Medical management of Hypercyanotic spells in neonates, infants with Tetralogy of Fallot

<table>
<thead>
<tr>
<th>Version:</th>
<th>3</th>
</tr>
</thead>
<tbody>
<tr>
<td>Approval Committee (e.g. Clinical network):</td>
<td>Children’s Services Review group</td>
</tr>
<tr>
<td>Date of Approval:</td>
<td>13 June 2018</td>
</tr>
<tr>
<td>Signature of approving Group Chair</td>
<td></td>
</tr>
<tr>
<td>Ratification Group:</td>
<td>Children’s Services Review Group, University Hospital Southampton</td>
</tr>
<tr>
<td>Date of Ratification:</td>
<td>13 June 2018</td>
</tr>
<tr>
<td>Signature of ratifying Group Chair</td>
<td>John Pappachan Chair of Children’s Services Review Group</td>
</tr>
<tr>
<td>Author’s and job titles</td>
<td>Katy Stimson Nurse Practitioner/Dr A. Magee Consultant Paediatric cardiologist</td>
</tr>
<tr>
<td>Date issued:</td>
<td>13 June 2018</td>
</tr>
<tr>
<td>Review date:</td>
<td>13 June 2021</td>
</tr>
<tr>
<td>Key words:</td>
<td>Medical management of Hypercyanotic spells in neonates, infants with Tetralogy of Fallot</td>
</tr>
<tr>
<td>Main areas affected:</td>
<td>Children’s services</td>
</tr>
<tr>
<td>Other stakeholders consulted e.g. other clinical networks, departments</td>
<td>Wessex PIER Regional Guideline Governance Group</td>
</tr>
<tr>
<td>Summary of most recent changes (if updated guideline):</td>
<td>19/10/17</td>
</tr>
<tr>
<td>Relevant national or international Guidance e.g. NICE, SIGN, BTS, BSPED</td>
<td>Resuscitation Council UK</td>
</tr>
<tr>
<td>Consultation document completed: see Appendix A</td>
<td>Paediatric Cardiologists</td>
</tr>
<tr>
<td>Total number of pages:</td>
<td></td>
</tr>
<tr>
<td>Is this document to be published in any other format?</td>
<td>On line</td>
</tr>
</tbody>
</table>

Does this document replace or revise an existing document? Yes....

Medical management of Hypercyanotic spells in neonates, infants and young children. UHS guide
Medical management of hyper-cyanotic spells in Tetralogy of Fallots

**STEP 1**
Always assess using ABCDE approach
Spells can be mild, treat according to response as follows

**STEP 2 - Position Knee to chest**
Call for help
Place baby in the knee to chest position either supine or over parent’s shoulder
(This calms the infant, increases systemic venous return and increases systemic vascular resistance)

**STEP 3 - Oxygen**
Administer 100% oxygen via non-re-breathing mask.
Monitor oxygen saturations and ECG

**STEP 4 - If no response to above**
If stable, give oral morphine 0.1mg/kg
Site IV cannula – check blood gas
Give IV 0.9% sodium chloride 20mls/kg in aliquots of 10mls/kg
Give IV morphine 0.1mg/Kg as a bolus (can be repeated)
Use IM or subcutaneous route if IV access not readily available.
Monitor neurological and respiratory status as morphine has respiratory depressant effects

**STEP 5 - If no response to above**
Give IV Propranolol 0.1mg/kg as a bolus
Monitor for bradycardia

**DETERIORATION**
Contact SORT/cardiology team
Phenylephrine, 0.02 mg/Kg IV (used to increase SVR)
IV Esmolol infusion may be another alternative before ventilating the baby as a last resort

**Symptoms & Triggers**
- may appear pale, grey or blue
- maybe ‘clammy’
- tachypnoea
- deepening of cyanosis
- decreased intensity of heart murmur
- loss of consciousness
- distress
- crying
- defecation or straining
- dehydration
- fever
- tachypnoea
- anaesthetic agents
- cardiac catheterisation

**Useful Contacts**
Contact SORT (Southampton Oxford retrieval team) for 24 hour advice on 02380 775502
Contact 02380 777222
Bleep 2811 On call Paediatric cardiology registrar for advice
1

1.1 Introduction
Hypercyanotic spells can occur in patients with uncorrected Tetralogy of Fallot and other abnormalities where there is dynamic right ventricular outflow tract obstruction and a ventricular septal defect. Such spells are often mild but are potentially very dangerous and patients can deteriorate rapidly. The cascade of therapeutic action is dependent upon the response to treatment.

