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Best practice critical cardiac care in the neonatal unit

Michael L. Rigby

Division of Paediatrics, Royal Brompton Hospital, London SW3 6NP, United Kingdom

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ABSTRACT

Major congenital or acquired heart disease in neonates presents with cyanosis, hypoxia, acute circulatory failure or cardiogenic shock. Antenatal diagnosis is made in up to 50% but heart disease is unanticipated in the remainder. The presence of significant heart disease in premature infants is also frequently not suspected at first; in general, whatever the underling cardiac anomaly, the clinical condition is worse, deteriorates more quickly and carries a poorer prognosis in premature and low birth weight infants. Although congenital cardiac malformations are the most likely, other important cardiac disorders are encountered. In general initial treatment options, often without a precise diagnosis, include diuretics, prostin, catecholamines, phosphodiesterase inhibitors, ventilation and occasionally ECMO but the key to successful treatment remains the correct diagnosis. Many conditions will only show significant improvement with treatment by the interventional cardiologist or cardiac surgeon.

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E-mail address: Michael.rigby@rbht.nhs.uk.

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1. Introduction

The presenting features of major congenital heart disease in the newborn are cyanosis and hypoxia or acute circulatory failure sometimes with cardiogenic shock. More generally available antenatal echocardiographic diagnosis now allows conditions to be diagnosed with remarkable accuracy in up to 50% but defacto heart disease is unanticipated in the remainder [1,2]. While increasing numbers of premature infants with major cardiac disease are surviving the first few days of life they are often too small to undergo major cardiac surgery. The presence of significant heart disease will frequently not be suspected at first and prematurity presents special problems in management. In general, whatever the underling cardiac anomaly, the clinical condition is worse, deteriorates more quickly and carries a poorer prognosis in premature and low birth weight infants. Although congenital cardiac malformations are the most likely not all cardiac disorders encountered in the neonate are congenital and as I will discuss may be a consequence of prenatal or perinatal events.

2. Management of hypoxia

Central cyanosis secondary to congenital heart disease may result from a right to left shunt, from common mixing situations or from complete transposition of the great arteries. In general the presence of associated pulmonary stenosis makes the hypoxia more severe. (See Table 1.).

Classical intrapulmonary shunting from focal or multiple pulmonary arteriovenous fistulas may cause profound cyanosis and hypoxia in the absence of any apparent congenital heart disease or lung pathology. Intrapulmonary shunting can also occur in parenchymal lung disease and pulmonary oedema. In cyanotic heart disease with severe hypoxia, positive pressure ventilation or just nasal oxygen delivery can increase oxygen saturations by up to 10% which can have a hugely beneficial effect. Persistent pulmonary hypertension of the newborn may cause severe hypoxia.

What should be the initial management of unexpected hypoxia particularly if there is no immediate access to a cardiology assessment? It is important to remember that an arterial oxygen saturation as low as 68–70% is extremely well tolerated by neonates. Acidosis begins to develop with lower saturations. Saturations of 80% and above are not a matter of immediate major concern and supplemental oxygen can be avoided and intubation and ventilation are not required. Initial examination, blood gases, CXR and ECG are essential and providing the x ray and gases do not indicate a pulmonary cause arrange a cardiology consult. If the oxygen saturation is persistently below 80% or falls further, start prostin at a dose of 5–20 μg/kg/min and add supplemental oxygen up to 80%. Persistent neonatal pulmonary hypertension will not be made worse by using prostin or oxygen. An improvement in saturations after starting prostin makes duct dependant pulmonary blood flow likely [3,4]. Insert umbilical venous and arterial lines to assist drug treatment and transport if required and inform the cardiology team.

Table 1
Causes of hypoxia.

Common mixing
Total anomalous pulmonary venous connection
Univentricular heart variants
Truncus arteriosus (Common arterial trunk)
Common atrium
Right to left shunts
Ebstein malformation with ASD
Tetralogy of Fallot
Severe pulmonary stenosis with ASD
Pulmonary atresia with intact ventricular septum

2.1. Complete transposition (TGA)

Simple TGA always causes marked systemic arterial desaturation and cyanosis [5]. An oxygen saturation of 70–75% is not unusual and this level of oxygenation, while being a warning of possible impending gloom, does not result in symptoms or acidosis. Although relatively unusual, the most alarming consequence of simple TGA (without VSD) is severe acute neonatal hypoxia, acidosis and collapse. When the diagnosis is known, emergency balloon atrial septostomy and intravenous prostin is the treatment of choice. Without a diagnosis and faced with a newborn infant with severe hypoxia, the logical immediate treatment is administration of prostin. Its effect is almost always to improve the clinical situation; by promoting ductal patency and allowing aortopulmonary mixing, oxygen saturations improve. The other benefit is an increase in pulmonary blood flow and consequently flow from the lungs to left atrium, causing an increase in left atrial pressure and as a result, an increased left to right shunt through a patent foramen ovale. These two mechanisms usually allow the arterial saturations to increase above 80%. Balloon atrial septostomy via a percutaneous femoral venous approach or via the umbilical vein is performed when the arterial oxygen saturation is persistently below 70%.

2.2. Duct dependant pulmonary blood flow

There is a group of congenital cardiac anomalies associated with severe pulmonary stenosis or pulmonary atresia in which all or the majority of pulmonary blood flow depends on patency of the arterial duct (Table 2). As a consequence of spontaneous closure of the duct soon after birth, severe hypoxia, and consequent metabolic acidosis will result in early neonatal death. The administration of intravenous prostin will maintain ductal patency or cause a small duct to dilate allowing urgent palliative or even 'corrective' surgery to be performed within hours or days. Prostin is given at a dose of 3–20 μg/kg/min but the lowest effective dose should be used because the drug may cause complications the most important being hypotension due to a fall in systemic vascular resistance and apnoea (Table 3). For these reason prostin should not be given unnecessarily.

In half the cases there will be an antenatal diagnosis and the foetal cardiologist will already have advised on whether or not intravenous prostin should be commenced at birth. Infants with mild to moderate pulmonary stenosis or other sources of pulmonary blood flow such as systemic artery to pulmonary artery collaterals will of course not require prostin. For premature infants considered too small for early surgical treatment the long term administration of low dose prostin (3–5 μg/kg/min) is relatively safe. In some instances an infant who is initially symptom free with mild to moderate central cyanosis and systemic arterial oxygen saturation > 78% will develop more profound cyanosis due to increasingly severe pulmonary stenosis so that early surgical treatment is needed. In cases of Tetralogy of Fallot or tricuspid atresia developing saturations below 80% or early cyanotic spells as well there is usually a good initial response to a B-blocking drug given regularly. This response is useful and safe in low birth weight premature infants who had initially been thought not to require prostin.

Table 2
Duct dependant pulmonary blood flow.

Tetralogy of Fallot with severe pulmonary stenosis or atresia
Other examples of pulmonary atresia with VSD
Pulmonary atresia with intact ventricular septum
Critical pulmonary stenosis
Complete AVSD with severe pulmonary stenosis or atresia
'Single' ventricle variants with severe pulmonary stenosis or atresia
Tricuspid atresia
Double inlet left ventricle
Transposition or DORV with severe pulmonary stenosis or atresia
Miscellaneous

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