University Hospitals of Leicester NHS
NHS Trust

UHL Paediatric Cardiology/EMCHC

Guideline for Cyanotic Spells (Tet spells) in Children within East Midlands Congenital Heart Centre and Leicester Children's Hospital

Staff relevant to:	Medical, nursing and allied health professional working within EMCHC & Leicester Children's Hospital
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Contents

Introduction and who this applies to:

This guidance can be used as an aid and learning tool by medical, nursing and allied health professional staff involved in the management of paediatric patients who present with cyanotic spells within East Midlands Congenital Heart Centre and Leicester Children's Hospital

Management flow chart

Suspected Cyanotic Spell?
(child with tetralogy of Fallot or similar)
Inconsolable cry, increased cyanosis, tachycardia and or tachypnoea/gasping
Low intensity or absent murmur

TREAT AS AN EMERGENCY

Inform on-call PICU and cardiology teams to attend immediately
2222 if patient in extremes or delay in arrival of assistance

Place child in knee chest position with pressure on the femorals

Give high flow oxygen (non-rebreathe mask)

Give Morphine 50 micrograms/kg SC/IV/IM

If spell resolves stop treatment

Repeat morphine once if required

Fluid bolus 10ml/kg 0.9% Sodium chloride IV

If spell resolves stop treatment

Repeat fluid bolus if required

If not on beta blockers regularly, try oral Propranolol 0.5-1mg/kg
If unwell for oral route: IV Esmolol 0.6mg/kg over 1 minute
(< 2months of age) or
IV Metoprolol 0.1mg/kg (≥2months of age), max 5mg per dose
over 5 min repeated every 5 min for total of 3 doses

No response to treatment

Call 2222

Repeat IV Esmolol or IV Metoprolol after 5 mins if required

IV Phenylephrine 5-10 micrograms/kg can be administered without dilution or diluted 10 times for easier administration. Administer over 5 minutes

Repeat phenylephrine dose after 20 mins if required

Arrange PICU admission

Title: Cyanotic Spells in Children
V: 5 Approved by: UHL Children's Services Quality & Safety Board: October 2023 Next Review: October 2026
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2. Background

Definition of Cyanotic spells ("Tet spells")

Cyanotic spells are a paediatric emergency requiring prompt recognition and treatment. Cyanotic spells are paroxysmal hypoxic events in a child due to decreased pulmonary blood flow and right to left shunting. They can occur in any heart condition involving VSD and a restriction to pulmonary blood flow. Spells are often associated with Tetralogy of Fallot as it is the commonest cyanotic congenital heart disease.

Pathogenesis of cyanotic spells

Cyanotic spells are caused by decreased pulmonary blood flow due to increased resistance in the pulmonary circuit leading to shunting of blood from right to left across the VSD.
Any increase in pulmonary vascular resistance (PVR) and/or decrease in systemic resistance (SVR) will cause right to left shunting and resulting cyanosis.
Potential precipitants include crying (increased pulmonary resistance), defecation (reduced systemic resistance), fever (reduced systemic resistance), awakening from naps, feeding, tachycardia (reduced preload) and ACE inhibitors (reduced afterload / systemic resistance).
During a spell the reduced oxygen saturations cause cerebral irritability leading to further crying; this increases pulmonary vascular resistance further exacerbating the problem

Recognising cyanotic spells

A spelling child will present with inconsolable crying, cyanosis, tachycardia, hyperpnoea (increased and deeper respiration due to stimulation of respiratory centre due to low pO2 and pH), gasping, reduced intensity or no murmur (as the murmur due to right ventricular outflow tract obstruction is proportional to the blood flow to the pulmonary circuit).
In later stages spells can lead to anoxic seizures, gasping respiration and apnoea. Spells can last from minutes to hours. They tend to occur more often early mornings but can occur at any time.

