Cardiology

In uncomplicated patent ductus arteriosus:

A	CXR shows evidence of pulmonary plethora	(True)
B	raised audibility of murmur when standing	(False)
C	there is evidence of LVH on ECG	(True)
D	decreased pulse volume	(False)
E	cyanosis is present	(False)

Comments:
The ductus arteriosus is a normal vascular channel during intrauterine life. It is a large vessel with a muscular wall which courses between the pulmonary artery and the aorta. The ductus arteriosus normally closes within the first 48 hours of life. If it remains patent longer than this it is unlikely to close spontaneously. The exception is in premature babies where closure should occur within 3 months. A persistently patent ductus is a common congenital heart lesion, occurring either singly or in combination with other defects.

It is more common in: girls than in boys, congenital rubella syndrome, premature babies

Features include increased cardiac output, increased pulmonary flow (plethora on CXR) LVH due to increased output, increased pulse volume. Cyanosis is not a feature.

At birth the following circulatory changes occur

A	A rise in right atrial pressure	(False)
B	Flap closure of the foramen ovale	(True)
C	Anatomical closure of the ductus arteriosus	(False)
D	Functional closure of the ductus venosus	(True)
E	A 20-fold increase in lung blood flow	(False)

Comments:

At the first breath, air fills the lungs and pulmonary vascular resistance falls. Blood flow to the lungs increases from 10 to 50% of cardiac output, and the increased pulmonary venous return raises left atrial pressure. There is functional closure of the ductus arteriosus and venosus. Ligation of the umbilicus increases systemic resistance.

Ostium secundum atrial septal defect:

A	often leads to atrial fibrillation during the second decade	(False)
B	is associated with left axis deviation on the ECG	(False)
C	is associated with mitral valve prolapse	(True)

D	accounts for the majority of cases of ASD	(True)
E	is generally thought to require surgical correction if the pulmonary blood flow is greater than twice the systemic	(True)

Comments:
a-Not that young. b-Long PR and RBBB. RAD with secundum, LAD with primum defect.

The following associations are recognised:

A	Down's Syndrome and SD	(True)
B	Maternal tyrosinaemia and congenital heart disease	(False)
C	Fragile X Syndrome and VSD	(False)
D	Noonan's Syndrome ad cardiomyopathy	(True)
E	Maternal phenylketonuria and congenital heart disease.	(True)

Comments:

Down's Syndrome is associated particularly with AV canal defects, VSDs and ASDs. Fragile X Syndrome is associated with mitral valve prolapse and aortic route dilatation. Noonan's Syndrome is associated with pulmonary stenosis (dysplastic pulmonary valve), ASD and cardiomyopathy. Maternal phenylketonuria is associated with VSD, ASD, PDA, and coarctation of the aorta.

The following cutaneous stigmata may be associated with congenital heart disease:

A	Steinberg's sign.	(True)
B	Hypotelorism	(True)
C	Absent radius.	(True)
D	Distal triradius.	(True)
E	Low hair-line	(True)

Comments:
A, B, C, D, E Steinberg's sign is where the thumb, when held in the clenched hand protrudes beyond the palm. This is characteristic of Marfan's Syndrome, and suggest arachnodactyly. Hypotelorism is found in Patau's Syndrome, which is associated with a number of cardiac defects including VSD, ASD, PDA, coarctation and bicuspid aortical pulmonary valves. TAR Syndrome (thrombocytopenia and absent radius) is associated with ASD and Tetralogy of Fallot. A distal triradius is found in Down's Syndrome which is associated with atrioventricular septal defects, and a low hairline in Turner's Syndrome which is associated with coarctation.

In the fetal circulation

A	Approximately each volumes of blood are pumped by the left and right ventricles.	(True)
B	The saturation of blood returning from the placenta is about 80%.	(True)
C	Approximately 25% of cardiac output traverses the fetal lung.	(False)
D	The ductus venosus is probe-patent in 75% of cases.	(False)
E	Pulmonary vascular resistance is suprasystemic.	(True)

Comments:
A, B, E In the fetal circulation, the vast majority of blood bypasses the lungs through the ductus arteriosus, but the right and left ventricles pump approximately equal volumes. The fetus is relatively hypoxaemic, with fetal haemoglobin being used to increase oxygen carriage to the tissues. Only 10% of cardiac output traverses the fetal lung, and the ductus venosus remains patent until the postnatal period. Approximately 50% of umbilical venous catheters can be passed through it into the right atrium postnatally. The pulmonary-vascular resistance remains suprasystemic until the time of the first breath, when the combination of lung aeration reducing pulmonary blood pressure, and umbilical ligation increasing systemic blood pressure reverses the situation.

Nitric oxide (endothelial relaxation factor):

A	Unlike prostacyclin, promotes platelet aggregation.	(False)
B	Can be enzymatically synthesised from glyceryl trinitrate.	(True)
C	Has a half life of 1-2 minutes in vivo.	(False)
D	Can be synthesised by macrophages.	(True)
E	Is synthesised from L-arginine.	(True)

Comments:
B, D, E Prostaglandin I2 inhibits platelet aggregation via cAMP, while nitric oxide works via cGMP. Glycerol trinitrate and nitroprusside are converted to nitric oxide before having their pharmacological effects. Nitric oxide is synthesised by macrophages, lymphocytes, endothelial cells, and is a neurotransmitter and vasodilator. When given by inhalation it is a specific pulmonary vasodilator because of its short half life.

In mitral stenosis caused by rheumatic heart disease, prophylaxis against endocarditis is unnecessary in:

A	cystoscopy	(False)
B	dental scaling	(False)
C	caesarean section	(True)
D	cardiac catherization	(True)
E	transesophageal echocardiogram	(True)

Comments:
The **American College of Cardiology guidelines** recommend antibiotic prophylaxis for dental work including scaling, cystoscopy, endoscopy, bronchoscopy and biliary tract surgery. It is not recommended

for Caesarian section, ToE (except high risk cases) and Cardiac catheterisation.

Which of the following are true of Wolf-Parkinson-White syndrome?

A	there is an abberant conduction between atria and ventricles	(True)
B	broad complex tachycardia is more frequent than narrow-complex tacchycardia	(True)
C	verapamil is the treatment of choice for an associated SVT	(False)
D	Amiodarone increases the refractory period in the accessory path.	(True)
E	Atrial fibrillation is a well recognised rhythm disturbance.	(True)

Comments:
a-Bundle of Kent. b-Although orthodromic tachycardia is the commonest it frequently has phasic aberrant conduction making it broad. Verapmil may exacerbate the rhythm disturbance associated with WPW and AF may result in VF.

Myocarditis is a recognised association of:

A	Salmonella infection.	(True)
B	Mycoplasma infection.	(True)
C	Lyme Disease.	(True)
D	Measles.	(True)
E	Hurler's Syndrome.	(False)

Comments:
A, B, C, D Hurler's is associated with a cardiomyopathy.

The following are features of aortic coarctation in infancy:

A	Rib-notching.	(False)
B	Inverted T wave in V6.	(True)
C	Ejection systolic murmur between the shoulder blades.	(True)
D	Radio-femoral delay.	(True)
E	Plateau pulses.	(False)

Comments:
B, C, D Rib notching is a late sign in adolescents, and is rarely seen these days. An inverted T wave in V6 suggests left ventricular strain. Plateau pulses are a feature of aortic stenosis.

Pulsus paradoxus is found with

A	a severe asthmatic attack	(True)
B	severe left ventricular failure	(False)
C	myocardial disease	(True)
D	constrictive pericarditis	(True)
E	cardiac amyloidosis	(True)

Comments:
Pulsus paradoxus is the exaggerated fall of pressure associated with inspiration due to disease process like asthma, constrictive pericarditis, tamponade and left ventricular disease (cardiomyopathy). It is not a feature of left ventricular failure.

Giant "a" waves in the jugular vein can occur in:

A	Constrictive pericarditis.	(True)
B	Pulmonary hypertension.	(True)
C	Aortic regurgitation.	(False)
D	Tricuspid stenosis.	(True)
E	Thyrotoxicosis.	(False)

Comments:
A, B, D "a" waves are the positive deflection in the jugular venous pulse following right atrial contraction. This becomes giant in situations where the atrium is hypertrophied or contracts against resistance, such as constrictive pericarditis, pulmonary hypertension, and tricuspid stenosis or atresia.

