Approach to a baby with cyanosis
Objectives

- Cyanosis: types
- Differentials: cardiac vs. non cardiac
- Approach
- Case scenarios
Cyanosis

- Greek word “kuaneos” meaning dark blue
- Bluish discolouration of skin, nail beds, and mucous membranes.
- Depends on absolute concentration of reduced haemoglobin (> 3 g/dl in arterial blood and >5 g/dl in capillary blood)
Types of cyanosis

**ACROCYANOSIS**
Physiological upto 72 hrs
Large arterio-venous oxygen difference

**CENTRAL CYANOSIS**
Pathological
Requires immediate evaluation

**DIFFERENTIAL CYANOSIS**
Definitive congenital heart anomalies (right-to-left shunt through PDA)
Differentials

Cyanotic heart disease
- Decreased pulmonary blood flow
- Increased pulmonary blood flow
- Severe pulmonary venous congestion

Non cardiac causes
- Respiratory disorders
- Persistent fetal circulation
- Central nervous system disorders
- Miscellaneous
Approach

- Confirm central cyanosis
- Pulse oximetry (preductal and postductal)
- Clinical evaluation
- Blood gas analysis
Approach contd

- Chest radiograph
- Hyperoxia test
- Cardiac or non cardiac
- Management
### Approach contd........

**Age at presentation of cyanosis**

<table>
<thead>
<tr>
<th>0-7 days</th>
<th>7-28 days</th>
<th>&gt;28 days</th>
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<tbody>
<tr>
<td>TGA</td>
<td>Truncus arteriosus</td>
<td>TOF like physiology</td>
</tr>
<tr>
<td>PS +IVS</td>
<td>TAPVC</td>
<td>TGA, ASD</td>
</tr>
<tr>
<td>HLHS</td>
<td>TGA, VSD</td>
<td>Truncus Arteriosus</td>
</tr>
<tr>
<td>Severe Ebstein Anomaly</td>
<td>TOF</td>
<td>PPHN group</td>
</tr>
<tr>
<td>TAPVC (obstructed)</td>
<td></td>
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</tbody>
</table>
Approach contd........

Pulse oximetry

- Simultaneous measurements from the right hand and a foot: flow patterns through the ductus arteriosus.
- Avoid left hand.
- Confirms/rejects central cyanosis
- R → L ductal shunting if differential cyanosis
Clinical evaluation: some pointers

- Tachypnea with distress
- Crepitations +
- Cyanosis mild/uniform
- Responsive to oxygen
- Improves with crying
- Age: usually at birth

- Tachypnea, no/less distress
- Crepts -, except with PVH
- Cyanosis variable/uniform
- No/minimal response to oxygen, Worsens with crying
- Usually after 24 hrs

NON CARDIAC

CARDIAC
What next? Hyperoxia Test

Pulse ox reading <85% in room air

Right radial artery ABG in room air

Repeat radial artery ABG

100% oxygen by hood for 15 min.

Jones, 1976
Interpret? Blood gas analysis

- Low pH
- Elevated PaCO2
- PaO2 >250 mm Hg after hyperoxia test (passed hyperoxia test)
- Respiratory acidosis predominantly

- Low pH
- Normal or low PaCO2
- PaO$_2$ < 100 mm Hg/ Rise <10-30 mm Hg (failed hyperoxia test)
- Metabolic acidosis predominantly

NON CARDIAC

CARDIAC
If still in dilemma?
Hyperoxia Hyperventilation Test

- Intubation & hyperventilation

**Rationale**: Pulmonary vasodilation, decreases right to left shunt at atrial or ductal level

Possible PPHN
Approach contd........... (X Ray)
Pulmonary vasculature (Normal)

Normal Vasculature - review

1. Gradual tapering of vessels from central to peripheral
2. Lower lobe vessels larger than upper lobe vessels
3. RDPA diameter
X-Ray: Decreased vascularity

- Dark Lung Field
- Thin peripheral vessels
- Small Hila
Cyanotic heart defects with decreased vascularity (examples)

Critical Pulmonary stenosis/pulmonary atresia with intact ventricular Septum

Tetralogy of Fallot physiology
- TOF (VSD/ PS)
- DORV/ VSD/ PS
- AVSD/ PS
- TGA/ VSD/ PS
- Single ventricle/ PS
- Tricuspid atresia with restrictive VSD and/ or PS
Increased vascularity

- Right des. PA dilated
- Prominent hilar vessels
- Pulm. vasculature traced till lateral 3rd of lung field
- End on vessels >4 in one lung field
Cyanotic heart defects with increased vascularity (examples)

