

Doctor the baby is blue: an approach to the diagnosis and management

Abstract

Cyanosis in the newborn is relatively common. Most cases are benign though a few like those due to respiratory distress or episodes of apnoea, are more serious and warrant careful review and appropriate management. Rarely cyanosis may be related to cyanotic heart disease in the newborn. That requires an early diagnosis and timely management to avoid potentially short or long term serious sequelae. This brief review highlights the common causes of cyanosis in the newborn. It suggests an approach to its diagnosis before concentrating on the potential serious cardiac causes that may require prompt action. Any blue baby needs to be carefully assessed, to arrive at an early diagnosis to facilitate management, and to achieve an optimal outcome.

Keywords: newborn, cyanosis, airway obstruction, respiratory distress, cyanotic heart disease

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Introduction

Cyanosis in the newborn is relatively common. While most of the causes are generally benign, a few are potentially serious and may lead to both short and long term morbidity and even mortality. There is therefore a need for an accurate diagnosis and timely management. This paper summarizes the not uncommon neonatal conditions that may lead to cyanosis before concentrating on the potentially more serious cardiac causes.

Initial assessment

Any suggestion of cyanosis warrants immediate medical review. It is essential to examine the infant in a bright light, preferably not a blue fluorescent light, as all infants look blue in the dark. Remember that the nursery lights are often turned down during the night.

Is the infant breathing normally?

The history here would be important. It is possible that the baby may be cerebrally depressed and hypo-ventilating from placental transfer of maternal medication – following a general anaesthetic, pain relief opiates, psychotropic medications which include tranquilisers, antidepressants and hypnotics.¹ Cerebral depression may also occur from traumatic deliveries such as a vacuum extraction, forceps and a precipitate labour which may also produce a “stunned” infant.

Is the baby having episodes of apnoea?

Again any cerebrally depressed baby may have brief periods of apnoea which may lead to cyanosis. Apnoea is not uncommon in the premature infant. It may also be a manifestation of seizure activity as a result of hypoxia, hypoglycaemia, hypoccalcaemia, a traumatic delivery, etc. One also needs to consider and exclude the possibility of sepsis particularly meningitis.²

Is there upper airway obstruction?

Newborns are obligatory nose-breathers. Therefore conditions

such as Pierre Robin Syndrome where there is a large cleft palate and micrognathia resulting in the tongue falling back into the cleft and obstructing the nasopharynx, will result in the baby becoming cyanosed at rest but pink on crying. The same would apply if there is bilateral choanal atresia where there are bony plates at the back of the nasal airway. A large lymphangioma or haemangioma compressing the trachea is readily observed.³

Is the cyanosis central or peripheral?

Peripheral cyanosis

Often babies appear blue after a bath in a cold or temperate climate. However the mucous membranes will tend to be pink. Post-term or dysmature infants with rather dry skin peripherally and potentially a high haematocrit arising from a low grade chronic hypoxia from placental insufficiency, commonly have blue peripheries so called acrocyanosis.¹ However once again the tongue and mucous membranes are pink.

A baby may develop bruising of the face following a precipitate labour, a finding more marked if there is a face presentation. Such infants will appear blue but again the mucous membranes will be pink. The bruising involving the face tends to take at least a few days to gradually resolve. The cutaneous oxygen saturation level will be normal in the above patients.

Polycythaemia

A high haematocrit with a haemoglobin of 22gm% or above will give the baby a ruddy blue appearance.⁴ That occurs because there will be a proportion of the haemoglobin that remains unsaturated. Such high haematocrits are not all that uncommon in babies that are post-term, small for dates or may have had a twin-twin transfusion with the recipient baby often looking dusky and plethoric.

Methhaemoglobinemia

Methhaemoglobinemia, a rare autosomal recessive inborn error of metabolism, where the iron in the haemoglobin molecule is oxidised

to ferric rather than remaining as ferrous, may result in the baby looking very blue.⁵ Occasionally oxidising drugs such as some of the older local anaesthetic agents inadvertently injected into the baby, for example via a pudendal block, may result in cyanosis which may be present for many hours.⁶ Again the oxygen saturations will be close to normal. A drop of blood onto the PKU blotting paper turns brown on drying and may be a simple test to carry out in such situations.

Is the baby tachypnoeic?