Immediately after a large haemorrhage:

A	pulse pressure is increased	(False)
B	thirst occurs	(True)
C	anaerobic glycolysis increases	(True)
D	coronary vasoconstriction occurs	(False)
E	carotid chemoreceptors are inhibited	(False)

Comments:
There is a precipitant fall in pulse pressure with narrowing. Thirst is a consequence of ADH secretion and stimulation of the thirst receptors. Anaerobic glycolysis occurs due to reduced perfusion with increased propensity to lactic acidosis. Coronary vasodilatation would be expected and chemoreceptors would of course be stimulated.

The following are true of cardiac transplantation:

A	the donor heart is provided to the recipient with the best HLA match	(False)
B	long term hypotension is a problem	(False)

C	recipients suffer less angina but accelerated coronary artery disease	(True)
D	the histology of atheroma in the transplanted heart is the same as that of typical atherosclerosis	(False)
E	75% 5 year survival	(True)

Comments:
a-This is true of kidney transplants but hearts go to the most needy. b-Hypertension is a significant problem related to use of cyclosporin and denervation of the heart. c-The heart is denervated. d-The pathology is different being more diffuse and symmetrical. e-For most centres with conventional selection criteria.

The following findings suggest that a murmur is innocent:

A	It is grade 3/6	(False)
B	It has a vibratory quality.	(True)
C	It is loudest at the apex.	(False)
D	It is only heard when the patient is febrile.	(True)
E	It changes in intensity with posture.	(True)

Comments:
B, D, E About 30% of children can have innocent murmurs. These are usually ejection generated by the outflow tracts of the left of right side of the heart, or venous hums due to turbulent flow in the head and neck veins. The hallmarks are their localised nature; the fact they are confined to systole; the fact they are soft; and that they are associated with no symptoms or other signs.

The following are recognised causes of cyanosis in the newborn:

A	Persistent fetal circulation.	(True)
B	Severe anaemia.	(False)
C	Congenital pneumonia.	(True)
D	Transient tachypnoea of the newborn.	(True)
E	Hypoplastic left heart syndrome.	(True)

Comments:
A, C, D, E Cyanosis can be caused by non-cardiac or cardiac lesions. The former include: persistent fetal circulation, hyaline membrane disease, congenital pneumonia and transient tachypnoea of the newborn. The latter include lesions due to abnormal mixing (TGA, univentricular heart) or to decrease pulmonary blood flow (pulmonary atresia, Fallot's Tetralogy). Hypoplastic left heart syndrome can lead to severe congestive cardiac failure, shock and secondary cyanosis.

The following are associated with sudden death in childhood:

A	Aortic stenosis	(True)
B	Primary pulmonary hypertension	(True)

C	Atrioventricular septal defect	(False)
D	Fallot's tetralogy	(True)
E	Cardiomyopathy	(True)

Comments:

Sudden death may be caused by tricyclic antidepressants, haemosiderosis, SIDS, metabolic disorders such as MCAD, child abuse, trauma, hyperthermia, asthma, meningitis/septicaemia, bacterial endocarditis, pertussis, cholera, RSV, gastro-oesophageal reflux, Reye Syndrome, unrecognised diaphragmatic hernia, upper respiratory obstruction, pulmonary thromboembolism, prolonged QT Syndrome, aortic stenosis, mitral valve prolapse, aspiration, anomalous right coronary artery or left coronary artery, Tetralogy of Fallot, pulmonary atresia intact septum, tricuspid atresia, transposition of the great arteries, arrhythmia, coronary artery disease in Hurler's Syndrome, calcinosis of the coronary arteries, viral myocarditis, primary cardiomyopathy, hypertrophic cardiomyopathy, Marfan's Syndrome, achondroplasia secondary to cervical cord compression, and volatile substance abuse.

A short PR interval on the ECG is associated with

A	hypertrophic obstructive cardiomyopathy	(True)
B	dystrophia myotonica	(False)
C	Lown-Ganong-Levine syndrome	(True)
D	rheumatic carditis	(False)
E	Duchenne muscular dystrophy	(True)

Comments:

a - In some cases a short PR interval may be seen the mechanism is not undertood. These cases are often associated with QRS changes suggestive of pre-excitation.

b+d = long PR interval

A short PR interval on the ECG is associated with:

A	Duchenne muscular dystrophy	(True)
B	dystrophia myotonica	(False)
C	Friedrich's ataxia	(True)
D	rheumatic carditis	(False)
E	hypertrophic cardiomyopathy	(True)

Comments:
a-It may be associated with accelerated AV conduction. b-Associated with a prolonged PR interval which may manifest before the overt clinical manifestations of the disease. (Am J Med 1978(61), 452). c-In a quarter of cases (JACC 1986(7), 1370-8). d-Long PR. e-In some cases HOCM is associated with a short PR. Commoner causes are WPW and Lown-Ganong-Levine syndromes.

The following are recognised treatments for Fallot's spell:

A	Noradrenaline infusion	(True)
B	Heel-chest position.	(True)
C	Morphine 0.1mg/kg.	(True)
D	Prostaglandin E2 50mg/kg/min.	(False)
E	Propranolol 0.1mg/kg IV.	(True)

Comments:
A, B, C, E The Fallot's spell is caused by a sudden increase in right ventricular outflow tract obstruction caused by spasm of the infundibulum. There is an acute decrease of blood circulating around the lungs and the child becomes cyanosed, hypoxic, and may loose consciousness. Treatment is 100% oxygen via face mask, Morphine intramuscularly or intravenously, and/or Propranolol. The heel-chest position is designed to increase systemic vascular resistance and decrease shunting in the heart, and Noradrenaline performs the same function. Although Prostaglandin infusion may be useful in maintaining duct patency during transfer to an acute cardiac centre, it is unlikely to be of benefit in the acute situation.

A 6 year old boy presents with fever. The following might contribute to a diagnosis of rheumatic fever:

A	The finding of target lesions on the hands.	(False)
B	The finding of tender nodules in the fingertips.	(False)
C	A prolonged PR interval on ECG.	(True)
D	A CRP of 10.	(False)
E	Positive Romberg's sign.	(False)

Comments:

The modified Jones Criteria include: Finding of preceeding streptococcal infection (recent scarlett fever, raised ASOT or other streptococcal antibodies, positive throat swab for Group A Strep). Plus:

- a) MAJOR CRITERIA:
  - Carditis
  - Polyarthritis
  - Chorea
  - Subcutaneous nodules
  - Erythema marginatum.
- b) MINOR CRITERIA:
  - Fever
  - Arthralgia
  - Previous history of rheumatic fever
  - Elevated acute phase reactions
  - Prolonged PR interval.

Erythema marginatum involves red circular lesions which gradually enlarge with central clearing. Sydenham's chorea consists of choreoathetoid movements with increased clumsiness, e.g. deteriorating handwriting. This is often associated with emotional lability. Target lesions suggest erythema multiforme. A CRP of 10 is not elevated much beyond the normal range. Erythema marginatum initially manifests as

non-specific pink macules seen over the trunk, with later blanching in the middle of the lesions and sometimes fusing of the borders resulting in a serpiginous (serpent-like) looking lesion. The rash is worsened with heat, but is characteristically evanescent. It does not itch, and can be mistaken for the rash of Lyme disease. Sub-cutaneous nodules are pea-sized, firm and non-tender. There is no associated inflammation and they are characteristically seen on the extensor surfaces of joints such as knees and elbows and also over the spine.

Diastolic dysfunction of the left ventricle may be a dominant feature of:

A	Hypertrophic obstructive cardiomyopathy.	(True)
B	Endocardial fibroelastosis.	(True)
C	Doxorubicin toxicity.	(True)
D	Anomalous left coronary artery.	(True)
E	Myocardial infarction after Kawasaki Disease.	(True)

Comments:
A, B, C, D, E Diastolic dysfunction is a particular feature of anthracycline chemotherapy, iron overload, and heart rejection following transplantation. It may also be seen in cardiac ischaemia.

The following are recognised causes of an infant cyanosed from day 2:

A	Eisenmenger's Syndrome.	(False)
B	Ebstein's anomaly.	(True)
C	Pulmonary atresia.	(True)
D	Transposition of the great arteries.	(True)
E	Total anomalous pulmonary venous drainage.	(True)

Comments:
B, C, D, E Eisenmenger's Syndrome is a late complication of left to right shunt, where pulmonary hypertension leads to shunt reversal. Ebstein's anomaly is due to a distally placed tricuspid valve with small ventricle and very large right atrium. This results in tricuspid regurgitation with right to left shunting at atrial level producing cyanosis. Usually patients are well into adult life, but newborn presentation is described. TAPVD refers to a situation where pulmonary veins drain via an aberrant confluence into the right side of the circulation. The aberrant vessel may become obstructed causing pulmonary plethora, but a normal cardio-thoracic ratio. Alternatively, venous return to the right side may be reduced such that the child becomes cyanosed.

