IAEM Clinical Guideline

Tetralogy of Fallot: Emergency Management of Hypercyanotic "Tet" Spells

Version 1.0

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DISCLAIMER

IAEM recognises that patients, their situations, Emergency Departments and staff all vary. These guidelines cannot cover all clinical scenarios. The ultimate responsibility for the interpretation and application of these guidelines, the use of current information and a patient's overall care and wellbeing resides with the treating clinician.

Revision History

Date	Version	Section	Summary of Changes	Author
June 2024	V1.0	All	Final version	G-PM/LJ/ KS/OF

CONTENTS

GLOSSARY OF TERMS

Tetralogy of Fallot (TOF)	A congenital heart defect diagnosed by echocardiography consisting of a malaligned ventricular septal defect (VSD), an overriding aorta, right ventricular outflow tract (RVOT) obstruction, and concentric right ventricular hypertrophy. Patients with TOF have an ejection systolic murmur over the left sternal edge which can vary with intensity depending on the degree of RVOT obstruction.[1]
Hypercyanotic Spells/ Tet Spells	Periodic worsening of cyanosis in a patient with TOF, due to shunting of deoxygenated blood across the VSD from worsening dynamic RVOT obstruction and decrease in pulmonary circulation.[1]
Neonate	Infant younger than 28 days

GLOSSARY OF ABBREVIATIONS

APLS	Advanced Paediatric Life Support
CHI	Children's Health Ireland
IM	Intramuscular
IO	Intraosseous
IV	Intravenous
O_2	Oxygen
PDA	Patent Ductus Arteriosus
PICU	Paediatric Intensive Care Unit
RVOT	Right Ventricular Outflow Tract

Tetralogy of Fallot: Emergency Management of Hypercyanotic “Tet” Spells

INTRODUCTION

Hypercyanotic spells also known as “Tet spells” can be recognised by worsening cyanosis and tachypnoea in patients with TOF.[1,2] In severe cases they can present with loss of consciousness.[3] Infants may also appear pale or grey in addition to cyanosis, and they are frequently crying and distressed. This is a medical emergency.[1]

The characteristic heart murmur may disappear or diminish in intensity as a result of reduced pulmonary blood flow due to dynamic RVOT obstruction.[1] Common triggers include fever, crying, feeding, stooling, stress, straining, and dehydration.[2,4]

