Cardiology

Theme : Emergency medicine

A. Adenosine
B. Adrenaline
C. Atropine
D. DC shock
E. Dobutamine
F. Diving reflex
G. Endotracheal intubation
H. Intraosseous line
I. IV Morphine
J. Naloxone

Select the most appropriate emergency treatment for the following children:

1) A 4-year-old child is rescued from a house fire. She is admitted tachypnoeic and tacchycardic. She has soot in her nostrils.

G. Endotracheal intubation

Note:
Item 1 relates to burns. Most deaths following house fires occur secondary to smoke inhalation. Amongst the indicators of inhaled smoke injury is deposits around the mouth and nose. Oedema follows thermal injury and therefore any suspicion of airway compromise should result in endotracheal intubation.

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2) A 3 month old baby is admitted with a history of poor feeding. On arrival he has a pulse rate of 220 beats per minute.

F. Diving reflex

Note:
Supraventricular tachycardia is the diagnosis in Item 3. Vagal stimulation is the treatment of choice and the diving reflex is the simple procedure elicited by submerging the baby's face in to ice or placing an ice bag over the face. The diving reflex increases vagal tone, slows AV conduction interrupting the tachycardia.

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3) A 13 year old boy is admitted with meningococcaemia. He is in shock. Peripheral cannulation is difficult.

H. Intraosseous line

Note:
Item 2 relates to a child in shock. In many life-threatening conditions venous cannulation is difficult. It is important to obtain vascular access very quickly and therefore intraosseous infusion is recommended.

Theme : Syndromes associated with congenital heart disease

A. Charcot-Marie- Tooth syndrome
B. Down syndrome
C. Fragile X syndrome
D. Marfan's syndrome
E. Noonan syndrome
F. Sturge-Weber syndrome
G. Tetrology of Fallot
H. Tourette syndrome
I. Turner's syndrome
J. William's syndrome

For each of the cardiac lesions described below, choose the most commonly associated syndrome from the above list of options. Each option may be used once or not at all.

1) Supravalvular aortic stenosis **J. William's syndrome**

Note:
William's syndrome ► is characterized by short stature, characteristic facies, supravalvular aortic stenosis, mild to moderate learning difficulties and transient neonatal hypercalcemia.

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2) Coarctation of Aorta **I. Turner's syndrome**

Note:
Turner's syndrome ► This is characterized by 45, X genotype, ovarian dysgenesis leading to infertility, short stature, webbing of the neck, wide carrying angles and wide spaced nipples. However they have normal intellectual development.

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3) Dilatation of aortic root/aortic regurgitation **D. Marfan's syndrome**

Note:
Marfan's syndrome ► This is an autosomal dominant disorder. The clinical features are tall stature, arachno-dactyly, high arched plate and increase in length of the lower segment of the body compared to the upper segment. The cardiac manifestations include dilated aortic root, aortic incompetence, mitral valve prolapse and mitral incompetence.

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4) Cardiac cushion defects (leading to ASD, VSD) **B. Down syndrome**

Note:
Down syndrome ► Features include characteristic facies, hypotonia, severe learning difficulties and small stature. About 40% of patients have cardiac anomalies mainly endocardial cushion defects leading to ASD and VSD.

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5) Infundibular pulmonary stenosis **G. Tetrology of Fallot**

Note:
Tetrology of Fallot This is a cyanotic heart disease and the cardinal features include:

1. Infundibular pulmonary stenosis
2. VSD
3. Right ventricular hypertrophy
4. Over-riding of the aorta

Comments:
Charcot-Marie-Tooth syndrome ► Autosomal dominant peroneal muscular dystrophy.
Fragile X syndrome ► Moderate learning difficulty, macrocephaly, characteristic facies (long face, large ears, prominent mandible and forehead)
Noonan syndrome► Facies, mild learning difficulties, short webbed neck, short stature and congenital heart disease (pulmonary valvular stenosis, ASD, left ventricular hypertrophy)
Sturge-Weber syndrome ► Haemangiomas in the distribution of trigeminal nerve and in the brain. Sometimes can have intractable epilepsy.
Tourette syndrome ► Tics, compulsive utterances of obscene words (coprolalia)

Theme : Emergency treatments

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Select the most appropriate emergency treatment for the following children:

1) A 5 year old boy is brought to the hospital with 15% scalds to his chest. **I. IV Morphine**

Note:
Item 1 relates to a child with serious burns. Children who have been burnt are in severe pain and therefore IV Morphine is the analgesic of choice.

