Duct-dependent Neonates Management

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Congenital heart defects with ductus-dependent circulation are defined as abnormalities, in which the permeability of the ductus arteriosus is mandatory in order to maintain systemic perfusion.
Anatomy of Ductus Arteriosus

Connects the main pulmonary artery to descending aorta.
Physiology of Ductus Arteriosus

- Carries 60% of combined ventricular output.
- Diverts blood from high resistance pulmonary circulation to low resistance descending aorta and placental circulation.
- PGE1 and PGI2 formed intra-murally and in placenta maintain ductal patency in fetal life.
Prenatal Physiology
Prenatal Physiology

- Severe left or right sided lesions do not affect fetal growth and development.
- Factors that mimic fetal circulation will stabilize the neonate.
- Ductus-dependent fetal cardiac defects are contraindications to the maternal use of prostaglandin inhibitors during pregnancy.
Post Natal Closure of PDA

Functional closure
- In 15 hr., contraction of medial smooth muscles due to $\uparrow$ PO2 & $\downarrow$ PGE1.

Anatomical closure
- In 3 wk., replacement of muscle fibres with fibrosis creating ligamentum arteriosus & the duct loses the ability to reopen
Duct dependent Physiology

- Pulmonary Circulation
- Systemic Circulation
- Both Circulation - Intercirculatory Mixing
Duct dependent Pulmonary circulation

• Pulmonary atresia- VSD or Intact Septum
• Severe TOF
• Neonatal Ebsteins
• Critical PS- Isolated or with complex CHD
• Severe Tricuspid Valve regurgitation
Duct dependent systemic circulation

- Left heart: Hypoplastic left heart
- Critical AS
- CoA
- Interrupted arch
Transposition of Great Arteries
Pulmonary Vs Systemic

Pulmonary
Cyanosis predominant- Hypoxia dominating the clinical picture
Relatively well at least initially- present in 24-48 hours of life

Systemic
Poor Perfusion- Decreased cardiac output
Sick infants- present in the first 2 weeks of life.
Effect of hypoxemia in duct-dependent CHD

- Decreased pulmonary blood flow
  - Decreased oxygen uptake
  - Systemic arterial hypoxemia
    - Decreased tissue oxygen delivery
      - Increased anaerobic glycolysis
        - Increased lactic acid production
        - Decreased CO₂ production

Sequence of events associated with inadequate oxygen supply to tissues resulting in metabolic acidemia and decreased CO₂ production.
• babies with higher fetal Hb level will have late visible cyanosis.

• Very sick babies usually have cyanotic spell or congestive heart failure and circulatory collapse without clinical cyanosis.

• An inaudible murmur must not be criteria for exclusion of CHD, and some times, the deterioration of the clinical condition with disappearance of murmur is a pointer for an urgent intervention.

• Involvement of multiple organs like kidney, brain or skeletal system, which may add up to the morbidity and mortality. Hence a detailed examination and parental counseling is required.
Duct-dependent pulmonary Circulation

- Lungs are underperfused in these babies.

- PDA diverts partially saturated systemic blood towards the pulmonary circulation to improve the overall saturation.

- Rarely, a widely open duct may raise the PaO2 > 49 mm Hg. Therefore, the concentration of oxygen, to start ductal constriction, is seldom achieved by oxygen supplementation.

- PaO2 remains in the range of 35 to 40 mm Hg

- Least benefitted by oxygen administration and the administration of 100 percent oxygen only increases the dissolved oxygen level.
Obstruction of Pulmonary Flow

Resistance Blood Flow

RVH

Fails

Right Atrial Pressure ↑

Persistent Opening Foramen Ovale

Shunting of unoxygenated blood from the right atrium into the left atrium

Systemic Cyanosis
Suspect and Rule out

- Any baby with shock or collapse after 24 hours - "Duct Dependent systemic circulation"
  - Poor perfusion, absent pulses, pallor, shock
- Cyanosis
- Differential cyanosis
- Respiratory symptoms but lungs clear
- No response to oxygen or worsening with oxygen
Differential Diagnoses

- Pulmonary hypertension of the New born (PPHN)
- Neonatal Sepsis
- Metabolic disorders
- Primary lung pathology
- Obstructed TAPVC
- Methemoglobinemia
- Other disorders - choanal atresia, Pierre Robin sequence, vascular ring, diaphragmatic hernia, acyanotic CHDs with shunt reversal, chest infections, hypothermia and hypoglycemia.
What to DO???
Neonatal Cardiac Evaluation

• General Evaluation
  Airway, Breathing, Circulation
  History and Examination

• Inspection and Palpation
  Activity
  Cyanosis
  Peripheral Perfusion
  Dysmorphism and System Review
Neonatal Cardiac Evaluation

