnosed children may adopt a characteristic **squatting position** during exertion to temporarily improve their oxygen saturation.

Fill in the blank: Squatting is a characteristic posture adopted by children with undiagnosed _____ during exertion.

2.3.3 diagnostic findings in tetralogy of Fallot

The **chest radiograph** in tetralogy of Fallot classically shows a **boot-shaped heart**, or coeur en sabot, reflecting right ventricular hypertrophy with an upturned cardiac apex and reduced pulmonary vascular markings due to decreased pulmonary blood flow. The **electrocardiogram** typically shows **right ventricular hypertrophy** and **right axis deviation**, consistent with the pressure-loaded right ventricle.

Echocardiography is diagnostic, directly demonstrating all four anatomical components of the tetralogy, and is essential for planning the timing and type of surgical repair. Most children now undergo complete **surgical correction** in infancy, closing the VSD and relieving the pulmonary outflow obstruction, which has transformed the long-term outlook for this condition.

Fill in the blank: The chest radiograph in tetralogy of Fallot classically shows a _____-shaped heart.

Component	Physiological Contribution
Ventricular Septal Defect (VSD)	**Allows mixing of oxygenated and deoxygenated blood between ventricles, contributing to systemic desaturation (cyanosis).**
Pulmonary Stenosis	**Obstruction to right ventricular outflow increases right-to-left shunting across the VSD, worsening hypoxemia.**
Overriding Aorta	**Aorta receives blood from both ventricles, further mixing oxygen-poor blood into systemic circulation.**
Right Ventricular Hypertrophy	**Thickened right ventricle develops due to chronic pressure overload from pulmonary stenosis, sustaining the pathophysiology of the defect.**

Table 2.3: The four anatomical components of tetralogy of Fallot.

Pilot questions 2.3

1. Define cyanotic congenital heart disease and name two examples besides tetralogy of Fallot. (Linked to SLO 2.5)
2. Explain the purpose of the hyperoxia test. (Linked to SLO 2.5)
3. List the four anatomical components of tetralogy of Fallot. (Linked to SLO 2.5)
4. Describe the chest radiograph and ECG findings typically seen in tetralogy of Fallot. (Linked to SLO 2.6)
5. Explain why squatting temporarily improves oxygen saturation in an undiagnosed child with tetralogy of Fallot. (Linked to SLO 2.6)

2.4 Hypercyanotic Spells and Heart Failure in Infancy and Childhood

2.4.1 clinical features and management of hypercyanotic spells

Recognising a spell before it becomes an emergency

A **hypercyanotic spell**, also called a **tet spell**, is a sudden, severe worsening of cyanosis occurring in infants with tetralogy of Fallot, typically triggered by crying, feeding, or defecation, which increase systemic oxygen demand and reduce systemic vascular resistance, worsening the right to left shunt across the VSD. The infant becomes acutely more cyanosed, breathless, and irritable, and severe spells can progress to loss of consciousness or, rarely, death if not managed promptly.

Emergency management includes placing the infant in the **knee-chest position**, which increases systemic vascular resistance and reduces the right to left shunt, providing **supplemental oxygen**, and ensuring the infant remains as calm as possible, since agitation worsens the spell. If the spell does not settle promptly, **morphine** may be given to reduce respiratory drive and agitation, and intravenous fluids together with a vasoconstrictor such as **phenylephrine** may be used to further raise systemic vascular resistance.

Fill in the blank: The knee-chest position helps terminate a hypercyanotic spell by increasing systemic vascular _____.

2.4.2 definition and pathophysiology of heart failure in children

Heart failure in children is defined as the inability of the heart to meet the metabolic demands of the body, or to do so only at the cost of abnormally elevated filling pressures. Unlike in adults, where **coronary artery disease** is the dominant cause, heart failure in children most commonly results from **congenital heart disease** with a large left to right shunt, or, less commonly, from **cardiomyopathy**, severe **arrhythmia**, or **acquired valvular disease**.

The underlying mechanism typically involves either **volume overload**, as seen with a large VSD or PDA, or **pressure overload**, as seen with severe outflow tract obstruction, both of which eventually exceed the heart's compensatory capacity, leading to the characteristic clinical picture of heart failure.

Fill in the blank: In children, heart failure most commonly results from congenital heart disease with a large _____ shunt.

2.4.3 causes by age and management of heart failure

The likely cause of heart failure in a child varies considerably by age. In the **neonatal period**, causes include severe left-sided obstructive lesions such as **hypoplastic left heart syndrome** and **coarctation of the aorta**. In **infancy**, large left to right shunt lesions such as **VSD** and **PDA** predominate, since pulmonary vascular resistance has fallen enough by this age to allow significant shunting. In **older children**, acquired causes such as **rheumatic heart disease**, **cardiomyopathy**, and **myocarditis** become relatively more important.

Management of heart failure includes treating the underlying cause where possible, alongside supportive medical therapy with **diuretics** to reduce fluid overload, **ACE inhibitors** to reduce afterload, and careful attention to **nutritional support**, since the increased metabolic demand of heart failure frequently causes significant growth faltering in affected infants.

Fill in the blank: Coarctation of the aorta is an important cause of heart failure presenting in the _____ period.

Step	Details
Calm and Comfort the Child	Reduce agitation and crying, as these worsen right-to-left shunting. Hold in knee–chest position to increase systemic vascular resistance.
Administer Oxygen	Provide high-flow oxygen by mask to improve arterial saturation.
Knee–Chest Position	Flex hips and knees to increase systemic vascular resistance, reducing right-to-left shunt.
Morphine Administration	Small dose (0.1–0.2 mg/kg IV/IM) to calm child, reduce respiratory drive, and decrease catecholamine surge.
IV Fluids	Administer isotonic fluids to improve preload and right ventricular filling.
Beta-Blocker (Propranolol)	IV or oral propranolol reduces infundibular spasm and improves pulmonary blood flow.
Correct Acidosis	Sodium bicarbonate if metabolic acidosis is present, as acidosis worsens pulmonary vasoconstriction.
Consider Phenylephrine	IV phenylephrine increases systemic vascular resistance, reducing right-to-left shunting.
Prepare for Surgical/Palliative Intervention	If spells are recurrent or refractory, urgent surgical evaluation (e.g., Blalock–Taussig shunt or definitive repair).

Box 2.4: Emergency management steps for a hypercyanotic spell.

Pilot questions 2.4

1. Describe the typical triggers and clinical features of a hypercyanotic spell. (Linked to SLO 2.7)
2. Explain why the knee-chest position helps terminate a hypercyanotic spell. (Linked to SLO 2.7)
3. Define heart failure as it applies to children. (Linked to SLO 2.8)
4. List two causes of heart failure typically presenting in the neonatal period. (Linked to SLO 2.8)
5. Describe the general principles of medical management of heart failure in children. (Linked to SLO 2.8)

Series Summary

This Series examined the common **congenital heart diseases** encountered in paediatric practice, beginning with the **acyanotic lesions**, ventricular septal defect, atrial septal defect, and persistent ductus arteriosus, together with the principles of their investigation and management.

We then turned to **cyanotic congenital heart disease**, focusing on tetralogy of Fallot and its diagnostic findings, before concluding with the recognition and emergency management of **hypercyanotic spells** and an overview of **heart failure** in infancy and childhood. Having worked through this Series, you should now be able to recognise and understand the common congenital heart diseases you will encounter in practice. The next Series turns to **acquired heart conditions**.

Glossary of Terms

1. **Left to right shunt:** flow of blood from the higher pressure left side of the heart to the lower pressure right side, characteristic of acyanotic congenital heart disease.
2. **Eisenmenger syndrome:** reversal of a left to right shunt to a right to left shunt due to longstanding pulmonary hypertension.
3. **Ventricular septal defect:** a defect in the interventricular septum allowing blood to shunt between the ventricles.
4. **Atrial septal defect:** a defect in the interatrial septum allowing blood to shunt between the atria.
5. **Persistent ductus arteriosus:** failure of the fetal ductus arteriosus to close after birth.
6. **Cyanotic congenital heart disease:** a group of congenital heart defects in which deoxygenated blood reaches the systemic circulation, producing cyanosis.
7. **Tetralogy of Fallot:** the most common cyanotic congenital heart disease, comprising VSD, pulmonary stenosis, overriding aorta, and right ventricular hypertrophy.
8. **Hyperoxia test:** a test comparing oxygen saturation before and after supplemental oxygen, used to distinguish cardiac from respiratory causes of cyanosis.
9. **Hypercyanotic spell:** a sudden, severe worsening of cyanosis in a child with tetralogy of Fallot, also called a tet spell.
10. **Knee-chest position:** a positioning manoeuvre that increases systemic vascular resistance, used to help terminate a hypercyanotic spell.

11. **Heart failure:** the inability of the heart to meet the metabolic demands of the body without abnormally elevated filling pressures.
12. **Volume overload:** excess blood volume presented to a cardiac chamber, as occurs with a large left to right shunt.

Concept Check Questions [Series 2]

Objective questions

- The most common congenital heart defect overall is _____.
- A fixed, widely split second heart sound is characteristic of _____.
- A persistent ductus arteriosus classically produces a continuous, _____-like murmur.
- The chest radiograph in tetralogy of Fallot classically shows a _____-shaped heart.
- The knee-chest position helps terminate a hypercyanotic spell by increasing systemic vascular _____.

Short-answer questions

1. Briefly explain what is meant by Eisenmenger syndrome.
2. Describe the classic auscultatory finding in atrial septal defect.
3. List the four anatomical components of tetralogy of Fallot.
4. Describe two emergency management steps for a hypercyanotic spell.
5. Name two causes of heart failure typically presenting in infancy.

Long-answer questions

6. Discuss the pathophysiology and clinical presentation of a large ventricular septal defect. (Linked to SLO 2.2)
7. Compare the clinical presentation of ventricular septal defect and atrial septal defect. (Linked to SLO 2.1, SLO 2.2)
8. Discuss the pathophysiology and diagnostic findings of tetralogy of Fallot. (Linked to SLO 2.5, SLO 2.6)
9. Describe the recognition and emergency management of a hypercyanotic spell. (Linked to SLO 2.7)
10. Discuss the causes of heart failure in children by age group and outline the principles of its management. (Linked to SLO 2.8)

Answers to Concept Check Questions [Series 2]

Objective questions

11. Ventricular septal defect.
12. Atrial septal defect.
13. Machinery.
14. Boot.
15. Resistance.

Short-answer questions

16. A longstanding, uncorrected left to right shunt causes pulmonary hypertension severe enough to reverse the shunt to right to left, producing cyanosis.
17. A soft ejection systolic murmur at the upper left sternal edge with a fixed, widely split second heart sound.
18. Ventricular septal defect, pulmonary stenosis, overriding aorta, and right ventricular hypertrophy.
19. Placing the infant in the knee-chest position and providing supplemental oxygen while keeping the infant calm.
20. Large left to right shunt lesions such as ventricular septal defect and persistent ductus arteriosus.

Long-answer questions

21. **Basic response:** A large VSD allows significant left to right shunting, leading to increased pulmonary blood flow, and presents with failure to thrive, tachypnoea, excessive sweating during feeds, and recurrent chest infections.

Value-added response: A palpable thrill and hepatomegaly may also be found, and the chest radiograph shows cardiomegaly with increased pulmonary vascular markings, reflecting the degree of shunting.

22. **Basic response:** VSD typically presents with a loud pansystolic murmur and, if large, symptoms of heart failure, while ASD is usually asymptomatic and detected through a soft ejection systolic murmur with fixed splitting of the second heart sound.

Value-added response: This difference arises because the pressure gradient across a VSD is much greater than across an ASD, producing a louder murmur and earlier symptoms despite both being left to right shunts.

23. **Basic response:** Tetralogy of Fallot comprises a VSD, pulmonary stenosis, overriding aorta, and right ventricular hypertrophy, and the degree of pulmonary stenosis determines the severity of cyanosis.

Value-added response: The chest radiograph classically shows a boot-shaped heart with reduced pulmonary vascular markings, while the ECG shows right ventricular hypertrophy and right axis deviation, and echocardiography confirms all four anatomical features.

24. **Basic response:** A hypercyanotic spell presents with sudden worsening cyanosis, breathlessness, and irritability, often triggered by crying or feeding, and is managed with the knee-chest position, oxygen, and keeping the infant calm.

Value-added response: If the spell does not settle, morphine and intravenous fluids with a vasoconstrictor such as phenylephrine may be required to further raise systemic vascular resistance and terminate the spell.

25. **Basic response:** Neonatal heart failure is often due to severe left-sided obstructive lesions, infancy heart failure to large left to right shunts, and heart failure in older children to acquired causes such as rheumatic heart disease and cardiomyopathy.

Value-added response: Management combines treating the underlying cause with supportive therapy, including diuretics, ACE inhibitors, and careful nutritional support to address the growth faltering commonly seen in affected infants.

Answers to Pilot Questions [Series 2]

Part 2.1

1. Acyanotic congenital heart disease involves a left to right shunt without significant mixing of deoxygenated blood; common examples are VSD, ASD, and PDA.
2. Eisenmenger syndrome is reversal of a left to right shunt to a right to left shunt due to longstanding pulmonary hypertension, causing cyanosis.
3. A large VSD presents with failure to thrive, tachypnoea, excessive sweating during feeds, and recurrent chest infections.
4. ASD classically produces a soft ejection systolic murmur with a fixed, widely split second heart sound.
5. Even asymptomatic ASDs are closed to prevent long-term complications such as right heart enlargement, arrhythmia, and pulmonary hypertension.

Part 2.2

6. The ductus arteriosus connects the pulmonary artery to the aorta in fetal life and normally closes functionally within the first day or two after birth.

