NHS
Greater Glasgow and Clyde

CLINICAL GUIDELINE

Hypercyanotic Episodes in Tetralogy of Fallot, Paediatrics

A guideline is intended to assist healthcare professionals in the choice of disease-specific treatments.

Clinical judgement should be exercised on the applicability of any guideline, influenced by individual patient characteristics. Clinicians should be mindful of the potential for harmful polypharmacy and increased susceptibility to adverse drug reactions in patients with multiple morbidities or frailty.

If, after discussion with the patient or carer, there are good reasons for not following a guideline, it is good practice to record these and communicate them to others involved in the care of the patient.

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Important Note:

The Intranet version of this document is the only version that is maintained.
Any printed copies should therefore be viewed as 'Uncontrolled' and as such, may not necessarily contain the latest updates and amendments.

HYPERCYANOTIC EPISODE

Objective
To provide a guideline for the recognition and management of paediatric hypercyanotic episodes.

Scope
This guideline is intended for all healthcare professionals involved in the care of paediatric patients with congenital heart disease susceptible to hypercyanotic episodes.

Background
Hypercyanotic episodes or as they are also referred to 'cyanotic spells' most frequently occur in young children with Tetralogy of Fallot, but may occur in children with other forms of pulmonary stenosis with ventricular septal defect.
Precipitating factors include crying, defecation, feeding, wakening from sleep, dehydration, fever, tachycardia, tachypneoa and events provoking agitation.

They are characterised by:

- Period of uncontrolled crying/panic/irritability
- Rapid and deep breathing
- Increased cyanosis
- Limpness and lethargy
- Reduced murmur intensity
  (signifying substantial blood flow reduction across right ventricular outflow tract)
- The older child squatting

Hypercyanotic spells require early recognition and management to prevent the development of complications from prolonged hypoxia. Whilst medical intervention may be required, many episodes are self-terminating.

Pathophysiology
Hypercyanotic episodes are the effect of an acute imbalance between pulmonary and systemic vascular resistance, resulting in an increased right to left blood flow through the ventricular septal defect and consequent attenuation of right ventricular outflow tract obstruction.

Emergency Management Of Hypercyanotic Episodes

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Initial management:
Calm the child, minimise stimuli
Knee to chest position
oxygen, if not causing distress
Cardiology registrar/ANP review

If no improvement or suboptimal:
Commence patient monitoring, ensure resuscitation equipment available
Morphine 100 micrograms/kg IM (IV if cannula insitu)
Caution in infants < 3 months of age

If no improvement or suboptimal:
Seek Cardiology Consultant advice
Insert IV/IO cannula
5-10mls/kg of 0.9% sodium chloride
(repeat once if required)

If no improvement or suboptimal:
Seek Cardiology Consultant and PICU review
The following should only be given in the presence of PICU team, within a monitored environment (ED Resus/PICU), intubation and ventilation equipment available and ready for use

Cardiology Consultant Preference

IV Phenylephrine

Bolus: 5-20 microgram/kg
slow push (max 500micrograms)
via peripheral or central venous access

Infusion: 0.1-0.5 microgram/kg/min
via central venous access

Preparation:
10mg phenylephrine added to 500mls
0.9% sodium chloride or 5% dextrose
(20 microgram/mL)

IV Esmolol

Bolus: 600 microgram/kg
slow push
via peripheral or central venous access

Infusion: 300-900 microgram/kg/min
via central venous access

Preparation: Neat solution (10mg/mL)

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