Tetralogy of Fallot – Hypercyanotic Spells

Background

The 'Tet spell' (also called ‘hypoxic spell’, ‘cyanotic spell’, ‘hypercyanotic spell’ or ‘paroxysmal dyspnea’) most frequently occurs in young infants with Tetralogy of Fallot but may occur with other congenital heart defects that have pulmonary or subpulmonary stenosis and a VSD, and at any age. Common precipitants include crying, defecation, feeding, waking from naps (low systemic resistance), fever, dehydration, tachypnea / tachycardia due to any cause, and medications (e.g. ACE inhibitors). They tend to occur in those with mild-to-moderate cyanosis at rest and are more common in children who are iron deficient.

They are characterised by:
- period of uncontrollable crying / panic,
- rapid and deep breathing (hyperpnea),
- deepening of cyanosis,
- decreased intensity of heart murmur,
- limpness, convulsions and rarely, death.

Hypercyanotic spells need to be recognised quickly and effectively controlled to prevent the development of serious complications from prolonged hypoxia. While medical intervention is indicated, many episodes are self-limiting. Those involved in the care of a child with Tetralogy of Fallot should be familiar with a spell’s presentation and early management.

Pathophysiology

Hypercyanotic spells may be best thought of as an imbalance between pulmonary and systemic vascular resistance favouring decreased pulmonary flow and increased right-to-left shunting. Hypoxemia, metabolic acidosis, hyperpnea, increased systemic venous return, catecholamines, and pulmonary vasoconstriction are thought to be involved in an interaction that results in a self-perpetuating cycle. Infundibular spasm is not required.
Management Details - Acute Care

Treatment of spells involves the following procedures in order of increasing complexity:

1. Knee-to-chest”/Squatting”:
Placing the child in the knee-chest position either lying supine or over the parent’s shoulder (see below). This calms the infant, reduces systemic venous return and increases systemic vascular resistance.

2. Oxygen (100%) can be administered but usually has minimal effect. If distressing the child it should be discontinued.

3. Morphine: 0.1-0.2 mg/kg IM. (Caution in infants under 3 months). Thought to act by respiratory centre suppression and sedation thereby reducing hyperpnea.

If the above procedures are ineffective or have suboptimal effect, the following treatments may need to be given in addition depending on the state of the child. Establish IV access and discuss with a senior colleague.

4. Crystalloid or colloid fluid bolus: 10-20ml/kg by rapid IV push.
This maximises preload and should be given prior to the following drugs which may induce hypotension.

5. Metaraminol or beta blocker (e.g. propranolol or esmolol) – see details below.
The availability of these medications may be limited in some centres.

These medications should be used only after discussion with a paediatric cardiologist or intensivist and should only be given in a monitored environment (e.g. HDU or ICU).

6. General anaesthesia and ventilation.

7. Emergency surgical intervention.

Metaraminol (ARAMINE®) 50 mcg/kg IV over 10-15 minutes. Then 0.25-1 mcg /kg/min as infusion
This is a peripheral vasoconstrictor which increases the systemic vascular resistance and reduces the right-to-left shunt.
Esmolol (BREVIBLOC®) 500 mcg/kg over one minute IV, then maintenance of 50 mcg/kg/min can be increased in steps of 50 mcg/kg/min to maximum dose of 300 mcg/kg/min. This is an ultra-short acting cardioselective beta blocker and is thought to work by reducing dynamic muscular stenosis of the right ventricular outflow tract and increasing pulmonary blood flow.

Note: Extravasation of Esmolol can cause severe tissue necrosis.

Ongoing Care

If a child presents to hospital with or following a TET spell, they should not be sent home before discussion with the Paediatrician on call.

If there is a single severe or frequent minor spells, commence oral Propranolol 0.5 mg/kg/dose TDS. This should be done in hospital, watching for hypotension and hypoglycaemia. Increase dose gradually to 1.0-1.5 mg/kg/dose TDS (3.0-4.5 mg/kg/day).

Discuss with Starship Paediatric Cardiology, as a TET spell is usually an indication for early surgery.

References


The Pediatric Cardiology Pharmacopoeia Pediatric Cardiology 2004;25(6) 623-646