7. A PDA classically produces a continuous, machinery-like murmur at the left infraclavicular area.
8. Echocardiography directly visualises the defect, measures its size, and quantifies shunt direction and velocity using Doppler.
9. A small VSD may be managed conservatively since many close spontaneously during early childhood.
10. Cyclo-oxygenase inhibitors such as indomethacin or ibuprofen can pharmacologically close a PDA in preterm infants.

Part 2.3

11. Cyanotic congenital heart disease causes deoxygenated blood to reach the systemic circulation; examples besides tetralogy of Fallot include transposition of the great arteries and tricuspid atresia.
12. The hyperoxia test helps distinguish cardiac from respiratory causes of cyanosis based on the degree of improvement in oxygen saturation with supplemental oxygen.
13. The four components are VSD, pulmonary stenosis, overriding aorta, and right ventricular hypertrophy.
14. Chest radiograph shows a boot-shaped heart with reduced pulmonary vascular markings, and ECG shows right ventricular hypertrophy with right axis deviation.
15. Squatting increases systemic vascular resistance, temporarily reducing the right to left shunt and improving oxygen saturation.

Part 2.4

1. Hypercyanotic spells are triggered by crying, feeding, or defecation and present with sudden worsening cyanosis, breathlessness, and irritability.
2. The knee-chest position increases systemic vascular resistance, reducing the right to left shunt across the VSD.
3. Heart failure is the inability of the heart to meet the metabolic demands of the body without abnormally elevated filling pressures.
4. Hypoplastic left heart syndrome and coarctation of the aorta are important neonatal causes of heart failure.
5. Management includes diuretics, ACE inhibitors, and careful nutritional support alongside treatment of the underlying cause.

References and Attributions [Series 2]

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Figures, Plates, Tables, and Boxes

1. Table 2.1: Comparison of ventricular septal defect and atrial septal defect. AI-generated using the prompt: "Create a clean reference table titled Ventricular Septal Defect versus Atrial Septal Defect, comparing location, murmur, and presentation. Simple clinical reference table style with outside border."
2. Box 2.2: Indications for intervention in acyanotic congenital heart disease. AI-generated using the prompt: "Create a simple single-column reference box titled Indications for Intervention in Acyanotic Congenital Heart Disease. Clean clinical reference box style."
3. Table 2.3: The four anatomical components of tetralogy of Fallot. AI-generated using the prompt: "Create a clean reference table titled The Four Components of Tetralogy of Fallot, listing each feature and its physiological contribution. Simple clinical reference table style with outside border."
4. Box 2.4: Emergency management steps for a hypercyanotic spell. AI-generated using the prompt: "Create a simple single-column reference box titled Emergency Management of a Hypercyanotic Spell, listing sequential steps. Clean clinical reference box style."

Course code: PAE 503

Course title: Cardiovascular and Respiratory Disorders

Course credit load: 2 Units

Series 3: Acquired Heart Conditions

- **Part 3.1: Acute Rheumatic Fever and Rheumatic Valvular Heart Disease**
  - Sub Part 3.1.1: Aetiology and pathogenesis of acute rheumatic fever
  - Sub Part 3.1.2: Clinical features and diagnosis of acute rheumatic fever
  - Sub Part 3.1.3: Rheumatic valvular heart disease and management of acute rheumatic fever
- **Part 3.2: Infective Endocarditis and Myocarditis**
  - Sub Part 3.2.1: Clinical features and diagnosis of infective endocarditis
  - Sub Part 3.2.2: Investigation and management of infective endocarditis
  - Sub Part 3.2.3: Clinical features and management of myocarditis
- **Part 3.3: Pericarditis and Cardiomyopathy**
  - Sub Part 3.3.1: Clinical features and management of pericarditis
  - Sub Part 3.3.2: Types and clinical features of cardiomyopathy
  - Sub Part 3.3.3: Investigation and management of cardiomyopathy
- **Part 3.4: Kawasaki Disease**
  - Sub Part 3.4.1: Aetiology and epidemiology of Kawasaki disease
  - Sub Part 3.4.2: Clinical features and diagnostic criteria of Kawasaki disease
  - Sub Part 3.4.3: Management and complications of Kawasaki disease

Introduction

In the last Series, we explored the common **congenital heart diseases** encountered in paediatric practice, conditions present from birth. We now turn to a different group of cardiac conditions, those that develop after birth in a heart that was initially structurally normal. Picture an eight

year old brought to the emergency department with fever, painful swollen joints that seem to move from one joint to another, and a new heart murmur, several weeks after a sore throat that was never treated. This presentation, strongly suggestive of **acute rheumatic fever**, opens our discussion of the **acquired heart conditions** that remain an important cause of childhood cardiac morbidity in Nigeria and much of sub-Saharan Africa.

The aim of this Series is to equip you with the knowledge required to recognise, diagnose, and understand the management of the common **acquired heart conditions** in children, including acute rheumatic fever, infective endocarditis, myocarditis, pericarditis, cardiomyopathy, and Kawasaki disease.

This Series begins with **acute rheumatic fever** and its long-term consequence, **rheumatic valvular heart disease**, before turning to **infective endocarditis** and **myocarditis**. Next, we examine **pericarditis** and **cardiomyopathy**, before concluding with **Kawasaki disease**, an important vasculitis with significant cardiac complications if untreated.

Series Learning Outcomes

At the end of this Series, you should be able to:

- **SLO 3.1:** describe the aetiology and pathogenesis of acute rheumatic fever,
- **SLO 3.2:** describe the clinical features and management of acute rheumatic fever,
- **SLO 3.3:** describe the pathophysiology and clinical features of rheumatic valvular heart disease,
- **SLO 3.4:** describe the clinical features, diagnosis, and management of infective endocarditis,
- **SLO 3.5:** describe the clinical features and management of myocarditis,
- **SLO 3.6:** describe the clinical features and management of pericarditis,
- **SLO 3.7:** describe the types and clinical features of cardiomyopathy, and
- **SLO 3.8:** describe the clinical features, diagnostic criteria, and management of Kawasaki disease.

3.1 Acute Rheumatic Fever and Rheumatic Valvular Heart Disease

3.1.1 aetiology and pathogenesis of acute rheumatic fever

An autoimmune reaction, not a direct infection

Acute rheumatic fever, abbreviated ARF, is a delayed, **autoimmune inflammatory reaction** that follows infection of the throat with **Group A beta-haemolytic streptococcus**, typically occurring two to four weeks after an untreated or inadequately treated pharyngitis. The underlying mechanism is thought to involve **molecular mimicry**, in which antibodies raised against streptococcal antigens cross-react with similar proteins in human cardiac, joint, and neural tissue, producing inflammation at these sites even though the bacteria themselves are no longer present.

ARF remains an important public health problem in Nigeria and much of the developing world, where crowded living conditions, limited access to healthcare, and delayed treatment of streptococcal pharyngitis all contribute to ongoing transmission and to a failure to prevent the initial episode before it triggers this autoimmune cascade.

Fill in the blank: Acute rheumatic fever typically occurs _____ to four weeks after untreated streptococcal pharyngitis.

3.1.2 clinical features and diagnosis of acute rheumatic fever

The diagnosis of ARF is guided by the **Jones criteria**, which divide clinical and laboratory findings into **major criteria**, comprising **carditis**, **polyarthritis**, **chorea**, **erythema marginatum**, and **subcutaneous nodules**, and **minor criteria**, comprising fever, elevated inflammatory markers, prolonged PR interval on ECG, and arthralgia. A diagnosis of ARF generally requires evidence of a preceding streptococcal infection together with either two major criteria or one major and two minor criteria.

The **polyarthritis** of ARF is classically **migratory**, affecting large joints such as the knees and ankles in succession rather than simultaneously, while **carditis** is the most serious manifestation, since it can permanently damage the heart valves, most commonly the **mitral valve**, even after the acute episode resolves.

Fill in the blank: The polyarthritis of acute rheumatic fever is classically described as _____.

3.1.3 rheumatic valvular heart disease and management of acute rheumatic fever

Repeated or severe episodes of carditis during ARF can lead to permanent **rheumatic valvular heart disease**, most commonly **mitral regurgitation** in the acute phase, progressing over years to **mitral stenosis** as the valve becomes progressively scarred and thickened. The **aortic valve**

may also be affected, usually in combination with mitral valve involvement rather than in isolation.

Management of an acute episode includes **bed rest**, **anti-inflammatory therapy** with aspirin or, in severe carditis, corticosteroids, and, most importantly, **secondary prophylaxis** with regular intramuscular benzathine penicillin, continued for many years, to prevent further streptococcal infections that would otherwise trigger recurrent, cumulatively damaging episodes of carditis.

Fill in the blank: Secondary prophylaxis for acute rheumatic fever is most commonly given as regular intramuscular _____.

Major Criteria	Minor Criteria
Carditis	Fever
Polyarthritis	Arthralgia
Chorea	Elevated ESR/CRP
Erythema Marginatum	Leukocytosis
Subcutaneous Nodules	Prolonged PR Interval

Table 3.1: The Jones criteria for the diagnosis of acute rheumatic fever.

Pilot questions 3.1

- Explain the mechanism by which acute rheumatic fever develops after streptococcal pharyngitis. (Linked to SLO 3.1)
- List the major criteria used in the Jones criteria for diagnosing acute rheumatic fever. (Linked to SLO 3.2)
- Describe the typical pattern of joint involvement in acute rheumatic fever. (Linked to SLO 3.2)
- Which cardiac valve is most commonly affected by rheumatic valvular heart disease? (Linked to SLO 3.3)
- Describe the role of secondary prophylaxis in the management of acute rheumatic fever. (Linked to SLO 3.3)

3.2 Infective Endocarditis and Myocarditis

3.2.1 clinical features and diagnosis of infective endocarditis

Why children with structural heart disease are at higher risk

Infective endocarditis is an infection of the **endocardium**, most often affecting the heart valves, and occurs disproportionately in children with pre-existing structural heart disease, such as unrepaired congenital heart defects or rheumatic valvular damage, since abnormal or turbulent blood flow across a diseased valve provides a surface on which circulating bacteria can more easily adhere and form a **vegetation**.

Clinical features include persistent **fever**, a **new or changing heart murmur**, and, in some cases, peripheral stigmata such as **splinter haemorrhages** under the nails, tender **Osler nodes** on the fingers or toes, and painless **Janeway lesions** on the palms or soles, reflecting small emboli or immune complex deposition arising from the infected valve.

Fill in the blank: Infective endocarditis occurs disproportionately in children with pre-existing _____ heart disease.

3.2.2 investigation and management of infective endocarditis

Diagnosis of infective endocarditis relies on the **modified Duke criteria**, which combine microbiological evidence, most importantly **positive blood cultures**, with echocardiographic evidence of a **vegetation** or new valvular regurgitation, alongside supporting clinical features. Multiple sets of blood cultures should be obtained before starting antibiotics wherever possible, since identifying the causative organism is essential for guiding targeted therapy.

Management requires **prolonged intravenous antibiotic therapy**, typically for four to six weeks, guided by the specific organism identified and its sensitivity pattern, and severe cases with significant valvular destruction or heart failure may ultimately require **surgical valve repair or replacement** in addition to antibiotic treatment.

Fill in the blank: Diagnosis of infective endocarditis relies on the modified _____ criteria.

3.2.3 clinical features and management of myocarditis

Myocarditis is inflammation of the heart muscle itself, most commonly caused by a **viral infection**, though it may also result from autoimmune or toxic processes. Clinical presentation is highly variable, ranging from mild, self-limiting illness with non-specific symptoms such as fatigue and chest discomfort, to fulminant **heart failure**, life-threatening **arrhythmias**, or **cardiogenic shock** in severe cases, making a high index of suspicion essential in a child with unexplained cardiac symptoms following a recent viral illness.

Management is largely **supportive**, focusing on treating heart failure or arrhythmias as they arise, with **intensive care support**, including mechanical circulatory support in the most severe cases, since no specific antiviral treatment reliably alters the course of most cases of viral myocarditis.

Fill in the blank: Myocarditis is most commonly caused by a _____ infection.

