Congenital Cardiac Defects

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Dr. Mikey Bryant

Introduction

- Congenital cardiac defects may be associated with other conditions or may be stand-alone conditions.
- They may be diagnosed by murmurs, but it is important to consider the complete clinical picture.
- These findings may be confirmed by echocardiography.
- There are interventions which can prolong a baby’s life.

When to suspect cardiac disease:

- Pulse oximetry not improving despite oxygen/CPAP.
- Loud murmur.
- Cyanosis.
- Poor weight gain.
- Recurrent chest infections.
- Reduced spO2 on lying flat.

Thinking about murmurs:

- Location:

- Lower left sternal edge: more likely to be a VSD, often loud.
- Upper sternal edge: Aortic Stenosis or pulmonary stenosis.
- Base of neck: consider aortic valve lesion.

Is it pathological?

- Characteristic features include:

- All diastolic murmurs.
- All pansystolic murmurs.
- Late systolic murmurs.
- Loud murmurs > 3/6.
- Other associated cardiac abnormalities.
- Other symptoms or signs e.g. SOB, fatigue, faltering growth, clubbing.

Classification:

- Cyanotic: Presence of bluish discolouration over the peripheries and central areas, may include the mouth.

- Acyanotic: absence of cyanosis.

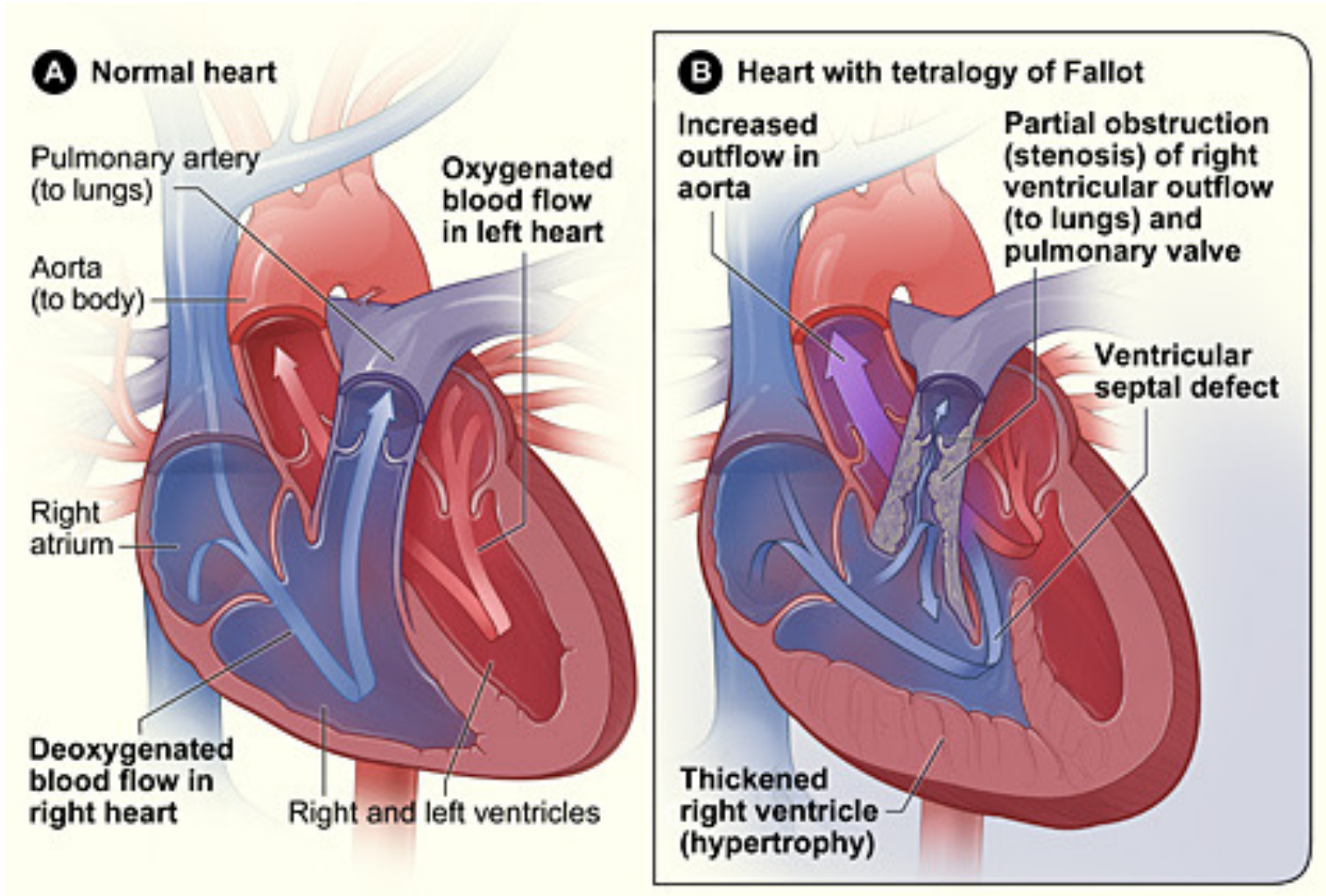
Cyanotic lesions: tetralogy of Fallot:

- 4 classic features:

1. Large VSD
2. Over-riding aorta.
3. Right Ventricular Hypertrophy.
4. Right Ventricular outflow obstruction.

Deoxygenated blood is shunted through the VSD and into the aorta as the pulmonary artery is stenosed. This leads to cyanosis.

Tetralogy of Fallot:



Tetralogy of Fallot:

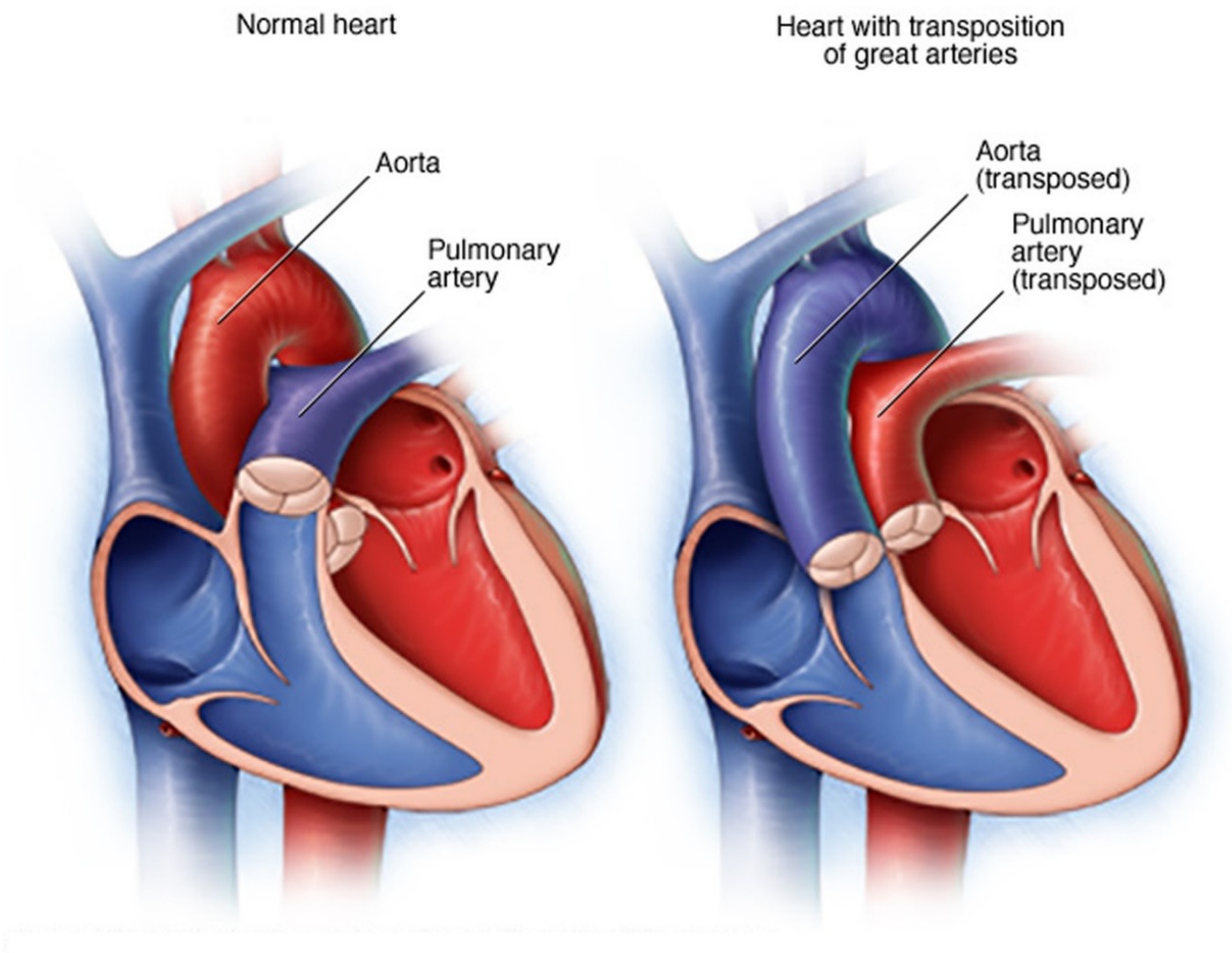
- Cyanosis, but not usually from birth.
- Clubbing.
- Paroxysmal hypercyanotic spells: Spontaneous and unpredictable in onset with tachycardia, tachypnoea and cyanosis. These are followed by episodes of becoming white and floppy, with syncopal symptoms.