A 7-year-old girl with Down's syndrome has central cyanosis. Examination of her chest shows only a soft mid-systolic murmur at the apex and in her abdomen there is a well healed scar in the epigastrium dating from the newborn period:

A	Her haematocrit is likely to be 30%	(False)
B	She has Fallot's tetralogy	(False)

C	Investigation of her blood gases shows carbon dioxide retention	(False)
D	The newborn period is likely to have been complicated by Hirschsprung's disease	(False)
E	Dental extraction should be covered by antibiotic prophylaxis	(True)

Comments:
Most probably she will have secondary polycythaemia and a raised haematocrit. She most likely has Eisenmonger's syndrome secondary to a reversed VSD shunt. She has cyanotic heart disease and the blood gases will show hypoxia rather than CO2 retention . Hirschprung's disease (congenital aganglionic megacolon) occurs more frequently in patients with Down's syndrome but it is still a relatively uncommon development. She is a high risk for the development of SBE and therefore she should receive antibiotic prophylaxis.

A 7 day old baby presents with cyanosis. This is pathophysiologically due to abnormal mixing in the following conditions:

A	Tetralogy of Fallot.	(False)
B	Transposition of the great arteries.	(True)
C	Total anomalous pulmonary venous drainage.	(True)
D	Univentricular heart.	(True)
E	Aortic coarctation.	(False)

Comments:
B, C, D 'Abnormal mixing' refers to those conditions where systemic and pulmonary circulations mingle. In Fallot's, cyanosis is caused by decreased pulmonary blood flow. Aortic coarctation does not cause cyanosis.
Question supplied by Colin Melville Consultant Paediatrician

Giant 'a' waves in the JVP occur in:

A	pulmonary hypertension	(True)
B	aortic regurgitation	(False)
C	thyrotoxicosis	(False)
D	constrictive pericarditis	(False)
E	tricuspid stenosis	(True)

Comments:
a - Giant 'a' waves occur when there is a poorly compliant right ventricle (or tricuspid stenosis) increasing the impedence against which the right atrium has to eject blood. d - In constrictive pericarditis the JVP is high with an abrupt fall in systole (x descent) and may rise with inspiration (Kussmaul's sign).

In a 2 year old child with persistent chest symptoms, the following should be considered:

A	Maternal smoking.	(True)
B	ASD.	(True)

C	VSD.	(True)
D	PDA.	(True)
E	Aortic coarctation.	(False)

Comments:
A, B, C, D In the neonatal period, aortic coarctation presents with shock. Later, an inter-scapular murmur or absent femoral pulses are commonest. In the older child, hypertension is likely. In a child with persistent chestiness, attendance at nursery, asthma, cystic fibrosis, and minor immune deficiency should also be considered.

In a child with patent ductus arteriosus, the following may be important aetiologically

A	Maternal SLE	(False)
B	Maternal alcoholism	(False)
C	Maternal Warfarin therapy	(True)
D	Maternal CMV infection	(False)
E	Maternal rubella infection	(True)

Comments:

SLE in the mother is associated with Ro antibody and congenital heart block. The mother may be presymptomatic. Fetal alcoholism is associated with ASD, VSD and Fallot's. CMV us not associated with cardiac defects, but rubella is associated with PDA and peripheral pulmonary stenosis. Congenital CMV causes IUGR, hepatitis, splenomegaly, jaundice, microcephaly, intra-cranial calcification and deafness.

Recognised toxic effects of Amiodarone include:

A	Hyperthyroidism	(True)
B	Peripheral neuropathy	(True)
C	Hepatitis	(True)
D	Hypothyroidism	(True)
E	Photosensitivity	(True)

Comments:

Amiodarone is a potent anti-arrhythmic useful in all forms of dysrhythmias. Caution is required in renal impairment and there is a risk of thyroid dysfunction with accumulation of iodine. It enhances the effects of Warfarin and increased Digoxin, Phenytoin and Cyclosporin levels. There is an increased risk of bradycardia and AV block and myocardial depression with betablockers and calcium channel blockers. Toxicity is increased if hypokalaemia occurs with diuretics. Reversible corneal microdeposits, optic neuritis, peripheral neuropathy and myopathy, bradycardia and conduction disturbances, phototoxicity,

and rarely a persistent slate grey discolouration, hypo and hyperthyroidism, diffuse pulmonary alveolitis in pneumonitis and fibrosis, disturbed liver function tests, jaundice, hepatitis and cirrhosis are all reported.

In patients with congenital heart disease:

A	Approximately 10-15% will have more than one cardiac abnormality.	(True)
B	The majority of lesions can be diagnosed antenatally using ultrasound.	(True)
C	The majority of infants will require cardiac catheterisation.	(False)
D	Complex defects usually require staged surgical procedures.	(False)
E	Approximately 25-30% will also have a non-cardiac abnormality.	(False)

Comments:
B, E The majority of patients with CHD can be diagnosed antenatally by specially trained operators. Postnatal echocardiography will diagnose most lesions, and catheterisation is less necessary. Most defects can be corrected in the neonatal period as a single stage procedure. About 10-15% have an associated non-cardiac abnormality.

The following are features of mitral stenosis:

A	right axis deviation on the ECG	(True)
B	increased pulmonary artery capillary wedge pressure	(True)
C	left parasternal heave	(True)
D	palpable 1st heart sound	(True)
E	palpable 2nd heart sound	(True)

Comments:
This lesion is almost always (99%) due to rheumatic heart disease although a clear history is obtained in only 50% of cases. 25% of patients with rheumatic heart disease have pure mitral stenosis, two-thirds are female. In parallel with the incidence of rheumatic fever, mitral stenosis is more common and generally more severe in developing countries. It causes elevated right sided pressures with a parasternal heave reflecting RVH. The tapping apex beat signifies a palpable 1st heart sound. With pulmonary hypertension, a palpable second sound may be felt.

The following may be found in atrioventricular septal defects:

A	RSR1 pattern in V1.	(False)
B	Mild cyanosis in the early stages.	(False)
C	A pan-systolic murmur at the apex.	(True)
D	Wide but variable splitting of the second heart sound.	(False)
E	An ejection murmur at the upper left sternal border.	(True)

Comments:
C, E An RSR pattern is caused by right bundle branch block, and is seen in secundum ASD. An apical pan-systolic murmur is common, and is caused by mitral regurgitation. The second heart sound has wide fixed splitting. A pulmonary flow murmur is usual.
Question supplied by Colin Melville Consultant Paediatrician

Cerebral abscess in the absence of endocarditis is a recognised complication of:

A	tetralogy of Fallot	(True)
B	persistant ductus arteriosus	(False)
C	tricuspid artresia	(True)
D	transposition of the great arteries	(True)
E	atrial septal defect of the ostium primum type	(False)

Comments:
This is paradoxical embolization seen in right to left shunts. Associated with ASD of patent foramen ovale and ostium secundum types.

The following lesions can be diagnosed antenatally on a routine 4 chamber view at 18 weeks gestation:

A	Tetralogy of Fallot.	(False)
B	Hypoplastic left heart syndrome.	(True)
C	VSD.	(False)
D	Pulmonary stenosis.	(False)
E	Transposition of the great arteries.	(False)

Comments:
B The routine 4 chamber view is likely to pick up only severe lesions causing small right or left ventricles, such as hypoplastic left heart syndrome or pulmonary atresia.

In the diagnosis of rheumatic fever, the following may be helpful:

A	Polyarthritis.	(True)
B	ASO titre of less than 1:200.	(False)
C	Staphylococcus aureus grown on throat culture.	(False)
D	A generalised macular-papular rash.	(False)
E	Splinter haemorrhages.	(False)

Comments:
A Jones criteria require two major or one major and two minor, and evidence of recent streptococcal infection for the diagnosis of rheumatic fever. MAJOR: - Pancarditis. - Polyarthritis - Erythema

marginatum - Chorea - Subcutaneous nodules - The rash is macular. MINOR: - Fever - Polyarthralgia - History of RF - Raised ESR/CRP - Prolonged PR interval on ECG.

