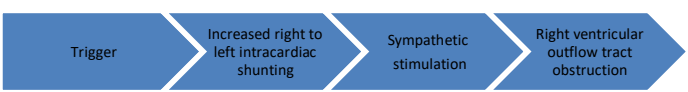
HYPERCYANOTIC/ "TET" SPELLS

Definition:
Tet spells also called a hyper cyanotic spells, hypoxic spells, or cyanotic spells, are a hallmark clinical manifestation of Fallot physiology characterized by a paroxysmal episode of hypoxia due to reduction in pulmonary blood flow .

Etiology:
Tet spells most commonly occur in Tetralogy of Fallot but other congenital heart diseases with Fallot physiology like double outlet right ventricle with pulmonary stenosis, pulmonary atresia with ventricular septal defect, tricuspid atresia with pulmonary stenosis, and transposition of great arteries with ventricular septal defect and pulmonary stenosis can develop cyanotic spells.

Precipitating factors:
Precipitating factors are events that result in sympathetic stimulation e.g. excessive crying, feeding or suckling, light plane of anaesthesia and increased physical activity. Other factors include hypovolaemia, metabolic or respiratory acidosis, higher airway pressures, and factors decreasing systemic vascular resistance.

Pathophysiology:



Presentation:
Hypoxia presents as tachypnea, deep breathing and increasing cyanosis. If not corrected this may end up as syncope, convulsions, cerebrovascular accidents and even death.

Objectives of management:
The objectives of management are

- To decrease the sympathetic surge and reduce catecholamines in blood.
- To increase blood oxygenation and reduce hypoxia.
- To increase systemic vascular resistance.
- To cause a reduction in right ventricular outflow tract obstruction for an increase in pulmonary blood flow.

Management of TET spells:
Prevention:

- Avoid triggers.
- Maintain adequate hydration.
- Premedication before anaesthesia with oral midazolam 0.5mg/kg 30 minutes preoperatively or chloral hydrate 50mg/kg 1 hour preoperatively.
- Awake children should have their parents close to them.
- Choice of anaesthesia with less effect on systemic vascular resistance e.g. Ketamine infusion as opposed to inhalational anaesthesia if appropriate.
- Use of regional or peripheral nerve blocks for better pain management.

Management:

- Stop the triggering agent or precipitating factors.
- Maintain airway and provide supplemental oxygen using facemask in a non-anaesthetized and 100% Oxygen in an anaesthetized patient.
- Anxiolysis or sedation in an awake patient (e.g. i.v. Morphine 0.1-0.2mg/kg) and increasing depth of anaesthesia and provision of analgesia in an anaesthetized patient. Use of Dexmedetomidine, Fentanyl and Ketamine have been described.
- Ensure volume status and give fluid bolus of 10ml/kg. Volume repletion may help with right ventricular outflow tract obstruction mechanically. Excessive fluids should be avoided to

avoid cerebral edema, pulmonary edema and dilutional effects in patients having compensatory polycythemia.

- Use of phenylephrine 1-2 µg/kg up to a dose of 5-10 µg/kg to increase SVR. If higher dose is needed then Norepinephrine can be used at the dose of 0.05 µg/kg/min to 1 µg/kg/min.
- Changing the posture of the patient to a knee chest position (with flexion of knees and hips so that the patient's knees tend to touch the patient's chest). This is to compress the femoral arteries, increase the systemic vascular resistance and decrease venous return.
- Use of Beta blockers e.g. i.v. Propranolol at the dose of 0.015mg/kg to 0.02mg/kg or Esmolol at the dose of 0.5mg/kg administered over 1 minute to relieve infundibular spasm and decrease right ventricular outflow tract obstruction.
- Use of Sodium Bicarbonate to treat metabolic acidosis or hyperventilation in an intubated patient for respiratory acidosis.
- Cardiology referral for definitive or temporizing procedures for management of anatomical cause leading to TET spells.

References:

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