Congenital cardiac emergencies

Joe Brierley

INTRODUCTION

Cardiac emergencies are rare in children in comparison to adults. The pathophysiology differs; ischaemic heart disease is virtually unknown, whereas most congenital cardiac emergencies occur secondary to structural heart disease. Of the myriad lesions many decompensate in the perinatal period, usually in association with arterial duct closure. Subsequent rapid haemodynamic deterioration may lead to a moribund infant in extremis at presentation and with more home deliveries and early discharges such infants may present not only to paediatricians, but to accident and emergency departments, GPs and community midwives.

As well as structural defects other 'congenital' cardiac conditions leading to perinatal emergencies will also be discussed, namely neonatal arrhythmias and perinatal myocarditis. For any practitioner considering a diagnosis of congenital cardiac emergency early liaison with the appropriate congenital heart centre is mandatory.

DUCT-DEPENDANT CONGENITAL (STRUCTURAL) CARDIAC EMERGENCIES

With a UK prevalence of 5.3 cases per 1000 live births congenital heart disease (CHD) is one of the commonest congenital anomalies (Wren et al, 1999). Structural critical congenital cardiac emergencies can present in the first hours, days or weeks of life with the transition from fetal to neonatal circulation exposing many of the morphological anomalies. The physiological effects of arterial duct closure in the presence of CHD can result in long-term morbidity or even death (Abu-Harb et al, 1994).

The North of England regional study (Wren et al, 1999) showed that one in ten infants with CHD presented before neonatal screening and that more than half were missed by routine neonatal screening. Furthermore 1.5% of babies with CHD died undiagnosed before the 6-week check, the majority with duct-dependant lesions.

Improvements in surgical technique and intensive care mean for some infants the greatest risk occurs at initial presentation and stabilization rather than during surgical correction or palliation. For infants with transposition of the great arteries surgical mortality rates approaching 0% are reported (Stark et al, 2000), whereas 4% of infants with simple transposition of the great arteries died before surgery in a Toronto series (Soongswang et al, 1998).

For those with duct-dependant CHD the restoration or maintenance of ductal patency is vital. The duct can then play one of three roles:

1. To maintain adequate pulmonary blood flow, e.g. in pulmonary atresia
2. To improve mixing of the systemic and pulmonary circulations, e.g. in transposition of the great arteries
3. To maintain adequate systemic blood flow, e.g. in left heart obstructive lesions.

The classic modes of presentation are increasing cyanosis in the first two cases and heart failure with inadequate systemic perfusion in the latter case. Significantly, lesions whose pathophysiology consists of left to right shunting, such as septal defects and truncus arteriosus, do not classically present until the pulmonary vascular resistance falls completely at around 4–6 weeks, and will not be discussed further.

General management

The management of critically ill children, focused by resuscitation courses, commences with recognition of the seriously ill child followed by resuscitation and appropriate treatment following the usual ABC (airway, breathing, circulation) approach. This is appropriate for suspected duct-dependant CHD neonates, not least because the diagnosis may be erroneous.

The airway must be assessed and secured if necessary. Breathing should be supported with supplemental oxygen and mechanical ventilation, if necessary. The circulation is resuscitated with 10–20 ml/kg of fluid and then inotropic support commencing with dopamine 5–10 μg/kg/min. In any sick neonate assessment and management of blood sugar abnormalities is mandatory.

Although this is the initial approach the clinician must change tack at some point and consider specific treatment with prostaglandin E_2 (PGE_2). The broad categorization of CHD into cyanotic and acyanotic, although simplistic, does offer a bedside management aid for these infants.

Assessment and specific management

Cyanotic CHD: *Suspicion*: The hallmark of cyanotic CHD is persisting cyanosis unresponsive to increasing oxygen treatment. Concerns about oxygen accelerating arterial duct closure are unimportant compared to mismanaging a hypoxic neonate without CHD. Indeed hyperoxia or nitrogen washout (*Figure 1*) is a long-used method of helping differentiate cyanotic neonates and the effect of PGE_2 on the arterial duct is far more potent than that of oxygen, therefore hyperoxia may be used with impunity if PGE_2 is available.

Figure 1. Nitrogen washout test. FiO_2 = fractional inspired oxygen concentration; PaO_2 = partial pressure of oxygen in arterial blood.

