Approach to a neonate with cyanosis

Dr Anil Khamkar

M.D. (Pediatrics) Fellow in Neonatology

Academic Head and Senior Consultant Neonatologist

Noble Hospital, Pune

Introduction:

- **Cyanosis** is appearance of skin, nail beds or mucus membranes as blue or purple. It is derived from greek word "kuaneos" which means dark blue. The term was coined by Baumes.
- Oxygenated hemoglobin is bright red, but the reduced hemoglobin is bluish to purple in colour. This colour of the reduced hemoglobin gives rise to the colour seen in cyanosis.
- At some instances it becomes difficult to detect cyanosis in dark-skinned individuals. Despite bruising or ecchymosis resembles cyanosis; the differentiation can be made easily by applying pressure over it which will blanch the cyanosis but not the bruising.
- In **central cyanosis** cyanosis is present throughout the body, including the mucous membranes and tongue. If cyanosis is limited to the extremities, it is called **peripheral cyanosis**, also known as acrocyanosis **(see table 1 for their differentiating features).**

Table. 1. Differentiating features of acrocyanosis and central cyanosis

	Peripheral Cyanosis (acrocyanosis)	Central Cyanosis
Evident in	extremities, nail beds	present throughout the body including tongue, buccal mucosa (warm mucus membrane)
Etiology	mostly physiological due to cold, vasoconstriction, resting baby	potentially life threatening and serious disease eg respiratory, cardiac etc
Mechanism	normal systemic arterial oxygen saturation but increased tissue oxygen extraction leads to a widened systemic arteriovenous oxygen difference of 60 percent (from the normal 40 percent) resulting in an increased concentration of reduced hemoglobin on the venous side of the capillarybed	Central cyanosis is caused by reduced arterial oxygen saturation
Relation with activity	improves	worsens
Relation with improvement in temperature	subsides or becomes better	no relation

Image		

Table. 2 Etiology and probable mechanism of central cyanosis

Disease	Underlying Mechanism
Airway obstruction	
Choanal atresia	Hypoventilation
Macroglossia	
Micrognathia or retrognathia (eg, Pierre-Robin syndrome)	
Laryngotracheomalacia	
Cardiac	
Congenital cyanotic heart disease	Right-to-left shunting
Heart failure/pulmonary edema	Impaired alveolar-arterial diffusion and V/Qmismatch
Hematologic	
Hemoglobinopathies (eg, methemoglobinemia)	Impaired oxygen saturation
Polycythemia	Elevated hemoglobin resulting in low oxygen saturation

Metabolic	
Severe hypoglycemia	Hypoventilation due to decreased or absent respiratory effort secondary to lethargy, seizures, and/or apnea
Inborn errors of metabolism	
Neurologic	
Central nervous system depression	
Apnea of prematurity	Hypoventilation
Infection (eg, meningitis, encephalitis)	
Intraventricular hemorrhage	
Maternal sedation	
Seizure	
Neuromuscular disorder	
Neonatal myasthenia gravis	Hypoventilation
Phrenic nerve injury	
Spinal muscular atrophy type 1 (Wernig-Hoffman disease)	
Pulmonary	
Parenchymal disease	
Atelectasis	V/Q mismatch
Respiratory distress syndrome (Hyaline membrane disease)	
Transient tachypnea of the newborn	
Pulmonary hemorrhage	
Pulmonary hypoplasia	
Pneumonia	
Pulmonary fibrosis	Impaired alveolar-arterial diffusion
Pulmonary edema	Impaired alveolar-arterial diffusion and V/Q mismatch

Nonparenchymal disease	
Pleural effusion	V/Q mismatch
Pneumothorax	
Other	
Persistent pulmonary hypertension of the newborn	Right-to-left shunting

Evaluation of a Neonate with Cyanosis

A. **Stabilisation:** While evaluating underlying cause of cyanosis stabilisation of critically ill baby remains the priority. Provide appropriate cardiorespiratory supportive care keeping in mind ABCs (airway, breathing andcirculation).

B. **Pulse oximetry:** After stabilisation or in a stable cyanosed neonate confirm cyanosis by pulse oximetry. Measure preductal (right hand) and post-ductal (foot) to determine if there is right-to-left shunting. Differential saturations are commonly encountered in conditions like persistent pulmonary hypertension of the newborn (PPHN) and in some cardiac lesions (eg, severe coarctation of the aorta). Many a times right to left shunting occurs through Patent ductus arterioles (PDA) but if shunting occurs at foramen ovale then differential saturations may not beobserved.