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2) A 14-year-old girl with a history of previous overdoses is admitted to A&E apnoeic and unconsciousness. ECG shows ventricular fibrillation. CPR is commenced. **D. DC shock**

Note:
Item 2 describes a child in ventricular fibrillation. This is uncommon in childhood although may occur as a result of tricyclic antidepressant overdose and hypothermia. If the arrest is witnessed a precordial thump is carried out otherwise electrical de-fibrillation at 2 joules per kilogram.

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3) A term baby is born in poor condition. Apgar scores 3 at 1 minute and 5 at 5 minutes. CPR is commenced. At 10 minutes he remains bradycardic. **B. Adrenaline**

Note:
Item 3 describes a baby born in poor condition. A bradycardia in an unstable newborn requires oxygenation, ventilation and cardiac compressions. IV adrenaline is administered as Atropine is ineffective in this age group.

Theme : Genetics - cardiac abnormalities in genetic disorders

A. Angelman's syndrome
B. Beckwith-Wiedemann syndrome
C. Congenital Rubella syndrome
D. Down syndrome
E. Foetal alcohol syndrome
F. Glycogen storage disease
G. Marfan's syndrome
H. Noonan's syndrome
I. Turner's syndrome
J. Williams syndrome

Match each of the following cardiovascular abnormalities to the single most likely associated genetic disorder.

1) Dilation of the aorta with aneurysms. **G. Marfan's syndrome**

Note:
In Marfan's syndrome dilatation of the ascending aorta is often seen with or without aneurysms. Less commonly the thoracic abdominal aorta or pulmonary arteries are affected with secondary aortic regurgitation and mitral valve prolapse.

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2) Supra-valvular aortic stenosis. **J. Williams syndrome**

Note:
In Williams syndrome supra-valvular aortic stenosis is the most common cardiac lesion. Septal defects also occur as well as peripheral branch pulmonary artery stenosis.

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3) Pulmonary stenosis. **H. Noonan's syndrome**

Note:
In Noonan's syndrome pulmonary valve stenosis due to a dysplastic or thick valve is seen often associated with left ventricular hypertrophy. Branch stenosis of the pulmonary artery also is found in Noonan's syndrome.

Theme : Congenital cardiac defects

A. Angelman's syndrome
B. Beckwith-Wiedemann syndrome

C. Congenital Rubella syndrome
D. Down syndrome
E. Foetal alcohol syndrome
F. Glycogen storage disease
G. Marfan's syndrome
H. Noonan's syndrome
I. Turner's syndrome
J. Williams syndrome

Match each of the following cardiovascular abnormalities to the single most likely associated disorder.

1) Endocardial cushion defect. **D. Down syndrome**

Note:
In Down syndrome approximately 40% of children have a congenital heart disease. The most common being endocardial cushion defects although VSDs, ASDs and PDA also occur.

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2) Coarctation of the aorta. **I. Turner's syndrome**

Note:
In Turner's syndrome cardiac defects are common. 30% include bicuspid aortic valves with the second most common heart defect being coarctation of the aorta. Aortic stenosis, mitral valve prolapse and hypertension are also found.

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3) Septal defects. **E. Foetal alcohol syndrome**

Note:
In foetal alcohol syndrome individuals have poor growth, developmental delay and usually characteristic facial features including microcephaly and a short smooth philtrum. The most common cardiac lesion in these children are septal defects primarily ventricular septal defects.

Theme : BREATHLESSNESS

A. Asthma
B. Hyperventilation
C. Tuberculosis
D. Cystic Fibrosis
E. Pneumocystis carinii

F. Ventricular septal defect
G. Gastroesophageal reflux
H. Atrial septal defect
I. Bronchiolitis
J. Mitral stenosis

For each of these patients with breathlessness, select the most likely diagnosis

1) A thirteen-year-old girl who has intermittent episodes of breathlessness, which tend to occur in crowded shops. She feels the need to take deep breaths and then breathes very quickly, complaining of pins & needles around her mouth and in her hands. Her chest is clear and her blood gases show a normal pO2 and low pCO2.

B. Hyperventilation

Note:
This description is one of anxiety. The blood gas picture is one of hyperventilation.

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2) A three-year-old boy who presents with worsening cough and breathlessness of 3 weeks' duration. His mother was an intravenous drug abuser. He has always been prone to infections. When he was 2 years old he had chicken pox for 4 weeks. On examination, he has an emaciated appearance, his weight is below the 0.4th centile, he has a temperature of 37.6°C and he has generalized crepitations on auscultation of his chest. A blood count shows severe lymphopenia.