FiO_2 1.0 for 10 minutes if remains cyanotic likely secondary to cyanotic heart disease

Defined in blood gases as follows:

PaO_2 <20 kPA → cyanotic heart disease likely

PaO_2 <27 kPa but >20 kPa → equivocal

PaO_2 >27 kPa → respiratory disease

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Any infant in which cyanosis persists must have pre- (right arm) and post-ductal (feet) oxygen saturations measured. Physiologically reduced pulmonary blood flow with a right to left shunt or common mixing is the cause of cyanosis in cyanotic CHD. Preductal arterial blood gases are measured as part of the hyperoxia test, chest X-ray findings, such as pulmonary artery bay and oligaemic lung fields (*Figure 2*), may suggest CHD. Electrocardiography (ECG) may aid discussion with the tertiary center (*Figure 3*) and can be faxed.

These lesions either have duct-dependant pulmonary blood flow, for example pulmonary atresia with intact ventricular septum, or critical pulmonary valve stenosis; or they are admixture lesions like transposition of the great arteries.

It is worth emphasizing that newborns receive relatively low partial pressures of oxygen in-utero and therefore other markers of systemic oxygen delivery are useful to guide management. These include serum lactate levels, acid–base status and end-organ dysfunction.

Figure 2. Pulmonary artery bay.

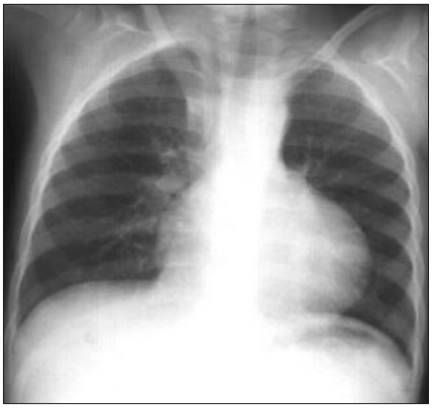
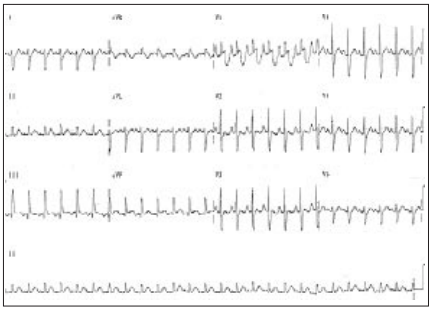


Figure 3. Electrocardiograph with Ebstein's anomaly – note right atrial hypertrophy.



Respiratory distress, chest X-ray changes or a history of either meconium aspiration or birth asphyxia may suggest pulmonary disease, often with secondary persistent pulmonary hypertension of the newborn (PPHN). Differentiating PPHN from CHD in the absence of frank parenchyma lung disease is difficult; the most helpful pointer to CHD in the cyanosed infant is the presence of a heart murmur (Danford et al, 1986). However, occasionally the only way of differentiating duct-dependant cyanotic CHD from PPHN or total anomalous pulmonary venous connection (TAPVC), which itself can masquerade as parenchymal lung disease, is echocardiography (Penny and Shekerdamian, 2001).

The detrimental effects of commencing PGE_2 in babies with alternative diagnoses are trivial, even in PPHN, as long as appropriate ventilation and inotropic support continues. Two CHD lesions may fail to respond to PGE_2, first transposition of the great arteries with intact ventricular septum and restrictive atrial septum where only an urgent balloon atrial septostomy can lead to stabilization, and second obstructed TAPVC where PGE_2-induced increased pulmonary blood flow may worsen the physiological condition. Failure to respond to PGE_2 must prompt urgent liaison with the tertiary centre.

The risk–benefit analysis in the undiagnosed baby supports the following statement: Any infant failing the classic nitrogen washout test with systemic markers of inadequate oxygen delivery should have PGE_2 commenced.

In otherwise well cyanosed infants, with no signs of inadequate oxygen delivery, PGE_2 may be withheld after liaison with a paediatric cardiologist depending on length of transfer and accessibility of PGE_2 en route. After transfer formal echocardiography enables diagnosis and for some lesions balloon atrial septostomy will follow.

