British Paediatric Neurology Association/Paediatric Critical Care Society

Guidelines for the Recognition and Management of Childhood Dystonia in the Critical Care Environment

Scope

1 Guidance title

Guidelines for the Recognition and Management of Childhood Dystonia in the Critical Care Environment

1.1 Short title

Childhood Critical Care Dystonia

2 The remit

At present, there is a lack of robust guidance on the recognition and management of childhood dystonia in the critical care environment. The BPNA and PCCS propose a Clinical Guideline Development Group to address this gap, aiming to develop evidence-informed guidelines developed through a transparent, representative consensus process. This guideline will be developed in line with RCPCH Guideline development principles/standards.

A systematic scoping review of the existing evidence will be undertaken to support the formulation of guidelines. Drawing on the Co-Chairs' expertise and previous scoping reviews on this subject[1,2]. It is anticipated that the available evidence will be sparse. Therefore, it will be essential to develop guidance through a comprehensive Delphi process. This guideline will explore the following questions:

1. How can childhood dystonia best be recognised in the Paediatric Critical Care environment?
2. How is childhood dystonia optimally managed in the Paediatric Critical Care environment (including, but not limited to SD)

3 Clinical need for the guideline

Dystonia is a movement disorder characterised by involuntary muscle contractions that force the affected body part into an abnormal posture. Dystonic movements can be painful and disrupt care and function. Dystonia is a common finding in children and young people (CAYP) with neurological disorders, often requiring pharmacological intervention.

Dystonic movements exhibit a wide range of severity, with the most extreme cases causing possible life-threatening situations. This is a condition called Status Dystonicus (SD)[3]. Although SD was once thought to be a rare occurrence, it is now recognised that severe dystonia is relatively common and frequently necessitates admission to a Paediatric Critical Care setting for effective management[1]. Presently, there is a significant gap in evidence-based guidelines for the management of SD. While treatment recommendations have been proposed by authors in the UK and North America, these are based on the insights of small groups of clinicians[4,5]. A more comprehensive and inclusive consensus is needed.

In addition to SD, CAYP with dystonia often requires admission into paediatric critical care for other elective or unplanned reasons[6]. Admissions complicated by dystonia have been correlated with extended hospital stays[6]. Challenges have been identified in recognising dystonia within the Paediatric Critical Care setting, particularly in distinguishing between abnormal movements attributable to dystonia and those resulting from epilepsy[7].

4 Guideline Content

- The guideline will be developed according to RCPCH standards for guideline development, which is NICE-accredited.

- This document is the scope. It defines exactly what this guidance will (and will not) examine, and what the guidance developers will consider.

- The areas to be addressed by the guideline are in the following sections.

4.1 Population

4.1.1 Groups that will be covered

CAYP aged 1 month up to 18 years of age admitted to the PCC environment

The definition of the PCC environment will include Paediatric Intensive Care (Level 3) and High Dependency Care (Level 2)

4.1.2 Groups that will not be covered

Patients in Neonatal Critical Care, or children < 1 month old

Age>18 years or patients aged 16-18 years within the adult intensive care.

4.2 Healthcare settings and services

The definition of the PCC environment will include Paediatric Intensive Care (Level 3) and High Dependency Care (Level 2). Immediate management prior to transfer to the PCC environment will also be included.

4.3 Key areas

4.3.1 Clinical issues that will be covered

This project will focus on the recognition and emergency management of severe dystonia (including Status Dystonicus) in the Paediatric Critical Care (PCC) environment. It will encompass the management of severe dystonia immediately before transfer to the PCC environment, as well as the management of less severe emergent and established dystonia in Children and Young People (CAYP) receiving care in the PCC environment due to other health conditions (e.g. an emergency admission for pneumonia or an elective admission following spinal surgery).

4.3.2 Clinical issues that will not be covered

Management of dystonia in CAYP not requiring admission to a PCC environment

Management of dystonia in CAYP following discharge from a PCC environment.

Management of GI dystonia in CAYP admitted to a PCC environment

5 Clinical

- When should CAYP be admitted to Level 2 PCC for management of dystonia?
- When should CAYP be admitted to Level 3 PCC for management of dystonia?
- When should CAYP be intubated as part of the management of dystonia?
- What infusion therapies should be used in the management of dystonia in the critical care setting, at what dose ranges should they be used, and in what order?
- When should paralytic agents be used in the management of dystonia?
- How can dystonia in CAYP be recognised in the critical care environment?
- What outcome measures/metrics should be used at i) the patient level and ii) the service level?