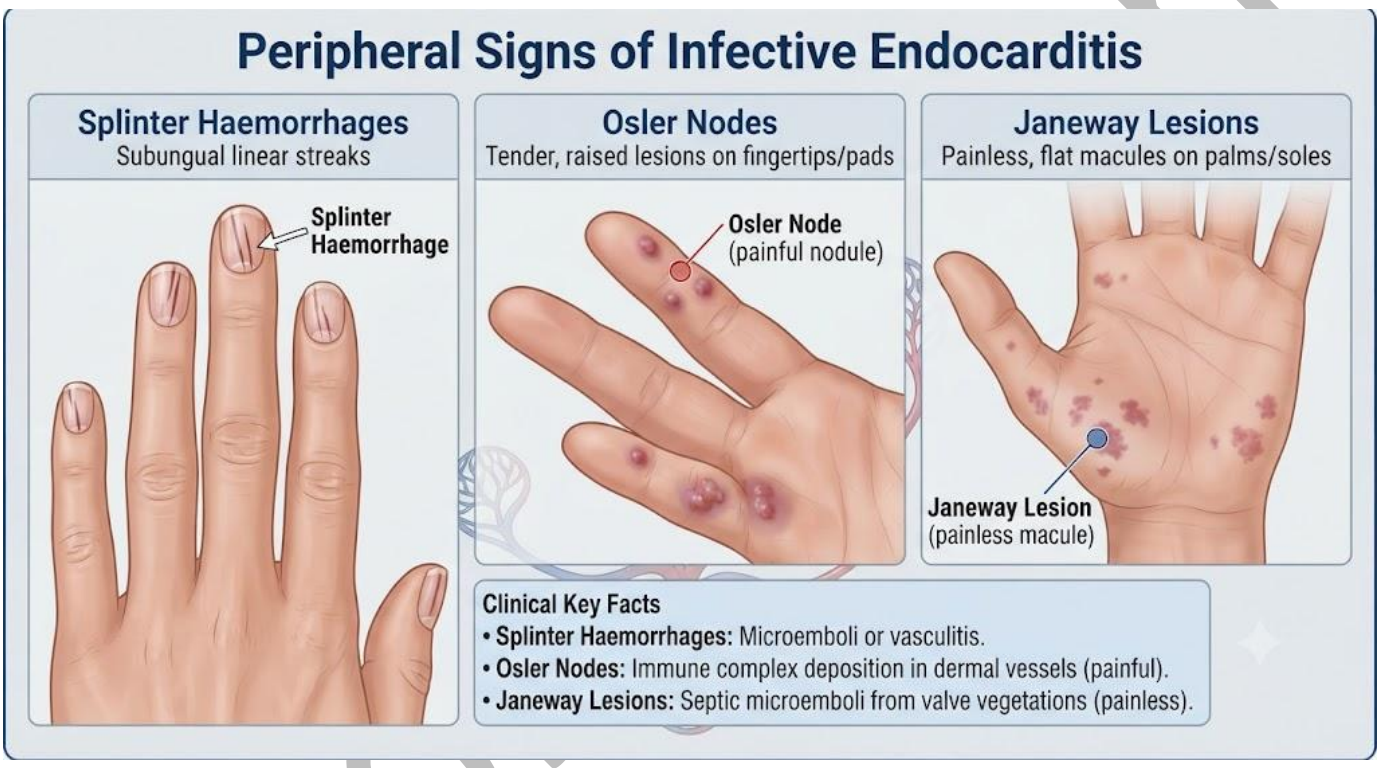

Figure 3.2: Peripheral clinical signs of infective endocarditis.

Pilot questions 3.2

1. Explain why children with structural heart disease are at higher risk of infective endocarditis. (Linked to SLO 3.4)
2. List two peripheral clinical signs of infective endocarditis. (Linked to SLO 3.4)
3. Describe the role of blood cultures in the diagnosis and management of infective endocarditis. (Linked to SLO 3.4)
4. What is the most common cause of myocarditis in children? (Linked to SLO 3.5)
5. Describe the range of clinical presentations of myocarditis. (Linked to SLO 3.5)

3.3 Pericarditis and Cardiomyopathy

3.3.1 clinical features and management of pericarditis

Recognising chest pain that changes with position

Pericarditis is inflammation of the **pericardium**, the fibrous sac surrounding the heart, most commonly caused by a **viral infection**, though bacterial, tuberculous, and autoimmune causes also occur. The classic clinical feature is **sharp, pleuritic chest pain** that characteristically worsens on lying flat and improves on sitting forward, often accompanied by a **pericardial friction rub** heard on auscultation, a scratchy sound produced by the inflamed pericardial surfaces rubbing against each other.

A significant complication of pericarditis is **pericardial effusion**, an accumulation of fluid within the pericardial sac, which, if it develops rapidly or becomes large, can progress to **cardiac tamponade**, a life-threatening emergency in which the fluid compresses the heart and impairs its ability to fill, requiring urgent **pericardiocentesis** to drain the fluid and relieve the pressure.

Fill in the blank: Pericarditis classically causes sharp chest pain that worsens on lying flat and improves on sitting _____.

3.3.2 types and clinical features of cardiomyopathy

Cardiomyopathy refers to a group of diseases affecting the heart muscle itself, broadly classified into **dilated cardiomyopathy**, in which the ventricles become enlarged and weakened, the most common type in children and a frequent cause of heart failure, **hypertrophic cardiomyopathy**, in which the ventricular wall becomes abnormally thickened and can be associated with sudden cardiac death during exertion, and **restrictive cardiomyopathy**, a rarer form in which the ventricular walls become stiff and impair filling despite normal wall thickness.

Clinical features depend on the type but often include symptoms of **heart failure**, such as breathlessness and poor growth, or, particularly in hypertrophic cardiomyopathy, **exertional syncope** or a family history of **sudden cardiac death**, which should always prompt careful cardiac evaluation of a child before they are cleared for competitive sporting activity.

Fill in the blank: The most common type of cardiomyopathy in children is _____ cardiomyopathy.

3.3.3 investigation and management of cardiomyopathy

Investigation of suspected cardiomyopathy includes **echocardiography**, which assesses chamber size, wall thickness, and ventricular function, an **electrocardiogram**, which may show chamber hypertrophy or arrhythmia, and, in selected cases, **cardiac MRI** or **genetic testing**, since many cardiomyopathies, particularly hypertrophic cardiomyopathy, have an identifiable inherited genetic basis with implications for screening family members.

Management is guided by the type and severity of disease, typically involving **medical therapy for heart failure** in dilated cardiomyopathy, activity restriction and, in some cases, an **implantable cardioverter-defibrillator** in hypertrophic cardiomyopathy at high risk of sudden death, and, in the most severe or refractory cases of any type, consideration of **cardiac transplantation**.

Fill in the blank: Genetic testing is particularly relevant in _____ cardiomyopathy, which often has an inherited basis.

Type
Dilated Cardiomyopathy – Characterized by dilated ventricles with impaired systolic function; presents with heart failure, arrhythmias, and risk of thromboembolism.
Hypertrophic Cardiomyopathy – Marked by thickened ventricular walls, often asymmetric septal hypertrophy; may cause outflow obstruction, syncope, or sudden cardiac death.
Restrictive Cardiomyopathy – Stiff ventricular walls with impaired diastolic filling; presents with hepatomegaly, ascites, and signs of right-sided heart failure.

Box 3.3: Distinguishing features of the three main types of cardiomyopathy.

Pilot questions 3.3

1. Describe the classic chest pain of pericarditis and the finding heard on auscultation. (Linked to SLO 3.6)
2. Explain the relationship between pericarditis, pericardial effusion, and cardiac tamponade. (Linked to SLO 3.6)
3. List the three main types of cardiomyopathy. (Linked to SLO 3.7)
4. Which type of cardiomyopathy is most associated with sudden cardiac death during exertion? (Linked to SLO 3.7)
5. Describe the general principles of investigating a child with suspected cardiomyopathy. (Linked to SLO 3.7)

3.4 Kawasaki Disease

3.4.1 aetiology and epidemiology of Kawasaki disease

A vasculitis of early childhood

Kawasaki disease is an acute **vasculitis** of unknown cause, predominantly affecting children under five years of age, that involves inflammation of medium-sized arteries throughout the body, with the **coronary arteries** being of greatest clinical concern because of the risk of **coronary artery aneurysm** if the condition is not recognised and treated promptly. Although the precise cause remains unknown, an abnormal immune response to an infectious trigger in a genetically susceptible child is the leading hypothesis.

Kawasaki disease is now recognised as one of the most common causes of **acquired heart disease** in children in many parts of the world, and prompt recognition is essential, since the risk of coronary artery complications is dramatically reduced when appropriate treatment is given within the first ten days of illness.

Fill in the blank: Kawasaki disease predominantly affects children under _____ years of age.

3.4.2 clinical features and diagnostic criteria of Kawasaki disease

The diagnosis of classic Kawasaki disease requires **fever for five days or more**, together with at least four of five principal clinical features: **bilateral non-purulent conjunctivitis**, changes in the **lips and oral cavity** such as redness, cracking, or a strawberry tongue, changes in the **extremities** such as redness or swelling of the hands and feet followed later by peeling, a **polymorphous rash**, and **cervical lymphadenopathy**, usually unilateral and involving a single enlarged node.

Some children present with **incomplete Kawasaki disease**, in which fever and fewer than four of the principal features are present, a pattern that is particularly common in infants and requires a high index of clinical suspicion together with supportive laboratory and echocardiographic findings to reach the diagnosis and avoid missing a child at risk of coronary complications.

Fill in the blank: Classic Kawasaki disease requires fever for _____ days or more, together with at least four principal features.

3.4.3 management and complications of Kawasaki disease

The mainstay of treatment for Kawasaki disease is **intravenous immunoglobulin**, given as a single infusion, together with **high-dose aspirin**, which is later continued at a lower, antiplatelet dose once the fever has settled. Treatment within the first ten days of illness significantly reduces the risk of **coronary artery aneurysm**, the most feared complication, which can lead to myocardial infarction or sudden death later in life if severe and unrecognised.

All children with Kawasaki disease require **serial echocardiography**, typically at diagnosis, at two weeks, and at six to eight weeks, to monitor for the development of coronary artery abnormalities, and children found to have significant aneurysms require long-term cardiology follow up and, in some cases, ongoing anticoagulation to reduce the risk of thrombosis within the aneurysm.

Fill in the blank: The mainstay of treatment for Kawasaki disease is intravenous _____.

Feature
Fever ≥ 5 days – **Persistent high-grade fever not explained by other causes.**
Conjunctival Injection – **Bilateral, non-purulent conjunctivitis.**
Oral Changes – **Red cracked lips, strawberry tongue, diffuse erythema of oral mucosa.**
Peripheral Changes – **Erythema of palms/soles, indurative edema, periungual desquamation.**
Polymorphous Rash – **Generalized rash, variable in appearance, not vesicular.**
Cervical Lymphadenopathy – **Usually unilateral, >1.5 cm diameter.**

Box 3.4: The principal diagnostic features of classic Kawasaki disease.

Pilot questions 3.4

1. Explain why prompt recognition of Kawasaki disease is clinically important. (Linked to SLO 3.8)
2. List the five principal clinical features used in the diagnosis of classic Kawasaki disease. (Linked to SLO 3.8)
3. Describe what is meant by incomplete Kawasaki disease. (Linked to SLO 3.8)
4. Describe the first-line treatment of Kawasaki disease. (Linked to SLO 3.8)
5. Explain why serial echocardiography is required in children with Kawasaki disease. (Linked to SLO 3.8)

Series Summary

This Series examined the common **acquired heart conditions** encountered in paediatric practice, beginning with **acute rheumatic fever** and its long-term consequence, rheumatic valvular heart disease, before turning to **infective endocarditis** and **myocarditis**.

We then examined **pericarditis** and **cardiomyopathy**, before concluding with **Kawasaki disease** and its potentially serious coronary complications. Having worked through this Series, you should now be able to recognise and understand the management of the common acquired heart conditions affecting children. The next Series turns to **respiratory system assessment**.

Glossary of Terms

1. **Acute rheumatic fever:** a delayed autoimmune inflammatory reaction following streptococcal pharyngitis, affecting the heart, joints, and central nervous system.
2. **Jones criteria:** the major and minor clinical and laboratory criteria used to diagnose acute rheumatic fever.
3. **Rheumatic valvular heart disease:** permanent valvular damage, most commonly of the mitral valve, resulting from repeated or severe carditis in acute rheumatic fever.
4. **Infective endocarditis:** infection of the endocardium, most often affecting the heart valves, particularly in children with pre-existing structural heart disease.
5. **Modified Duke criteria:** the diagnostic criteria combining microbiological, echocardiographic, and clinical findings used to diagnose infective endocarditis.
6. **Myocarditis:** inflammation of the heart muscle, most commonly caused by viral infection.
7. **Pericarditis:** inflammation of the pericardium, classically causing chest pain that worsens on lying flat.
8. **Cardiac tamponade:** compression of the heart by a rapidly accumulating or large pericardial effusion, impairing cardiac filling.
9. **Cardiomyopathy:** a group of diseases affecting the heart muscle, classified as dilated, hypertrophic, or restrictive.
10. **Kawasaki disease:** an acute vasculitis of early childhood affecting medium-sized arteries, particularly the coronary arteries.
11. **Coronary artery aneurysm:** abnormal dilation of a coronary artery, the most feared complication of untreated Kawasaki disease.

12. **Intravenous immunoglobulin:** the first-line treatment for Kawasaki disease, used to reduce the risk of coronary artery complications.

Concept Check Questions [Series 3]

Objective questions

- Acute rheumatic fever typically follows untreated infection with Group A beta-haemolytic _____.
- The polyarthritis of acute rheumatic fever is classically described as _____.
- Diagnosis of infective endocarditis relies on the modified _____ criteria.
- The most common type of cardiomyopathy in children is _____ cardiomyopathy.
- Classic Kawasaki disease requires fever for _____ days or more, together with at least four principal features.

Short-answer questions

1. List the major criteria of the Jones criteria for acute rheumatic fever.
2. Describe two peripheral clinical signs of infective endocarditis.
3. Explain the relationship between pericarditis, pericardial effusion, and cardiac tamponade.
4. List the three main types of cardiomyopathy.
5. Describe the first-line treatment of Kawasaki disease.

Long-answer questions

6. Discuss the pathogenesis, diagnosis, and management of acute rheumatic fever. (Linked to SLO 3.1, SLO 3.2)
7. Describe the clinical features, diagnosis, and management of infective endocarditis. (Linked to SLO 3.4)
8. Compare the clinical features and typical presentation of pericarditis and myocarditis. (Linked to SLO 3.5, SLO 3.6)
9. Discuss the classification and clinical features of cardiomyopathy. (Linked to SLO 3.7)
10. Discuss the diagnostic criteria, management, and potential complications of Kawasaki disease. (Linked to SLO 3.8)

Answers to Concept Check Questions [Series 3]

Objective questions

11. Streptococcus.
12. Migratory.
13. Duke.
14. Dilated.
15. Five.

Short-answer questions

16. Carditis, polyarthritis, chorea, erythema marginatum, and subcutaneous nodules.
17. Splinter haemorrhages, Osler nodes, and Janeway lesions.
18. Pericarditis can cause a pericardial effusion, which, if large or rapidly accumulating, can progress to cardiac tamponade, a life-threatening compression of the heart.
19. Dilated, hypertrophic, and restrictive cardiomyopathy.
20. Intravenous immunoglobulin together with high-dose aspirin.