Causes of cyanotic heart disease with oligaemia include

A	Tricuspid atresia	(True)
B	Pulmonary atresia with ventricular septal defect and multiple aorto-pulmonary collateral arteries	(False)
C	Fallot's tetralogy	(True)
D	Transposition of the great arteries	(False)
E	Pulmonary stenosis	(True)

Comments:

Cyanotic heart disease can be divided into 2 groups: that with increased, or that with decreased pulmonary blood flow. The former include conditions such as transposition of the great arteries while the latter include tricuspid atresia and Fallot's Tetralogy.

The following are recognised causes of secondary hypertension:

A	Henoch Schonlein Purpura	(True)
B	Nephroblastoma	(True)
C	Neuroblastoma	(True)
D	Addison's Disease	(False)
E	Hypothyroidism	(False)

Comments:

Recognised secondary causes include:

1. RENAL:
    - Congenital: Dysplastic or polycystic kidneys.
    - Acquired: Glomerulonephritis, chronic pyelonephritis, reflux nephropathy, HUS, HSP, Wilm's tumour.
2. VASCULAR:
    - Coarctation.
    - Renal artery stenosis or embolisation, renal vein thrombosis.
3. ESSENTIAL..
4. ENDOCRINE:
    - Pheochromocytoma.
    - Neuroblastoma.
    - Cushing's Disease.
    - Conn's Syndrome.
    - Diabetes mellitus.
    - Adrenogenital syndrome.
    - Hyperthyroidism.
5. NEUROLOGICAL:
    - Neurofibromatosis.

- Raised intracranial pressure.
- Encephalitis.

6. MISCELLANEOUS:
   - lead poisoning.
   - Obesity.
   - Porphyria.
   - Drugs, e.g. steroids.

Which of the following procedures are appropriate?

A	Rastelli procedure for Taussig-Bing heart (double-outlet right ventricle with sub-pulmonary VSD).	(True)
B	Modified Blalock-Taussig shunt for critical aortic stenosis.	(False)
C	Balloon valvotomy for critical pulmonary stenosis.	(True)
D	Stage 1 Norwood procedure for hypoplastic left heart syndrome.	(True)
E	Fontan procedure for tricuspid atresia.	(True)

Comments:

A Rastelli operation involves using a pulmonary of aortic homograft conduit to relieve pulmonary obstruction in double outlet right ventricle with pulmonary stenosis (Taussig-Bing heart).

A Blalock-Taussig shunt is used to increase pulmonary blood flow in duct dependent cyanotic conditions, e.g. pulmonary atresia.

A Stage 1 Norwood Procedure for hypoplastic left heart syndrome involves atrial septectomy and transection and ligation of the distal main pulmonary artery. The paroximal pulmonary artery is then connected to the hypoplastic aortic arch, while the coarcted segment of the aorta is repaired. An aorto-pulmonary shunt is created to connect the aorta to the main pulmonary artery to provide pulmonary blood flow. A Fontan procedure is used to direct blood flow from the systemic veins to the pulmonary artery directly, bypassing the hypoplastic ventricle.

Recognised features of abetalipoproteinaemia include:

A	a high serum cholesterol	(False)
B	palmar xanthomas	(False)
C	advanced atherosclerotic vascular disease	(False)
D	abnormal red blood cell morphology	(True)
E	severe mental retardation	(False)

Comments:
Abetalipoproteinaemia causes neurodegenerative nervous system changes - ataxia with retinitis pigmentosa. IQ is usually normal. Other clinical manifestations of steatorrhoea and malabsorption which may lead to osteopenia and skeletal fractures. Radiologically, there is thickening of the mucosal folds of

the duodenum and jejunum. Inheritance is autosomal recessive.

The following are compatible with a secundum ASD:

A	Mid-diastolic rumble at the left sternal edge.	(True)
B	QRS axis of -30 degrees.	(False)
C	RSR pattern in V1.	(True)
D	Wide splitting of the second heart sound.	(True)
E	Pulmonic ejection click	(False)

Comments:
A, C, D Secundum ASD usually presents with a murmur due to increased blood flow through the pulmonary valve. If the shunt is large, then a mid-diastolic rumble may be heard at the apex due to increased mitral valve flow, but a tricuspid rumble does not occur. A superior axis deviation suggests a primum ASD, but an RSR pattern (M pattern) in V1 is characteristic. Wide fixed splitting of the second heart sound occurs because of equal filling of the left and right atria during all phases of the respiratory cycle. A pulmonic ejection click is characteristic of pulmonary valve stenosis.
Question Supplied by Colin Melville Consultant Paediatrician

The following features suggest Tetralogy of Fallot rather than transposition of the great arteries:

A	Presentation at 2 months of age with a murmur.	(True)
B	Absence of clinical cyanosis	(True)
C	Increased pulmonary vascular markings on chest x-ray.	(False)
D	Single second heart sound.	(False)
E	Absence of S wave in V1.	(True)

Comments:
A, B, E On chest x-ray, a 'boot-shaped' heart with pulmonary artery bay and uptilted apex suggests Fallot's, while an 'egg on side' appearance and pulmonary plethora suggests TGA. In Fallot's lung blood flow is decreased, and the absence of an S wave in V1 suggests right ventricular hypertrophy. The second heart sound is single in both conditions. Transposition usually presents in the first few days of life, and the patient is invariably clinically cyanosed.

The following are features of Fallot's tetralogy:

A	prominent systolic murmur of ventricular septal defect	(False)
B	pulmonary plethora	(False)
C	squatting	(True)

D	paradoxical embolus	(True)
E	presentation in late teens	(False)

Comments:
a-The systolic murmur comes from the pulmonary infundibular stenosis. b-There is usually pulmonary oligaemia as there is a right to left shunt. c-Squatting relieves the dyspnoea. d-Because of the right to left shunt at ventricular level. e-Presentation is invariably in childhood. Read here for more on **Fallot's**

Regarding cardiogenic shock:

A	Increased firing of neurones in the carotid body stimulates the vagus nerve.	(False)
B	Renal cortical necrosis is a recognised complication.	(True)
C	Constriction of the renal afferent arterioles stimulates renin production.	(True)
D	Angiotensin II production is increased in the liver.	(False)
E	ANP production is increased.	(False)

Comments:
B, C The vagus slows the heart, and angiotensin II is produced in the lungs. ANP production is increased in response to atrial stretching, and generally the effects are opposite to those of the renin-angiotensin-aldosterone system.
Question supplied by Colin Melville Consultant Paediatrician

Broad complex tachycardias:

A	May be associated with shock.	(True)
B	May be associated with pulselessness	(True)
C	Are usually treated with asynchronous DC shock.	(True)
D	Are uncommon in childhood.	(True)
E	May be due to supraventricular tachycardia.	(True)

Comments:
Wide complex tachycardias may be due to ventricular fibrillation, ventricular tachycardia, or supraventricular tachycardia with associated ventricular conduction defect. It may therefore be associated with shock and pulselessness. Unstable patients should therefore be treated as if they have ventricular tachycardia. Adenosine will distinguish the rare SVT with conduction defect, and Lignocaine can be tried in VT if no shock is present. Otherwise (and usually) asynchronous DC shock 0.5J/kg is used.

The following are recognised associations with pulmonary hypertension:

A	An apgar of 3 at 5 minutes.	(True)
B	Meconium aspiration	(True)

C	Hyaline membrane disease	(True)
D	Hypoglycaemia	(True)
E	Oligohydraminos	(True)

Comments:

Persistent fetal circulation occurs with all of these. It is usually treated using nitric oxide, a specific pulmonary vasodilator.

The following drugs are contra-indicated for the treatment of atrial fibrillation associated with the WPW syndrome:

A	Digoxin	(True)
B	Adenosine	(False)
C	Amiodarone	(False)
D	Verapamil	(True)
E	Sotalol	(False)

Comments:
In WPW digoxin or calcium channel blockers may increase anterograde conduction through the bypass tract, so they should be avoided.

The following are found in VSD:

A	Upright T wave in V1 in pulmonary hypertension.	(True)
B	Pulmonary plethora and a cardio-thoracic ratio of 60%.	(True)
C	Mild cyanosis in the first few days of life.	(False)
D	A loud second heart sound.	(True)
E	A parasternal thrill.	(True)

Comments:
A, B, D, E An upright T wave in V1 suggests right ventricular hypertrophy. Pulmonary plethora and increased CTR suggests right heart failure, usually due to left to right shunt. A loud second sound is caused by increased pulmonary artery pressure from the shunt. Cyanosis is not a feature of VSD.

Infective endocarditis rarely occurs with:

A	mitral valve prolapse	(False)
B	patent ductus arteriosus	(False)

C	bicuspid aortic valve	(False)
D	atrial septal defect	(True)
E	tight mitral stenosis	(True)

Comments:
a - Regurgitant mitral valve is most frequent site of endocarditis. Mitral valve prolapse has a high prevalence (7.6% females, 2.5% males in Framingham study). b - However early surgery has virtualy abolished patent ductus arteriosus. e - The rate is 1 - 5%. (**Read more ...**)

The following findings may occur with a cyanotic episode in an individual with Fallot's tetralogy:

A	bradycardia	(False)
B	pallor	(False)
C	increased systolic murmur	(False)
D	loss of consciousness	(True)
E	continuous murmur at the upper left sternal border	(True)

Comments:
Usually polycythaemic and cyanosed. The Murmur may becoma softer during a cyanotic episode and syncope can occur. The continuous murmur may still be audible. (Dr Jacob Easaw)

Rheumatic chorea

A	does not occur in conjunction with rheumatic fever	(False)
B	is not associated with muscular weakness	(False)
C	may be unilateral	(True)
D	responds rapidly to ACTH	(False)
E	movements cease during sleep	(True)

Comments:
Sydenham's Chorea/St Vitus dance is associated with Rheumatic fever and frequently seen in children. Unilateral chorea may be a feature and muscular weakness is associated. ACTH is not used for treatment and is treated with appropirate therapy of underlying condition.

An Early Diastolic Murmur at the left sternal edge is characteristic in:

A	Mitral stenosis	(False)
B	Ankylosing spondylitis	(True)
C	Pulmonary stenosis	(False)
D	SLE	(True)
E	Eisenmenger's syndrome	(True)

Comments:

A - There will be a rumbling low pitched mid diastolic murmur best heard in the mitral area. B - Due to AR. Other causes of AR include Rheumatic heart disease, Congenital bicuspid aortic valves, Hypertension, Dilated aortic root as in Marfan's, SBE, Syphilitic aortitis, Degenerative valve disease, SLE, Prolapse of a aortic cusp due to a VSD, Rheumatoid arthritis, Trauma, Takayasu's Disease. C - There will be a loud ejection murmur usually in the pulmonary area best heard on inspiration. E - Due to PR (Graham Steell murmur)due to pulmonary hypertension.

In the interpretation of ECGs, the following are correct:

A	Pulmonary hypertension id the P wave in lead II is greater than 2mm.	(False)
B	Inverted T waves in V5 and V6 are occasionally found in infants.	(False)
C	Mean QRS axis in the newborn lies between 60 and 160 degrees.	(True)
D	Slightly elevated ST segments in V3 may occur in normal teenagers.	(True)
E	An inverted T wave in lead I may indicate situs inversus.	(True)

Comments:
C, D, E In pulmonary hypertension the P wave in lead II is greater than 2.5mm. Inverted T waves may be found across to V3 in normal infants, and may remain so beyond the second decade. The mean QRS axis in the newborn lies between 60 degrees and 160 degrees. Dextracardia with dextraversion may result in inverted ECG appearances.
Question supplied by Colin Melville Consultant Paediatrician

The following drug side effects and interactions are recognised:

A	Adenosine and sinus arrest.	(True)
B	Verapamil and asystole.	(True)
C	Amiodarone and hyperthyroidism.	(True)
D	Amiodarone and hypothyroidism.	(True)
E	Adenosine and bronchospasm.	(True)

Comments:
A, B, C, D, E Verapamil is rarely used in childhood. In combination with betablockers, it is particularly lethal.

In cardiac tamponade, the following may be recognised:

A	Pulsus alternans	(False)
B	Raised jugular venous pulse	(True)
C	Muffled heart sounds	(True)
D	Cannon waves	(False)
E	Diminished pulse volume	(True)

Comments:

Cardiac tamponade is caused by fluid in the pericardial sac constricting the heart. This results in pulsus paradoxis, raised jugular venous pulse, muffled heart sounds, and if severe, diminished pulse volume and shock. Cannon waves occur because of atrial contraction against the closed tricuspid valve as in atrial fibrillation, and pulsus alternans suggests a severely diseased ventricular muscle. Electrical alternans, with variable QRS complex amplitude, may be present in pericarditis. (See example on **ecglibrary.com**)

Cyanotic attack in Tetralogy of Fallot is characterised by

A	Pallor	(False)
B	Bradycardia	(True)
C	Increase in severity of systolic murmur	(False)
D	Apnoea	(False)
E	Impaired consciousness	(True)

Comments:

Paroxysmal hypercyanotic attacks (blue or 'tet' spells) are a problem in the first 2 years of life. The infant hyperventilates and becomes restless with cyanosis, gasping and finally syncope. Spells occur most often in the early morning or after vigorous crying. The pulmonary murmur disappears or quietens during the episodes as RVOT flow decreases. They last from minutes to hours, but are rarely fatal. Complications include severe hypoxia, metabolic acidosis, convulsions or hemiparesis.

The following are recognised complications of cyanotic congenital heart disease:

A	Psychomotor delay.	(True)
B	Cerebral thrombosis.	(True)
C	Infective endocarditis.	(True)
D	Arthritis.	(True)
E	Cerebral abscess.	(True)

Comments:
A, B, C, D, E Complications of cyanotic congenital heart disease in Eisenmenger physiology include: - Polycythemia - CNS abscess - Thromboembolic stroke - Low grade DIC with thrombocytopenia - Haemoptysis - Gum disease - Gout - Arthritis - Clubbing - Failure to thrive - Psychomotor delay Janeway lesions are painless, small, erythematous or haemorrhagic lesions on the soles of the palms or soles, that occur in infective endocarditis.

The calibre of arterioles is affected as follows:

A	angiotensin II causes marked vasodilatation	(False)
B	epinephrine causes vasodilatation in skeletal muscles	(True)
C	decreased oxygen tension causes vasodilatation	(True)
D	locally released serotonin causes vasoconstriction	(True)
E	increased noradrenergic discharge causes vasodilatation	(False)

Comments:
Adrenaline produces vasodilatation of arterioles within muscles but constriction within the skin. Noradrenaline causes vasoconstriction. Decreased O_2 produces vasodilatation but serotonin/5HT causes vasoconstriction except for vasodilatation of muscle arterioles.

Regarding supraventricular tachycardias:

A	Rates of 120080 beats per minute are typical in adolescence.	(False)
B	They are usually associated with structural cardiac defects.	(False)
C	Lown-Ganong-Levine Syndrome is the most commonest cause.	(False)
D	J waves may be seen.	(False)
E	DC shock is most commonly required.	(False)

Comments:
All answers are false. SVT is usually due to re-entry within the AV node, and is characterised clinically by abrupt onset and cessation. The heart rate exceeds 180 beats per minute, and can occasionally be as fast as 300 beats per minute. The heart is usually anatomically normal, but there may be an associated bypass tract in WPW or Lown-Ganong-Levine Syndromes. It may also occur in relation to Ebstein's anomaly or corrected transposition of the great arteries. In older children it can be precipitated by sympathomimetics such as cold cures. Vagal manoeuvres or facial immersion in ice may abort the attack. Adenosine may be used in the non-shocked patient, or DC cardio-aversion 0.5J/kg in the child with congestive heart failure.

A 3 day old baby, discharged following a normal routine postnatal examination, is rushed to hospital with a thready pulse and heart rate of 180/min, a 5 cm liver edge, and a capillary refill time of 6 seconds. The following cardiac lesions should be considered:

A	Transposition of the great arteries	(False)
B	Double outlet right ventricle	(False)
C	Pulmonary atresia	(False)
D	Hypoplastic left heart syndrome	(True)
E	Truncus arteriosus	(True)

Comments:

Neonatal shock may be caused by infection, cardiac lesions or metabolic abnormalities. TGA, double outlet right ventricle, and pulmonary atresia cause cyanosis. Hypoplastic left heart syndrome, truncus arteriosus, coarctation, or critical aortic stenosis are the likely cardiac causes of this presentation.

A murmur is exaggerated during inspiration in:

A	pulmonary regurgitation	(True)
B	aortic regurgitation	(False)
C	tricuspid regurgitation	(True)
D	patent ductus arteriosus	(False)
E	mitral stenosis	(False)

Comments:
a - All right sided events increase on inspiration except a pulmonary click.

A 6 week old girl presents with respiratory distress. The following suggests cardiac failure rather than bronchiolitis:

A	Wheezing.	(False)
B	Poor weight gain for the past 3 weeks.	(True)
C	A respiratory rate of 60 breaths/min.	(False)
D	Cardio-thoracic ratio on AP x-ray of 60%.	(True)
E	A 4cm liver.	(True)

Comments:
B, D, E Tachypnoea and wheezing may occur in both heart failure and bronchiolitis, though crepitations are commoner in the latter. Bronchiolitis rarely lasts as long as 3 weeks. A normal AP cardio-thoracic ratio is less than 55%. H Heart failure is rare in bronchiolitis, so an increased CTR and very large liver suggest heart failure. Not uncommonly, a 2-3cm liver is found in bronchiolitis because of hyperinflation of the lungs.

Recognised features of a large uncomplicated patent ductus arteriosus include:

A	Narrow pulse pressure.	(False)
B	Pulmonary plethora.	(True)
C	Decreased noise of murmur on squatting.	(False)
D	Left ventricular hypertrophy on ECG.	(True)
E	Soft pulmonary second heart sound.	(False)

Comments:
B, D A large uncomplicated patent ductus arteriosus results in a bounding pulse and wide pulse pressure, because of increased cardiac output on the left side of the heart draining to a low resistance pulmonary circulation. Because of increased left to right shunt, pulmonary plethora and cardiomegaly are characteristic. Squatting will increase systemic arterial resistance, but this is unlikely to have any effect on the murmur. Because the left ventricle pumping at excessive volume, left ventricular hypertrophy and strain may be seen on the ECG. The second pulmonary heart sound is loud because of pulmonary hypertension, but may not be audible beneath a loud murmur.

Regarding systemic hypertension in childhood:

A	Sodium nitroprusside is useful for the long-term treatment of severe cases.	(False)
B	Headache is the usual presenting feature.	(False)
C	It is defined as systolic blood pressure above the 99th centile for age.	(False)
D	Abnormalities are frequently seen on DMSA scan.	(True)
E	Aortic coarctation is the commonest secondary cause.	(False)

Comments:
D Sodium nitroprusside is useful only in the short term, as cyanide levels accumulate with time. Hypertension is usually diagnosed incidentally, and is defined as systolic blood pressure >95th centile for age. Secondary causes are usually due to renal abnormalities, with reflux associated scarring being the commonest renal disease. This will cause abnormalities on DMSA scan. Coarctation of the aorta is the commonest non-renal cause, with pheochromocytoma/neuroblastoma, congenital adrenal hyperplasia, Cushing Syndrome and steroid therapy being rarer causes.

Interventional catheterisation can be used to repair the following defects:

A	Ostium primum ASD.	(False)
B	Ostium secundum ASD.	(True)
C	Muscular VSD.	(False)
D	PDA.	(True)
E	Dysplastic pulmonary valve.	(False)

Comments:
B, D Ostium primum ASD is at the minor end of the spectrum of AV canal defects. Perimembranous VSDs require surgical closure, but muscular ones often close themselves. Dysplastic pulmonary valves can be palliated by balloon dilatation, but usually require surgical repair later. Secundum ASDs and PDA, can by closed using umbrella devices.

The carotid body chemoreceptors are:

A	stimulated by a decrease in pO_2 of arterial blood	(True)
B	inhibited by a decrease in pH of arterial blood	(False)

C	stimulated in an hypotensive subject at rest	(True)
D	responsible for production of reflex peripheral vasoconstiction	(True)
E	have a blood flow of over 50ml/100g per minute	(True)

Comments:

a - discriminating question

c+d-Hard.

e-Highest of any tissue.

carotid sinus is baroreceptor responding to hypotension & mediating vasoconstriction, while carotid body is chemoreceptor. Respiratory arrest and circulatory shock dramatically increase chemoreceptor activity leading to enhanced sympathetic outflow to the heart and vasculature via activation of the vasomotor centre.

The following are indications for immediate transfer to a tertiary cardiology centre in infancy:

A	Suspected large VSD.	(False)
B	Suspected ASD.	(False)
C	A diagnosis of Down's Syndrome.	(False)
D	A diagnosis of Kawasaki Disease.	(False)
E	Suspected aortic coarctation.	(True)

Comments:
E Urgent transfer to a tertiary cardiac centre is required for life-threatening conditions, particularly those dependent on the ductus arteriosus for survival. These include: 1. Hypoplastic left heart syndrome. 2. Critical aortic valve stenosis. 3. Severe coarctation of the aorta. 4. Interruption of the aortic arch. The other conditions mentioned require more routine cardiac evaluation.

Myocardial infarction is a recognised feature of the following

A	Kawasaki Disease	(True)
B	Viral myocarditis	(True)
C	Hypertrophic obstructive cardiomyopathy	(True)
D	Mitral stenosis	(False)
E	Anomolous left coronary artery	(True)

Comments:

In mitral stenosis, large p-mitrale is first seen, with AF being a late sign. In hypertrophic obstructive

cardiomyopathy, the heart muscle outgrows the blood supply, causing angina or myocardial infarction.

The following are characteristic of tricuspid atresia:

A	P wave of >2.5mm which is narrow.	(True)
B	Increased pulmonary blood flow.	(False)
C	Co-existing ASD.	(True)
D	Left axis deviation on the ECG.	(True)
E	Small right ventricle on echocardiography.	(True)

Comments:

In tricuspid atresia there is a fibromuscular membrane in place of the tricuspid valve, a variably small right ventricle, a VSD, and a large left ventricle and aorta. Since there is no outlet from the right atrium, the entire systemic venous return enters the left heart via the foramen ovale or an associated ASD. Left ventricular blood is then pumped into the right ventricle by the VSD into the pulmonary circulation. The pulmonary blood flow is therefore decreased and a characteristic presentation is cyanosis with decreased pulmonary blood flow. Since the right ventricle is small, there is usually left axis deviation on ECG. Treatment is often with a modified Fontan procedure where the systemic venous return is diverted using a right atrial baffle to the pulmonary arteries with subsequent closure of the ASD and VSD.

An opening snap is found in:

A	mitral stenosis due to rheumatic heart disease	(True)
B	congenital mitral stenosis	(True)
C	mitral incompetence associated with a rigid posterior valve leaflet but a normal anterior leaflet	(True)
D	left atrial myxoma	(True)
E	severe aortic incompetence	(False)

Comments:
d-10% of cases secondary to increasing left atrial pressure. Soft late snap - actually a 'tumour plop' that mimics the opening snap of Mitral Stenosis. (Wassermil M, Warkentin DL, Ravin A: Myxoma of the left atrium: Phonocardiographic study of three cases. Circulation 1962;25:50–56)

A loud first heart sound may be due to:

A	a long preceding diastolic interval	(False)
B	mitral stenosis	(True)
C	rupture of a papillary muscle	(False)
D	increased systemic arterial pressure	(False)

E	increrased pulmonary arterial pressure	(False)

Comments:
A loud first heart sound is due to abrupt closure of the mitral valve against a high left atrial pressure. MR occurs with papillary muscle rupture and thereby 1st heart sound is soft.A2 and P2 are loud in systemic HT and pulmonary hypertension respectively.

Cyanosis occurs in:

A	patent ductus arteriosus	(False)
B	Fallot's tetralogy	(True)
C	ostium primum Atrial Septal Defect	(False)
D	tricuspid atresia	(True)
E	Eisenmenger's complex	(True)

Comments:
Cyanosis defined as greater than 5g% reduced haemoglobin typically occurs in association with right to left shounts as is Fallot's, Transposition, hypoplastic left heart, tricuspid atresia and Eisenmenger's. PDA and ostium primum are not associated with cyanosis unless the latter converts to eisenmenger's complex with time.

The following can cause bradycardia:

A	hypothermia	(True)
B	hypothyroidism	(True)
C	severe anaemia	(False)
D	subdural haematoma	(True)
E	shock	(True)

Comments:
a-One of the main features - can be extreme bradycardia, wide QRS, long QT interval and prominent J wave. b-Sinus tachycardia / AF with hyperthyroidism. c-Usually associated with sinus tachycardia especially if severe. d-From raised intracranial pressure (Cushing's reflex - raised ICP leads to increased blood pressure and decreased heart rate). e-This is arguable as it is usually associated with sinus tachycardia however there may be a bradycardic response to shock.