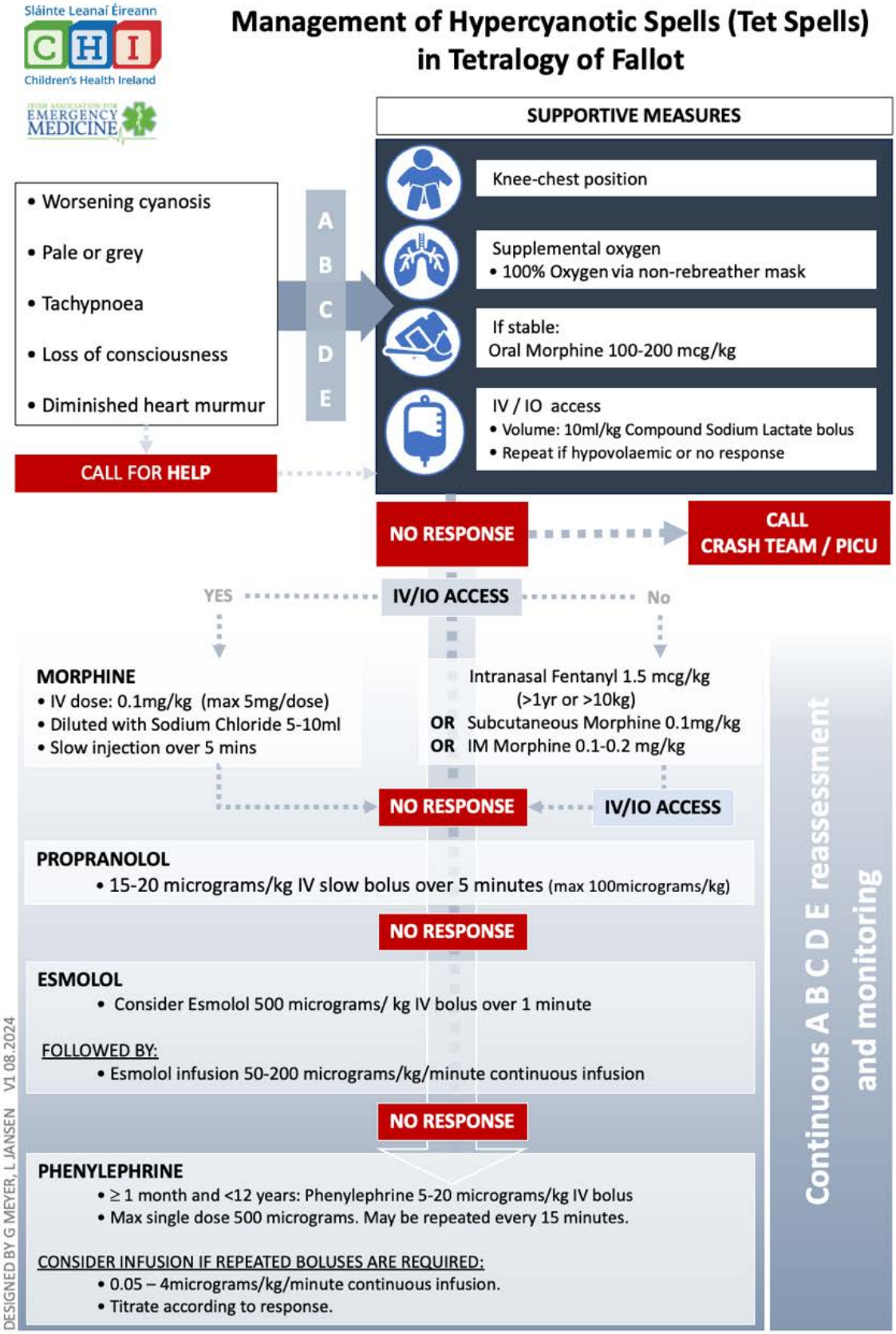
PARAMETERS

Target users	This guide is directed at healthcare professionals engaged in the acute care of children presenting with hypercyanotic spells and should be utilised in collaboration with the paediatric cardiology team.
Target Patient population	This evidence summary applies to paediatric patients with suspected or known Tetralogy of Fallot. It does not cover APLS principles in depth, which should be applied in all cases.
Exclusion criteria	Clinical judgement should prevail, as this guideline does not cover in-depth management of other causes of cyanosis, which should be considered in all cases.

AIMS

To provide an evidence-based guide for the emergency medical management of a hypercyanotic spells in infants with known or suspected Tetralogy of Fallot.

Figure 1. Management of Hypercyanotic Spells (Tet Spells)



ASSESSMENT

The following describes the assessment plan for patients with known or suspected TOF presenting with cyanosis.

Initial acute assessment

- Recognise hypercyanotic spells: may present with cyanosis, and deeper and faster breathing.[4]
- Apply APLS principles and perform a primary survey and assess the airway, breathing and circulation.
- Take note that patients with known TOF and hypercyanotic spells may be on oral Propranolol and may be due a dose or have already had a dose administered prior to presenting.[1]
- When left untreated and in severe cases there may be haemodynamic compromise and shock from tissue hypoxia and metabolic acidosis. Call for help early.[1]

Ongoing assessment

- **It is essential to perform continuous APLS assessment during treatment and act accordingly.**

INVESTIGATIONS

- Investigations should not delay medical treatment.
- In the event of acute deterioration, check blood glucose and a capillary gas.
- Baseline bloods including full blood count, urea and electrolytes and group and cross-match are appropriate investigations if a cannula is sited and it does not delay medical treatment.

MANAGEMENT

First-line management

- Call for help early, as supportive measures and timely medical management can prevent further deterioration.
- Involve the Paediatric and CHI Cardiology team early.
- Initial management focuses on calming the child, minimal handling, and on manoeuvres that increase pulmonary blood flow.[1,3]
- If possible, manage the patient in the caregiver's arms.[1]
- Place the baby in a knee-chest position, either supine or lying over the parent's shoulders.[2] This increases systemic vascular resistance and reduces the shunt across the VSD, thereby increasing pulmonary blood flow.[1,2]
- Administer supplemental O_2 but take care not to worsen agitation.[1,2,3]
- If stable, administer **oral Morphine: 100-200 micrograms/kg.**[5]
- IV access and volume administration: **10ml/kg Compound Sodium Lactate solution (Hartmann's Solution) bolus** to increase preload to the heart. This can be repeated if no response or in cases of hypovolaemia.[2]

If not yet done, call the crash team and for PICU involvement as intubation and ventilation may be required with the following treatments. The following drugs should be administered by the attending doctor.