Long-answer questions

21. **Basic response:** Acute rheumatic fever follows streptococcal pharyngitis through a molecular mimicry mechanism, is diagnosed using the Jones criteria, and is managed with anti-inflammatory therapy and secondary prophylaxis.

Value-added response: Secondary prophylaxis with regular intramuscular benzathine penicillin is the most important long-term measure, since it prevents further streptococcal infections that would otherwise cause cumulative valvular damage.

22. **Basic response:** Infective endocarditis presents with persistent fever and a new or changing murmur, is diagnosed using the modified Duke criteria combining blood cultures and echocardiography, and is managed with prolonged intravenous antibiotics.

Value-added response: Children with pre-existing structural heart disease are at particular risk, and severe cases with significant valvular destruction may require surgical repair or replacement in addition to antibiotics.

23. **Basic response:** Pericarditis causes sharp chest pain worsened by lying flat with a pericardial friction rub, while myocarditis presents more variably, from mild fatigue to fulminant heart failure or arrhythmia.

Value-added response: Both are most commonly viral in origin, but pericarditis can progress to tamponade requiring urgent pericardiocentesis, whereas myocarditis management is largely supportive, focused on treating heart failure and arrhythmia as they arise.

24. **Basic response:** Cardiomyopathy is classified as dilated, the most common type in children and associated with heart failure, hypertrophic, associated with sudden cardiac death during exertion, and restrictive, the rarest form.

Value-added response: Investigation includes echocardiography, ECG, and, in selected cases, genetic testing, since hypertrophic cardiomyopathy in particular often has an identifiable inherited basis with implications for family screening.

25. **Basic response:** Kawasaki disease is diagnosed using fever for five days or more with at least four of five principal features, and is treated with intravenous immunoglobulin and high-dose aspirin within the first ten days of illness.

Value-added response: Serial echocardiography at diagnosis, two weeks, and six to eight weeks is required to monitor for coronary artery aneurysm, the most feared complication, which can cause myocardial infarction or sudden death if severe and unrecognised.

Answers to Pilot Questions [Series 3]

Part 3.1

1. Molecular mimicry causes antibodies raised against streptococcal antigens to cross-react with cardiac, joint, and neural tissue.
2. The major criteria are carditis, polyarthritis, chorea, erythema marginatum, and subcutaneous nodules.
3. The polyarthritis is classically migratory, affecting large joints such as the knees and ankles in succession.
4. The mitral valve is most commonly affected by rheumatic valvular heart disease.
5. Secondary prophylaxis with regular intramuscular benzathine penicillin prevents further streptococcal infections that would otherwise cause recurrent, cumulative valvular damage.

Part 3.2

6. Abnormal or turbulent blood flow across a diseased valve provides a surface for circulating bacteria to adhere and form a vegetation.
7. Splinter haemorrhages, Osler nodes, and Janeway lesions are peripheral clinical signs.
8. Blood cultures identify the causative organism, which is essential for guiding targeted antibiotic therapy.
9. Viral infection is the most common cause of myocarditis in children.
10. Presentation ranges from mild, self-limiting illness to fulminant heart failure, arrhythmia, or cardiogenic shock.

Part 3.3

11. Pericarditis causes sharp, pleuritic chest pain that worsens on lying flat and improves on sitting forward, with a pericardial friction rub on auscultation.
12. Pericarditis can cause a pericardial effusion, which, if large or rapid, can progress to cardiac tamponade, requiring urgent pericardiocentesis.
13. The three main types are dilated, hypertrophic, and restrictive cardiomyopathy.
14. Hypertrophic cardiomyopathy is most associated with sudden cardiac death during exertion.
15. Investigation includes echocardiography, ECG, and, in selected cases, cardiac MRI or genetic testing.

Part 3.4

1. Prompt treatment within the first ten days of illness dramatically reduces the risk of coronary artery aneurysm.
2. The five features are conjunctivitis, oral or lip changes, extremity changes, rash, and cervical lymphadenopathy.
3. Incomplete Kawasaki disease refers to fever with fewer than four of the principal features, common in infants.
4. First-line treatment is intravenous immunoglobulin together with high-dose aspirin.
5. Serial echocardiography monitors for the development of coronary artery abnormalities, the most feared complication.

References and Attributions [Series 3]

1. Kliegman, R. M., St Geme, J. W., Blum, N. J., Shah, S. S., Tasker, R. C. and Wilson, K. M. (2020). Nelson Textbook of Pediatrics (21st ed.). Elsevier.
2. World Health Organization (2018). Rheumatic Fever and Rheumatic Heart Disease. WHO Press.
3. McCrindle, B. W. et al. (2017). Diagnosis, Treatment, and Long-Term Management of Kawasaki Disease. Circulation, 135(17), e927 to e999.
4. Baltimore, R. S. et al. (2015). Infective Endocarditis in Childhood. Circulation, 132(15), 1487 to 1515.

Figures, Plates, Tables, and Boxes

1. Table 3.1: The Jones criteria for the diagnosis of acute rheumatic fever. AI-generated using the prompt: "Create a clean reference table titled The Jones Criteria for Acute Rheumatic Fever, listing major criteria and minor criteria in two columns. Simple clinical reference table style with outside border."
2. Figure 3.2: Peripheral clinical signs of infective endocarditis. AI-generated using the prompt: "Create a professional medical illustration titled Peripheral Signs of Infective Endocarditis, showing splinter haemorrhages, Osler nodes, and Janeway lesions with clear labels. Clean clinical educational diagram style."
3. Box 3.3: Distinguishing features of the three main types of cardiomyopathy. AI-generated using the prompt: "Create a simple single-column reference box titled Types of Cardiomyopathy Compared, listing dilated, hypertrophic, and restrictive cardiomyopathy with key features. Clean clinical reference box style."
4. Box 3.4: The principal diagnostic features of classic Kawasaki disease. AI-generated using the prompt: "Create a simple single-column reference box titled Principal Diagnostic Features of Kawasaki Disease, listing the five clinical features. Clean clinical reference box style."

Course code: PAE 503

Course title: Cardiovascular and Respiratory Disorders

Course credit load: 2 Units

Series 4: Respiratory System Assessment

- **Part 4.1: Anatomy, Physiology, and History Taking in Respiratory Assessment**
  - Sub Part 4.1.1: Brief anatomy and physiology of the respiratory system
  - Sub Part 4.1.2: Basic principles of history taking for respiratory complaints
  - Sub Part 4.1.3: Communication considerations when taking a respiratory history
- **Part 4.2: General Physical Examination of the Respiratory System**
  - Sub Part 4.2.1: Steps in general physical examination
  - Sub Part 4.2.2: Recognising signs of respiratory distress
  - Sub Part 4.2.3: Examination of the upper airway and associated structures
- **Part 4.3: Systematic Examination Technique of the Chest**
  - Sub Part 4.3.1: Inspection and palpation of the chest
  - Sub Part 4.3.2: Percussion of the chest
  - Sub Part 4.3.3: Auscultation and breath sounds
- **Part 4.4: Investigations and Communication in Respiratory Assessment**
  - Sub Part 4.4.1: Investigations used in respiratory assessment
  - Sub Part 4.4.2: The role of communication in respiratory assessment
  - Sub Part 4.4.3: Integrating history, examination, and investigation findings

Introduction

In the last Series, we explored the common **acquired heart conditions** encountered in paediatric practice. We now turn to a different organ system, the respiratory system, beginning with its evaluation. Picture a two year old brought to the emergency department breathing rapidly, with visible chest indrawing and a soft grunt at the end of each breath. Recognising the significance of these findings, and knowing how to systematically evaluate a child with a respiratory complaint, is the focus of this Series.

The aim of this Series is to equip you with the knowledge and clinical skills required to take a focused respiratory history, perform a systematic general and chest examination, and understand the investigations used in the assessment of a child with a respiratory complaint.

This Series begins with the **anatomy, physiology, and history taking** relevant to respiratory assessment, before turning to the **general physical examination** of the respiratory system, including the recognition of respiratory distress. Next, we examine the **systematic technique** of chest examination, before concluding with the **investigations and communication principles** relevant to respiratory assessment.

Series Learning Outcomes

At the end of this Series, you should be able to:

- **SLO 4.1:** describe the basic anatomy and physiology of the respiratory system in children,
- **SLO 4.2:** describe the basic principles of history taking for a child with a respiratory complaint,
- **SLO 4.3:** list the steps in general physical examination relating to the respiratory system,
- **SLO 4.4:** describe the signs of respiratory distress found on general examination,
- **SLO 4.5:** describe the systematic technique of chest examination, including inspection, palpation, percussion, and auscultation,
- **SLO 4.6:** differentiate normal from abnormal breath sounds,
- **SLO 4.7:** describe the investigations used in respiratory assessment, and
- **SLO 4.8:** discuss the principles of communication when assessing a child with respiratory illness.

4.1 Anatomy, Physiology, and History Taking in Respiratory Assessment

4.1.1 brief anatomy and physiology of the respiratory system

Why a child's airway behaves differently from an adult's

The respiratory system comprises the **upper airway**, consisting of the nose, pharynx, and larynx, and the **lower airway**, consisting of the trachea, bronchi, bronchioles, and alveoli, where **gas exchange** takes place. In children, particularly infants, the airway is proportionally **narrower**, the tongue is relatively **larger**, and the chest wall is more **compliant**, or flexible, than in adults,

meaning children are more prone to airway obstruction and show more visible chest wall movement, such as indrawing, when work of breathing increases.

Children also have a higher **metabolic rate** and oxygen consumption relative to body weight than adults, and a correspondingly faster **normal respiratory rate**, which falls progressively with age. Understanding these normal anatomical and physiological differences is essential before you can accurately interpret respiratory findings in a child of any age.

Fill in the blank: In children, the chest wall is more _____ than in adults, producing more visible movement during increased work of breathing.

4.1.2 basic principles of history taking for respiratory complaints

A focused respiratory history should establish the nature of the presenting complaint, whether **cough**, **breathlessness**, **wheeze**, **stridor**, or **noisy breathing**, together with its **onset**, **duration**, and **pattern**, for example whether cough is worse at night or with exertion. Associated symptoms such as **fever**, **poor feeding**, or **cyanotic episodes** should be actively sought, as should any history of choking, suggesting a possible **foreign body aspiration**.

A relevant **past medical history**, including prematurity, previous respiratory illnesses, and any known **atopic conditions** such as eczema or allergic rhinitis, together with a **family history** of asthma or atopy and details of the child's **immunisation status** and **home environment**, including exposure to tobacco smoke or biomass fuel, all contribute important context to the assessment.

Fill in the blank: A history of choking should raise suspicion for possible foreign body _____.

4.1.3 communication considerations when taking a respiratory history

As with any paediatric history, most of the information will come from the **parent or caregiver**, particularly in younger children, but older children and adolescents should still be given the opportunity to describe their own symptoms wherever possible, since this both respects their autonomy and can reveal details a parent may not have noticed, such as chest tightness or difficulty keeping up with peers during play.

Clinicians should remain alert to the **emotional impact** of chronic or severe respiratory symptoms on the family, particularly in conditions such as asthma that require ongoing management, and should use **clear, jargon-free language** when explaining findings and next steps, checking the caregiver's understanding rather than assuming that technical terms such as wheeze or stridor are already familiar to them.

Fill in the blank: Older children and adolescents should be given the opportunity to describe their own _____ wherever possible.

Table 4.1: Normal respiratory rate ranges in children by age.

Age group	Normal respiratory rate (breaths per minute)
Newborn to 1 year	30 to 60
1 to 3 years	24 to 40
3 to 6 years	22 to 34
6 to 12 years	18 to 30
Over 12 years	12 to 20

Pilot questions 4.1

- Describe two anatomical differences between a child's airway and an adult's airway. (Linked to SLO 4.1)
- Why do children have a faster normal respiratory rate than adults? (Linked to SLO 4.1)
- List the key features to establish when taking a history of cough in a child. (Linked to SLO 4.2)
- Why should a history of choking raise particular concern? (Linked to SLO 4.2)
- Explain why older children should be given the opportunity to describe their own respiratory symptoms. (Linked to SLO 4.2)

4.2 General Physical Examination of the Respiratory System

4.2.1 steps in general physical examination

Starting with the general impression

General physical examination of a child with a suspected respiratory complaint begins, as always, with a calm, structured **general inspection**, noting the child's **level of activity and alertness**, **colour**, including the presence or absence of **cyanosis**, and overall **work of breathing** before any hands-on examination begins. Counting the **respiratory rate** for a full minute, ideally while the child is calm and undisturbed, is an essential first step, since a rate obtained while the child is crying or agitated can be misleadingly elevated.

The examiner should also assess the child's **growth and nutritional state**, since chronic respiratory illness, such as poorly controlled asthma or chronic lung disease of prematurity, can impair growth over time, and should note any **finger clubbing**, which, while uncommon in acute illness, is an important sign of chronic hypoxaemia or suppurative lung disease when present.

Fill in the blank: Counting the respiratory rate for a full _____ is recommended for accuracy.

4.2.2 recognising signs of respiratory distress

Signs of **respiratory distress** should be actively sought during general examination, including **nasal flaring**, **tracheal tug**, **intercostal and subcostal recession**, or indrawing, **use of accessory muscles**, and audible sounds such as **grunting**, a sign of significant distress in infants representing an attempt to maintain positive airway pressure at the end of expiration, and **stridor**, a harsh, high-pitched sound suggesting upper airway obstruction.