C. **History** — Meticulous history taking provides crucial clues to arrive at diagnosis ofcyanosis (see **Table 3**)

Table 3. Pointers in history and probable etiology

History	Possible Diagnosis
GDM in Mother	CCHD like TGA, Polycythemia, Hypoglycemia
Oligohydramnios	Pulmonary hypoplasia
Polyhydramnios	CDH, Airway abnormalities, atresias (eg TEF, Oesophageal atresia), Chromosomal and Genetic disorders
Maternal fever, UTI, PROM	EOS, PPHN
Meconium stained liquor, Foetal Distress	MAS, PPHN, Perinatal asphyxia
Family history of CHD	CCHD
Family history of Hemoglobinopathy	Methemoglobinemia

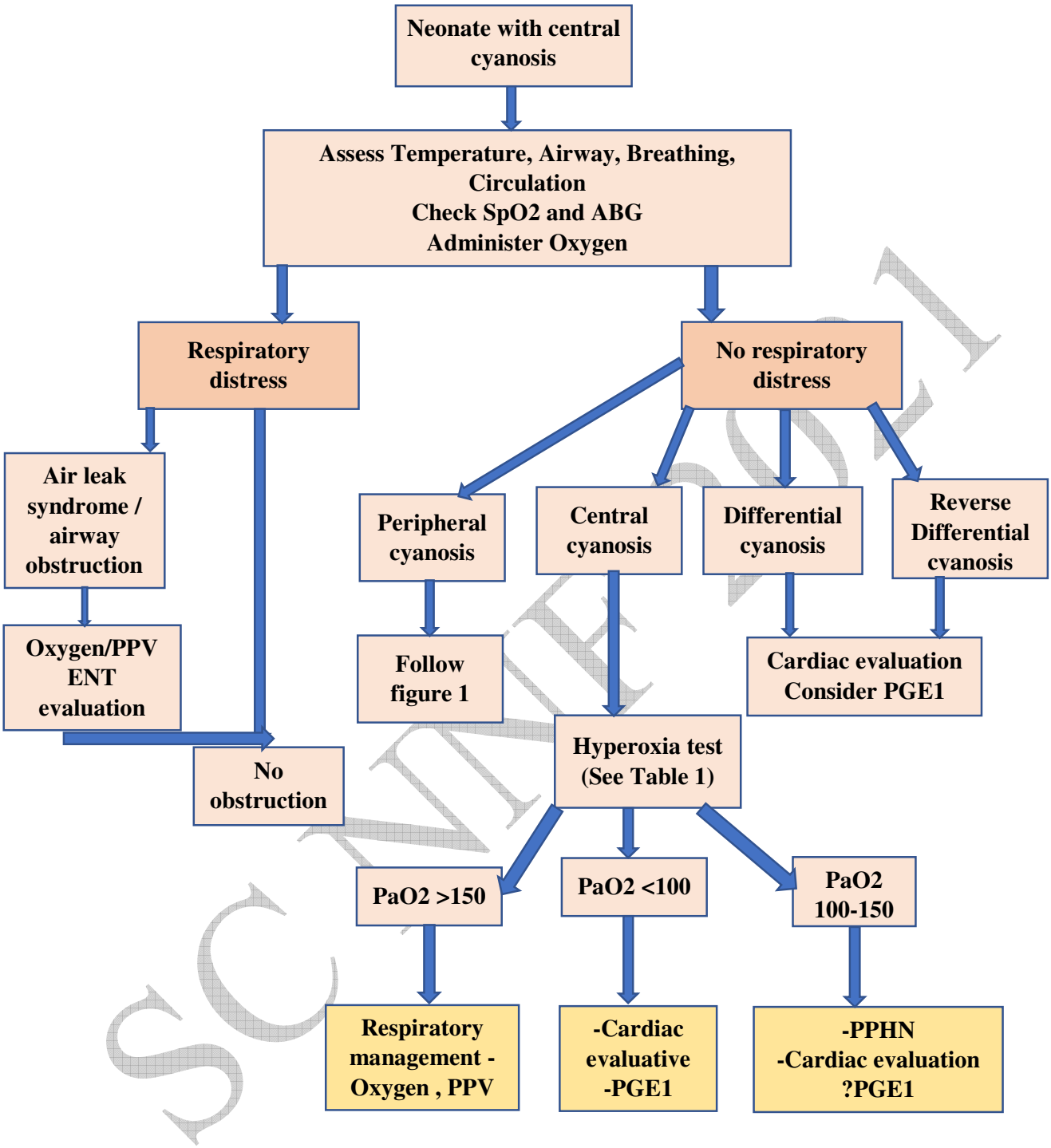


Fig. 1 Approach to a neonate with central cyanosis:

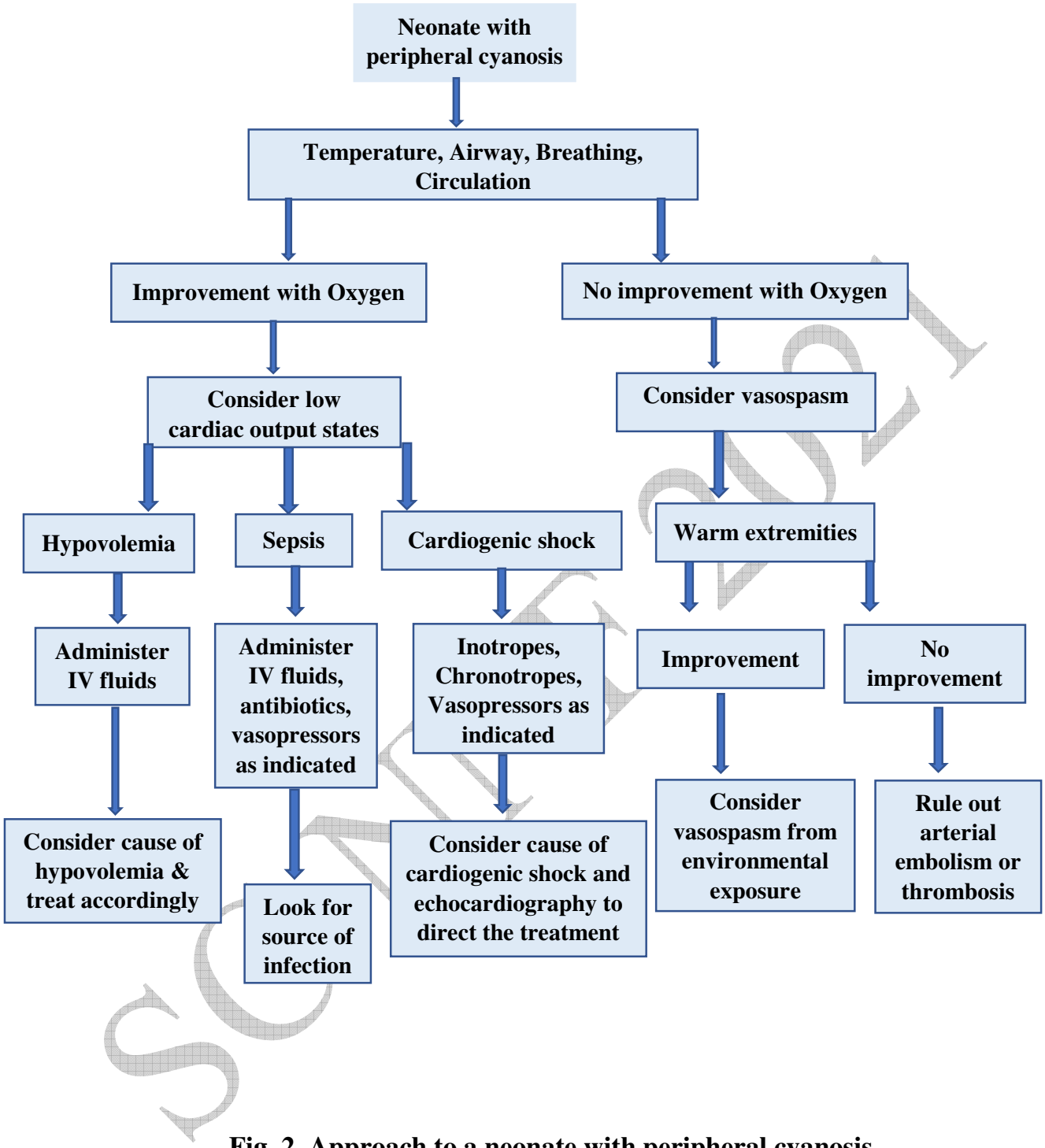


Fig. 2. Approach to a neonate with peripheral cyanosis

Table 4. Hyperoxia test results in neonates:

	PaO2 (percent saturation) when FiO2 = 0.21		PaO_2 (percent saturation) when FiO_2 = 1	$PaCO_2$
Normal	>70 (>95)		>300 (100)	35
Pulmonary disease	50 (85)		>150 (100)	50
Neurologic disease	50 (85)		>150 (100)	50
Methemoglobinemia	>70 (<85)		>200 (<85)	35
Cardiac disease				
• Parallel circulation	<40 (<75)		<50 (<85)	35
• Mixing with reduced pulmonary blood flow	<40 (<75)		<50 (<85)	35
• Mixing without restricted pulmonary blood flow	40 to 60 (75 to 93)		<150 (<100)	35
	Preductal	**Postductal**		
Differential cyanosis	70 (95)	<40 (<75)	Variable	35 to 50
Reverse differential cyanosis	<40 (<75)	>50 (>90)		

Marino BS, Bird GL, Wernovsky G. Diagnosis and management of the newborn with suspected congenital heart disease. Clin Perinatol 2001; 28:91.

Laboratory Evaluation:

The initial laboratory testing compriseof

1. Arterial blood gas (ABG) – See table 3 for detailed interpretation

2. Chest X ray (CXR) – Help in differentiating lung and cardiac cause

3. Septic screen

4. **Hyperoxia test :**

   Traditionally the hyperoxia test is used to differentiate cyanotic congenital heart disease (CCHD) from pulmonary pathology and other causes of cyanosis (Table 3). Albeit this aids in differentiation of respiratory versus cardiac conditions in resource limited settings awaiting 2D Echo; if there is strong suspicion of cardiac disease and facility exists urgent 2D Echo is done. This will ensure prompt diagnosis and unnecessary exposure to harmful effects of 100 percent oxygen.

5. **Echocardiography:**

   It not only confirms the structural lesions in heart but also provides information of volume status, myocardial contractility, shunts, hemodyanamic significance of PDA and pulmonary pressures. Functional echocardiography provides rational approach towards management of perfusion problems and deranged hemodyanamics . It also aids in serial monitoring and improvement of outcome if done by experienced neonatologist.

Treatment:

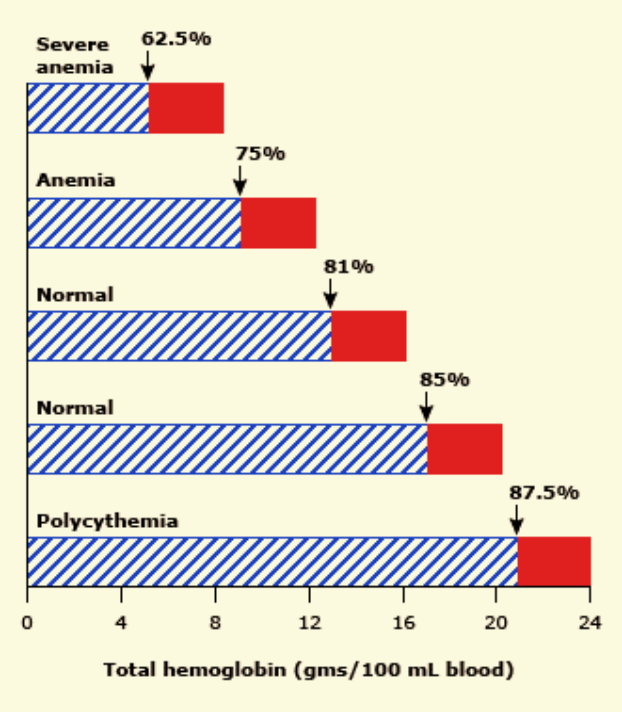
- Initial stabilisation is done by improving temperature, securing position (eg prone position inpierre robin sequence), opening the airway (intubation if required), free flow oxygen or byhood
- To improve saturation, ventilation (mechanical or CPAP), establishing iv access, treatment of shock and to improve perfusion (fluid resuscitation, inotropes).
- In suspected sepsis sending appropriate blood investigations including blood culture and starting antibiotics as per unit protocol
- Correction of hypoglycemia, acid base imbalance, dyselectrolytemia
- Urgent **Echocardiography** in suspected cases of congenital heart disease (mainly cyanotic).
- **Inj prostaglandin E1 (alprostadil)** is initiated at 0.05 to 0.1 mcg/kg/minute as a continuous iv infusion (preferably via a large vein). Sometimes maintenance dose required may be as low as 0.01 mcg/kg/min. Watch for apnea (reported in 10 to 12% neonates with CHDs, particularly if weight is less than 2 kg and more common in first hour of drug infusion). Depending on underlying oetiology, succeeding therapy is directed towards its correction and its consequences.

Summary:

- ***The evaluation of the cyanotic neonate includes assessment for respiratory, cardiac, neurologic, and hematologic disease, with respiratory disease being the most common.***
- ***History and physical examination will guide the initial diagnosis.***
- ***Chest radiograph, the hyperoxia test, and pulse oximetry screening may add information,***

though the definitive diagnostic test for congenital heart disease is echocardiography.

- ***Prompt identification of ductal dependent lesions is essential for effective treatment, which will be discussed in the accompanying article.***



The arterial oxygen saturation level at which cyanosis is detectable at different total hemoglobin concentrations is illustrated above. The solid, red portion of each bar represents 3 g/dL reduced hemoglobin.

Lees MH. Cyanosis of the newborn infant. J Pediatr 1970; 77:484.