

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

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Medical management of Hypercyanotic spells in neonates, infants and young children. UHS guide

Contents

Paragraph		Page
	Flowchart	3
1	Introduction	4
1.2	Scope	4
1.3	Aim/Purpose – outline objectives and intended outcomes	4
1.4	Definitions	4
2	Additional headings	5
3	Implementation (including training and dissemination)	5
4	Process for Monitoring Compliance/Effectiveness of this policy	6
5	References	6
Appendices		
Appendix A	Consultation signatures	6
Appendix B	[relevant documents, eg audit forms/patient information leaflets}	6

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 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

Always assess infant using ABCDE principles

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

Contact SORT Southampton Oxford retrieval team 02380775502

Cardiology team Contact 02380 777222 Bleep 2811 Paediatric cardiology registrar for advice

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

BNFC (2016-2017) BNF for Children BMJ group.

Knuf, M et al (2010) 'Significance of patient categorization for perioperative management of children with Tetralogy of Fallot, with special regard to co-existing malformations'. *Cardiology Journal*. 1:20-28

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

Appendix A

Paediatric Regional Guideline Consultation Documentation:

Trust	Name of person consulted* (print)	Designation of signatory	Signature
Chichester	Nick Brennan		
Dorchester	Dr D. Shenoy		
Hampshire Hospitals Foundation Trust	Dr L. Winckworth		
Poole	Dr S Bokhandi		
Portsmouth	Dr. R Sievers		
Salisbury	Dr J Baird		
Southampton	Dr Alan Magee		
IOW	Dr E. Blake		

*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.