The severity and combination of these signs help gauge the overall severity of respiratory illness, and their trend over time, whether improving or worsening, is often more clinically informative than a single assessment at one point in time, making serial reassessment an essential part of managing any child with significant respiratory distress.

Fill in the blank: Grunting represents an attempt to maintain positive airway pressure at the end of _____.

4.2.3 examination of the upper airway and associated structures

Examination should also include the **upper airway and associated structures**, since pathology here frequently contributes to or mimics lower respiratory symptoms. This includes inspecting the **nose** for discharge or obstruction, the **throat** for tonsillar enlargement or exudate, and the **ears**, since otitis media commonly coexists with respiratory infections in young children, together with palpation of the **cervical lymph nodes** for any associated lymphadenopathy.

In a child presenting with **stridor**, careful assessment of the severity, and avoidance of unnecessarily distressing examination manoeuvres such as forcing a young child to lie flat or attempting to examine the throat with a tongue depressor, is important, since agitation can worsen upper airway obstruction in conditions such as **croup** or, rarely, **epiglottitis**.

Fill in the blank: Otitis media commonly coexists with respiratory infections in young _____.

Box 4.2: Key signs of respiratory distress to assess on general examination.

Key signs of respiratory distress
Nasal flaring: widening of the nostrils with each breath, reflecting increased effort.
Tracheal tug: visible downward tug of the trachea with inspiration.
Intercostal and subcostal recession: indrawing of the chest wall between and below the ribs.
Use of accessory muscles: visible use of neck and shoulder muscles to assist breathing.
Grunting: an expiratory sound reflecting an attempt to maintain positive airway pressure.
Stridor: a harsh, high-pitched inspiratory sound suggesting upper airway obstruction.

Pilot questions 4.2

1. List three components of the general inspection of a child with a respiratory complaint. (Linked to SLO 4.3)
2. Why should the respiratory rate be counted while the child is calm? (Linked to SLO 4.3)
3. List four signs of respiratory distress. (Linked to SLO 4.4)
4. Explain the significance of grunting in an infant. (Linked to SLO 4.4)
5. Why should examination of a child with stridor avoid unnecessarily distressing manoeuvres? (Linked to SLO 4.4)

4.3 Systematic Examination Technique of the Chest

4.3.1 inspection and palpation of the chest

What the chest wall can tell you before you touch it

Chest **inspection** should assess the **shape and symmetry** of the chest wall, noting any deformity such as **pectus excavatum** or **pectus carinatum**, and observe the **pattern and symmetry of chest movement** with breathing, since asymmetrical movement may indicate unilateral pathology such as a pneumothorax or large pleural effusion on the affected side. The **respiratory rate**, **rhythm**, and any visible signs of distress described earlier should also be reassessed at this point.

Palpation of the chest assesses **chest expansion**, comparing both sides for symmetry, and **tactile vocal fremitus**, the palpable vibration felt when a child says a word such as "ninety-nine", which

is increased over areas of **consolidation** and decreased over a **pleural effusion** or **pneumothorax**. The **trachea** should also be palpated in the suprasternal notch to assess whether it is central or deviated, an important sign of significant unilateral chest pathology.

Fill in the blank: Tactile vocal fremitus is increased over areas of lung _____.

4.3.2 percussion of the chest

Percussion of the chest, performed by tapping the middle finger of one hand placed firmly against the chest wall with the tip of the middle finger of the other hand, assesses the underlying density of lung tissue. A normal lung produces a **resonant** note, while a **dull** note suggests underlying **consolidation** or **collapse**, and a **stony dull** note is characteristic of a significant **pleural effusion**. Conversely, a **hyperresonant** note may be heard over a **pneumothorax**, reflecting the presence of trapped air rather than fluid or solid tissue.

Percussion should be performed systematically, comparing corresponding areas on both sides of the chest anteriorly, laterally, and posteriorly, and should also include assessment of **cardiac and liver dullness** as reference points, helping to confirm that the technique is being performed correctly and consistently.

Fill in the blank: A stony dull percussion note is characteristic of a significant pleural _____.

4.3.3 auscultation and breath sounds

Auscultation of the chest should be performed systematically, comparing corresponding areas on both sides, and should assess the **quality** of breath sounds, normally described as **vesicular**, and the presence of any **added sounds**. **Crackles**, or crepitations, are discontinuous sounds suggesting fluid in the small airways or alveoli, as in pneumonia or pulmonary oedema, while **wheeze** is a continuous, musical sound produced by narrowed airways, typically heard on expiration in conditions such as asthma or bronchiolitis.

Bronchial breathing, a harsher, more tubular sound normally heard only over the trachea, is abnormal when heard over lung fields distant from the trachea, and suggests underlying **consolidation** with a patent airway, since consolidated lung tissue transmits the sound of large airway breathing more readily than normal aerated lung.

Fill in the blank: Bronchial breathing heard over a peripheral lung field suggests underlying _____.

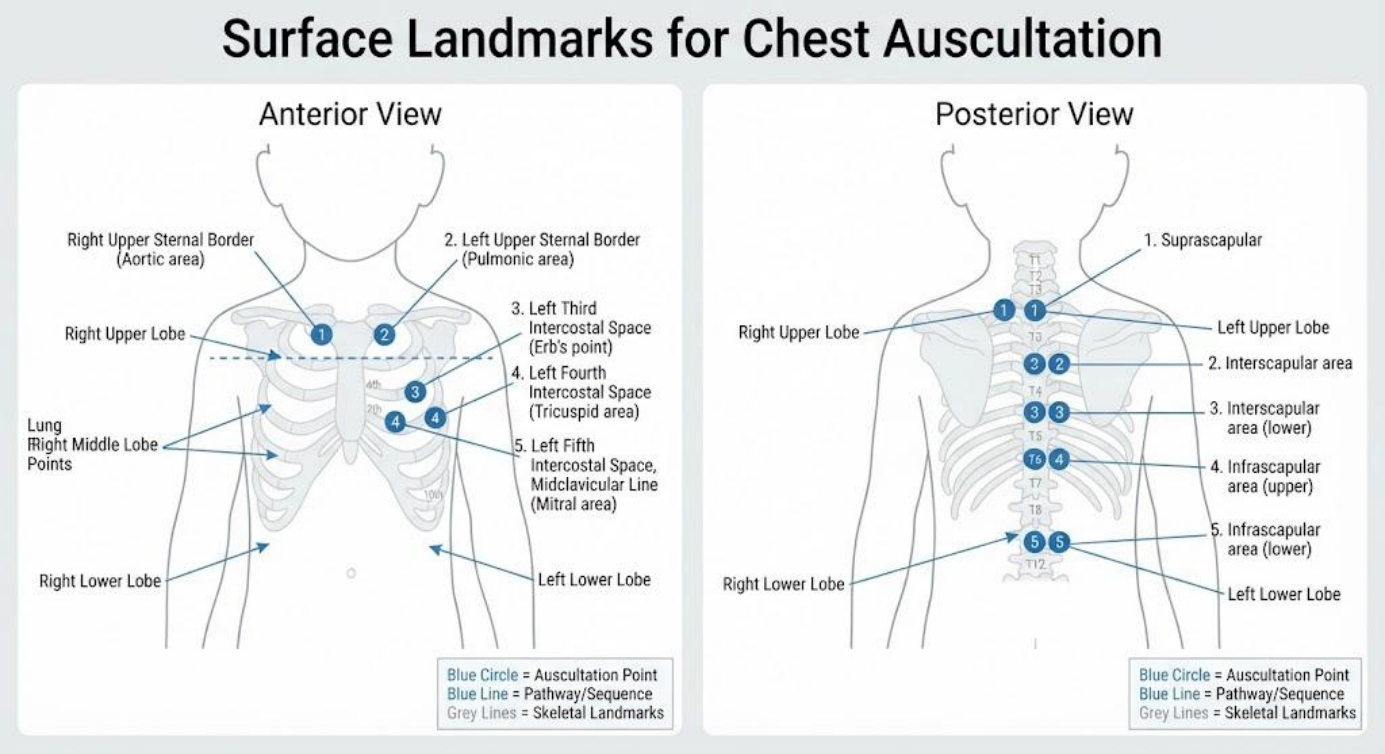

Figure 4.3: Surface landmarks for chest auscultation.

Pilot questions 4.3

1. Describe what is assessed during palpation of the chest. (Linked to SLO 4.5)
2. What does a dull percussion note suggest, and what does a hyperresonant note suggest? (Linked to SLO 4.5)
3. Distinguish crackles from wheeze on auscultation. (Linked to SLO 4.6)
4. What does bronchial breathing heard over a peripheral lung field suggest? (Linked to SLO 4.6)
5. Describe the correct technique for percussing the chest. (Linked to SLO 4.5)

4.4 Investigations and Communication in Respiratory Assessment

4.4.1 investigations used in respiratory assessment

Choosing the right test for the clinical question

Pulse oximetry provides a rapid, non-invasive estimate of **oxygen saturation** and should be performed in any child with suspected respiratory distress, since hypoxaemia may not always be clinically obvious, particularly in a child with anaemia. **Chest radiograph** remains useful for

identifying **consolidation**, **pleural effusion**, **pneumothorax**, or **foreign body** related changes, though it is not required for every child with a straightforward, mild respiratory illness.

Additional investigations are selected according to the clinical picture, and may include **full blood count** and **inflammatory markers** where a bacterial infection is suspected, **blood culture** in a child unwell enough to warrant intravenous antibiotics, and **peak expiratory flow rate** or **spirometry** in older children with suspected or known asthma, to objectively assess the degree of airway obstruction and response to treatment.

Fill in the blank: Pulse oximetry provides a rapid, non-invasive estimate of oxygen _____.

4.4.2 the role of communication in respiratory assessment

Clear **communication** is essential throughout respiratory assessment, both to gather an accurate history and to explain findings and next steps to an often anxious parent, particularly when a child is visibly distressed. Explaining what you are looking for and finding as you examine the child, in simple language, helps reduce parental anxiety and improves cooperation, both from the child and the accompanying adult.

Where a chronic respiratory condition such as **asthma** is diagnosed, communication extends beyond the single consultation to **structured education** about trigger avoidance, correct **inhaler technique**, and a clear, written **action plan** for managing worsening symptoms at home, since poor understanding of these elements is a well recognised contributor to preventable emergency presentations and hospital admissions.

Fill in the blank: A written action plan for managing worsening asthma symptoms at home is an important part of structured _____.

4.4.3 integrating history, examination, and investigation findings

Effective respiratory assessment ultimately depends on **integrating** the history, examination, and any relevant investigation findings into a coherent clinical picture, rather than relying on any single element in isolation. A normal chest radiograph, for example, does not exclude significant respiratory illness such as early bronchiolitis, just as a mild examination finding does not always correlate neatly with the severity of the underlying history.

Developing this integrative clinical judgement takes deliberate practice, and forms the essential platform for the remaining Series of this course, which will apply these assessment skills to the specific respiratory disorders of the upper airway and to asthma and other chronic respiratory conditions.

Fill in the blank: A normal chest radiograph does not always _____ significant respiratory illness such as early bronchiolitis.

Table 4.4: Common investigations used in the assessment of a child with respiratory illness.

Investigation	Main clinical use
Pulse oximetry	Rapid, non-invasive assessment of oxygen saturation.
Chest radiograph	Identifies consolidation, effusion, pneumothorax, or foreign body changes.
Full blood count and inflammatory markers	Supports assessment of suspected bacterial infection.
Peak expiratory flow rate or spirometry	Objectively assesses airway obstruction, mainly in older children with asthma.

Pilot questions 4.4

1. Why should pulse oximetry be performed in any child with suspected respiratory distress? (Linked to SLO 4.7)
2. List two situations in which a chest radiograph would be particularly useful. (Linked to SLO 4.7)
3. Describe the components of structured asthma education for a family. (Linked to SLO 4.8)
4. Why is a written asthma action plan important? (Linked to SLO 4.8)
5. Explain why respiratory assessment requires integrating history, examination, and investigation findings together. (Linked to SLO 4.8)

Series Summary

This Series introduced the systematic **assessment of the respiratory system** in children, beginning with relevant anatomy, physiology, and the principles of history taking, before turning to the **general physical examination**, including the recognition of respiratory distress.

We then examined the **systematic technique** of chest examination, covering inspection, palpation, percussion, and auscultation, before concluding with the **investigations and**

communication principles relevant to respiratory assessment. Having worked through this Series, you should now be able to conduct a systematic respiratory assessment of a child. The next Series turns to **respiratory disorders and management**.

Glossary of Terms

1. **Tracheal tug:** a visible downward tug of the trachea with inspiration, a sign of respiratory distress.
2. **Intercostal recession:** indrawing of the chest wall between the ribs during inspiration, a sign of increased work of breathing.
3. **Grunting:** an expiratory sound reflecting an infant's attempt to maintain positive airway pressure, a sign of significant respiratory distress.
4. **Stridor:** a harsh, high-pitched inspiratory sound suggesting upper airway obstruction.
5. **Tactile vocal fremitus:** the palpable vibration felt on the chest wall when a child speaks, altered by underlying lung pathology.
6. **Resonant:** the normal percussion note heard over aerated lung tissue.
7. **Stony dull:** the percussion note characteristic of a significant pleural effusion.
8. **Hyperresonant:** the percussion note heard over a pneumothorax, reflecting trapped air.
9. **Crackles:** discontinuous added breath sounds suggesting fluid in the small airways or alveoli.
10. **Wheeze:** a continuous, musical added breath sound produced by narrowed airways.
11. **Bronchial breathing:** a harsh, tubular breath sound, abnormal when heard away from the trachea, suggesting consolidation.
12. **Pulse oximetry:** a non-invasive method of estimating oxygen saturation.