Fixed splitting of the second heart sound occurs in the following conditions

A	Ostium primum defect	(True)
B	Fallot's tetralogy	(False)
C	Chronic constrictive pericarditis	(False)
D	Aortic stenosis	(False)
E	Ostium secundum defect	(True)

Comments:

Fixed splitting of the second heart sound occurs in atrial septal defects because of equal pressures in the atria throughout the respiratory cycle.

Fixed splitting of the second heart sound is found in:

A	Ostium secundum ASD	(True)
B	Perimembranous VSD	(False)
C	Ebstein's anomaly	(False)
D	Ostium primum ASD	(True)
E	Pulmonary valve stenosis	(False)

Comments:

Fixed splitting of the second heart sound occurs with ASDs. VSD is associated with a loud second heart sound, and Ebstein's anomaly with a widely split variable second heart sound. Pulmonary valve stenosis is associated with a soft or absent pulmonary component to the second heart sound.

The following associations are recognised:

A	Pulmonary plethora and pulmonary atresia with intact ventricular septum.	(False)
B	Prominent right heart border and ASD.	(True)
C	Pericardial calcification and endocardial fibroelastosis.	(False)
D	'Cottage loaf' heart and total anomalous pulmonary venous drainage.	(True)
E	Selenium deficiency and cardiomyopathy.	(True)

Comments:
B, D, E In pulmonary atresia with intact ventricular septum there is decreased pulmonary blood flow. The prominent right heart border occurs in ASD becasue of right atrial hypertrophy. In endocardial fibroelastosis the interior of the ventricles appear echogenic, but there is no pericardial calcification. Pericardial calcification is found in constrictive pericarditis. A 'cottage loaf' heart is characteristic of TAPVD, and selenium deficiency is the cause of Keshan Disease, a rare nutritional cardiomyopathy that is frequently fatal in the Keshan Province of China.

In the differential diagnosis of cyanosis in the newborn:

A	A ground glass appearance confirms hyaline membrane disease.	(False)
B	An arterial PO_2 above 13 kPa suggests non-cyanotic congenital heart disease.	(False)
C	A normal cardiothoracic ratio excludes total anomalous pulmonary venous	(False)

drainage.

D	Persistent hypoxia in a child of 34 weeks gestation suggests transposition of the great arteries.	(False)
E	A saturation of 91% in the right hand and 85% in the right leg suggests significant shunting.	(True)

Comments:

A ground glass appearance is not specific for hyaline membrane disease: Group B Streptococcus can also present in this way. In the nitrogen wash-out test, an arterial PO_2 <13kPa in 100% oxygen suggests cyanotic congenital heart disease. Other causes of cyanosis, such as pneumonia or hyaline membrane disease, can normally be oxygenated beyond this level. A normal cardio-thoracic ration is characteristic of total anomalous pulmonary venous drainage. This is becasue the obstruction is extra-cardiac. The usual reason for hypoxia in a pre-term baby is hyaline membrane disease or infection, with cardiac causes being rarer. A significant difference in oxygen saturation between the pre-ductal and post-ductal circulations suggests that hypoxic blood is shunting from the pulmonary artery to the aorta.

The following are normal findings on the neonatal ECG

A	A frontal axis of +50°	(False)
B	Larger R wave than S wave in V1	(True)
C	An RS ratio in V6 of 2	(False)
D	An inverted T wave in V4R	(True)
E	A small Q wave in V1	(False)

Comments:

In the neonate the ECG reflects the transitional circulation. Since in-utero the right ventricle is the systemic ventricle, the right sided forces of greater than the left, gradually changing postnatally. The right sided chest leads in the neonates, therefore show larger positive R waves than negative waves, and this may remain for several months or years. The left sided leads (V5 and V6) also reflect the RV dominance, and the RS ratio in these leads may be <1. The QRS axis usually lies between 110 and 180°. The T waves are inverted in V4R, V1, V2 and V3 during infancy, and this may remain until the middle of the second decade and beyond. Q waves are always abnormal.

The following are characteristic of acute pericardial effusion:

A	Giant "a" waves in the jugular venous pulse.	(True)
B	Pulsus paradoxus.	(True)
C	Right ventricular end-diastolic collapse on echocardiography.	(True)
D	Low blood pressure.	(True)

E	Cold extremities.	(True)

Comments:
A, B, C, D, E Acute pericardial effusion causes cardiac tamponade with all these signs. In addition the heart sounds may be muffled and there may be signs of shock.

Regarding the ductus arteriosus:

A	It usually closes functionally by 6 hours post delivery in a term infant.	(True)
B	May close prematurely due to maternal aspirin in pregnancy.	(True)
C	Has a right to left shunt in utero.	(True)
D	Closure can be delayed by the use of prostaglandin E2.	(True)
E	Closure is encouraged by hypoxaemia.	(False)

Comments:
A, B, C, D The ductus arteriosus functionally closes within the first few hours after birth. Aspirin antenatally is associated with impaired platelet function and risk of haemorrhage, delayed onset and increased duration of labour, and fetal ductus arteriosus closure in high doses. It may also be related to persistent pulmonary hypertension and kernicterus in jaundiced neonates. Prostaglandin E2 is used in the emergency management of duct dependent cardiac lesions prior to transfer to cardiac units. Hypoxaemia tends to keep the duct open.

An infant is noted at postnatal examination to have a slow pulse rate. ECG confirms congenital complete heart block. The following statements are true:

A	Echocardiography is usually normal.	(True)
B	It is usually associated with a presence of anti-Ro antibodies in the mother.	(True)
C	Heart failure is uncommon.	(True)
D	Mother's thyroid function should be checked.	(False)
E	The underlying defect may be due to prolonged QT Syndrome.	(False)

Comments:
The commonest cause is maternal SLE, which may be asymptomatic. Echocardiography is usually normal (>60% of cases), and heart failure is uncommon. Maternal hyperthyroidism can cause abortion, preterm labour, and fetal tachycardia. Prolonged QT may occur with hypokalaemia, hypocalcaemia, antihistamines (e.g. Terfenadine). Congenital prolongation presents with syncope in late childhood.

The following suggest pulmonary rather than aortic valve stenosis:

A	An ejection murmur radiating to the neck.	(False)
B	An ejection click best heard in the 2nd and 3rd left intercostal spaces.	(True)
C	An upright T wave in V1.	(True)
D	A deep S wave in V2.	(False)

E	Prominence in the left upper mediastinum.	(False)

Comments:
B, C An ejection murmur radiating to the back (along the direction of the pulmonary arteries) is found in pulmonary stenosis. The ejection click is found at the apex in aortic stenosis. A deep S wave in V2 and large R wave in V6 suggests LVH, while an upright T wave in V1 suggests RVH. A prominent left upper mediastinum on chest x-ray may be caused by post-stenotic aortic dilatation in aortic stenosis.

Cannon waves may be seen in the jugular veins in:

A	constrictive pericarditis	(False)
B	first degree AV heart block	(False)
C	tricuspid stenosis	(False)
D	ventricular pacing	(True)
E	nodal tachycardia	(True)

Comments:
Canon waves are seen with unsynchronised closure of the tricuspid valve and right atrial contraction. Giant V waves are seen with constrictive pericarditis and tall a waves but not cannon a waves are seen with Tricuspid stenosis.

Nitric oxide:

A	is generated from glutamine	(False)
B	is produced by both inducible and constitutive forms of nitric oxide synthetase	(True)
C	raises systemic vascular resistance	(False)
D	is inactivated by oxygen free radicals	(True)
E	is increased by cyclic AMP activation.	(False)

Comments:
Nitric oxide is produced from l-arginine by nitric oxide synthase and is produced by the vascular endothelium in response to haemodynamic stress and produces smooth muscle relaxation and reduced vascular resistance. Nitric oxide is a free radical and may be inactivated through interaction with other oxygen free radicals e.g. oxidised LDL. It causes the production of cGMP as a second messenger.

The following conditions are inherited as an autosomal dominant trait:

A	Friedrich's ataxia	(False)
B	Ehlers-Danlos syndrome	(True)
C	Noonan's syndrome	(True)
D	Romano-Ward syndrome	(True)
E	Marfan's syndrome	(True)

Comments:
a-Autosomal recessive and associated with cardiomyopathy (Modes of inheritance questions are very common in the MRCP part 1 exam). b-Associated with aortic aneurysm. c-Chromosome 12. Associated with pulmonary stenosis, short stature, webbing of the neck, undescended testes in boys, HOCM, ASD (**more ...**) d-Associated with long QT (Jervell-Lange-Neilson syndrome is Autosomal Recessive). e-Associated with mitral valve prolapse, aortic aneurysm and aortic regurgitation.

