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| Clinical Guideline**hypercyanotic spell emergency management (‘spelling’)** |
| **SETTING** | Paediatric wards and High Dependency Units, South West & South Wales Congenital Cardiology Network |
| **FOR STAFF** | Nursing, medical and pharmacy staff on paediatric wards and High Dependency Units  |
| **PATIENTS** | Infants/young children |
| **Disclaimer:** **This has been produced as a guide; each individual patient should be considered whether treatment is clinically appropriate. If needed, please discuss further with specialist teams.**\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_ |
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| **BACKGROUND** Hypercyanotic episodes or “spelling” typically occurs in patients with Tetralogy of Fallot or any anatomy with dynamic right ventricular outflow tract obstruction (RVOTO) and a ventricular septal defect (VSD). A hypercyanotic spell may be triggered as the result of some form of stimuli such as exercise or feeding or may occur in situations leading to vasodilatation. It can also happen without any obvious precipitant. **IT IS AN EMERGENCY SITUATION**  |

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| **PRESENTATION** The patient may look pale, blue or grey, and may be clammy. They may have a cry which is said to be “different” from their normal cry. If systemic output is significantly compromised they may lose consciousness/appear to go to ‘sleep’. Oxygen saturations if being monitored will fall and the outflow tract murmur shorten or disappear. **PREDISPOSING FACTORS** If a child is dehydrated or has had anaesthetic or sedative drugs they are at increased risk of having an episode. Patients who have had a cardiac catheter are also at increased risk. Any patient with the above cardiac anatomy at risk of intravascular depletion should be treated early with intravenous fluids to attempt to prevent the onset of a spell. **MANAGEMENT**1. Call for help
2. Keep child calm with parent
3. Assess airway patency, respiratory effort and circulatory status
4. Knee to chest position (squatting)
5. Give 100% oxygen via non-rebreather face mask
6. Continuous ECG, regular blood pressure (BP), oxygen saturation monitoring
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 Correct potential hypovolaemia: Give 10ml/kg Sodium Chloride 0.9% **OR**10ml/kg Plasmalyte148 (max 2 boluses)No improvement in 5 minutesPropranolol 15-20 micrograms/kg (max 100 micrograms/kg/dose)slow IV with ECG & continuous BP **OR**Esmolol bolus 0.5mg/kg IV over 1 min then start 50micrograms/kg/minute infusion.**REVIEW** 1. Observe and keep calm
2. Start / review propranolol dose (0.5 – 1.5mg/kg every 6-8 hourly) PO)

Morphine 50-100 micrograms/kg slow IV**↑∕****Immediately contact Paediatric Middle Grade & Consultant**Does the patient have IV access?Morphine 100 microgram/kg **subcutaneous/intramuscular** x 1 dose**OR****Buccal** midazolam 300 micrograms/kg (max 2.5mg) x 1 dose onlySet up for Intravenous orIntraosseous accessNo resolution in 5 minutes - **Paediatric 2222 CALL****Hypercyanotic episode****Terminated ?**Yes.**Refer to Review section above.**No |
| Consider increasing Esmolol infusion by 50microgram/kg/min every 5 minutes until resolution (max 200microgram/kg/min)**OR**Phenylephrine 5 microgram/kg IV**OR**Metaraminol 10 microgram/kg IV.Consider correcting metabolic acidosis with 8.4.% sodium bicarbonate.Yes**Hypercyanotic Episode Persists****Contact WATCh** (0300 0300 789), who will also include paediatric cardiology consultant on call. Consider Rapid Sequence Induction after discussion with WATCh & paediatric cardiology. WATCh to urgently transfer to cardiac surgical centre (Bristol). **References**British National Formulary for Children (BNFC) 2017-2018The Paediatric Cardiology Handbook 3rd Edition 2003Pediatric Cardiac Intensive Care Williams and Wilkins 1998Pediatric Acute Care 2nd Edition Williams and Wilkins 2001Royal Brompton and Harefield NHS Foundation Trust. Hypercyanotic Spell Algorithm\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_ |
| **RELATED**No**DOCUMENTS** | None |
| **AUTHORISING BODY** | Paediatric cardiac governance committee Bristol Royal Hospital for Children |
| **SAFETY** | Any safety concerns should be directed to the on call paediatric cardiology consultant and/or the paediatric cardiology clinical governance group |
| **QUERIES** | Contact Dr Michael Yeong (Consultant Paediatric Cardiologist) via Bristol Royal Hospital for Children switchboard if there are queries about this guideline. |