Concept Check Questions [Series 4]

Objective questions

- A history of choking should raise suspicion for possible foreign body _____.
- Grunting represents an attempt to maintain positive airway pressure at the end of _____.
- Tactile vocal fremitus is increased over areas of lung _____.

- A stony dull percussion note is characteristic of a significant pleural _____.
- Bronchial breathing heard over a peripheral lung field suggests underlying _____.

Short-answer questions

1. List three anatomical or physiological differences between a child's and an adult's respiratory system.
2. List four signs of respiratory distress.
3. Distinguish crackles from wheeze on auscultation.
4. Describe two clinical uses of pulse oximetry in respiratory assessment.
5. Describe the components of a written asthma action plan.

Long-answer questions

6. Discuss the basic principles of history taking for a child with a respiratory complaint. (Linked to SLO 4.2)
7. Describe the signs of respiratory distress and their clinical significance. (Linked to SLO 4.3, SLO 4.4)
8. Discuss the systematic technique of chest examination, including inspection, palpation, percussion, and auscultation. (Linked to SLO 4.5, SLO 4.6)
9. Discuss the investigations used in the assessment of a child with respiratory illness. (Linked to SLO 4.7)
10. Discuss the role of communication in the assessment and ongoing management of a child with a chronic respiratory condition. (Linked to SLO 4.8)

Answers to Concept Check Questions [Series 4]

Objective questions

11. Aspiration.
12. Expiration.
13. Consolidation.
14. Effusion.
15. Consolidation.

Short-answer questions

16. A narrower airway, a proportionally larger tongue, and a more compliant chest wall.

17. Nasal flaring, tracheal tug, intercostal and subcostal recession, and grunting.
18. Crackles are discontinuous sounds suggesting fluid in the small airways or alveoli, while wheeze is a continuous, musical sound produced by narrowed airways.
19. Rapid assessment of oxygen saturation in a distressed child, and ongoing monitoring during treatment.
20. Trigger avoidance, correct inhaler technique, and a clear plan for managing worsening symptoms at home.

Long-answer questions

21. **Basic response:** A focused respiratory history establishes the nature, onset, duration, and pattern of the presenting complaint, together with associated symptoms and relevant past medical, family, and environmental history.

Value-added response: A history of choking should specifically raise suspicion for foreign body aspiration, and older children should be given the opportunity to describe their own symptoms wherever possible.

22. **Basic response:** Signs of respiratory distress include nasal flaring, tracheal tug, intercostal and subcostal recession, use of accessory muscles, grunting, and stridor.

Value-added response: The trend of these signs over time, whether improving or worsening, is often more clinically informative than a single assessment, making serial reassessment essential in significant respiratory distress.

23. **Basic response:** Chest examination proceeds through inspection, palpation for chest expansion and fremitus, percussion to assess underlying lung density, and auscultation for breath sounds and added sounds.

Value-added response: Specific findings localise pathology, for example dullness and increased fremitus suggest consolidation, while a stony dull note and reduced fremitus suggest a pleural effusion.

24. **Basic response:** Pulse oximetry rapidly assesses oxygen saturation, chest radiograph identifies structural findings such as consolidation or effusion, and peak flow or spirometry objectively assesses airway obstruction.

Value-added response: Investigations should be selected according to the clinical picture rather than requested indiscriminately, since a normal chest radiograph, for example, does not exclude significant respiratory illness such as early bronchiolitis.

25. **Basic response:** Clear communication helps gather an accurate history, reduces parental anxiety during examination, and supports structured education for chronic conditions such as asthma.

Value-added response: Structured asthma education, including trigger avoidance, inhaler technique, and a written action plan, helps prevent avoidable emergency presentations and hospital admissions.

Answers to Pilot Questions [Series 4]

Part 4.1

1. The airway is narrower, the tongue is proportionally larger, and the chest wall is more compliant.
2. Children have a higher metabolic rate and oxygen consumption relative to body weight than adults.
3. Onset, duration, pattern, associated symptoms, and any history of choking.
4. It may indicate foreign body aspiration, requiring urgent evaluation.
5. It respects their autonomy and can reveal details a parent may not have noticed.

Part 4.2

6. Level of activity and alertness, colour, and overall work of breathing.
7. A rate obtained while the child is crying or agitated can be misleadingly elevated.
8. Nasal flaring, tracheal tug, intercostal and subcostal recession, and grunting.
9. Grunting reflects an attempt to maintain positive airway pressure and signifies significant distress.
10. Agitation can worsen upper airway obstruction in conditions such as croup or epiglottitis.

Part 4.3

11. Palpation assesses chest expansion, tactile vocal fremitus, and tracheal position.
12. A dull note suggests consolidation or collapse, while a hyperresonant note suggests a pneumothorax.
13. Crackles are discontinuous sounds suggesting fluid in the airways or alveoli, while wheeze is a continuous, musical sound from narrowed airways.

14. It suggests underlying consolidation with a patent airway.
15. Tap the middle finger of one hand against the chest wall with the tip of the middle finger of the other hand, comparing both sides systematically.

Part 4.4

1. Hypoxaemia may not always be clinically obvious, particularly in a child with anaemia.
2. Suspected consolidation, pleural effusion, pneumothorax, or foreign body aspiration.
3. Trigger avoidance, correct inhaler technique, and a written plan for managing worsening symptoms.
4. It helps families recognise and respond appropriately to worsening symptoms at home, reducing avoidable admissions.
5. No single element reliably reflects severity alone, so all three must be considered together for an accurate clinical picture.

References and Attributions [Series 4]

1. Kliegman, R. M., St Geme, J. W., Blum, N. J., Shah, S. S., Tasker, R. C. and Wilson, K. M. (2020). Nelson Textbook of Pediatrics (21st ed.). Elsevier.
2. World Health Organization (2014). Handbook: IMCI Integrated Management of Childhood Illness. WHO Press.
3. Talner, N. S. and Carboni, M. P. (2019). Physical examination of the respiratory system in children. Pediatrics in Review, 40(5), 213 to 225.
4. Global Initiative for Asthma (2023). Global Strategy for Asthma Management and Prevention.

Figures, Plates, Tables, and Boxes

1. Table 4.1: Normal respiratory rate ranges in children by age. AI-generated using the prompt: "Create a clean reference table titled Normal Respiratory Rate Ranges in Children by Age, listing age group and breaths per minute. Simple clinical reference table style with outside border."

2. Box 4.2: Key signs of respiratory distress to assess on general examination. AI-generated using the prompt: "Create a simple single-column reference box titled Key Signs of Respiratory Distress, listing each sign with a brief description. Clean clinical reference box style."
3. Figure 4.3: Surface landmarks for chest auscultation. AI-generated using the prompt: "Create a professional medical diagram titled Surface Landmarks for Chest Auscultation, showing anterior and posterior auscultation points on a paediatric chest outline. Clean clinical educational diagram style."
4. Table 4.4: Common investigations used in the assessment of a child with respiratory illness. AI-generated using the prompt: "Create a clean reference table titled Common Investigations in Respiratory Assessment, listing investigation and main clinical use. Simple clinical reference table style with outside border."

Course code: PAE 503

Course title: Cardiovascular and Respiratory Disorders

Course credit load: 2 Units

Series 5: Respiratory Disorders and Management

- **Part 5.1: Disorders of the Upper Respiratory Tract**
  - Sub Part 5.1.1: Classification of upper airway disorders
  - Sub Part 5.1.2: Croup and acute epiglottitis
  - Sub Part 5.1.3: Foreign body aspiration and bacterial tracheitis
- **Part 5.2: Asthma: Definition, Pathophysiology, and Clinical Features**
  - Sub Part 5.2.1: Definition and mechanism of asthma
  - Sub Part 5.2.2: Clinical features of asthma
  - Sub Part 5.2.3: Diagnosis of asthma
- **Part 5.3: Asthma: Treatment, Prevention, and Communication**
  - Sub Part 5.3.1: Pharmacological treatment of asthma
  - Sub Part 5.3.2: Inhaler technique and delivery devices
  - Sub Part 5.3.3: Prevention and communication in asthma management
- **Part 5.4: The Wheezing Child and Chronic Respiratory Disorders**
  - Sub Part 5.4.1: Differential diagnosis of the wheezing child
  - Sub Part 5.4.2: Clinical approach to the wheezing child
  - Sub Part 5.4.3: Other chronic respiratory disorders of childhood

Introduction

In the last Series, we explored the systematic **assessment of the respiratory system**, building the history taking and examination skills you will now apply directly. Picture a three year old brought to clinic with a harsh, barking cough and noisy breathing that started suddenly overnight, and compare this with a six year old who has had recurrent nighttime cough and wheeze for the

past two years. These two presentations, croup and asthma, illustrate the range of **respiratory disorders** this Series will introduce, together with their management.

The aim of this Series is to equip you with the knowledge required to recognise and understand the management of common **upper respiratory tract disorders**, **asthma**, and the broader differential diagnosis of the **wheezing child**.

This Series begins with the **disorders of the upper respiratory tract**, before turning to **asthma**, covering its definition, pathophysiology, and clinical features, and then its **treatment, prevention, and communication**. Finally, we conclude with the **wheezing child** and a brief overview of other chronic respiratory disorders of childhood.

Series Learning Outcomes

At the end of this Series, you should be able to:

- **SLO 5.1:** classify the disorders of the upper respiratory tract in children,
- **SLO 5.2:** describe the clinical presentation of common upper airway disorders,
- **SLO 5.3:** define asthma and describe its underlying mechanism and pathophysiology,
- **SLO 5.4:** describe the clinical features and diagnosis of asthma,
- **SLO 5.5:** describe the pharmacological treatment of asthma, including the stepwise approach,
- **SLO 5.6:** describe the principles of asthma prevention and communication with the family,
- **SLO 5.7:** describe the differential diagnosis and clinical approach to the wheezing child, and
- **SLO 5.8:** describe other chronic respiratory disorders of childhood and their general management.

5.1 Disorders of the Upper Respiratory Tract

5.1.1 classification of upper airway disorders

Defining where the upper airway ends

The **upper airway** extends from the nose to the level of the **vocal cords**, and disorders affecting it commonly present with **stridor**, a harsh, high-pitched inspiratory sound reflecting turbulent airflow through a narrowed segment. Common causes include **croup**, or laryngotracheobronchitis, typically caused by **parainfluenza virus**, **acute epiglottitis**, a rare but

life-threatening bacterial infection now much less common following **Haemophilus influenzae type b** vaccination, **foreign body aspiration**, and **bacterial tracheitis**, a less common but serious secondary bacterial infection.

Distinguishing between these causes relies heavily on the **history**, particularly the **speed of onset** and any preceding symptoms, together with the **child's overall appearance**, since a toxic-looking, drooling child with stridor raises far greater concern for epiglottitis than a child with a barking cough and mild stridor typical of croup.

Fill in the blank: Stridor reflects turbulent airflow through a narrowed segment of the _____ airway.

5.1.2 croup and acute epiglottitis

Croup typically affects children between six months and six years of age, presenting with a **barking cough**, **hoarse voice**, and **inspiratory stridor**, often preceded by a day or two of mild coryzal symptoms, and characteristically **worse at night**. Most cases are mild and self-limiting, managed with a single dose of oral **dexamethasone**, which reduces airway inflammation, while more severe cases with stridor at rest may additionally require **nebulised adrenaline** and close observation.

Acute epiglottitis, by contrast, presents with a **rapid onset** of high fever, severe sore throat, drooling due to painful swallowing, and a **toxic, anxious appearance**, with the child often preferring to sit upright, leaning forward in a **tripod position**. Epiglottitis is a medical emergency, since attempting to examine the throat or lay the child flat can precipitate complete airway obstruction, and management requires **urgent, controlled airway assessment**, typically in an operating theatre, by the most experienced available anaesthetist and otolaryngologist.

Fill in the blank: Croup is characteristically worse at _____.

5.1.3 foreign body aspiration and bacterial tracheitis

Foreign body aspiration should be suspected in a child with a **sudden onset** of coughing, choking, or stridor, particularly if there is a witnessed or reported choking episode, most commonly occurring in **toddlers** exploring their environment orally. Depending on the location of the foreign body, presentation can range from immediate, severe respiratory distress to a more subtle, persistent cough or recurrent chest infection if the object lodges lower in the airway and is not immediately recognised.

Bacterial tracheitis is an uncommon but serious condition in which a child with apparent croup fails to improve, or deteriorates, developing high fever and increasing respiratory distress,

reflecting secondary bacterial infection of the trachea, often with **Staphylococcus aureus**, and requiring **intravenous antibiotics** and, frequently, **airway support** in a high dependency or intensive care setting.

Fill in the blank: Foreign body aspiration is most common in _____ exploring their environment orally.

Table 5.1: Comparison of croup and acute epiglottitis.

Feature	Croup	Acute epiglottitis
Onset	Gradual, over 1 to 2 days	Rapid, over hours
Cough	Barking cough present	Cough usually absent
Appearance	Generally well between episodes of stridor	Toxic, anxious, drooling
Position	No specific preferred position	Prefers to sit forward in tripod position
Typical cause	Parainfluenza virus	Bacterial, classically Haemophilus influenzae type b

Pilot questions 5.1

- Define the upper airway and list two common causes of stridor. (Linked to SLO 5.1)
- Describe the typical clinical presentation of croup. (Linked to SLO 5.2)
- Describe the typical clinical presentation of acute epiglottitis. (Linked to SLO 5.2)
- Explain why examining the throat of a child with suspected epiglottitis is dangerous. (Linked to SLO 5.2)
- When should foreign body aspiration be suspected in a child with stridor or cough? (Linked to SLO 5.2)

5.2 Asthma: Definition, Pathophysiology, and Clinical Features

5.2.1 definition and mechanism of asthma

Three processes that together narrow the airway

Asthma is a chronic inflammatory disorder of the airways characterised by **reversible airway obstruction**, **bronchial hyperresponsiveness**, and underlying **chronic airway inflammation**.

