Ashford and St. Peter's Hospitals
NHS Foundation Trust

Management of Hypercyanotic spells

Author: Dr Adekunle Matthew Sobowale (Specialty Doctor)

Supervisors: Dr Lynda Ehizode (Consultant Paediatrician)

Dr Allison Groves (Consultant Paediatrician with Expertise in Cardiology)

Contact details: adekunle.sobowale@nhs.net

Guideline History		
Date	**Comments**	**Approved By**
15/03/2021	New guideline by Dr Matthew Sobowale	Paediatric Guideline Group

Patients first • Personal responsibility • Passion for excellence • Pride in our team

Contents **Page**

1. Guideline

1.1 Introduction……3

1.2 Scope……3

1.3 Purpose……3

1.4 Definitions……4

1.5 Presentation……4

1.6 Potential triggers or predisposing factors……4

1.7 Management……5

1.8 Algorithm……6

1.9 References……7

2. Guideline Governance

a. Scope……8

b. Purpose……8

c. Duties and Responsibilities……8

d. Approval and Ratification……8

e. Dissemination and Implementation……8

f. Review and Revision Arrangements……8

g. Equality Impact Assessment……9

h. Document Checklist and Approval……10-12

1. Guideline of Management of Hypercyanotic spells

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 structural changes; ventricular septal defect, stenosis of the right ventricular outflow tract (RVOT), stenosis of the pulmonary valve, an aortic root that overrides both ventricular outflow tracts and a secondary right ventricular hypertrophy.

1.2 Scope

This guideline applies to all infants with uncorrected Tetralogy of Fallot and other abnormalities where there is a dynamic obstruction to flow of blood from the right ventricle to the lungs.

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 obstruction of flow of blood from the right ventricle to the lungs.

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.

1.5 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.

1.6 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.

1.7 Management

Mild Hypercyanotic spell

Always assess infant using the 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. 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, inform consultant.
If stable, give oral Morphine 0.1 mg/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 STRS retrieval Team 020 7188 5000
Contact the Paediatric cardiology registrar at the Royal Brompton Hospital 020 7352 8121(switchboard)

IV phenylephrine 0.02mg/kg (This is used to increase SVR)
IV Esmolol infusion maybe another alternative before ventilating as a last resort

1.8 Algorithm

1.9 References

1: Christian Apitz, Gary D Webb, Andrew N Redington. Seminar Tetralogy of Fallot. The Lancet. Volume 374, Issue 9699, 24–30 October 2009, Pages 1462-1471

2: BMJ best practice. Tetralogy of Fallot. http://bestpractice.bmj.com/topics/en-gb/701/pdf/701.pdf. Accessed 16/01/2021

3: K. Stimson, A. Magee. Medical management of Hypercyanotic Spells in neonates, infants with tetralogy of Fallot. Children's Services Review Group, University Hospital Southampton. Guideline. 2018.

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

2. Guideline Governance

a. Scope

This guideline is relevant to all staff caring for all children from 0-18 years old across the emergency department, inpatient ward and outpatient department.

b. Purpose

i. This guidelines aims to facilitate a common approach to the management of children. At times deviation from the guideline may be necessary, this should be documented and is the responsibility of the attending consultant.

ii. This guideline is subject to regular review to ensure ongoing evidence based practice.

c. Duties and Responsibilities

All healthcare professionals responsible for the care of of all children 0-18years should be aware of practice according to this guideline.

d. Approval and Ratification

This guideline will be approved and ratified by the Paediatric Guidelines Group.

e. Dissemination and Implementation

i. This guideline will be uploaded to the trust intranet 'Paediatric Guidelines' page and thus available for common use.
ii. This guideline will be shared as part of ongoing education within the Paediatric Department for both medical and nursing staff.
iii. All members of staff are invited to attend and give comments on the guideline as part of the ratification process.

f. Review and Revision Arrangements

a. This policy will be reviewed on a 5 yearly basis by the appropriate persons.
b. If new information comes to light prior to the review date, an earlier review will be prompted.
c. Amendments to the document shall be clearly marked on the document control sheet and the updated version uploaded to the intranet. Minor amendments will be ratified through the Paediatric Guidelines Group. A minor amendment would consist of no major change in process, and includes but is not limited to, amendments to documents within the appendices.

g. Equality Impact Assessment

Background • Who was involved in the Equality Impact Assessment
Author and the supervising consultants.
Methodology • A brief account of how the likely effects of the policy was assessed (to include race and ethnic origin, disability, gender, culture, religion or belief, sexual orientation, age) • The data sources and any other information used • The consultation that was carried out (who, why and how?)
All groups of staff and patients were taken into consideration and there is no bias towards or against any particular group.
Key Findings • Describe the results of the assessment • Identify if there is adverse or a potentially adverse impacts for any equalities groups
There is no evidence of discrimination.
Conclusion • Provide a summary of the overall conclusions
There is no evidence of discrimination.
Recommendations • State recommended changes to the proposed policy as a result of the impact assessment • Where it has not been possible to amend the policy, provide the detail of any actions that have been identified • Describe the plans for reviewing the assessment
This guideline is appropriate for use.

h. Document Checklist

To be completed (electronically) and attached to any document which guides practice when submitted to the appropriate committee for approval or ratification.

Title of the document: Management of Hypercyanotic spells

Policy (document) Author: Dr Matthew Sobowale

Executive Director: N/A

		Yes/No/ Unsure/NA	Comments
1.	**Title**		
	Is the title clear and unambiguous?	**Yes**	
	Is it clear whether the document is a guideline, policy, protocol or standard?	**Yes**	
2.	**Scope/Purpose**		
	Is the target population clear and unambiguous?	**Yes**	
	Is the purpose of the document clear?	**Yes**	
	Are the intended outcomes described?	**Yes**	
	Are the statements clear and unambiguous?	**Yes**	
3.	**Development Process**		
	Is there evidence of engagement with stakeholders and users?	**Yes**	
	Who was engaged in a review of the document (list committees/ individuals)?		**Royal Brompton Cardiology Dept**
	Has the policy template been followed (i.e. is the format correct)?	**Yes**	
4.	**Evidence Base**		
	Is the type of evidence to support the document identified explicitly?	**Yes**	

		Yes/No/ Unsure/NA	Comments
	Are local/organisational supporting documents referenced?	**Yes**	
5.	**Approval**		
	Does the document identify which committee/group will approve/ratify it?	**Yes**	
	If appropriate, have the joint human resources/staff side committee (or equivalent) approved the document?		
6.	**Dissemination and Implementation**		
	Is there an outline/plan to identify how this will be done?	**Yes**	
	Does the plan include the necessary training/support to ensure compliance?	**Yes**	
7.	**Process for Monitoring Compliance**		
	Are there measurable standards or KPIs to support monitoring compliance of the document?	**No**	
8.	**Review Date**		
	Is the review date identified and is this acceptable?	**Yes**	
9.	**Overall Responsibility for the Document**		
	Is it clear who will be responsible for coordinating the dissemination, implementation and review of the documentation?	**Yes**	
10.	**Equality Impact Assessment (EIA)**		
	Has a suitable EIA been completed?	Yes	

Committee Approval (Paediatric Guidelines Group)			
If the committee is happy to approve this document, please complete the section below, date it and return it to the Policy (document) Owner			
Name of Chair	**Dr Claire Mitchell**	**Date**	**06/05/2021**
Ratification by Management Executive (if appropriate)			
If the Management Executive is happy to ratify this document, please complete the date of ratification below and advise the Policy (document) Owner			
Date: n/a			