The most common cause of central cyanosis in the newborn arises from respiratory distress.² That includes such conditions as hyaline membrane disease especially noted in the premature infant, transient tachypnoea of the newborn, meconium aspiration, or the onset of pneumonia. Any large chest occupying lesion such as a diaphragmatic hernia, pneumothorax, a lung cyst, congenital cystic adenomatoid malformation (CCAM), etc, may lead to cyanosis, with the additional factor of a possibly reduced pulmonary vascular bed contributing to pulmonary hypertension. In addition there may be a right to left shunt within the lungs with blood bypassing the pulmonary capillary bed, further contributing to the cyanosis - a ventilation/perfusion mismatch. Tachypnoea occurring on day 1 or 2 is usually respiratory in origin. Very rarely is there a cardiac cause, as for example if the infant is in failure from a familial cardiomyopathy resulting in poor LV function, or a peripheral AV fistula, for example involving the great vein of Galen, resulting in a large left to right shunt. In contrast an intracardiac left to right shunt from example a ventricular septal defect (VSD) will be limited in the early newborn period because of the high pulmonary vascular resistance.

A chest X-ray may be helpful, particularly to exclude a space occupying lesion within the chest. At times urgent treatment is required as for example with a large pneumothorax. Delivery at or transfer to an appropriate tertiary centre may be in order, particularly in those where it has been possible to make a prenatal diagnosis of such conditions as a diaphragmatic hernia or a large CCAM. Any cyanosed neonate with tachypnoea requires a clear diagnosis and appropriate management usually oxygen therapy±respiratory support±antibiotics depending on the risk factors.

Cardiac causes

These fall into three broad groups.^{7,8}

Separate circulations

Transposition of the great vessels especially those with an intact ventricular septum which occurs in approximately 70% of cases, leads to early cyanosis of the newborn. As the duct closes, which may occur rapidly over the first hour or so of life, or more slowly over the first day or so, increasing cyanosis is noted. If there is no pulmonary stenosis and/or VSD there is no murmur to hear. The unsaturated systemic venous blood returns to the right atrium and from there into the right ventricle which ejects it into the aorta while the saturated pulmonary venous blood returns to the left ventricle to be ejected back into the pulmonary artery (Figure 1). Mixing only occurs if there is no ventricular septal defect, at duct level if it is still open, or at patent foramen ovale level, with a left to right shunt at both sites. The only saturated blood that reaches the systemic circulation is through the foramen ovale which if small results in a very blue baby. An urgent diagnosis is required as a balloon atrial septostomy via a cardiac catheter at the bedside under ultrasound guidance will allow

for greater mixing at atrial level. Transfer to a tertiary surgical centre may then be undertaken at a convenient time. If facilities are not available for an ultrasound diagnosis and/or balloon atrial septostomy, then maintaining the duct patency via a prostaglandin E1 infusion will be helpful and may stabilize the baby for transfer to a tertiary centre.

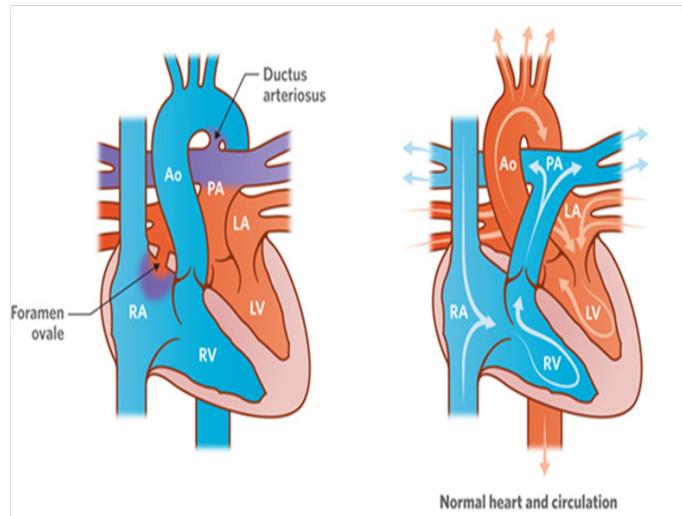


Figure 1 Transposition of the great arteries.