Three processes together contribute to airway narrowing during an asthma exacerbation: **bronchoconstriction**, contraction of the smooth muscle surrounding the airways, **mucosal oedema**, swelling of the airway lining due to inflammation, and increased **mucus production**, all of which combine to reduce the calibre of the airway and increase resistance to airflow.

This inflammation is typically driven by an **allergic**, or atopic, immune response in many children, though **non-allergic triggers** such as viral respiratory infections, cold air, exercise, and irritants including tobacco smoke can also provoke symptoms, sometimes in the same child who also has allergic triggers, reflecting the genuinely multifactorial nature of the condition.

Fill in the blank: The three processes contributing to airway narrowing in asthma are bronchoconstriction, mucosal oedema, and increased _____ production.

5.2.2 clinical features of asthma

Children with asthma classically present with **recurrent episodes** of **wheeze**, **cough**, particularly at **night** or with exercise, **chest tightness**, and **breathlessness**, often triggered by identifiable factors such as viral infections, exercise, cold air, or allergen exposure. Symptoms are typically **variable**, both over time and in response to treatment, a pattern that helps distinguish asthma from other causes of chronic respiratory symptoms.

On examination during an acute exacerbation, findings may include **audible wheeze**, a **prolonged expiratory phase**, and, in more severe episodes, the signs of respiratory distress discussed in Series 4, including intercostal recession and accessory muscle use. Between exacerbations, examination findings are frequently entirely **normal**, which is why the history of recurrent, variable, trigger-related symptoms is so central to making the diagnosis.

Fill in the blank: Between exacerbations, examination findings in a child with asthma are frequently entirely _____.

5.2.3 diagnosis of asthma

The diagnosis of asthma in children, particularly younger children, is largely **clinical**, based on a compatible history of recurrent, variable, trigger-related wheeze and cough, together with a **positive response to bronchodilator treatment**. In older children who can perform the manoeuvre reliably, usually from around five to six years of age, **spirometry** demonstrating reversible airway obstruction, defined as a significant improvement in **forced expiratory volume in one second** after bronchodilator administration, provides objective supporting evidence.

It is important to actively consider and exclude **alternative diagnoses** in a child with atypical features, such as symptoms present from birth, poor growth, or a wet, productive cough, since conditions such as **cystic fibrosis**, **primary ciliary dyskinesia**, or an inhaled **foreign body** can mimic asthma but require an entirely different diagnostic and management pathway.

Fill in the blank: A significant improvement in forced expiratory volume in one second after bronchodilator use supports a diagnosis of _____.

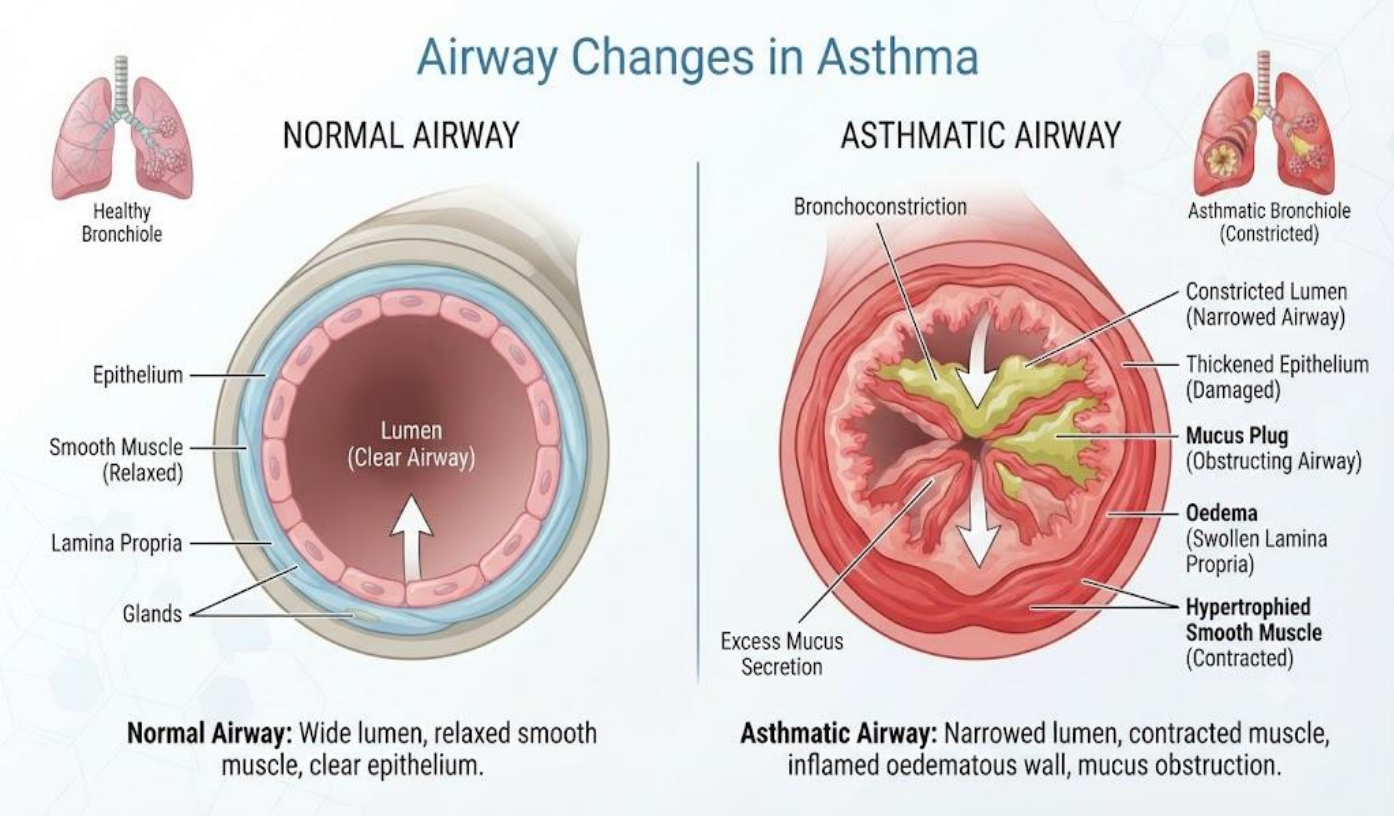

Figure 5.2: The three processes contributing to airway narrowing in asthma.

Pilot questions 5.2

1. Define asthma and list the three processes that contribute to airway narrowing. (Linked to SLO 5.3)
2. Describe the typical clinical features of asthma. (Linked to SLO 5.4)
3. Explain why examination findings may be normal between asthma exacerbations. (Linked to SLO 5.4)
4. Describe how spirometry can support a diagnosis of asthma. (Linked to SLO 5.4)
5. List two atypical features that should prompt consideration of an alternative diagnosis to asthma. (Linked to SLO 5.4)

5.3 Asthma: Treatment, Prevention, and Communication

5.3.1 pharmacological treatment of asthma

Relievers and controllers serve different purposes

Asthma treatment is broadly divided into **reliever medication**, most commonly a **short-acting beta-2 agonist** such as salbutamol, which rapidly relaxes airway smooth muscle to relieve acute symptoms, and **controller medication**, most commonly an **inhaled corticosteroid**, which reduces the underlying chronic airway inflammation and is taken regularly, regardless of symptoms, to prevent exacerbations rather than to treat them once they occur.

Treatment generally follows a **stepwise approach**, starting with an as-needed reliever alone for infrequent, mild symptoms, and adding a regular low-dose inhaled corticosteroid, then increasing the dose or adding additional controller medications such as a **long-acting beta-2 agonist** or **leukotriene receptor antagonist**, as needed to achieve good symptom control, before stepping back down once control has been well maintained for a sustained period.

Fill in the blank: A short-acting beta-2 agonist such as salbutamol is used as a _____ medication for acute asthma symptoms.

5.3.2 inhaler technique and delivery devices

Correct **inhaler technique** is essential for effective drug delivery, and poor technique is a common, correctable cause of apparently poorly controlled asthma. A **pressurised metered-dose inhaler** used with a **spacer device** is generally recommended for young children, since coordinating a hand-held inhaler activation with inhalation is difficult for a young child to master reliably, and a spacer allows the medication to be inhaled over several breaths without this coordination requirement.

For infants and very young children, a spacer with an attached **face mask** is used, transitioning to a mouthpiece as the child grows older and can form an adequate seal around it. **Inhaler technique should be checked at every review**, since technique can deteriorate over time even in children who were initially taught correctly, and this is a simple, high-value intervention that is often overlooked in busy clinical practice.

Fill in the blank: A pressurised metered-dose inhaler is generally used with a _____ device in young children.

5.3.3 prevention and communication in asthma management

Prevention in asthma centres on identifying and, where possible, avoiding individual **triggers**, such as tobacco smoke exposure, specific allergens, or, in some children, particular irritants, together with ensuring regular controller medication is taken consistently rather than only during symptomatic periods. **Annual influenza vaccination** is also recommended for children with asthma, since respiratory viral infections are a common trigger for exacerbations.

Effective **communication** with the family is central to good long-term asthma control, including structured **education** about the difference between reliever and controller medication, correct **inhaler technique**, recognition of **worsening symptoms**, and a clear, written **asthma action plan** describing what to do at each level of symptom severity, since poor understanding of these elements remains one of the most common, preventable contributors to asthma-related emergency presentations.

Fill in the blank: Annual _____ vaccination is recommended for children with asthma.

Box 5.3: The stepwise approach to asthma pharmacological treatment.

Stepwise approach to asthma treatment
Step 1: As-needed short-acting beta-2 agonist alone for infrequent, mild symptoms.
Step 2: Add a regular low-dose inhaled corticosteroid as a controller.
Step 3: Increase inhaled corticosteroid dose or add a long-acting beta-2 agonist.
Step 4: Add a leukotriene receptor antagonist or further increase controller therapy.
Step 5: Refer for specialist assessment and consider additional add-on therapy.
At every step: check inhaler technique, adherence, and trigger avoidance before stepping up.

Pilot questions 5.3

1. Distinguish between reliever and controller medication in asthma. (Linked to SLO 5.5)
2. Describe the stepwise approach to asthma pharmacological treatment. (Linked to SLO 5.5)
3. Why is a spacer device recommended for young children using a metered-dose inhaler? (Linked to SLO 5.5)
4. List two components of asthma prevention. (Linked to SLO 5.6)
5. Describe the components of a written asthma action plan. (Linked to SLO 5.6)

5.4 The Wheezing Child and Chronic Respiratory Disorders

5.4.1 differential diagnosis of the wheezing child

Not every wheeze is asthma

While **asthma** is the most common cause of recurrent wheeze in children, particularly older children, it is important to recognise that **not every wheezing child has asthma**, and the differential diagnosis varies considerably by age. In **infants**, **viral bronchiolitis**, most commonly caused by **respiratory syncytial virus**, is the most common cause of a first episode of wheeze, typically presenting with a preceding coryzal illness followed by increasing respiratory distress and wheeze, usually in the first two years of life.

Other important causes to consider include **foreign body aspiration**, particularly with a sudden onset of wheeze or asymmetrical findings on examination, **cystic fibrosis**, particularly if accompanied by poor growth or a chronic wet cough, **gastro-oesophageal reflux**, and, rarely, a structural airway or vascular abnormality such as a **vascular ring** compressing the airway.

Fill in the blank: The most common cause of a first episode of wheeze in infants is viral _____.

5.4.2 clinical approach to the wheezing child

A structured clinical approach to the wheezing child should establish the **age of onset**, since bronchiolitis is far more likely in a young infant while asthma becomes relatively more likely with increasing age, the **pattern** of wheeze, whether episodic and variable, as in asthma, or persistent and progressive, which raises concern for a structural or chronic underlying cause, and any **red flag features**, such as poor growth, symptoms from birth, or a wet, productive cough, suggesting an alternative diagnosis.

Response to bronchodilator treatment is also informative, since a wheeze that responds well to a trial of bronchodilator supports a diagnosis of asthma or a reactive airway process, while a wheeze that fails to respond at all should prompt reconsideration of the diagnosis and, where appropriate, referral for further specialist evaluation, including consideration of a chest radiograph or, in selected cases, bronchoscopy.

Fill in the blank: A wheeze that fails to respond at all to bronchodilator treatment should prompt reconsideration of the _____.

5.4.3 other chronic respiratory disorders of childhood

Beyond asthma, several other **chronic respiratory disorders** affect children and require general awareness even outside specialist practice. **Cystic fibrosis**, an inherited condition affecting chloride channel function, causes thick, sticky mucus that predisposes to recurrent chest infections and, over time, progressive lung damage, alongside pancreatic insufficiency affecting growth and nutrition. **Chronic lung disease of prematurity**, also called bronchopulmonary dysplasia, affects some infants born very preterm and can cause ongoing respiratory symptoms requiring long-term follow up.

Children with any chronic respiratory disorder benefit from a coordinated, **multidisciplinary approach**, involving paediatricians, specialist nurses, physiotherapists, and dietitians where relevant, together with the same principles of clear **communication**, structured **education**, and a written **management plan** discussed earlier in this Series, since these general principles apply across the full range of chronic respiratory conditions, not only asthma.

Fill in the blank: Cystic fibrosis is an inherited condition affecting _____ channel function.

