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Paroxysmal Hypercyanotic Attack

Standard Operating Procedure (SOP)



Union of Myanmar Special region - 1
Pediatric (SOP)

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Union of Myanmar Special region - 1
Pediatric (SOP)

CARDIOLOGY

Paroxysmal Hypercyanotic Attack

- *Sudden severe episodes of intense cyanosis caused by reduction of pulmonary flow in patients with underlying Tetralogy of Fallot or other cyanotic heart lesions that have pulmonary stenosis and ventricular septal defect*
- *Due to an imbalance between pulmonary and systemic vascular resistance or spasm of the right ventricular outflow tract favouring decreased pulmonary flow and increased right-to-left shunting across the ventricular septal defect*
- *Tend to occur in those with mild-to-moderate cyanosis at rest and are more common in children who are iron deficient*

Diagnosis

History

- *Age of onset of cyanosis, history of previously diagnosed congenital heart disease, history of similar attacks and hospitalization before*
- *History of squatting (a hallmark of TOF although occasionally occurs in other cyanotic lesions)*
- *Common precipitants include crying, defecation, feeding, waking from naps fever, dehydration, tachypnea/tachycardia due to any cause and medications; vasodilators (e.g. ACEI)*

Clinical presentation

- *Spell - starts crying, becomes dyspnoeic, bluer than before, convulsion, may lose consciousness, stroke or death may occur*
- *Frequency-Once in a few days or numerous attacks everyday*
- *Physical examination*
 - *Cyanosis, clubbing*
 - *Cardiac size - Normal, may be enlarged in other cyanotic lesions*
 - *In TOF: Normal 1s* sound, single and soft 2nd sound, in other cyanotic lesions-loud 2nd sound may be heard*
 - *Ejection systolic murmur (murmur intensity will be reduced during spells)*

Treatment

Treat as a medical emergency to prevent the development of serious complications from prolonged hypoxia. The followings are listed in the order of least to most invasive

- *Have parent hold and calm child*
- *Knee-chest/squatting position*
 - *Place the baby on the mother's shoulder with the knees tucked up. underneath.*
 - *This provides a calming effect, reduces systemic venous return and increases systemic vascular resistance*
- *Avoid iatrogenic agitation*
 - *Limit examination, venipuncture, etc.*
 - *No inotropes (e.g. no digoxin, dopamine or dobutamine, adrenaline) and no diuretics*
- *100% oxygen can be administered (Use least aggravating method)*
 - *It will help to decrease peripheral pulmonary vasoconstriction and pulmonary vascular resistance*
 - *However, it is of limited value as the primary abnormality is reduced pulmonary blood flow and therefore, if it distresses the child, it should be discontinued*
- *Morphine - CS/IM 0.1-0.2 mg/kg*
 - *It can cause sedation and also decreases sympathetic tone, decreases oxygen consumption, decrease tachypnea and decreases pulmonary vascular resistance (Caution in infants under 3 months)*
 - *Diazepam and other sedations are contraindicated in this stage*

If the above measures are ineffective, the following treatments may need to be given with IV access

- *Fluid bolus if needed (Crystalloid or colloid fluid bolus: 10-20 ml/kg by rapid IV push to increase preload)*
- *Beta-blockers - IV Propranolol 0.05 -0.25 mg/kg >5 - 10 minutes*
 - *It may lessen infundibular spasm, decrease oxygen consumption and decrease tachycardia leading to greater diastolic filling and reduce right to left shunting*
- *IV Sodium bicarbonate 1-2 mEq/kg/dose, repeat in 10-15 minutes if needed.*
 - *It reduces the respiratory centre stimulating effect of metabolic acidosis*

In resistant cases, consider

- *Phenylephrine: Bolus - 0.1 mg/kg IM/SC or IV 0.01 mg/kg slowly*
Start infusion: 0.1-0.5 µg/kg/min IV OR
- *Methoxamine 0.1 mg/kg IV (It increases systemic vascular resistance) OR*
- *Noradrenaline infusion (It increases systemic vascular resistance)*
 - *0.02-0.1 µg/kg/min of noradrenaline base**How to prepare noradrenaline infusion*

$$\frac{\text{mg of Noradrenaline in 100 ml}}{(5\%D/W \text{ or } D/S)} = \frac{6 \times \text{body weight (kg)} \times \text{Dose } (\mu\text{g/kg/min})}{\text{Rate (ml/hr)}}$$

(Noradrenaline base 1 mg is equivalent to noradrenaline acid tartrate 2 mg)

- *Dose should be titrated according to response to a maximum of 1.0 4μg/kg/min (titrate to effect- reflex bradycardia, raised BP, increased pulse oximeter saturation and monitor ECG and haemodynamic status)*
- *IV infusion dilute in 5% glucose or D/S and administer preferably via a central line as local extravasation into tissue may cause necrosis.*
Noradrenaline is incompatible with sodium bicarbonate and alkaline solution
- *The availability of these medications may be limited in some centers and these should only be given in a monitored environment (e.g. HDU or ICU)*
- *Intubate/paralyse/anesthetize or heavy sedation and mechanical ventilation under supervision of Anesthesiologist*
 - *IM Ketamine 1-3 mg/kg or General Anesthesia*
 - *It causes sedation and increases systemic vascular resistance*
 - *It reduces oxygen consumption to minimum*
- *Surgical shunt or RVOT stent emergently. (Refer to Cardiac Center)*

Other notes

- *A single episode of hypercyanotic spell is an indication for early surgical referral (either total repair or Blalock Taussig shunt)*
- *Oral Propranolol 0.2-1 mg/kg/dose 8 to 12 hourly should be started soon after stabilization while waiting for surgical shunt*
- *Correct iron deficiency anaemia*