Management of Hypercyanotic Spells in Tetralogy of Fallot

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1 Scope

This guideline is intended for use in children with a background of Tetralogy of Fallot presenting to Newcastle Hospitals with a hypercyanotic spell.

2 Background

Tetralogy of Fallot (TOF) is characterised by the Tetralogy of overriding aorta, ventricular septal defect, infundibular / Right Ventricular Outflow Tract / pulmonary stenosis and right ventricular hypertrophy. Tetralogy of Fallot is the most common type of cyanotic congenital heart disease and occurs in 3 in 10,000 live births (1). The actual cause is not known, but it may be associated with other conditions such as 22q11 deletion (DiGeorge) Syndrome (1). Babies with TOF will eventually require surgical correction. Prior to surgical repair, these children are at risk of hypercyanotic spells characterised by a sudden significant reduction in oxygen saturation, hyperpnoea (rapid and deep respiration) as well as decreased intensity / disappearance of the heart murmur. This is a clinical diagnosis and does not rely on additional investigations.

Spells can self-resolve, but non-resolving spells can cause progressive hypoxia and can, in fact, be life threatening. As such, it is a medical emergency, and this guideline has been developed to support safe timely management.

3 Diagnosis and Pathophysiology

Hypercyanotic spells typically begin in the first few months of life; the exact mechanism remains uncertain, however key features that result in the vicious circle that underpins this presentation include:

1. A reduction in pulmonary blood flow secondary to:
    - Increased right ventricular outflow tract obstruction (infundibular spasm)
    - Increased pulmonary vascular resistance
    - A decrease in systemic vascular resistance
2. An increase in the degree of shunting from right to left through the VSD (secondary to point 1 above, as well as other factors such as tachycardia and hypovolaemia).

By stimulating the respiratory centre, the resulting hypoxia and acidosis increase the efficiency of the negative thoracic pump. This ultimately results in an increase in systemic venous return, which further worsens the degree of cyanosis.

Common triggers, in keeping with the above mechanism include agitation, sudden dehydration, defecation, waking from a nap and increased physical activity (2).
Hypercyanotic spells can present with the following features:

- Reduced oxygen saturations
- Hyperpnoea (ensuing metabolic acidosis from poor oxygen delivery to tissues can further deepen respiration)
- Reduced intensity of, or absent murmur (typical murmur in Tetralogy of Fallot is due to turbulent blood flow across the pulmonary outflow tract, but in a hypercyanotic spell the pulmonary outflow blood flow is significantly reduced)
- Inconsolable cry
- Tachycardia
- Squatting in older children

4 Management

Management is outlined in a stepwise approach in Appendix 1 with recommended doses (3)(4). Prevention is always better than treatment and ensuring children are well hydrated, particularly during peri-anaesthetic periods, is essential.

The management of hypercyanotic spells focusses on promoting left to right shunting across the VSD and increasing pulmonary blood flow to allow sufficient oxygenation. The initial management involves bringing the child's knees to the chest to increase systemic vascular resistance as well as to increase systemic venous return (5). Next, giving high flow oxygen reduces pulmonary vascular resistance (2).

Subsequent management if resolution is not achieved, involves administration of IV fluid and morphine. An IV fluid bolus treats any underlying dehydration, maximises preload and should be given ahead of any drugs which may cause hypotension (e.g. beta-blocker). Morphine suppresses the respiratory centre and treats hyperpnea. It also reduces agitation, decreases catecholamine release and slows the heart rate (1).
Intraosseous (IO) access should be secured if difficulty obtaining IV access and this can be utilised for administration of fluids and drugs.

If the child is stable, oral beta blockers such as Propranolol (0.25mg/kg-1mg/kg) may be advised as an additional management step with specialist input.

Further advanced management is required if the above measures fail and require involvement of paediatric intensive care and expert senior support from paediatric cardiology consultant.

Beta blockers reduce the heart rate and may reduce RVOT spasm. They may also increase systemic vascular resistance by counteracting the systemic vasodilatory effects of beta adrenergic stimulation. Intravenous phenylephrine and metaraminol are used in the intensive care setting, and work to further increase systemic vascular resistance (2).

Although uncommon, in situations where hypercyanotic spells persist despite medical therapy, general anaesthesia and ventilation become necessary. Depending on the child's clinical status, options to consider at this stage include emergency transcatheter (RVOT stent/ductal stent) or surgical (BTT shunt) interventions as well as ECMO.

5 References

(1) Bailliard F, Anderson RH. Tetralogy of Fallot. Orphanet Journal of Rare Diseases 2009;4(1):2.

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(3) Joint Formulary Committee. British National Formulary (online). 2024; Available at: https://bnfc.nice.org.uk. Accessed 1st August, 2024.

(4) Shann F. Drug Doses. 17th Edition ed.: Collective Pty, Limited; 2017.

(5) Gossett J. BMJ Best Practice: Tetralogy of Fallot. 2023; Available at: https://bestpractice.bmj.com/topics/en-gb/701. Accessed 19th June, 2024.

Appendix 1 – Treatment Algorithm