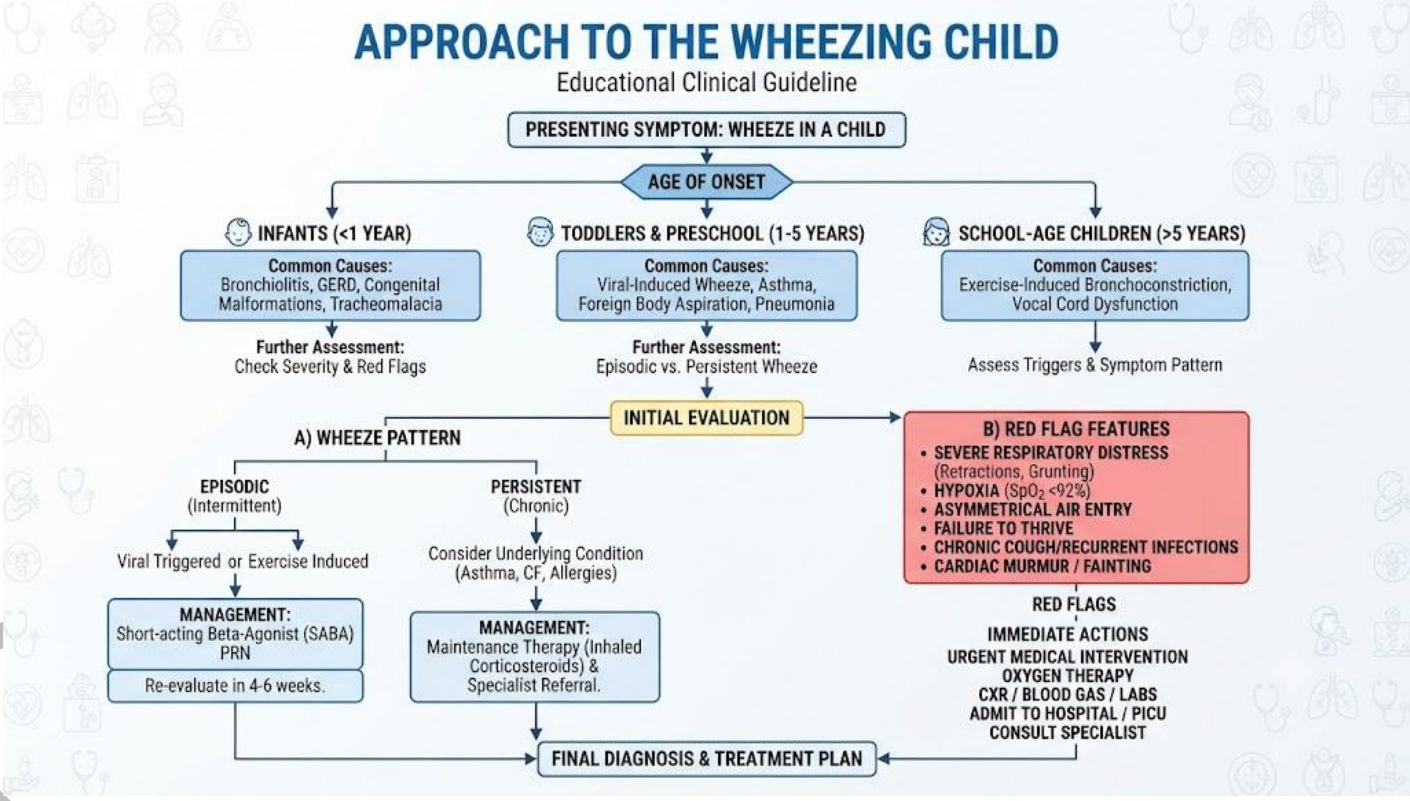

Figure 5.4: A clinical approach to the differential diagnosis of the wheezing child.

Pilot questions 5.4

1. List three causes of wheeze in children other than asthma. (Linked to SLO 5.7)
2. Describe the most common cause of a first episode of wheeze in infancy. (Linked to SLO 5.7)
3. List two red flag features that should prompt consideration of a diagnosis other than asthma. (Linked to SLO 5.7)
4. Describe the basic pathophysiological defect in cystic fibrosis. (Linked to SLO 5.8)
5. Describe the general principles of managing a child with a chronic respiratory disorder. (Linked to SLO 5.8)

Series Summary

This Series examined the common **respiratory disorders** of childhood, beginning with **disorders of the upper respiratory tract**, including croup, acute epiglottitis, foreign body aspiration, and bacterial tracheitis, before turning in depth to **asthma**, covering its definition, pathophysiology, clinical features, treatment, and prevention.

We then concluded with a structured approach to the **wheezing child** and a brief overview of other **chronic respiratory disorders** of childhood. Having worked through this Series, and indeed this course, you should now be equipped with a systematic approach to the evaluation and management of the common cardiovascular and respiratory conditions encountered in paediatric practice.

Glossary of Terms

1. **Croup:** laryngotracheobronchitis, typically viral, presenting with a barking cough, hoarse voice, and stridor.
2. **Acute epiglottitis:** a rare, life-threatening bacterial infection of the epiglottis causing rapid-onset stridor and a toxic appearance.
3. **Bacterial tracheitis:** a serious secondary bacterial infection of the trachea in a child with apparent croup who fails to improve.
4. **Asthma:** a chronic inflammatory airway disorder characterised by reversible airway obstruction and bronchial hyperresponsiveness.

5. **Bronchoconstriction:** contraction of airway smooth muscle, one of the processes narrowing the airway in asthma.
6. **Reliever medication:** medication, typically a short-acting beta-2 agonist, used to rapidly relieve acute asthma symptoms.
7. **Controller medication:** medication, typically an inhaled corticosteroid, taken regularly to reduce underlying airway inflammation and prevent exacerbations.
8. **Spacer device:** a chamber attached to a metered-dose inhaler that removes the need to coordinate activation with inhalation.
9. **Asthma action plan:** a written plan describing what a family should do at each level of asthma symptom severity.
10. **Bronchiolitis:** a viral lower respiratory tract infection, commonly caused by respiratory syncytial virus, and the most common cause of wheeze in infancy.
11. **Cystic fibrosis:** an inherited condition affecting chloride channel function, causing thick mucus and recurrent chest infections.
12. **Bronchopulmonary dysplasia:** chronic lung disease of prematurity affecting some infants born very preterm.

Concept Check Questions [Series 5]

Objective questions

- Croup is characteristically worse at _____.
- The three processes contributing to airway narrowing in asthma are bronchoconstriction, mucosal oedema, and increased _____ production.
- A pressurised metered-dose inhaler is generally used with a _____ device in young children.
- The most common cause of a first episode of wheeze in infants is viral _____.
- Cystic fibrosis is an inherited condition affecting _____ channel function.

Short-answer questions

1. Describe two features that distinguish acute epiglottitis from croup.
2. Describe the typical clinical features of asthma.
3. Distinguish between reliever and controller medication in asthma.
4. List two red flag features that should prompt consideration of a diagnosis other than asthma in a wheezing child.

5. Describe the components of a written asthma action plan.

Long-answer questions

6. Discuss the clinical presentation and management of croup and acute epiglottitis. (Linked to SLO 5.1, SLO 5.2)
7. Discuss the pathophysiology and clinical features of asthma. (Linked to SLO 5.3, SLO 5.4)
8. Describe the stepwise pharmacological treatment of asthma. (Linked to SLO 5.5)
9. Discuss the principles of asthma prevention and communication with the family. (Linked to SLO 5.6)
10. Discuss the differential diagnosis and clinical approach to a wheezing child. (Linked to SLO 5.7)

Answers to Concept Check Questions [Series 5]

Objective questions

11. Night.
12. Mucus.
13. Spacer.
14. Bronchiolitis.
15. Chloride.

Short-answer questions

16. Epiglottitis has a rapid onset, high fever, drooling, and a toxic appearance, whereas croup has a gradual onset with a barking cough.
17. Recurrent wheeze, cough particularly at night or with exercise, chest tightness, and breathlessness, often with identifiable triggers.
18. Relievers, such as short-acting beta-2 agonists, rapidly relieve acute symptoms, while controllers, such as inhaled corticosteroids, are taken regularly to reduce underlying inflammation.
19. Symptoms from birth, poor growth, or a wet, productive cough.
20. Clear description of reliever and controller use, recognition of worsening symptoms, and specific actions to take at each severity level.

Long-answer questions

21. **Basic response:** Croup presents with a gradual onset barking cough and stridor, managed with oral dexamethasone, while epiglottitis presents with rapid onset fever, drooling, and a toxic appearance, requiring urgent controlled airway assessment.

Value-added response: Examining the throat or laying a child with suspected epiglottitis flat can precipitate complete airway obstruction, so airway assessment must occur in a controlled setting with experienced anaesthetic and ENT support.

22. **Basic response:** Asthma involves bronchoconstriction, mucosal oedema, and increased mucus production, producing reversible airway obstruction with recurrent, variable, trigger-related wheeze and cough.

Value-added response: Examination findings are frequently normal between exacerbations, making the history of recurrent, variable symptoms central to diagnosis, supported where possible by spirometry showing reversible obstruction.

23. **Basic response:** Treatment follows a stepwise approach, starting with an as-needed reliever, then adding regular low-dose inhaled corticosteroid, and stepping up further therapy as needed to achieve control.

Value-added response: Inhaler technique and adherence should be checked at every step before escalating therapy, since poor technique is a common, correctable cause of apparently poor control.

24. **Basic response:** Prevention involves identifying and avoiding triggers and ensuring consistent controller medication use, while communication involves structured education on technique, symptom recognition, and a written action plan.

Value-added response: Annual influenza vaccination is also recommended, since viral infections are a common trigger for exacerbations, and poor family understanding remains a leading preventable cause of asthma emergencies.

25. **Basic response:** The differential diagnosis of wheeze includes bronchiolitis in infants, foreign body aspiration, cystic fibrosis, gastro-oesophageal reflux, and, rarely, structural airway abnormalities.

Value-added response: A structured approach considers age of onset, pattern of wheeze, red flag features, and response to bronchodilator treatment, with a poor response prompting reconsideration of the diagnosis.

Answers to Pilot Questions [Series 5]

Part 5.1

1. The upper airway extends from the nose to the vocal cords; causes of stridor include croup and acute epiglottitis.
2. Croup presents with a barking cough, hoarse voice, and inspiratory stridor, often worse at night.
3. Epiglottitis presents with rapid-onset high fever, drooling, and a toxic, anxious appearance, often in a tripod position.
4. Examining the throat can precipitate complete airway obstruction in a child with epiglottitis.
5. Foreign body aspiration should be suspected with sudden-onset stridor or cough, particularly with a witnessed choking episode.

Part 5.2

6. Asthma involves bronchoconstriction, mucosal oedema, and increased mucus production.
7. Recurrent wheeze, cough, chest tightness, and breathlessness, often triggered by identifiable factors.
8. Symptoms and airway obstruction are variable rather than constant, so the airway can appear normal between episodes.
9. Spirometry demonstrating a significant improvement in forced expiratory volume in one second after bronchodilator supports the diagnosis.
10. Symptoms from birth, poor growth, or a wet, productive cough should prompt consideration of an alternative diagnosis.

Part 5.3

11. Relievers rapidly relieve acute symptoms; controllers are taken regularly to reduce underlying airway inflammation.
12. Treatment starts with an as-needed reliever, then adds regular controller therapy, stepping up as needed for control.
13. A spacer removes the need to coordinate inhaler activation with inhalation, which young children find difficult.
14. Trigger avoidance and consistent controller medication use are key components of prevention.
15. A written action plan describes what to do at each level of symptom severity, supporting timely home management.

Part 5.4

1. Bronchiolitis, foreign body aspiration, and cystic fibrosis are causes of wheeze other than asthma.
2. Viral bronchiolitis, most commonly due to respiratory syncytial virus, is the most common cause in infancy.
3. Poor growth and a wet, productive cough are red flag features.
4. Cystic fibrosis affects chloride channel function, causing thick, sticky mucus.
5. Management involves a coordinated multidisciplinary approach with clear communication, education, and a written plan.

References and Attributions [Series 5]

1. Kliegman, R. M., St Geme, J. W., Blum, N. J., Shah, S. S., Tasker, R. C. and Wilson, K. M. (2020). Nelson Textbook of Pediatrics (21st ed.). Elsevier.
2. Global Initiative for Asthma (2023). Global Strategy for Asthma Management and Prevention.
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4. Bjornson, C. L. and Johnson, D. W. (2013). Croup in children. Canadian Medical Association Journal, 185(15), 1317 to 1323.

Figures, Plates, Tables, and Boxes

1. Table 5.1: Comparison of croup and acute epiglottitis. AI-generated using the prompt: "Create a clean reference table titled Croup versus Acute Epiglottitis, comparing onset, cough, appearance, position, and cause. Simple clinical reference table style with outside border."
2. Figure 5.2: The three processes contributing to airway narrowing in asthma. AI-generated using the prompt: "Create a professional medical diagram titled Airway Changes in

Asthma, comparing a normal airway cross section with an asthmatic airway showing bronchoconstriction, oedema, and mucus. Clean clinical educational diagram style."

3. Box 5.3: The stepwise approach to asthma pharmacological treatment. AI-generated using the prompt: "Create a simple single-column reference box titled Stepwise Approach to Asthma Treatment, listing steps 1 through 5. Clean clinical reference box style."
4. Figure 5.4: A clinical approach to the differential diagnosis of the wheezing child. AI-generated using the prompt: "Create a professional medical flowchart titled Approach to the Wheezing Child, branching by age of onset, wheeze pattern, and red flag features. Clean clinical educational diagram style."