The following findings may occur with a cyanotic episode in an individual with Fallot's tetralogy:

A	bradycardia	(False)
B	pallor	(False)
C	increased systolic murmur	(False)
D	loss of consciousness	(True)
E	continuous murmur at the upper left sternal border	(True)

Comments:
B- Usually polycythaemic and cyanosed. The Murmur may becoma softer during a cyanotic episode and syncope can occur. The continuous murmur may still be audible. (Dr Jacob Easaw)

W hich of the following features would be expected in a 17 year old male with pre-excitation tachycardia of Wolff-Parkinson-White syndrome?

A	Narrow complex tachycardia is usually regular	(True)
B	Amiodarone is contraindicated	(False)
C	Digoxin is contraindicated	(True)
D	Verapamil is useful in irregular broad-complex tachycardia	(False)
E	VF is always stopped by amiodarone	(False)

Comments:
There is typically a narrow complex tacchycarida with a characterisitic short PR and delta wave present between episodes. Amiodarone can be used as a treatment but the most appropriate therapy is destruction of the accessory pathway with frequency ablation. Digoxin is not useful as a single drug since it shortens refractory period in the accessory pathway and speeds up ventricular response during AF and like verapamil may precipitate VF.

Hypertrophic obstructive cardiomyopathy is associated with:

A	a double apical impulse	(True)
B	decreased left ventricular ejection fraction	(False)
C	left ventricular diastolic dysfunction	(True)
D	increased left ventricular outflow obstruction	(True)

E	mitral regurgitation	(True)

Comments:
Hypertrophic cardiomyopathy is defined as the unexplained, asymmetical or concentric hypertrophy of the undilated left ventricle. There is also hypertrophy of the right ventricle. It may be inherited as an autosomal dominant condition, but at least half of cases may be the result of sporadic mutation. Double apical impulse may be felt, with diastolic dysfunction being typical. Outflow obstruction develops over time associated with increasing hypertrophy and associated mitral regurgitation is common.

The following are recognised determinants of systemic blood pressure in childhood:

A	Systolic blood pressure increases with age.	(True)
B	Systemic hypertension is commoner in girls.	(False)
C	Diastolic blood pressure correlates with height.	(True)
D	Hypertension is defined as systolic blood pressure greater than the 99th percentile for age.	(False)
E	In children under the age of 5, the majority will have secondary hypertensions.	(True)

Comments:

Blood pressure is the product is the product of peripheral vascular resistance and cardiac output. Accurate measurement is dependent on the quality of the equipment and the skill of the observer. Anxiety may raise the initial level. Careful attention is required to cuff size. The bladder of the pressure cuff should nearly encircle the upper arm, but the ends should not overlap. The cuff should cover at least two thirds of the length of the arm. Systolic blood pressure is indicated by the first carotid sound, and in pre-adolescence the diastolic by the fourth sound. Systemic blood pressure gradually increases with age and correlates with weight and height. Hypertension is defined as a blood pressure consistently above the 95th centile for age. The younger the child the more likely there is to be a secondary cause for the raised blood pressure. In the majority of these cases, this will be due to renal disease (e.g. renal artery stenosis, reflux nephropathy). Coarctation of the aorta and neuroendocrine causes should also be considered.

The following are recognised features of cardiac failure in the infant:

A	Jaundice.	(False)
B	Splenomegaly.	(True)
C	Raised jugular venous pulse.	(False)
D	Pallor.	(True)
E	Grunting.	(True)

Comments:
B, D, E Cardiac failure in the infant usually presents with feeding difficulties and respiratory distress. On examination, there may be sweating, tachypnoea, nasal flaring, recession, grunt, hepatomegaly, and splenomegaly. Pallor may reflect poor peripheral perfusion, and there may be a murmur from the underlying heart lesion. The squat neck of the infant makes the jugular venous pulse impossible to

assess reliably, and jaundice does not occur.

Paradoxical (reverse) splitting of the second heart sound is typical of:

A	Ostium primum ASD.	(False)
B	Pulmonary stenosis.	(False)
C	Aortic stenosis.	(True)
D	Left bundle branch block.	(True)
E	Ostium secundum ASD.	(False)

Comments:
C, D Paradoxical (reverse) splitting of the second heart sound occurs when splitting sounds larger in expiration than inspiration. Normally, inspiration increases right-sided filling and delays pulmonary valve closure. In situations such as aortic stenosis, or left bundle branch block, delayed closure of the aortic valve results in reverse splitting.

The following are characteristic of supraventricular tachycardia in a 6 month old child:

A	A cardiac rate of 190/min	(True)
B	Sudden collapse	(False)
C	Hepatomegaly	(True)
D	A cardiac rate of 200/min that increases with crying	(True)
E	Irritability	(True)

Comments:

SVT is usually caused by a re-entry within the AV node. The attacks are characteristically abrupt in onset and cessation, and may be precipitate by acute infection. They usually occur when the patient is at rest. In infants, the heart rate is usually above 200, but in older children usually exceeds 180/min. The only complaint may be awareness of a rapid heart rate, and this is usually well tolerated. If the rate is exceptionally rapid or the attack is prolonged then precordial discomfort in congestive heart failure may supervene. In the infant whose attack lasts 6-24 hours with an extreme heart rate, the child may become acutely ill with a poor colour, irritability, tachypnoea, and hepatomegaly. There may be fever and leukocytosis. In the fetus, there may be hydrops fetalis. SVT is usually associated with an anatomically normal heart, or may be associated with a bypass tract in one of the pre-excitation syndromes (Wolff-Parkinson-White, Lown-Ganong-Levine). Occasionally, it occurs in association with Ebstein's anomaly, or corrected transposition of the great arteries. It may be precipitated by exposure to sympathomiemeticamines (e.g. pseudoephedrine in cold cures).

The following are found in ostium secundum ASD:

A	Cardio-thoracic ratio of 60% with pulmonary bay.	(False)
B	Superior axis deviation on ECG.	(False)
C	Grade 1/6 diastolic murmur at the lower left sternal edge.	(True)
D	Cardiac arrhythmias in later life.	(True)
E	A fixed widely split second heart sound.	(True)

Comments:

A pulmonary bay suggests small pulmonary arteries, as in Fallot's. Superior axis deviation may be found in primum ASD. A diastolic tricuspid rumble is due to increased blood flow, and suggest a large shunt. Fixed splitting is due to equal left and right atrial pressure throughout the respiratory cycle.

Cardiac output is altered by the following factors:

A	reduction during sleep	(True)
B	cardiac output is altered by moderate changes in environmental temperature	(False)
C	standing from a lying position decreases the cardiac output	(False)
D	histamine increases the cardiac output	(True)
E	eating decreases the cardiac output	(False)

Comments:
The fundamental equation is BP=COxTPR. Sleeping is associated with reduced metabolic requirements, decreased HR, BP and hence reduced CO. Output is affected by changes in the temp of the individual but not by small changes in environmental temp. The orthostatic response is associated with an increase in CO. Histamine is associated with vasodilatation and hence increased CO. Post-prandially CO is increased by 30%

Regarding infective endocarditis:

A	The commonest causative organism is Group A Streptococcus.	(False)
B	Lesions are most frequent in areas of high velocity blood flow.	(True)
C	Amoxycillin is first choice for prophylaxis in procedures done under local anaesthetic.	(True)
D	Presentation in infancy is extremely rare.	(True)
E	Splenomegaly is a subtle early sign.	(False)

Comments:
B, C, D Infective endocarditis is usually caused by Strep. Viridans (Group D), but Staphylococcus aureus is becoming increasingly common. The portal of entry is usually the teeth. The organisms settle usually on congenital or rheumatic heart lesions, particularly in areas of high velocity blood flow. Early symptoms and signs are usually mild, and include prolonged fever and weight loss, which may last for several months before diagnosis. A rare presentation is with high fever and prostration, but the usual course is

somewhere between these two. Fever, fatigue, myalgia, changing heart murmurs, heart failure, splenomegaly and petechia are common. Serious complications include cerebral abscess and mycotic aneurysms. Osler's nodes, Janeway lesions and splinter haemorrhages may occur due to vasculitis.