Tetralogy of fallot (TOF) is a congenital heart defect. TOF is characterized by four morphological changes; ventricular septal defect, stenosis of the right ventricular outflow tract (RVOT), stenosis of the pulmonary valve, an ante- and dextro-positioned overriding aortic root and a secondary right ventricular hypertrophy (Knuf et al 2010).

1.2 Scope
This guideline applies to all infants with uncorrected Tetralogy of Fallot and other abnormalities where there is a dynamic right ventricular outflow tract obstruction infants in the Oxford Southampton network.

1.3 Purpose
The purpose of this guideline is to provide a standardised approach to the management of hypercyanotic spelling in neonates/infants with uncorrected Tetralogy of Fallot and other abnormalities where there is dynamic right ventricular outflow tract obstruction.

1.4 Definitions
The cascade of therapeutic action is dependent upon the response to treatment.

MILD: the baby may appear pale, grey blue and may be clammy, with tachypnoea. Oxygen saturations will be lower than usual.

SEVERE: On examination the right ventricular outflow tract murmur may be absent or short in duration. Loss of consciousness or ‘dropping off to sleep’ may be seen due to poor systemic perfusion.

2 Related Policies
Resuscitation Council UK  https://www.resus.org.uk/resuscitation-guidelines/

Presentation:
The baby may appear pale, grey blue and may be ‘clammy’
Tachypnoea (rapid shallow breathing)
Peripheral oxygen saturations will be significantly lower than the baby’s usual measurement.
On examination the RVOT murmur may be absent or short in duration.
Loss of consciousness or ‘dropping off to sleep’ can be seen if there is poor systemic perfusion.

Potential triggers or predisposing factors:
Can be triggered by a variety of stimuli (distress, crying, dehydration, defecation or straining, fever, tachypnoea, anaesthetic agents, cardiac catheterisation) but may also occur without any reason.
### Mild hypercyanotic spell

<table>
<thead>
<tr>
<th>Always assess infant using ABCDE principles</th>
</tr>
</thead>
</table>

Call for help. In mild cases position on parent's/carer's shoulder with the knees tucked up underneath.  
(This calms the infant, increases systemic venous return and increases systemic vascular resistance)

Reassess using ABCDE principles. Call for help. Administer 100% oxygen via non re-breath bag valve mask if necessary. Monitor oxygen saturations and ECG

---

### If no Response to above

#### Call for help

- If stable, give oral morphine 0.1mg/Kg
- Site IV cannula – check venous blood gas
- Give IV 0.9% sodium chloride 20mls/Kg in aliquots of 10mls/Kg
- Give IV morphine 0.1mg/Kg as a bolus (can be repeated)
- Use IM or subcutaneous route if IV access not readily available

**Monitor neurological and respiratory status as morphine has respiratory depressant effects**

#### If no Response to above

- Give IV Propranolol 0.1mg/Kg as a bolus
- Monitor for bradycardia

### Deterioration

<table>
<thead>
<tr>
<th>Contact SORT Southampton Oxford retrieval team 02380775502</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cardiology team Contact 02380 777222 Bleep 2811 Paediatric cardiology registrar for advice</td>
</tr>
</tbody>
</table>

Give IV Phenylephrine 0.02mg/Kg IV (This is used to increase SVR)

IV Esmolol infusion maybe another alternative before ventilating as a last resort.

---

2  **Additional headings as required e.g. procedures to be followed or chart titles**

None.

3  **Implementation**

This guideline will be made available regionally on the PIER Website. Local leads for critically ill children will disseminate guideline and raise awareness locally.

4  **Process for Monitoring Effectiveness**

The Wessex Paediatric Critical Care Network will review problems associated with a failure to comply with this guideline through its regional governance process.

5  **References**


Resuscitation Council UK https://www.resus.org.uk/resuscitation-guidelines/

Appendix A

Paediatric Regional Guideline Consultation Documentation:

<table>
<thead>
<tr>
<th>Trust</th>
<th>Name of person consulted* (print)</th>
<th>Designation of signatory</th>
<th>Signature</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chichester</td>
<td>Nick Brenan</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Dorchester</td>
<td>Dr D. Shenoy</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hampshire Hospitals</td>
<td>Dr L. Winckworth</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Foundation Trust</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Poole</td>
<td>Dr S Bokhandi</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Portsmouth</td>
<td>Dr. R Sievers</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Salisbury</td>
<td>Dr J Baird</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Southampton</td>
<td>Dr Alan Magee</td>
<td></td>
<td></td>
</tr>
<tr>
<td>IOW</td>
<td>Dr E. Blake</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

*this person agrees they have read the guidelines, consulted with relevant colleagues and members of MDT, managers and patients, young people & their families as appropriate. Any queries raised during consultation and review process should be documented with responses and any changes made to guideline.