- Management generally requires surgery in addition to prostaglandin E infusions.
- Hypercyanotic spells require management with oxygen, IV phenylephrine, morphine and propranol with urgent cardiac intervention.

Transposition of the Great Arteries:

- 2 separate circulatory systems with blood from the right side of the heart returned straight into the systemic circulation via the aorta.
- Blood from left side of heart goes back to the lungs via pulmonary artery.
- Only survivable if associated with a large VSD or PDA, otherwise the child will die early.
- Present with cyanosis from birth, duct dependent.
- Requires urgent surgical intervention.

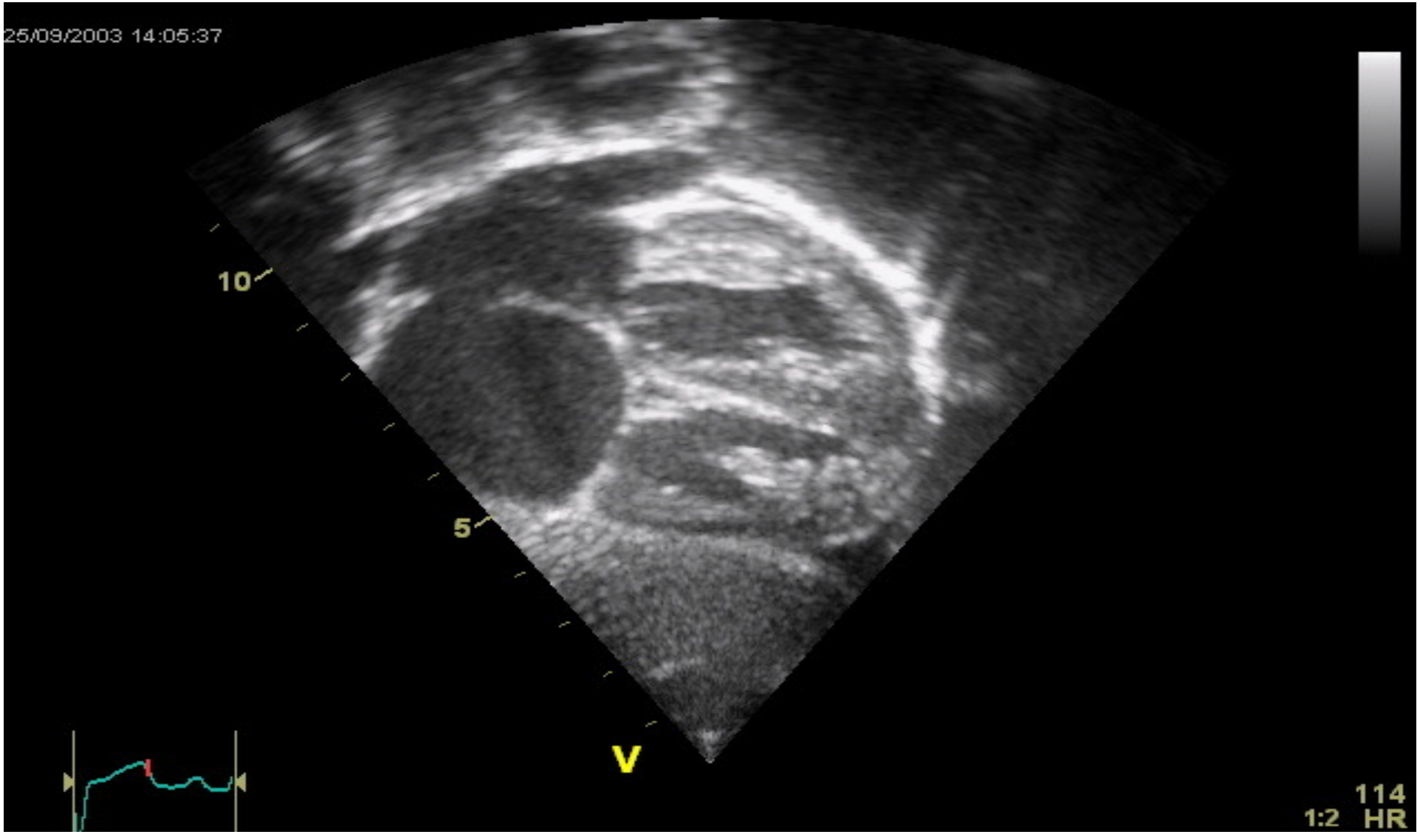
Transposition of the Great Arteries:



Acyanotic lesions: Atrial septal defect:

- This occurs when the septum between the atria is defective or absent.
- Patent Foramen Ovale is a form of this defect, which occurs when the blood continues to flow between atria after birth.
- Ostium secundum is the more common of the two defects, may be asymptomatic.
- Ostium primum defects are more serious and may well be associated with AVSDs or other cardiac defects, and also are more likely to be symptomatic.

Ostium Secundum defect:

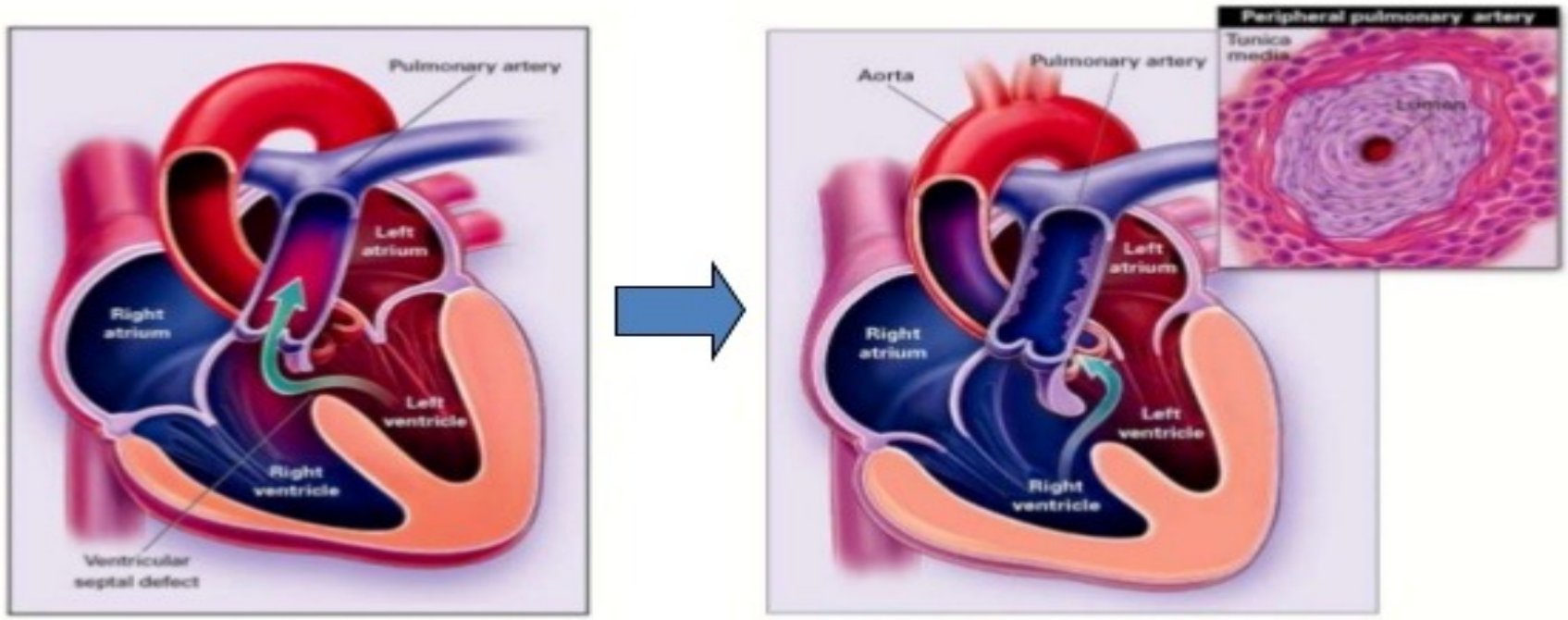


Presentation:

- ASDs may present with symptoms of shortness of breath on exertion, in addition to symptoms and signs of cardiac failure.
- There may also be symptoms of chest pain and sudden collapse.

- If not treated, eisenmenger's syndrome may occur, where the shunt from left to right causes chronic increased pressure in the pulmonary vasculature. This leads to reversal of the shunt so that it is from right to left. This means that deoxygenated blood returning to the right atrium from the heart is shunted into the body, leading to cyanosis.

Eisenmenger Syndrome:



NEJM 2000; 342(5); 334-342

Ventricular Septal Defect:

- 0.25% population.
- May initially be asymptomatic.
- Followed by worsening breathlessness in addition to recurrent chest infections.
- Cyanosis if eisenmengers (more rare).
- Babies often not able to feed, with poor weight gain.
- Complications include endocarditis.
- Classic finding is a loud pan-systolic murmur left lower sternal edge.

Management and prognosis:

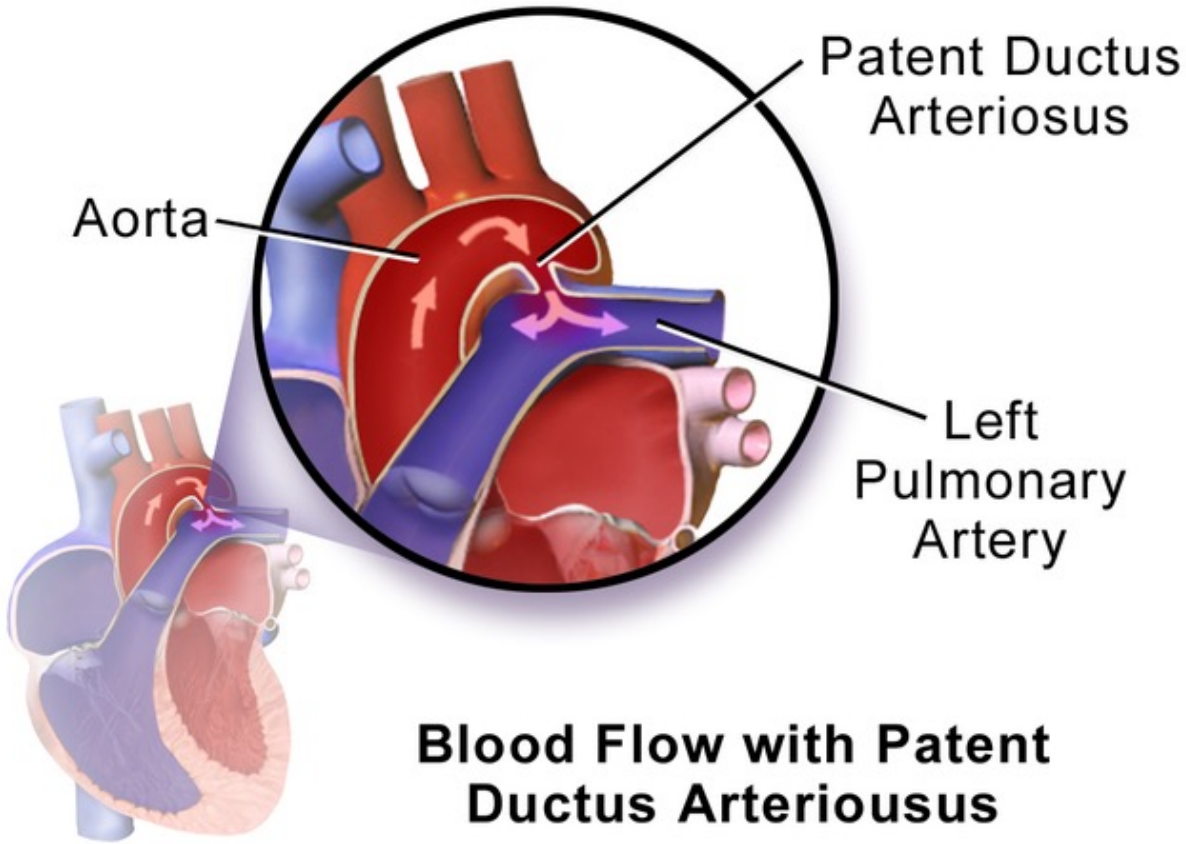
- Ostium secundum defects all require closure surgically, even if not symptomatic.
- Ostium primum defects usually require surgery in order to prevent congestive cardiac failure or pulmonary hypertension.
- VSDs are more likely to close by age 1 year, but require surgery if there are signs of cardiac failure or pulmonary hypertension.

Patent Ductus Arteriosus

- 1-2/1000 live births.
- Common in preterms.
- Blood flows back into the pulmonary artery from the aorta.

- This leads to low diastolic pressure with a wide pulse pressure or bounding peripheral pulses.
- Continuous machinery murmur.
- Managed initially with diuretics, cardiac catheterisation or surgical ligation.

PDA:

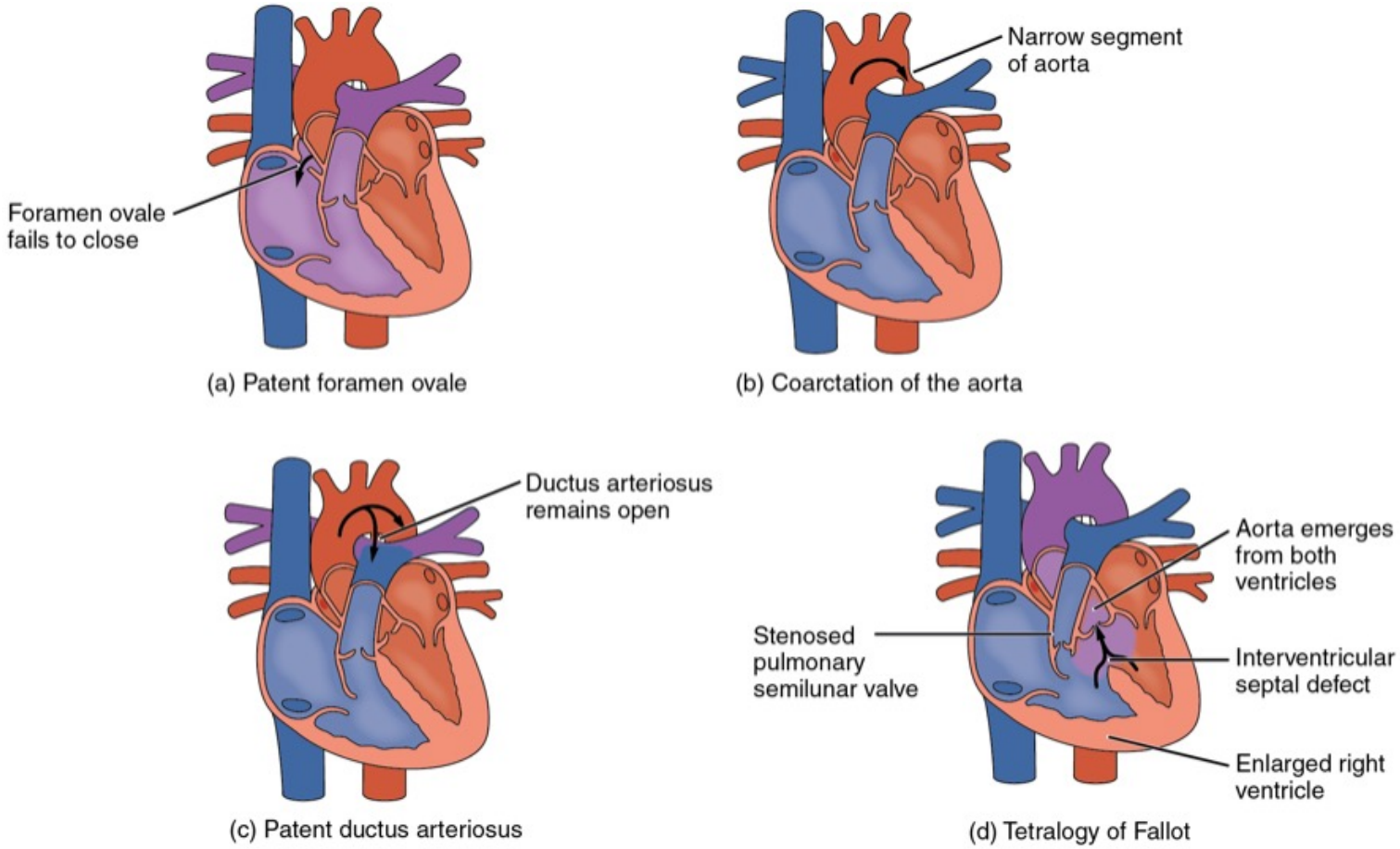

Patent Ductus Arteriosus
Aorta
Left Pulmonary Artery
Blood Flow with Patent Ductus Arteriousus

Co-arctation of the Aorta:

- Constriction of the aorta may occur at any point.
- Usually distal to the left subclavian artery.
- Commoner in boys, associated with Turner’s syndrome.
- May present with collapse in the neonatal period and cardiac failure.

- Hallmark is weak or absent femoral pulses. Different pulse pressure seen.
- Murmur in the upper left sternal edge.
- Management requires early surgery.

Overview:


Foramen ovale
fails to close
(a) Patent foramen ovale
Narrow segment
of aorta
(b) Coarctation of the aorta
Ductus arteriosus
remains open
(c) Patent ductus arteriosus
Aorta emerges
from both
ventricles
Stenosed
pulmonary
semilunar valve
Interventricular
septal defect
Enlarged right
ventricle
(d) Tetralogy of Fallot

Advice for cardiac failure:

- Early oxygen and nurse at 45 degrees.
- Use fluids sparingly, avoid boluses.
- Consider furosemide at 1-2mg per kg.
- Consider digoxin if tachycardic.
- Monitor for signs of renal failure or shock.
- Consider ace inhibitors if long term management possible.
- Monitor for signs of endocarditis.

Summary:

- Consider the whole cardiac picture, including the impact on the respiratory system.
- Assess for cyanosis.
- Continue with ongoing treatment of other conditions.
- Ensure that patients followed up well and options explored.