- What non-pharmacological measures should be used in the management of dystonia in the PCC Environment?
- What non-infusion pharmacological treatments should be used for the management of dystonia in the critical care environment, at what dose ranges, and in what order?
- What are the side effects of pharmacological managements for dystonia in the PCC environment which should be actively monitored for?
- What are the acceptable range of physiological parameters for CAYP receiving treatment for dystonia in the PCC environment?
- What analgesics strategies should be used in the management of dystonia in the PCC environment?
- What are the common triggers (and perpetuating factors) for worsening dystonia in the PCC environment which should be actively monitored and managed?
- How commonly does GI Dystonia contribute to worsening dystonia in the PCC Environment?
- What are the common co-morbidities of CAYP receiving treatment for dystonia in the PCC environment, which may complicate this management?
- What should be included in ongoing dystonia management plans for CAYP following the management of dystonia in the PCC environment?
- When can CAYP receiving treatment for dystonia in the Level 3 PCC environment be stepped down to Level 2 care?
- When can CAYP receiving treatment for dystonia in the Level 2 PCC environment be discharged from the Critical Care environment?
- When should parallel planning and ceilings of care be discussed in CAYP receiving treatment for dystonia in the PCC environment?
- How should the MDT providing management for dystonia in the PPC Environment be structured?
- What psychological support should be offered to i) CAYP receiving treatment for Dystonia in the PCC Environment, ii) their family/carers and iii) staff involved in their care?

- What is the role of transdermal clonidine patches in the management of dystonia in the PCC environment?
- What novel/emerging therapies may play a role in the management of dystonia in the PCC environment?

6 Related guidance

Nil relevant to this project

7 References

1 Lumsden DE, Cif L, Capuano A, Allen NM. The changing face of reported status dystonicus - A systematic review. *Parkinsonism Relat Disord* 2023; **112**: 105438.

2 Lumsden DE, Papandreou A, Allen NM, Lin JP. The utility of creatine kinase in status dystonicus and pre-status dystonicus. *Eur J Paediatr Neurol* 2025; **57**: 57-63.

3 Lumsden DE, King MD, Allen NM. Status dystonicus in childhood. *Curr Opin Pediatr* 2017; **29**: 674-82.

4 Lumsden DE. Fifteen-minute consultation: Management of acute dystonia exacerbation and status dystonicus. *Archives of disease in childhood. Education and practice edition* 2025; **110**: 8-14.

5 Vogt LM, Yang K, Tse G, Quiroz V, Zaman Z, Wang L, Srouji R, Tam A, Estrella E, Manzi S, Fasano A, Northam WT, Stone S, Moharir M, Gonorazky H, McAlvin B, Kleinman M, LaRovere KL, Gorodetsky C, Ebrahimi-Fakhari D. Recommendations for the Management of Initial and Refractory Pediatric Status Dystonicus. *Mov Disord* 2024; **39**: 1435-45.

6 Ahmed R, Griffiths B, Lumsden DE. Dystonia in paediatric intensive care: A retrospective prevalence study. *Archives of disease in childhood*: BMJ Publishing Group; 2020. 912-4.

7 Stephen CD, Dy-Hollins M, Gusmao CM, Qahtani XA, Sharma N. Dystonias: Clinical Recognition and the Role of Additional Diagnostic Testing. *Seminars in neurology* 2023; **43**: 17-34.

Appendix 1: Guideline Development Group Expertise and Stakeholders Organisations

The Guideline Development Group (GDG) will include multidisciplinary clinical experts, methodologists, and parent/CYP representatives. Stakeholder organisations such as relevant professional bodies, specialty groups, and associations whose practice or care may be affected will be identified by the working group. They will be formally invited and engaged at key stages of development, including the scope and draft consultations.

Working Group	Stake Holder Group
• Paediatric Neurology • Paediatric Neurodisability	• British Paediatric Neurology Association • Paediatric Critical Care Society

• Paediatric Critical Care Medicine • Paediatric Intensive Care • Paediatric Intensive Care CNS • Paediatric Palliative Care • Paediatric Pharmacy • Paediatric Intensive Care Grid Trainee • Occupational Therapy • Advocates with Lived Experience of Dystonia in Childhood	• Association of Paediatric Palliative Care Medicine • Royal College of Anaesthetists • Royal College of Paediatrics and Child Health • British Academy of Childhood Neurodisability • Neonatal and Paediatric Pharmacy Group • Association of Paediatric Anaesthetists of Great Britain • Together for Short Lives • Roald Dahl's Marvellous Children's Charity • Association of Paediatric Emergency Medicine • Dystonia UK • British Association of General Paediatrics