E. Pneumocystis carinii

Note:
This has resulted from congenitally acquired HIV. PCP has an insidious onset and often there are no chest signs in children. Lymphopenia is consistent. Treatment is with septrin or nebulised pentamidine as second line.

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3) A 4-month-old baby has not gained much weight since birth and only takes small milk feeds, as he appears to become breathless on feeding. He is tachypnoeic, sweaty and has a tachycardia. His liver is enlarged and he has a harsh grade 2-pansystolic murmur at the left lower sternal edge.

F. Ventricular septal defect

Note:
Poor feeding is a symptom of heart failure in babies. A VSD has a pan systolic murmur and if large may have a lower grade as there is less resistance to flow. Hepatomegaly is an early sign of heart failure in infants.

Theme : CONGENITAL HEART DISEASE

A. Ostium secundum atrial septal defect
B. Ventricular septal defect
C. Transposition of the great arteries
D. Total anomalous pulmonary venous drainage
E. Atrioventricular septal defect
F. Patent ductus arteriosus
G. Pulmonary valve stenosis

H. Coarctation of the aorta
I. Tetralogy of Fallot
J. Hypoplastic left heart syndrome

Which of the above is the most likely diagnosis in the following cases.

1) A 13-year-old girl is referred for evaluation of her short stature. She is prepubertal. On auscultation she has an ejection systolic murmur in the second and third left intercostals spaces radiating to the back, but is a symptomatic.

G. Pulmonary valve stenosis

Note:
The murmur describes pulmonary stenosis, which could also be a left peripheral pulmonary stenosis. She is short and has delayed puberty and coupled with the cardiac findings would suggest Noonan's syndrome.

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2) A 7-week-old infant presents with breathlessness on feeding and failure to thrive. On examination his femoral pulses are difficult to feel but present. Chest radiograph shows cardiomegaly and increased vascular markings.

H. Coarctation of the aorta

Note:
Absent or weak femoral pulses suggest coarctation. Remember association with Turner's syndrome.

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3) An infant is seen for his 6-week check and found to have a loud ejection systolic murmur in the third left intercostal space and a single second heart sound on examination. There is no obvious cyanosis but a suggestion of mild desaturation. On the chest X ray there is a concavity on the left heart border and decreased pulmonary vascular markings.

I. Tetralogy of Fallot

Note:
Tetralogy of Fallot may present later than in the neonatal period. The ejection systolic murmur is from the infundibular stenosis. The desaturation results from the right to left shunt across the VSD.

Theme : Congenital heart disease

A. Atrioventricular septal defect
B. Coarctation of the aorta
C. Hypoplastic left heart syndrome
D. Ostium secundum atrial septal defect
E. Patent ductus arteriosus
F. Pulmonary valve stenosis
G. Tetralogy of Fallot
H. Total anomalous pulmonary venous drainage
I. Transposition of the great arteries

J. Ventricular septal defect

Which is the most likely diagnosis in the following cases?

1) An infant is found profoundly cyanosed and lethargic in his cot on day 2. On auscultation there is a soft systolic murmur heard inconsistently at the left sternal edge and a single second sound. The chest X ray shows a narrow upper mediastinum, hypertrophied right ventricle and increased pulmonary vascular markings. The ECG shows a normal neonatal pattern.

I. Transposition of the great arteries

Note:
Cyanosis on the second day is suggestive of a duct-dependent lesion. The rest of the answer describes TGA

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2) A 3-week-old premature infant born at 27 weeks gestation remains ventilated following surfactant deficient respiratory distress syndrome. On auscultation of his chest a systolic murmur is heard at the left sternal edge and pulses are very easy to feel. There is pulmonary plethora on chest X ray.

E. Patent ductus arteriosus

Note:
PDA is a relatively common problem in premature babies. The left to right shunt results in excess blood flow through the lungs and frequently oxygen dependency and difficulty in weaning from the ventilator. A loud systolic murmur radiating to the back with easily palpable pulses are typical.

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3) A 7-year-old boy is examined for a chest infection. An incidental finding of a short systolic murmur with fixed splitting of the second heart sound is detected. His blood pressure is normal and all pulses are normal.

D. Ostium secundum atrial septal defect

Note:
Ostium primum defects are unlikely to present incidentally but rather with heart failure or pulmonary hypertension. Fixed splitting is typical of ostium secundum defects

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