Acyanotic CHD: *Suspicion*: More challenging than cyanotic CHD to diagnose, the classic presentation is shock in the presence of abnormal pulses, usually weaker femoral pulses (Danford et al, 1986), and a murmur may also be present. The progression from poor feeding, tachypnoea and lethargy to overt congestive heart failure and shock can be rapid. Furthermore, in the moribund infant with left heart obstruction, all the pulses can be weak as a result of secondary myocardial dysfunction so clinical differentiation from septic shock may be extremely difficult. ECG may aid diagnosis and helps rule out rhythm abnormalities as relative tachycardia may be present. Chest X-ray may show cardiomegaly or plethoric lung fields. Progressive multiorgan impairment evolves secondary to poor perfusion and hypotension with persisting lactic acidosis, renal failure and neurological impairment.

***Specific management*:** Lesions with duct-dependant systemic blood flow include critical aortic stenosis, coarctation of the aorta, interruption of the aortic arch and hypoplastic left heart syndrome.

The initial management of any shocked neonate should again follow the ABC algorithm. Hence initial resuscitation is with high flow oxygen, intubation and ventilation if indicated. Stabilization of the circulation may require 10 ml/kg of volume but this should be limited unless septic shock remains the primary diagnosis. As the differential diagnosis includes septic shock, birth asphyxia and metabolic conditions, antibiotics and blood sugar assessment are vital.

The presence of abnormal pulses in a shocked neonate should lead to the commencement of PGE_2 and early liaison with a congenital heart centre is mandatory.

In a persistently shocked infant with generally weak pulses and no murmur the side effects of PGE_2 are trivial in comparison to not commencing it as the benefits of PGE_2 far outweigh the risks (Brierley, 2001).

Once the duct is patent the predominant task is manipulation of the parallel circulations which are in dynamic competition. An increase in systemic, and therefore coronary, cerebral and aortic flow, at the

expense of pulmonary flow, is achieved by decreasing the inspired content of oxygen, even to air, and in ventilated infants the partial pressure of carbon dioxide (pCO_2) is allowed to rise. Suggested goals to optimize systemic circulation are saturations of 75–85% and a pCO_2 of 4–5 kPa. With a confident diagnosis more aggressive treatment with systemic vasodilators can also be considered.

Many of these infants have an element of myocardial dysfunction at presentation and so inotropes and diuretics to offload the heart may also be indicated. Dopamine 5–10 μg/kg/min and frusemide 1 mg/kg are appropriate.

Prostaglandin

Available preparations: PGE_1 alprostadil and PGE_2 dinoprostone are equally effective in maintaining the duct (Burch and Archer, 1998), but alprostadil is licensed for this use while dinoprostone is not. The latter, however, remains the drug of choice in the author's unit, and many others.

PGI_2 (prostacyclin) or epoprostenol is ineffective for ductal patency and should not be used.

PGE_2 is given by continuous infusion into a dedicated intravenous line. The short half life necessitates a second cannulae to enable rapid re-introduction of drug should the PGE_2 line stop working. The usual dose range is 5–20 ng/kg/min – higher doses are no more effective and carry greater risks of adverse effects (Kramer et al, 1995).

Preparation of drug: Low concentrations are safer in the event of inadvertent line flushing and delivery problems are detected earlier with faster flow rates (*Figure 4*).

Figure 4. Suggested preparation of prostaglandin E_2.

To deliver 5–10 ng/kg/minute (usual starting dose)

15 μg/kg/50 ml at 1–2 ml/hour

or

50 μg/50ml at 0.3–0.6 ml/kg/hour

(compatible with 5% dextrose or normal saline)