Right to left shunt

Congenital heart defect

Fallot's Tetralogy is the classical example where there is a large ventricular septal defect and severe pulmonary stenosis thereby allowing some desaturated blood to cross the ventricular septal defect into the systemic circulation because of the high resistance caused by the pulmonary stenosis. Newborns so affected will have a murmur from the pulmonary stenosis which will draw attention to a probable cardiac cause for the cyanosis. However there are other abnormalities where like transposition of the great vessels with an intact ventricular septum, there may be no murmurs. For example pulmonary atresia with an intact ventricular septum will lead to cyanosis in the newborn with blood that enters the right ventricle being sent back to the right atrium because of the lack of any forward flow, and then passing across the foramen ovale into the left atrium and out into the systemic circulation (Figure 2). Maintaining duct patency is essential in such infants as once the duct closes the infant will die. Commencement of a Prostaglandin E1 infusion is lifesaving and needs to be maintained until a surgical systemic arterial shunt is created usually between the right subclavian artery and the right pulmonary artery.

Obstructed total anomalous pulmonary venous drainage will cause severe cyanosis in the newborn period again without the infant having a murmur. The intracardiac anatomy appears to be normal echocardiographically but the shunt across the foramen ovale is all right to left with the observation of a channel arising from the pulmonary venous confluence being directed caudally into the portal circulation (Figure 3).

Persistent foetal circulation

At birth if all goes well the circulation rapidly changes from a foetal circulation to a so called "adult" circulation with a left to right shunt at duct level and at foramen ovale level. That occurs because of

a dramatic fall in the pulmonary vascular resistance with the first few breaths that the baby takes. The lung expands, the alveoli fill with air and the capillary beds rapidly open filling with blood exponentially expanding the pulmonary vascular bed. That is followed by the expulsion of alveoli fluid over the next few hours, a growth of further lung tissue over the next few days and weeks and a gradual thinning out of the thick walls of the branch pulmonary arteries and arterioles. Over the first few hours and days of life, the circulation is labile or "transitional" so that anything that causes a sustained rise in the pulmonary vascular resistance may result in reduced pulmonary blood flow. That causes a drop in the left atrial pressures because of a reduced pulmonary venous return. The right atrial pressures may increase following a rise in the right ventricular end diastolic pressure resulting in a bidirectional shunt at atrial level. In addition there will be bidirectional shunting at duct level confirmed by a drop in the lower extremity oxygen saturations compared to the saturations in the right upper limb. The right to left shunting at duct level occurs because the pulmonary vascular resistance at times will be higher than the systemic vascular resistance causing the duct to reverse. The development of a persistent foetal circulation (PFC) or so-called persistent pulmonary hypertension, results in the baby becoming cyanosed despite at times a normal or mildly increased respiratory rate.

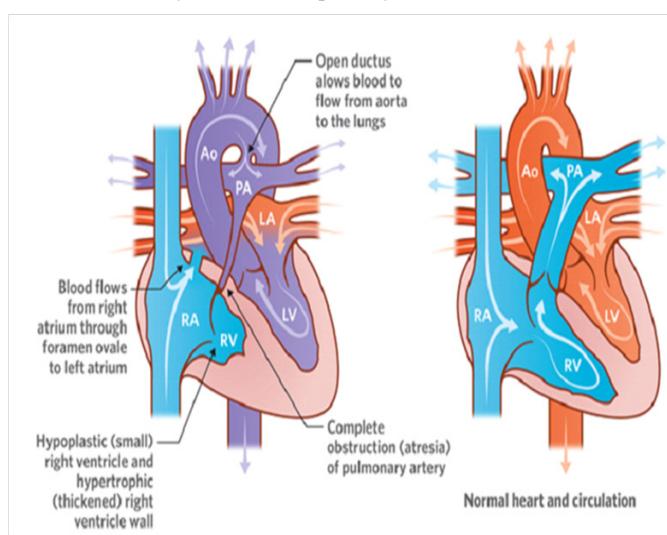


Figure 2 Pulmonary atresia with intact ventricular septum.

The causes are multiple and may include the baby getting cold and/or being over handled, becoming dehydrated from vomiting and/or diarrhoea, having a very high haematocrit, respiratory distress from whatever cause for example a diaphragmatic hernia (see above) or transient tachypnoea of the newborn. In addition it is essential to consider and treat for sepsis. However at times a cause may not be found despite extensive investigation. The management of PFC is to treat the cause wherever possible. Dropping the pulmonary vascular resistance by oxygen ± respiratory support ± nitric oxide will help reverse the process back to an “adult” circulation.

Obligatory mixing

Infants with a truncus arteriosus (Figure 4) or an unobstructed total anomalous pulmonary venous drainage (Figure 5), will have reduced oxygen saturations which run at about an 88-92% level, a level difficult to tell clinically. Such infants tend not to present in the newborn period but later on either with a murmur and cardiac failure

as with a truncus, or recurrent chest infections and a soft murmur with unobstructed total anomalous pulmonary venous drainage. Obligatory mixing is only mentioned to be complete and to help explain some infants who may be monitored by regular oxygen saturations and found to have persistently low oxygen saturation despite essentially normal findings. An echocardiogram will clarify the diagnosis.

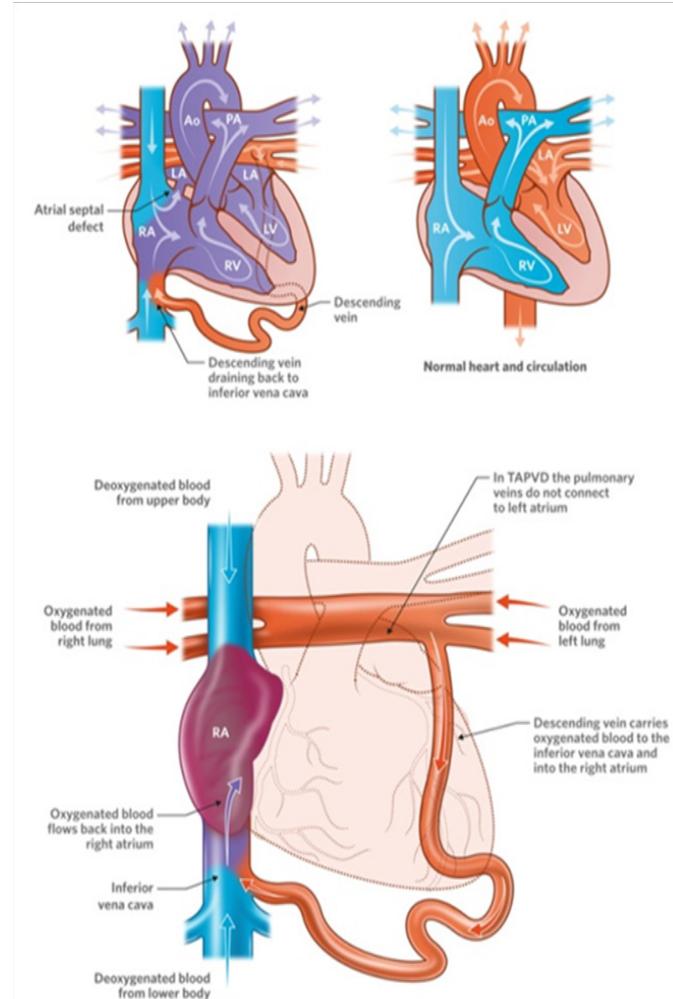


Figure 3 Infracardiac TAPVD

The very sick baby

Finally any sick baby may be blue, whether from cardiac failure as for example arising from critical aortic valve stenosis or myocarditis, respiratory failure for example from meconium aspiration or neonatal pneumonia, or severe sepsis for example septicaemia or meningitis, or from an acute surgical abdomen for example necrotising enterocolitis. While the cyanosis will tend to be peripheral, if the illness is severe, there will also be a drop in the oxygen saturation following a drop in cardiac output with the baby at times looking pale and constricted from acidosis and a high lactate. Early recognition and immediate treatment is required.

Newborn screening

Newborn screening for critical congenital heart disease helps pick up duct dependent systemic circulation abnormalities where there is little or no murmur, for example from a tight coarctation of the aorta or

an interrupted aortic arch.⁹ The peripheral circulation is maintained by the right ventricle ejecting blood through the duct into the descending aorta so that the saturations in the lower limb will be less than the saturations in the right upper limb (pseudotral). Such screening is becoming more widespread and is compulsory in some centres. Duct dependent pulmonary circulations such as pulmonary atresia and an intact ventricular septum will have uniform low saturations

