

# PAEDIATRIC EMQs



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# CONGENITAL HEART DISEASE

## Questions

The following are all forms of congenital heart disease:

- A. Aortic stenosis
- B. Atrial septal defect
- C. Atrioventricular septal defect
- D. Coarctation of the aorta
- E. Patent ductus arteriosus
- F. Pulmonary stenosis
- G. Tetralogy of Fallot
- H. Transposition of the great arteries
- I. Tricuspid atresia
- J. Ventricular septal defect

Select the most likely diagnosis for each of the following children.

Note: Each answer may be used more than once.

1. A 2 week old baby was born at 26 weeks gestation. It has not been possible to take the baby off the ventilator. There is a loud, continuous murmur heard best under the left clavicle.
2. A 6 day old baby is brought by ambulance to hospital. He is breathless, pale and only responding to painful stimuli. He has weak radial pulses and absent femoral pulses. No murmur is heard. He has hepatomegaly.
3. A 3 year old boy is seen by a paediatrician for a chest infection. During the examination a loud systolic murmur is heard. It is loudest at the left sternal edge. The boy's mother reports that he is under follow-up with a cardiologist for his murmur and that no treatment is required as it is likely to resolve.
4. A 3 day old baby becomes severely unwell with profound cyanosis. There is no cardiac murmur but a chest x-ray shows increased pulmonary vascular markings, a narrow upper mediastinum and "egg on side" cardiac shadow. The child has an emergency cardiac catheterisation procedure.
5. A newborn baby is found to have Trisomy 21 (Down Syndrome). At birth the baby is cyanosed and remains so 12 hours later. There is no cardiac murmur but an ECG shows a superior QRS axis. After an echocardiogram the baby's parents are told that the baby will need a heart operation in 3 – 6 months time.

# CONGENITAL HEART DISEASE

## Answers

Congenital heart disease comprises the most common group of childhood structural malformations; almost 1% of newborns have a significant malformation. Hence, it is important to maintain a high index of suspicion for these conditions.

1. E – Patent ductus arteriosus

In the majority of children the ductus arteriosus closes in the first few days after birth. Failure to close means that blood can flow from the aorta to the pulmonary artery (especially when the pulmonary vascular resistance falls after birth). When the pulmonary arterial blood pressure is sufficiently low shunting of blood will occur throughout the cardiac cycle giving rise to the characteristic continuous murmur. A bounding or collapsing pulse may be palpable. Prematurity significantly increases the risk of PDA.

2. D – Coarctation of the aorta

Coarctation of the aorta (CoA) often presents in later life due the consequences of hypertension or hypertension is found incidentally. However, it is important to recognise that in some cases it can present in a severe fashion in the neonatal period, typically when the ductus arteriosus closes. Symptoms include poor feeding, tachypnea, and lethargy before progressing to overt heart failure. Cardiac output may be too low to produce a murmur but differential arm and leg pulses may be a clue to the diagnosis.

3. J – Ventricular septal defect

Ventricular septal defect (VSD) is a relatively common problem and accounts for around a third of congenital heart disease. Small lesions typically have a louder murmur than larger lesions. The majority of small VSDs will close spontaneously.

4. H - Transposition of the great arteries

In this condition the aorta receives blood from the right ventricle and the pulmonary artery receives blood from the left ventricle. Without other frequently occurring abnormalities such as VSD, ASD or PDA. Cyanosis is the predominant feature. The so-called “egg on side” cardiac outline on CXR with a narrow pedicle and increase pulmonary vascular markings are features of transposition. If there is insufficient mixing of the two circulations when the ductus arteriosus closes it may be necessary to perform an emergency life-saving atrial septostomy (a hole is made between the two atria by a cardiac catheterisation procedure).

5. C - Atrioventricular septal defect

An atrioventricular septal defect (AVSD) is the most common form of heart disease found in association with Trisomy 21. There is often no murmur but there is almost invariably a superior QRS axis on ECG. It is important to understand the relationship between certain conditions and congenital heart disease as this should prompt screening by echocardiography.

## Further Reading

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