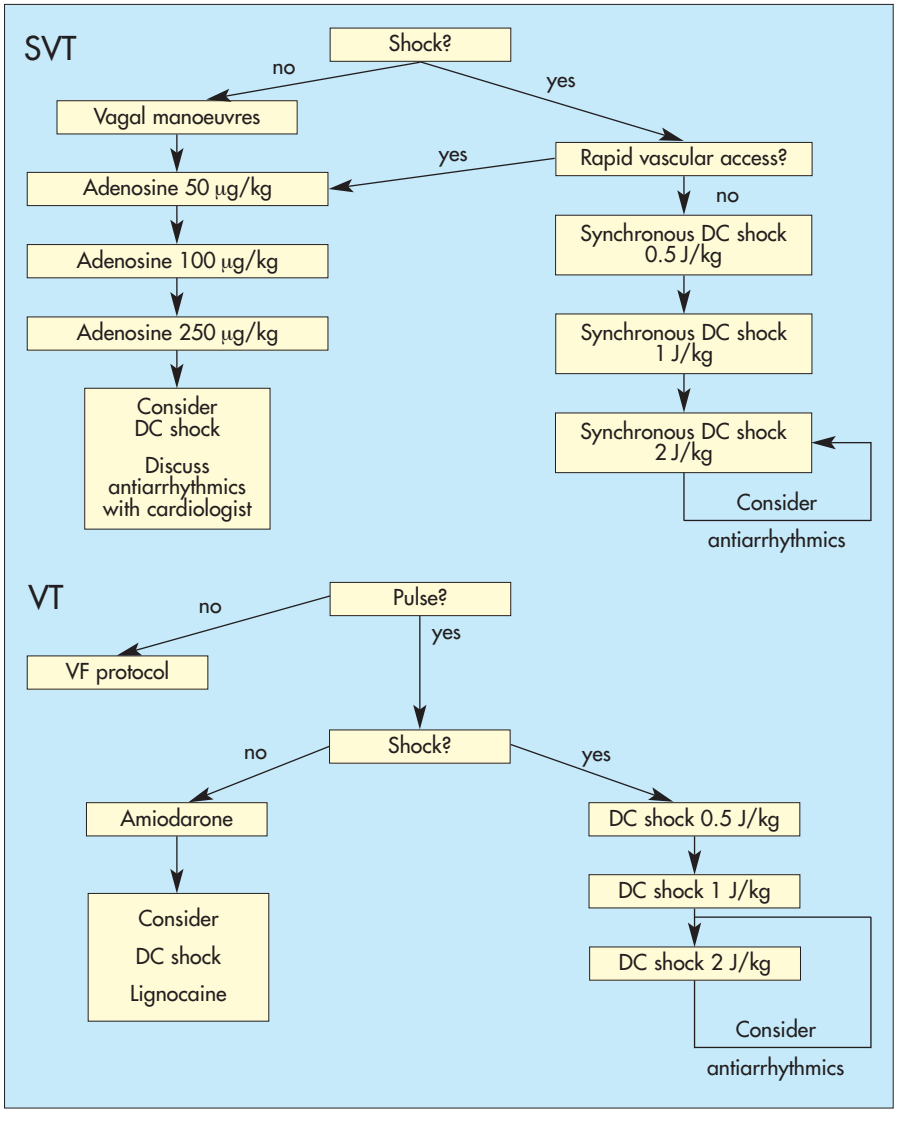
Side effects of PGE_2: The most significant of these, apnoea necessitating ventilatory support, is more common at higher doses, lower birth weight and if PGE_2 is flushed, but can be avoided with respiratory stimulants (Lim et al, 2003). If higher doses are needed prophylactic ventilation is reasonable. Pyrexia, flushing, bradycardia, gastric outlet obstruction and seizures are described. Side effects are never an indication to stop PGE_2.

Antenatal diagnosis

Antenatal diagnosis has the potential to avoid the circulatory deterioration associated with duct closure. Planned care in a congenital heart centre should provide a physiologically well infant for surgical intervention in all but rare cases. Surprisingly survival advantage has been difficult to demonstrate (Bull, 1999; Sullivan, 2002), but more recent literature now supports this (Bonnet et al, 1999; Tworetzky et al, 2001; Franklin et al, 2002). Most infants born with CHD, however, have not been diagnosed antenatally, although this is improving in the better centres (Carvalho et al, 2002).

In essence, if diagnosed in fetal life PGE_2 unresponsive lesions, such as transposition of the great arteries with intact ventricular septum and restrictive atrial septum, should be delivered where urgent treatment, e.g. balloon atrial septostomy, is feasible. Unfortunately the majority of duct-

Figure 5. Simplified algorithms for common tachyarrhythmias. SVT = supraventricular tachycardia; VT = ventricular tachycardia.



dependant CHD infants still present de-novo in the perinatal period.

NON-DUCT-DEPENDANT CONGENITAL CARDIAC EMERGENCIES

Arrhythmia

Arrhythmias can present with shock in the perinatal period, as bradyarrhythmia or tachyarrhythmia may cause inadequate cardiac output.

Palpation of peripheral pulses may lead to suspicion of arrhythmia, with bradycardia easier to detect as the rapid rates associated with supraventricular tachycardia (SVT) may not be accurately palpable. The normal neonatal heart rate is 100 to 160 beats per minute. All neonates presenting in cardiogenic shock should have a 12-lead ECG, and for those with shock and rate outside this range ECG is mandatory. Deviation from the usual finding of a P wave in front of every narrow QRS should lead to liaison with a cardiologist.