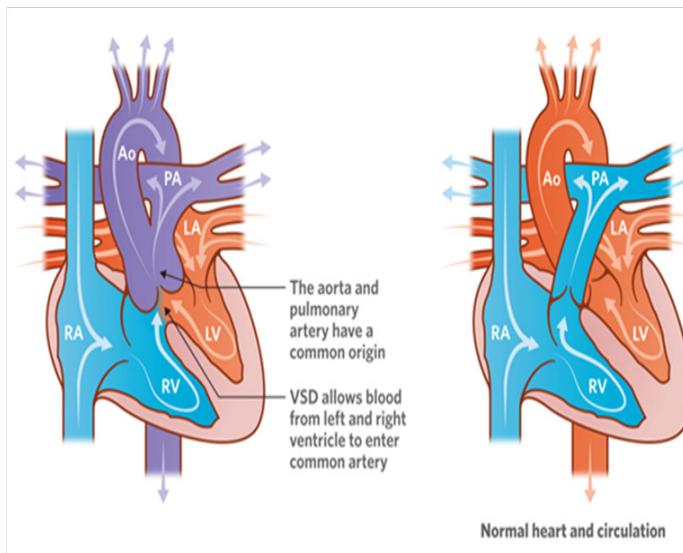


Figure 4 Truncus arteriosus.

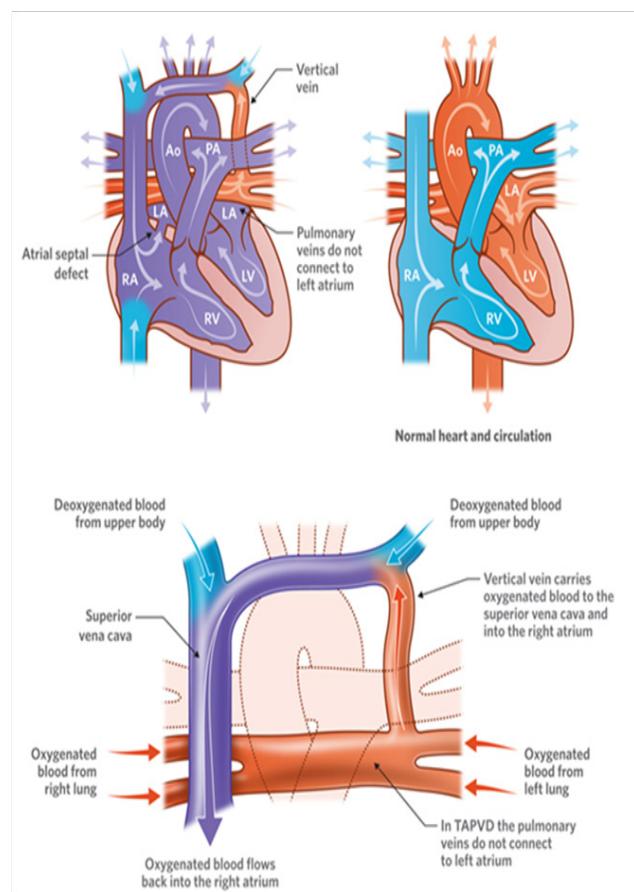


Figure 5 Total anomalous pulmonary venous drainage (TAPVD).

What should one do if there are no echo cardiographic facilities?

Fortunately many of the more serious congenital heart abnormalities are picked up prenatally by initial ultrasound screening and wherever possible subsequent referral to established fetal centres. However it is not unusual for a baby to be born who becomes cyanosed with the question arising as to whether the cause is cardiac or respiratory. A hyperoxemic (hyperoxia) test¹⁰ may be helpful in such situations. Measuring the pO (not oxygen saturation) in air and then after 5-10 minutes in 100% oxygen via a head box, will give some guidance as to the underlying diagnosis. Respiratory problems generally respond dramatically to 100% oxygen with a sharp rise in the pO to 150mmHg or more, at times reaching 250 or even 300mmHg. Transposition of the great vessels start off with a low pO of 28-30mmHg with only a slight rise to 30-32mmHg. Right to left shunts rise from 35-40mmHg to 45-50mmHg. Unobstructed obligatory mixing have a pO of about 75-80mmHg in air rising to 120-130mmHg in 100% oxygen. Please note that the oxygen saturation will be 100% and therefore will not be discriminating.

Once a cardiac cause is suspected or diagnosed and there are no local facilities for treatment, transfer to a surgical centre on Prostaglandin E1 to maintain duct dependency is essential. Delay in the diagnosis may lead to increasing cerebral hypoxia/seizures and possible long term disability and even death. Timely treatment will result with generally excellent outcomes. For example an arterial switch for transposition of the great arteries in best centres results in a mortality of about 1% with a 5% risk for re-operation with good long term survival.

Conclusion

Early diagnosis and timely treatment for any blue baby is essential to obtain the best results. It is mandatory to exclude a serious cardiac cause to avoid short or long term sequelae and/or mortality.

Acknowledgements

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Conflict of interest

Author declares that there is no conflict of interest.

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