Transposition physiology
- Complete TGA
- DORV/ subpulmonic VSD (Taussig Bing)

Admixture physiology without PS
- At systemic or right atrial level: TAPVR, Mitral/ Aortic atresia with IVS
- At left atrial level: Tricuspid atresia
- At ventricle/ great artery level: Single ventricle, Complete AVSD with straddling AV valve, DORV/ subaortic or inlet VSD, Persistent truncus arteriosus
Pulmonary venous hypertension

- Perihilar Haze
- Fluid in fissures
- Kerley’s Lines

Causes
- Obstructed TAPVR
- HLHS/ Mitral atresia with restrictive ASD

Cephalization
Questions which need to be answered

- Is there an imminent risk of death?
- What group of cardiac lesion?
- What further investigations?
- When to intervene?

Making an exact diagnosis may not always be possible.
ECG: INTERPRETATION

**Axis:** Leads I and aVF are used

1. **P axis:** The P wave must be upright in leads I and aVF.
   - 0 to +90 degree = normal
   - +90 to +180 degree = Atrial inversion
   - 0 to -90 degree = Ectopic atrial pacemaker/ AV junctional rhythm

2. **QRS axis:** The QRS axis is perpendicular to the lead with equiphasic QRS complex (R=S)

3. **T axis:** T waves must be upright in lead I and aVF
## Normal QRS axis

<table>
<thead>
<tr>
<th>Age</th>
<th>Mean ( Range )</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;1 wk</td>
<td>ard +135</td>
</tr>
<tr>
<td>1 week - 1 month</td>
<td>+110 (+30 to +180)</td>
</tr>
<tr>
<td>1-3 months</td>
<td>+70 (+10 to +125)</td>
</tr>
<tr>
<td>3 month - 3 years</td>
<td>+60 (+10 to 110)</td>
</tr>
<tr>
<td>&gt;3 years</td>
<td>+60 (+20 to 120)</td>
</tr>
<tr>
<td>Adult</td>
<td>+50 (-30 to +105)</td>
</tr>
</tbody>
</table>
Abnormal QRS axis

- LAD – QRS axis is less than lower limit of normal for age.
  (a) LVH   (b) LBBB
  (c) Left anterior hemiblock
- RAD – QRS axis is greater than upper limit of normal for age.
  (a) RVH   (b) RBBB
- Superior QRS axis: S>R in aVF
  (a) Endocardial cushion defect (ECD)
  (b) Tricuspid atresia
  (c) RBBB
Further Evaluation

- Echocardiography: To confirm the type of lesion
- Cardiac catheterisation studies
- Angiography: confirmation, haemodynamics, oxygenation, intervention
- MRI: diagnostic for anomalies in pulmonary arteries, aorta, and vena cava
Management: Role of PG E1

Indications:
- Cyanotic newborn suspected to have duct dependent lesion
- Echo proven duct dependent cardiac lesions

Dose: 0.01 mcg/kg/min to 0.1 mcg/kg/min; gradually dec. to 0.025 mcg/kg/min before stopping (Neofax 2010)

Side effects: Apnea, pulmonary congestion, fever, hypotension, seizures, and diarrhea
Case 1

A neonate is profoundly cyanosed and lethargic in his cot at 22 hours of life.

- Clinical examination reveals a soft systolic murmur heard at the left sternal edge and a single second heart sound
- Blood gas: unavailable
- ECG: normal neonatal pattern
- Chest X ray: available
Cardiomegaly with typical egg on side appearance, increased pulmonary blood flow

Transposition of great arteries
CASE 2

A 3 mo infant presented with bluish discoloration of lips on crying since past 2 weeks

- No H/o suck-rest –suck cycle/ sweating/ cough or breathlessness
- Clinical examination reveals HR:110/min, RR:28/min. Central cyanosis+ worsening on crying. Apex beat in 4th ICS inside MCL . ESM Grade 3/6 best heard in Pulmonary area. S1 N S2 single
- ECG and chest X ray is available
Tetralogy of Fallot

Boot shaped heart with right sided aortic arch

RAD with RVH
Case 3

Preterm (34 wks) neonate born by normal vaginal delivery with mild respiratory distress and cyanosis

- Put on CPAP
- Spo2 decreased from 95% on room air to 78% on 45% Fio2
- RR=60/min with Intercostal recession with decreased air entry on the left
- CVS: S1 S2 normal. No murmur
Air fluid levels in chest with defect in diaphragm

Congenital diaphragmatic hernia