Bradycardia: Bradycardia as a result of CHD in the neonate is rare, therefore other causes, such as vagal stimulation in intubated infants or endocrine disturbance, must be excluded. Autoimmune congenital heart block, as a result of transplacental passage of maternal antibodies, is the commonest cause of complete perinatal heart block. With symptomatic bradycardia initial management consists of the standard ABC approach, however, urgent liaison with a paediatric cardiologist must follow. Historically isoprenaline infusion has been used but as the drug is now withdrawn alternatives suggested include inotropes such as adrenaline or salbutamol, which are used in fetal heart block (Groves et al, 1995). Ultimately temporary then permanent pacing may be necessary (Castilla et al, 2004).

Tachycardia: Tachycardia is more common and appropriate algorithms should be readily available in areas receiving sick infants (*Figure 5*) (Advance Life Support Group, 2004). Suffice to say that the presence or absence of shock, whether the heart rate is fast or slow, regular or irregular, and whether the QRS complex on the ECG is broad or narrow are the vital pointers to successful management. Antenatal hydrops fetalis as a result of fetal arrhythmia should have prompted a clear postnatal management plan.

For resuscitation purposes narrow complex tachycardia is SVT, and broad complex is ventricular tachycardia unless proven otherwise by a cardiologist reviewing the ECG. ABC management takes priority unless there is a rhythm that will rapidly respond to DC cardioversion, i.e. ventricular tachycardia, in the presence of shock or ventricular fibrillation. However, in most clinical settings the ABCs will be being attended to long before the defibrillator is found and set up.

For the non-shocked infant, if vagal manoeuvres are unsuccessful, adenosine can transiently ablate atrioventricular node conduction and is extremely safe (Till et al, 1989). As this facilitates diagnosis as well as often terminating the arrhythmia an ECG must be continually running during adenosine administration. In the shocked child appropriate defibrillation is used.

Myocarditis

Neonatal myocarditis presents with tachypnoea, poor feeding and progressive congestive cardiac failure (Inwald et al, 2004). Any neonate presenting with congestive cardiac failure must have a chest X-ray and ECG and although the diagnosis may be suggested by these investigations, by gross cardiomegaly and ST segment depression, formal echocardiography is required. Specific cardiac enzymes are useful, however, early and in-depth liaison with a congenital heart centre is fundamental to guide investigation, management and transfer of these infants (Burch, 2002). Structural anomalies amenable to surgery must be excluded, notably anomalous left coronary artery from the pulmonary artery.

TRANSFER OF THE INFANT WITH CHD AND TELEMEDICINE

Improvements in the transfer process for critically ill children (Britto et al, 1995) mean all congenital heart centres in the UK should have a reliable retrieval service trained in the transfer of babies with congenital cardiac emergencies with clear protocols, e.g. www.cats.nhs.uk/downloads/CATS_CHD.pdf

The major differences from general retrieval are an understanding of the pharmacology of PGE_2 together with parallel circulation manipulation detailed above, the need for rapid access to an appropriate defibrillator and the ability to ventilate in air. Importantly, if stabilization is not possible rapid retrieval to the cardiac centre for balloon atrial septostomy should be considered.

In the future telemedicine offers great promise for children with congenital cardiac emergencies. Indeed transmitted bedside echo reviewed at the tertiary centre has been shown to enable accurate diagnosis and improve management (Mulholland et al, 1999).

CONCLUSIONS

Regarding congenital cardiac emergencies an understanding of the basic physiology and therapeutic manoeuvres enables stabilization of the majority of infants. This vital task, usually performed at local hospital level with tertiary support, is crucial before transfer to the lead centre. Protocols, referral pathways and transport services must therefore be in place for congenital cardiac emergencies in any centre receiving sick infants. HM

Abu-Harb M, Hey E, Wren C (1994) Death in infancy from unrecognised congenital heart disease. *Arch Dis Child* **71**(1): 3–7

Advance Life Support Group (2004) *Advanced Paediatric Life Support: The Practical Approach.* BMJ Books, London

Bonnet D, Coltri A, Butera G et al (1999) Detection of transposition of the great arteries in fetuses reduces neonatal morbidity and mortality. *Circulation* **99:** 916–18