3. Management of Cyanotic Spells (See FLOW CHART page 2)

Management of spells is by intervening to increasing systemic vascular resistance, reducing systemic blood flow (increasing afterload) and at the same time reducing pulmonary vascular resistance and increasing pulmonary blood flow.

1. Inform on-call Cardiology and PICU SpR

2. Knee-chest posture with pressure on femoral pulses

3. High flow oxygen with non-rebreathe mask if available

4. Fluid bolus 10ml/kg of 0.9% sodium chloride IV:

 Repeat once if necessary after reviewing fluid status.

5. Morphine 50 micrograms/kg IV/SC/IM, repeat once if necessary:

- **For IV administration:** Morphine 50 micrograms/kg diluted to 5-10ml with 0.9% sodium chloride. Administer as slow IV injection over at least 5 minutes.
- **SC administration:** do not need to dilute (can be diluted if volume of dose is too small to administer)
- **IM administration:** (avoid this route if possible): do not need to dilute (can be diluted if volume of dose is too small to administer)

6. Propranolol PO:

If child is not on a full dose of regular beta blockers then give propranolol.

- **Oral administration:** if possible give orally starting with 0.5mg/kg TDS, increasing up to 1mg/kg TDS as tolerated.
- **IV administration: Not available**

7. IV Esmolol (< 2 months of age) or IV Metoprolol (≥ 2months of age)
Both Kept on Ward1, CPICU, CICU
Esmolol 600 micrograms/kg given over1-2 minutes can be repeated after 3 minutes for maximum of 3 doses then (consider intravenous infusion) 300–900 micrograms/kg/minute if required.
or Intravenous Metoprolol (minimum age 2 months) 100 micrograms /kg (maximum 5mg per dose) slowly over 5 minutes, can be repeated after 5 minutes (maximum of 3 doses) followed by infusion at 1-2mcg/kg/min

8. IV Phenylephrine 5-10 mcg/kg: Kept on Ward 1, CPICU, CICU
(For IV injection use 100 microgram/ml solution without further dilution. (Can be diluted using sodium chloride 0.9% or glucose 5% if required to assist administration)

- Administer as a slow injection over 3-5 minutes. Repeat every 20 minutes if necessary.

9. Transfer to PICU for intubation and ventilation:

- Phenylephrine infusion could be considered at this stage.

4. Education and Training

No new training is required to implement this guideline.

5. Monitoring Compliance

None

6. Supporting References

Daubeney, P., Rigby, M., Gatzoulis,M. and Niwa,K. Pediatric Heart Disease: A clinical guide. London: John Wiley and Sons.
2. Van Roekens, C. and Zuckerberg, A (1995). Emergency management of hypercyanotic crises in tetralogy of fallot. *Annals of Emergency medicine*1995; 25:256-8.
3. Emergency Dept Royal hospital for sick children Yorkhill. Emergency management of hypercyanotic spells guideline. URL: http://www.clinicalguidelines.scot.nhs.uk/
4. Weil ,J., Bertram,H. and Sachweh, J. Guideline: Tetralogy of Fallot. URL:http://www.kinderkardiologie.org/
5. Atit A Gawalkar*, Y S Shrimanth, Akash Batta, Manoj Kumar Rohit Management of Tet spell-Updated Review. Current Research In Emergency Medicine Vol1 Issue 1 2021
6. Rola Saqan1 & Hanan Thiabat1 Evaluation of the safety and efficacy of metoprolol infusion for children and adolescents with hypertensive crises: a retrospective case series Pediatr Nephrol (2017) 32:2107–2113

7. Key Words

Cyanotic spells, Tet spells.

The Trust recognises the diversity of the local community it serves. Our aim therefore is to provide a safe environment free from discrimination and treat all individuals fairly with dignity and appropriately according to their needs.
As part of its development, this policy and its impact on equality have been reviewed and no detriment was identified.

Contact and review details	
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Details of Changes made during review: August 2023- Changes made to B-Blockers introducing Esmolol and Metoprolol instead of iv Propranalol	