Second-line management

Opioids

Opioids decreases heart rate and hyperpnoea and therefore decreases the right to left shunting of blood flow in the heart.[1-3]

- **IV Morphine** if IV access is readily available: **0.1mg/kg** (not to exceed a maximum single dose of 5mg) diluted using Sodium Chloride 0.9% to an appropriate volume for administration (5-10ml) should be given as a slow intravenous injection over at least 5 minutes.[5,6]
- In patients >1 years of age and >10kg, intranasal opioids can be considered if patient is unstable and IV access is unavailable.[3,5,6,7]
    - **Intranasal Fentanyl: 1.5 micrograms/kg**, (not to exceed a maximum single dose of 100 micrograms).[5,6]
- Consider subcutaneous opioids if IV access is unavailable
    - **Subcutaneous Morphine: 0.1mg/kg** subcutaneous injection (not to exceed a maximum single dose of 5mg).
- **IM Morphine** is an alternative if no IV access is obtained: **0.1 – 0.2 mg/kg** IM (not to exceed a maximum single dose of 5mg).[2]

Continuous monitoring of heart rate, oxygen saturations, respiratory rate, temperature and blood pressure is advised.

Third-line management

The following medications should be administered following discussion with the tertiary cardiology team, or intensive care team or consultant and administered in a monitored environment; Resuscitation Room, HDU, or PICU.

Beta blocker

Beta blockers decrease RVOT obstruction by reducing infundibular muscle spasm, and decrease heart rate thereby prolonging diastolic filling and systole for increased flow across outflow tract. These actions improve pulmonary blood flow.[1]

- Administer **Propranolol 15 – 20 micrograms/kg** IV slow bolus over 5 minutes, increasing to a maximum of 100 microgram/kg IV.[5]
- If no response after Propranolol:
    - **Esmolol** should be considered while awaiting transfer to PICU: **500 micrograms/kg** IV bolus over 1 minute initially, followed by Esmolol continuous infusion 50 – 200 micrograms/kg/minute infusion.[2,5]

Continuous monitoring of heart rate, oxygen saturations, respiratory rate, monitor temperature, blood pressure and blood glucose is advised.

Phenylephrine

If the above measures fail, Phenylephrine should be considered. The aim is to increase the systemic vascular resistance to greater than pulmonary vascular resistance, and to redirect blood through the RVOT instead of across the VSD to the systemic circulation.[1]

- ≥ 1 month to <12 years old: **Phenylephrine 5 - 20 micrograms/kg** by slow IV bolus (not to exceed a maximum single dose of 500 micrograms). This may be repeated as necessary after at least 15 minutes.
- Consider continuous infusion if repeated boluses are required:

    0.05 – 4 micrograms/kg/minute continuous intravenous infusion.[1,5,8]
- Monitor for hypertension, and titrate dose according to response.[5]

RESCUE THERAPIES

Patients with severe pulmonary stenosis and non-remitting cyanosis will require surgical intervention with a Blalock-Taussig shunt to increase pulmonary blood flow as a palliation procedure until definitive surgery can be performed.[1]

SPECIAL CONSIDERATIONS

- In a neonate (<28 days of life) with cyanosis and an undiagnosed suspected congenital heart lesion, treatment with Dinoprostone (Prostin®) could be lifesaving as it could maintain patency of the PDA and improve pulmonary blood flow.[1]
    - Initially 5 nanograms/kg/minute by continuous IV infusion.[5]
    - Increase by 5 nanograms/kg/minute as required.[5]
    - Doses higher than 20 nanograms/kg/minute have increased side effects (apnoea) and will require consideration for intubation.[5] A resuscitation trolley with intubation equipment should be readily available for emergency airway management upon commencement of Prostin.
    - Take care not to interrupt an infusion and have secure intravenous access in place. A second back-up IV line is recommended.
    - Requires monitoring: advise continuous monitoring of heart rate, oxygen saturations and respiratory rate; monitor temperature, and blood pressure.
- The use of intranasal Fentanyl in patients under 1 years of age is currently not recommended by CHI, except in PICU in special circumstances.[5]
- Intravenous Ketamine is the induction agent of choice for intubation, as it increases the peripheral vascular resistance thereby improving flow across RVOT.[10]
- Avoid Pancuronium due to its chronotropic effect.[11]

LINKS TO USEFUL WEBSITES

Clinibee app for CHI formulary download link:

https://app.clinibee.com/signin?returnUrl=%2F

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