Brierley J (2001) Coarctation of the aorta and interrupted aortic arch. In: Anderson R, ed. *Paediatric Cardiology.* Churchill Livingstone, Edinburgh: 1523–57

Britto J, Nadel S, Maconochie I, Levin M, Habibi P (1995) Morbidity and severity of illness during interhospital transfer: impact of a specialised paediatric retrieval team. *BMJ* **311**(7009): 836–9

Bull C, on behalf of the British Paediatric Cardiac Association (1999) Current and potential impact of fetal diagnosis on the prevalence and spectrum of serious congenital heart disease at term. *Lancet* **354:** 1242–7

Burch M (2002) Heart failure in the young. *Heart* **88:** 198–202

Burch M, Archer N (1998) *Paediatric*

Cardiology, an Introduction. Chapman & Hall, London: 83
Carvalho J, Mavrides E, Shinebourne E, Campbell S, Thilaganathan B (2002) Improving the effectiveness of routine prenatal screening for major congenital heart defects. *Heart* **88:** 387–91
Castilla M, Jerez M, Llacer M, Martinez S (2004) Anaesthetic management in a neonate with congenital complete heart block. *Pediatr Anesth* **14:** 172–5
Danford DA, Gutgesell HP, McNamara DG (1986) Application of information theory to decision analysis in potentially prostaglandin-responsive neonates. *J Am Coll Cardiol* **8:** 1125–30
Groves A, Allan L, Rosenthal E (1995) Therapeutic trial of sympathomimetics in three cases of complete heart block in the fetus. *Circulation* **92**: 3394–6
Franklin O, Burch M, Manning N, Sleeman K, Gould S, Archer N (2002) Prenatal diagnosis of coarctation of the aorta improves survival and reduces morbidity. *Heart* **87:** 67–9
Inwald D, Franklin O, Cubitt D, Peters M, Goldman A, Burch M (2004) Enterovirus myocarditis as a cause of neonatal collapse. *Arch Dis Child Fetal Neonatal Ed* **89**(5): F461–2
Kramer HH, Sommer M, Rammos S, Krogmann O (1995) Evaluation of low dose prostaglandin E1 treatment for ductus dependent congenital heart disease. *Eur J Pediatr* **154**(9): 700–7
Lim DS, Kulik TJ, Kim DW, Charpie JR, Crowley DC, Maher KO (2003) Aminophylline for the prevention of apnea during prostaglandin E1 infusion. *Pediatrics* **112**: e27–9
Mulholland HC, Casey F, Brown D et al (1999) Application of a low cost telemedicine link to the diagnosis of neonatal congenital heart defects by remote consultation. *Heart* **82:** 217–21
Penny DJ, Shekerdemian LS (2001) Management of the neonate with symptomatic congenital heart disease. *Arch Dis Child Fetal Neonatal Ed* **84**(3): F141–5
Soongswang J, Adatia I, Newman C et al (1998) Mortality in potential arterial switch candidates with transposition of the great arteries. *J Am Coll Cardiol* **32:** 753–7
Stark J, Gallivan S, Lovegrove J, Hamilton JR, Monro JL, Pollock JC, Watterson KG (2000) Mortality rates after surgery for congenital heart defects in children and surgeons' performance. *Lancet* **355**(9208): 1004–7
Sullivan ID (2002) Prenatal diagnosis of structural heart disease: does it make a difference to survival? *Heart* **87:** 405–6
Till J, Shinbourne EA, Rigby ML, Clarke B, Ward DE, Rowland E (1989) Efficacy and safety of adenosine in the treatment of supraventricular tachycardia in infants and children. *Br Heart J* **62:** 204–11
Tworetzky W, McEhlinney DB, Reddy V et al (2001) Improved surgical outcome after fetal diagnosis of hypoplastic left heart syndrome. *Circulation* **103:** 1269–73
Wren C, Richmond S, Donaldson L (1999) Presentation of congenital heart disease in infancy: implications for routine examination. *Arch Dis Child Fetal Neonatal Ed* **80:** F49–F53

KEY POINTS

- Management must precede formal diagnosis.
- Initial management of infants with congenital heart disease must follow standard guidelines for the critically ill child.
- Prostaglandin E_2 treatment should be commenced with a low threshold.
- Early and continuing liaison with a congenital heart centre is mandatory.