- Auscultation
  
  Single S2
  
  Murmurs +/-
Neonatal Cardiac Evaluation

- Saturations
- 4limb BP
- Liver Size
- Auscultation
- CXR
- Hyperoxia Test
- ABG- pH, PCO2, pO2, HCO3, Blood Sugar
- Septic Workup
**Hyperoxia Test**

100% Oxygen (or as close as possible) for 10 minutes
- by Blow by Mask, intubation
- Repeat pO2 assessments by ABG
- Pulseoximeter is Unacceptable

**Measure right arm pO2**

- pO2 > 250 – Unlikely to be Cyanotic CHD
- pO2 100-250 – possible cyanotic CHD
- pO2 <100 – Cyanotic CHD
Pulmonary Oligemia
Diagnosis of duct dependence

Echo is the gold standard
- Delineate the anatomy
- Assess the size of PDA, degree of restriction/constriction
- Assess PVR/SVR ratio on the flow pattern across the duct
Management Principles

Back to Basics

A, B, C and Drugs (Prostaglandin E1)
Management of Sick Neonate

- Ensure Airways
- Optimise Oxygen
- Breathing
  - Good Now
  - Will it Sustain?
  - Fatigue an issue
  - When to ventilate?
- Ventilate:
  Off load work of breathing
  Ensure good Oxygenation
  Secure Lines
  Secure airways
  Concomitant respiratory issues
When in doubt

- Blue baby
- Baby in shock

Start.................. Prostaglandin
PGE\textsubscript{1} Preparation

- Available as PROSTIN VR from Upjohn pharmacia as \textbf{1 ml ampoules} containing \textbf{500 mcg/ml}.
- We dilute \textbf{0.1 ml (50 mcg)} in \textbf{50 cc 5\% Dextrose} under laminar air flow to avoid sepsis.
- The remaining PGE\textsubscript{1} is stored in a fridge at \textbf{2-8\textdegree C}.
- Dilute \textbf{0.1 ml (50 mcg)} in \textbf{50 ml 5\%D}; in this dilution, each ml equals \textbf{1 mcg of PGE}\textsubscript{1}.
PGE₁ infusion protocol

- PGE1 infusion is started at 0.05 - 0.1 mcg/kg/min
- Depending on clinical response (Saturations, femoral Pulse, urine output) infusion rates can be increased up to 0.4 mcg/kg/min
- Once a desired response is obtained, (stable sats of 70 - 80%) maintain patency by continuing infusions at 0.01 - 0.05 mcg/kg/min
- It is often possible to bring down the rate to low levels to maintain effect
How long does one Ampoule last?

<table>
<thead>
<tr>
<th>Weight (kg)</th>
<th>0.01 mcg/kg</th>
<th>0.025 mcg/kg</th>
<th>0.05 mcg/kg</th>
<th>0.1 mcg/kg</th>
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</thead>
<tbody>
<tr>
<td>2 kg</td>
<td>17 days</td>
<td>7 days</td>
<td>3.5 days</td>
<td>2 days</td>
</tr>
<tr>
<td>3 kg</td>
<td>12 days</td>
<td>5 days</td>
<td>3 days</td>
<td>1.5 days</td>
</tr>
<tr>
<td>4 kg</td>
<td>8 days</td>
<td>3.5 days</td>
<td>2 days</td>
<td>1 day</td>
</tr>
</tbody>
</table>
Monitoring PGE₁

Hypotension: Manifests as cold extremities, delayed capillary refill > 3 min tachycardia, low urine

Rx: IV fluid bolus 5-10 ml/kg N. Saline in

Apnea: Often noted in first hours, noted by apnea monitor or a watchful nurse (ideal). common in < 2 kg, preterms

Rx: Stimulation, Hand Bagging, Ventilation
Monitoring PGE₁

Fever: often in first 24 hours, needs tepid sponging
  Rx: Ensure that insensible fluid losses of a febrile neonate are met adequately

Fluid retention:
  Often a common problem

Seizures:
  Rare, may need cessation of PGE₁ infusion
  Rx: Rule out other metabolic and infective causes in a sick neonate
Monitoring PGE₁

Bradycardia: if severe stop infusion
Flushing: often noted in first few hours
Others: Diarrhea, sepsis, DIC, Low K⁺.

> 7 DAYS USE: Long bone hyperostosis
  Gastric outlet (antral)obstruction
Goals of management

- To establish the diagnosis after initial stabilization or resuscitation
- Intubation whenever indicated
- To minimize hypoxemia
- Ensure balance between Qp & Qs
The key to successful outcomes in duct dependent lesions is......

to realize that the patient IS duct dependent........!!

Thank you