Guideline for Cyanotic Spells (Tet spells) in paediatric patients within East Midlands Congenital Heart Centre and Leicester Childrens Hospital

1. Scope

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

Index

<table>
<thead>
<tr>
<th>Section</th>
<th>Page</th>
</tr>
</thead>
<tbody>
<tr>
<td>Scope</td>
<td>1</td>
</tr>
<tr>
<td>Background</td>
<td>2</td>
</tr>
<tr>
<td>Management flowchart</td>
<td>3</td>
</tr>
<tr>
<td>Management</td>
<td>4</td>
</tr>
<tr>
<td>Monitoring and audit criteria</td>
<td>5</td>
</tr>
<tr>
<td>Equality Statement</td>
<td>5</td>
</tr>
<tr>
<td>References and supporting information</td>
<td>5</td>
</tr>
<tr>
<td>Keywords</td>
<td>6</td>
</tr>
<tr>
<td>Review record</td>
<td>6</td>
</tr>
<tr>
<td>Appendix 1 (incl Electronic drugs calculator link)</td>
<td>7</td>
</tr>
</tbody>
</table>
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), 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 apnoeas. Spells can last from minutes to hours. They tend to occur more often early mornings but can occur at any time.
Suspected Cyanotic Spell?
(child with tetralogy of Fallot)
Insoluble cyanosis, increased cyanosis, tachycardia and or tachypnoea
Low intensity or absent murmur

TREAT AS AN EMERGENCY
Inform on-call PICU and cardiology teams immediately

Place child in the knee-chest position with pressure on the femoralis

Give high flow oxygen (non-rebreath mask)

Give Morphin 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 Propanolol 0.5mg/kg
(If oral route not possible IV Propanolol 0.1mg/kg)

No response to treatment
Repeat IV Propanolol after 15mins if required

IV Phenylephrine 5-10micrograms/kg diluted 10 times. Administer over 5 minutes.

Repeat phenylephrine dose after 20mins if required

Arrange PICU admission

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3. Management of Cyanotic Spells

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/IV:
   - 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:** If oral route is not possible, use IV. Give 100 microgram/kg (0.1mg/kg) as a slow bolus over at least 3-5 minutes. Rate should not exceed 1mg/minute. May be diluted with sodium chloride 0.9% or glucose 5% if required to aid administration. ECG monitoring is required. Repeat after 15 minutes if required.
7. IV Phenylepherine 5-10 mcg/kg:
   - 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.
8. Transfer to PICU for intubation and ventilation:

- Phenylepherine infusion could be considered at this stage.

4. Education and Training

No new training is required to implement this guideline.

5. Monitoring and Audit Criteria

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<th>Key Performance Indicator</th>
<th>Method of Assessment</th>
<th>Frequency</th>
<th>Lead</th>
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6. Equality Statement

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/guideline and its impact on equality have been reviewed and no detriment was identified.

7. Supporting Documents and Key References


8. Key Words
Cyanotic spells, Tet spells.

<table>
<thead>
<tr>
<th>DEVELOPMENT AND APPROVAL RECORD FOR THIS DOCUMENT</th>
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<tbody>
<tr>
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<td>Executive Lead: Simon Robinson</td>
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<tr>
<td>June 2016</td>
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<td>Feb 2017</td>
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Appendix 1

**TOF cyanotic Spells - emergency drugs**

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<tr>
<th>Name</th>
<th>Weight (kg):</th>
<th>10</th>
<th>Doctor's signature</th>
<th>Doctor's name (PRINT)</th>
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Fluid bolus 0.9% sodium chloride IV (10ml/kg) : 100 ml

Morphine (50 micrograms/kg): 500 micrograms IM/SC/IV

If child not on full-dose regular beta blockers then

<table>
<thead>
<tr>
<th>PO Propranolol (0.5mg/kg) TDS</th>
<th>5mg</th>
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<tr>
<td>(If oral route not possible) IV Propranolol (0.1mg/kg)</td>
<td>1mg over 3-5 mins</td>
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IV phenylephrine (10micrograms/kg) 100 micrograms

Dilute to ten times volume to give total volume 1 ml 100 micrograms/ml solution 10 ml slow push over 5 minutes

Please follow this link to access the electronic drugs calculator: