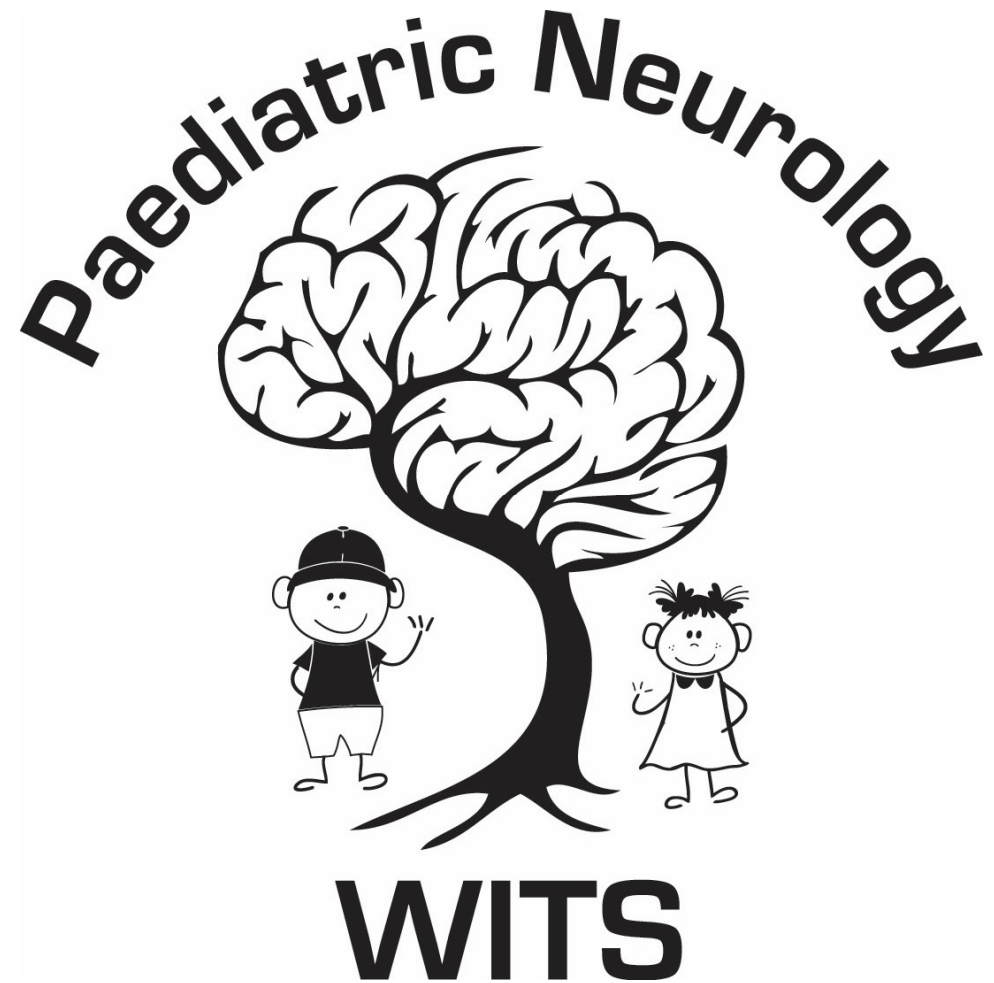


STATUS DYSTONICUS

Marc Hauptfleisch

Consultant Paediatric Neurologist

CHBAH and WDGMC



- Dystonia is a disorder of tone, movement and posture
- Dystonia is common in childhood
- Dystonia, at it's most severe, can be life threatening

WHAT IS TONE

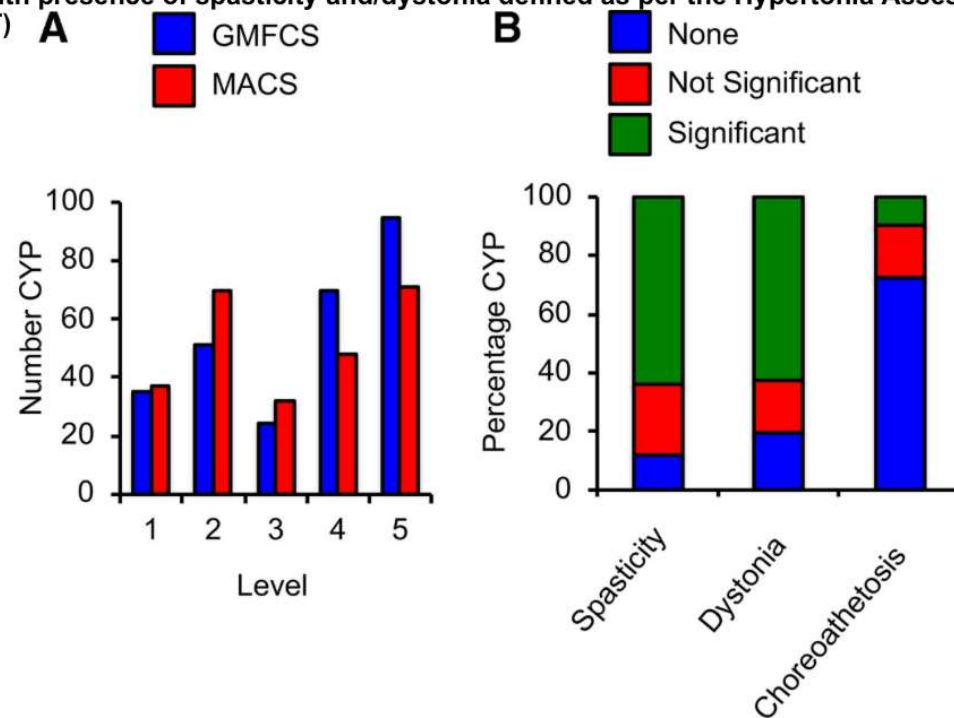
- We know what it is but not easy to explain
- Resistance to passive stretch while the patient is attempting to maintain a relaxed state of muscle activity.
- Often requires multiple assessments

SPASTICITY VS DYSTONIA

- Spasticity:
 - Velocity dependent increase in tone
 - Component of UMN complex
 - Implies dysfunction of the corticospinal tract/descending motor pathways and a loss of inhibition at the spinal cord level
- Dystonia
 - Disorder of involuntary sustained or intermittent muscle contractions causing abnormal movements or postures
 - Traditionally “basal ganglia” disorder – now appreciated to be due to a dysfunction across a broader motor network
 - CST not involved

MIXED TONE IS THE
GENERAL RULE
RATHER THAN THE
EXCEPTION

(A) Distribution of Gross Motor Function Classification System (GMFCS) and Manual Ability Classification System (MACS) levels across the cohort. (B) Motor phenotype of CYP in the cohort, with presence of spasticity and/dystonia defined as per the Hypertonia Assessment Tool (HAT) orders.



Daniel E Lumsden et al. Arch Dis Child 2019;104:775-780

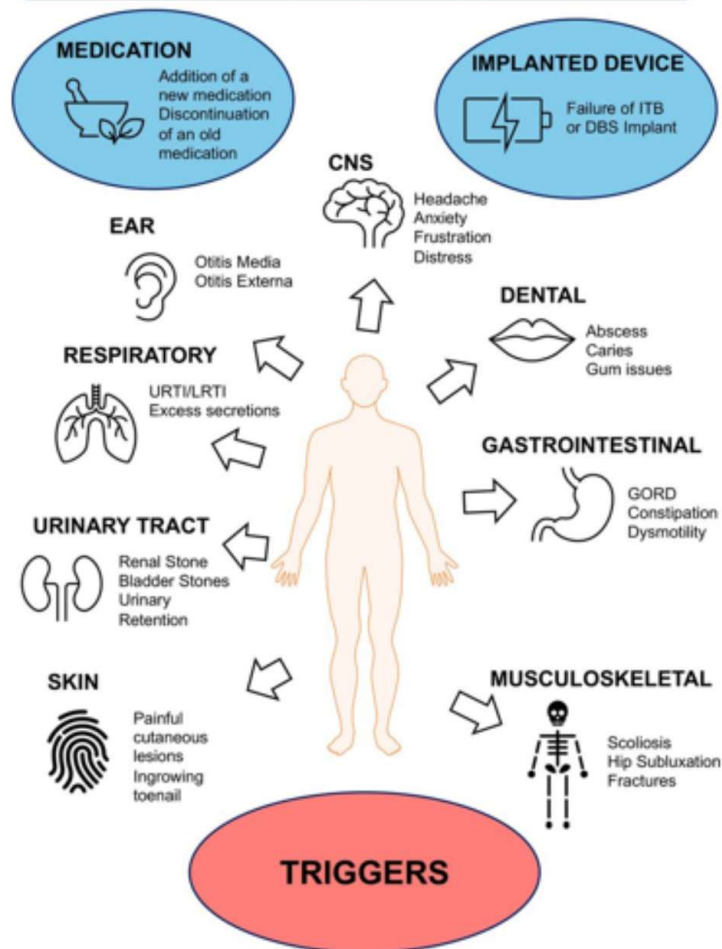
WHY TREAT DYSTONIA IN GENERAL?

Medications have very significant side effects, so it is very important to have a clear rationale for treatment, as well as some way of measuring if the desired outcome has been achieved

- Pain
- Disabling
- Interference with care
- Musculoskeletal deformity
- Respiratory compromise

Ahmed et al 2020
doi.org/10.1136/archdischild-2018-316421

PROVOKERS OF ACUTE DYSTONIA EXACERBATION



Daniel E Lumsden Arch Dis Child Educ Pract Ed
2025;110:8-14

Avoid Triggers

- Good pressure/skin care
- Good sleep hygiene
- Nutrition
- Vaccination
- Emotional/psychological support

Treat Triggers

- Analgesia
- Laxatives
- Urinary retention
- Orthopaedic input
- Antibiotics

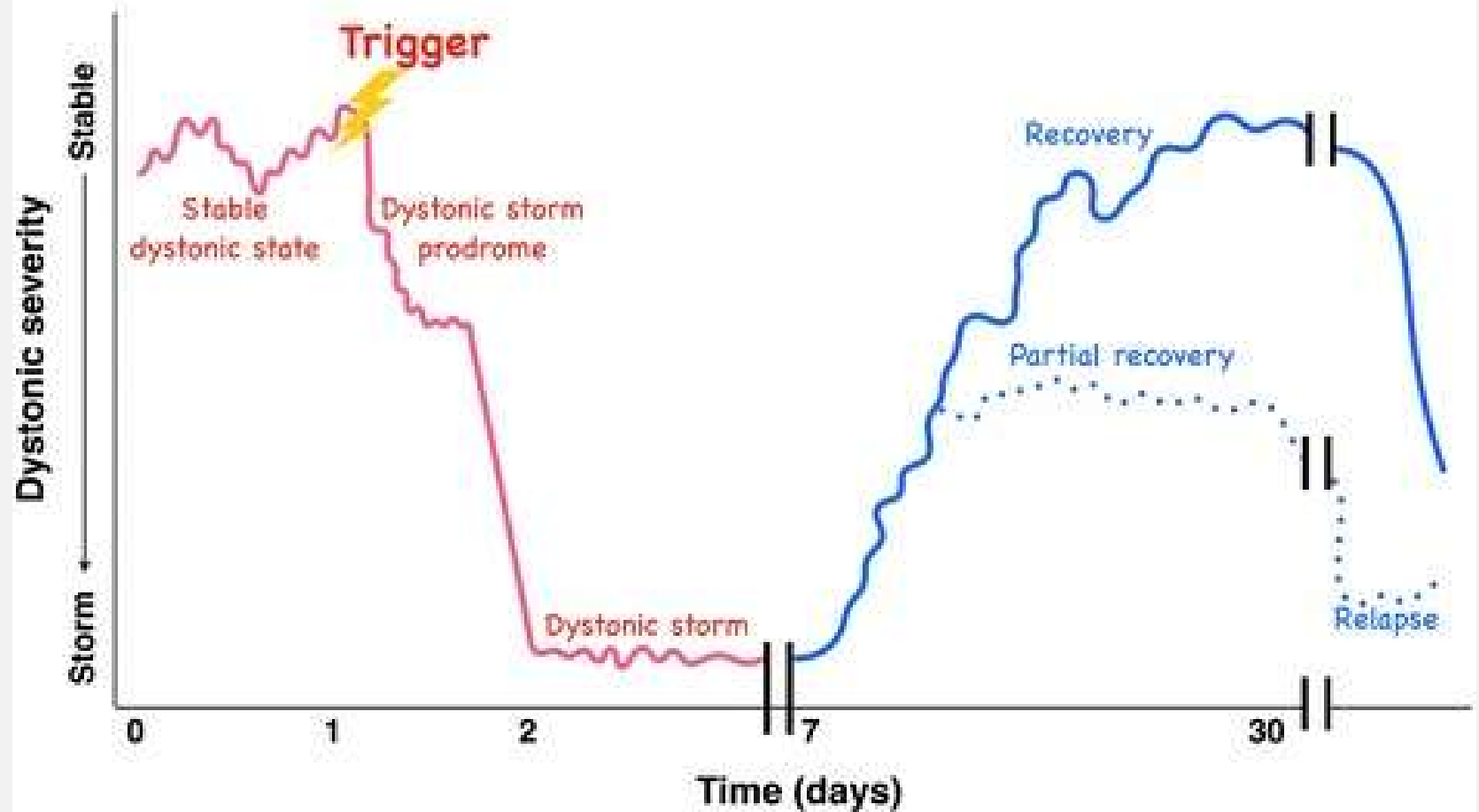
Treat Dystonia

- Consider
- Back ground meds
 - Urgency of treatment

STATUS DYSTONICUS

- Dystonic Crisis Dystonic Storm Life Threatening Dystonia
- First recognized in 1982 with numerous cases since and roughly $\frac{3}{4}$ of all published cases are in children
- NOT a discrete event with a clear starting point, but more of the severe end of a progressive scale of worsening

Typical clinical course of dystonic storm patient



- Termsarasab and Frucht 2017

CAN WE DEFINE “STATUS DYSTONICUS”

“Increasingly frequent and severe episodes of generalized dystonia, which necessitate urgent hospital admission”

Manji et al 1998

”A malignant hyperkinetic movement disorder emergency in which rapid deterioration of dystonia requires emergent intervention”

Termsarasab 2017

“A movement disorder emergency characterized by severe episodes of generalized or focal dystonic with or without other hyperkinetic movements that have necessitated urgent hospital admission because of the direct life-threatening complications of these movements, regardless of the patient’s neurological condition at baseline”

Lumsden and Allen 2018

RECOGNIZING SD

- Generalized dystonia – phasic or tonic
- Fever Tachycardia Hypertension
Tachypnoea Sweating Autonomic instability
- Bulbar involvement with dysarthria and dysphagia

DIFFERENTIAL DIAGNOSIS OF SD

	Trigger	Time Course	Movements	Altered mental status	Autonomic instability
Dystonic storm	+/-	Hours-days	Dystonia +/- chorea	-	+
Choreic storm	+/-	Hours-days	Chorea	-	-
Neuroleptic malignant synd	+++	Days-weeks	Parkinsonism	+	+
Serotonin syndrome	+++	Hours-days	Myoclonus	+	+
Malignant hyperthermia	+++	Acute	nil	-	-
Drug intoxication	+++	Acute	-	+ with psychosis	+/-
Auto-immune encephalitis	-	Days-weeks	Chorea	+ with psychosis	+
Oculogyric crisis	+++	Acue	Dystonia	-	+/-

CLASSIFYING SD

1. Phenomenology

- Phasic
- Tonic

2. Temporal Profile

- Monophasic
- Relapsing

3. Recovery of episode

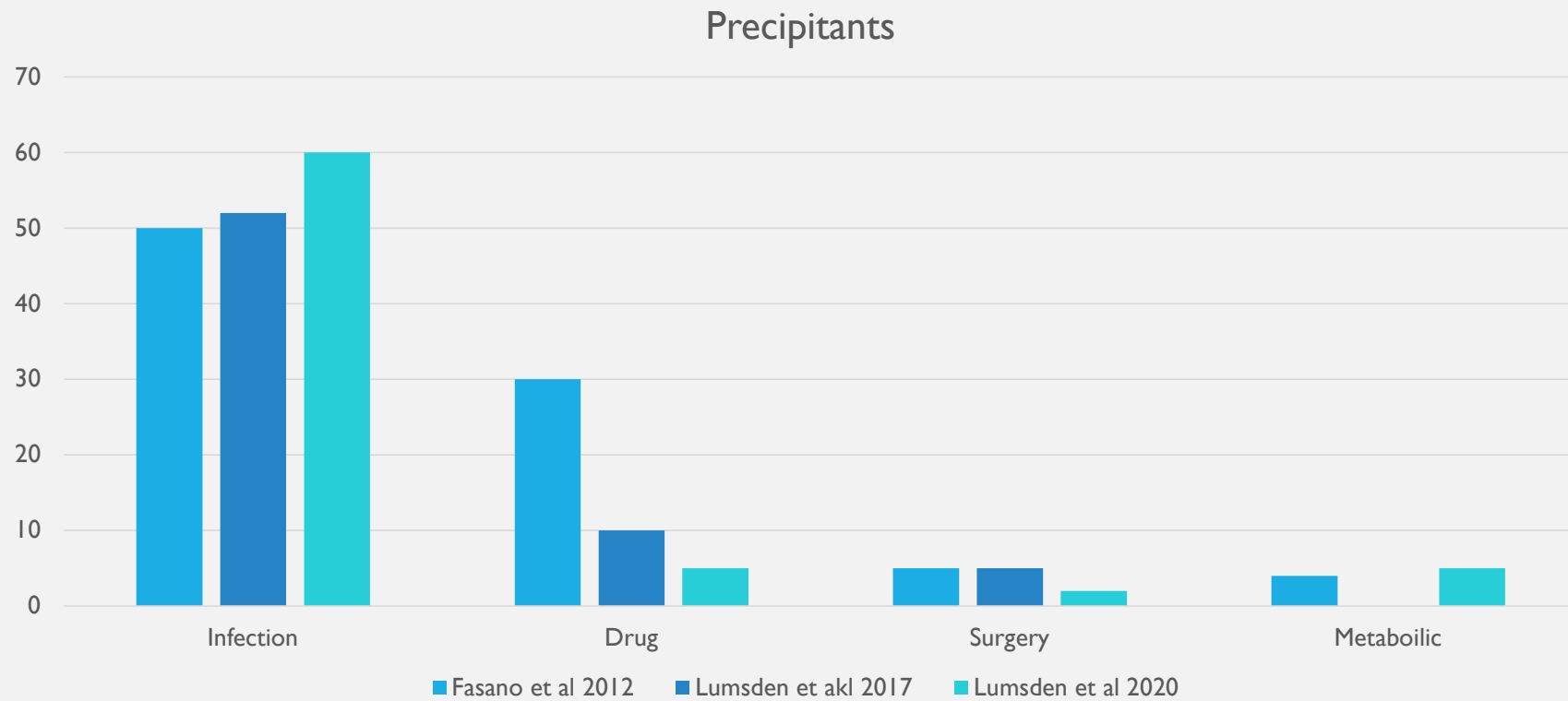
- Total
- Partial
- No recovery

WHO GETS STATUS DYSTONICUS?

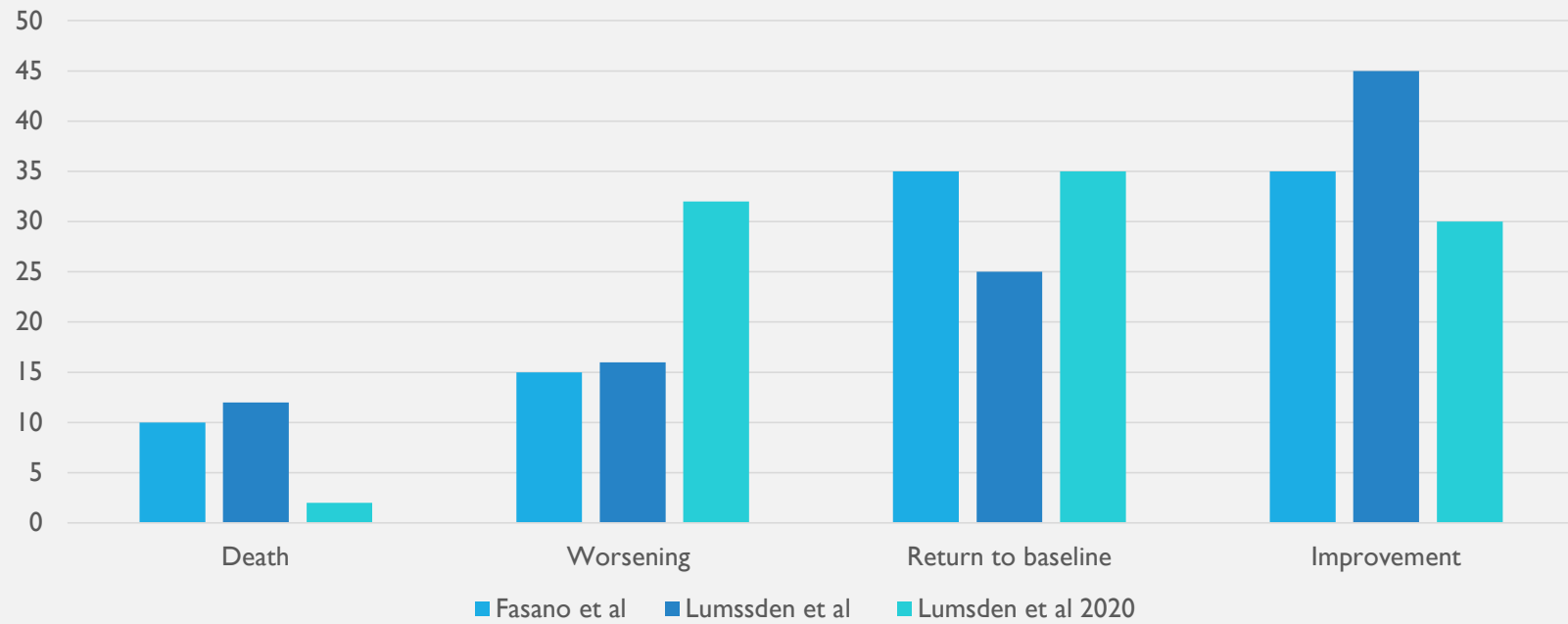
Any child with dystonia is potentially at risk of developing status dystonicus

Dystonia is a fluctuating condition with the very worst end being status dystonicus – children typically progress through a period of worsening dystonia before they get to that point

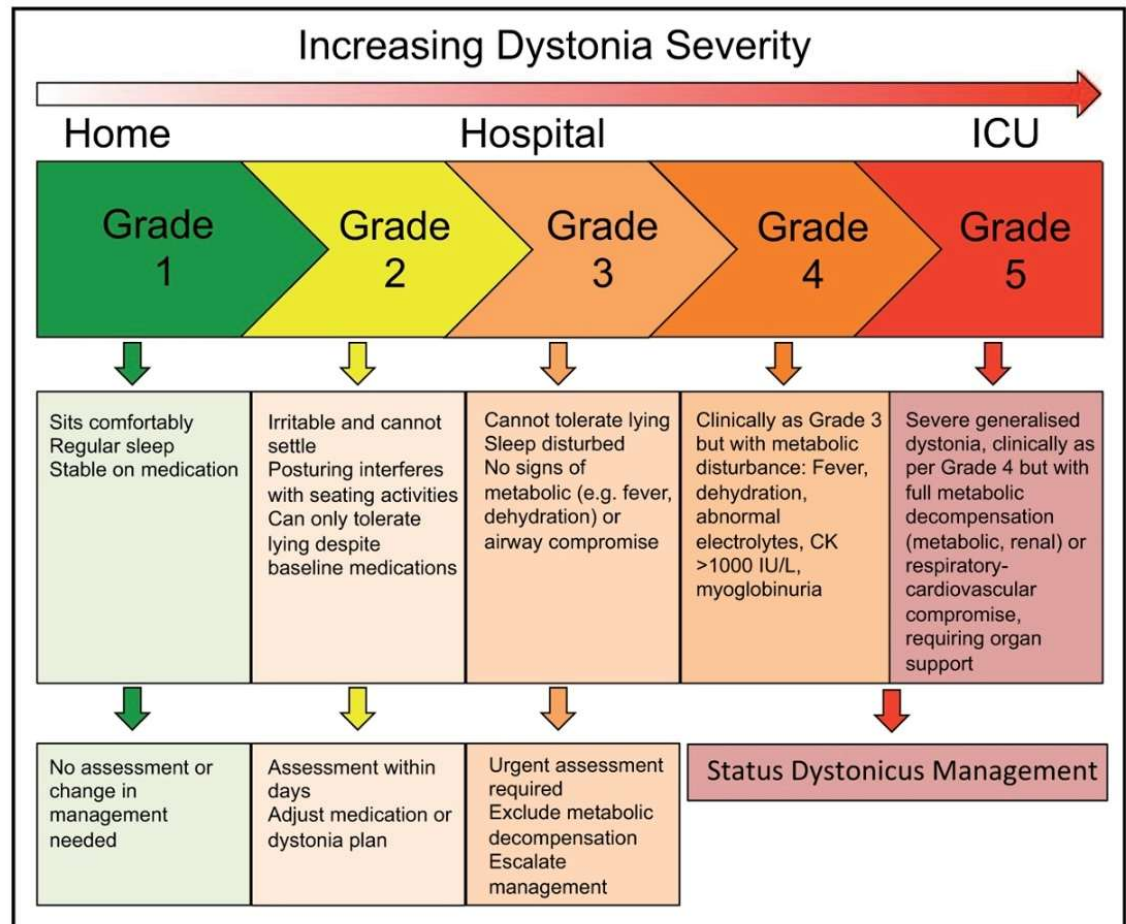
WHAT DO WE KNOW?



Outcomes



MEASURING DYSTONIA

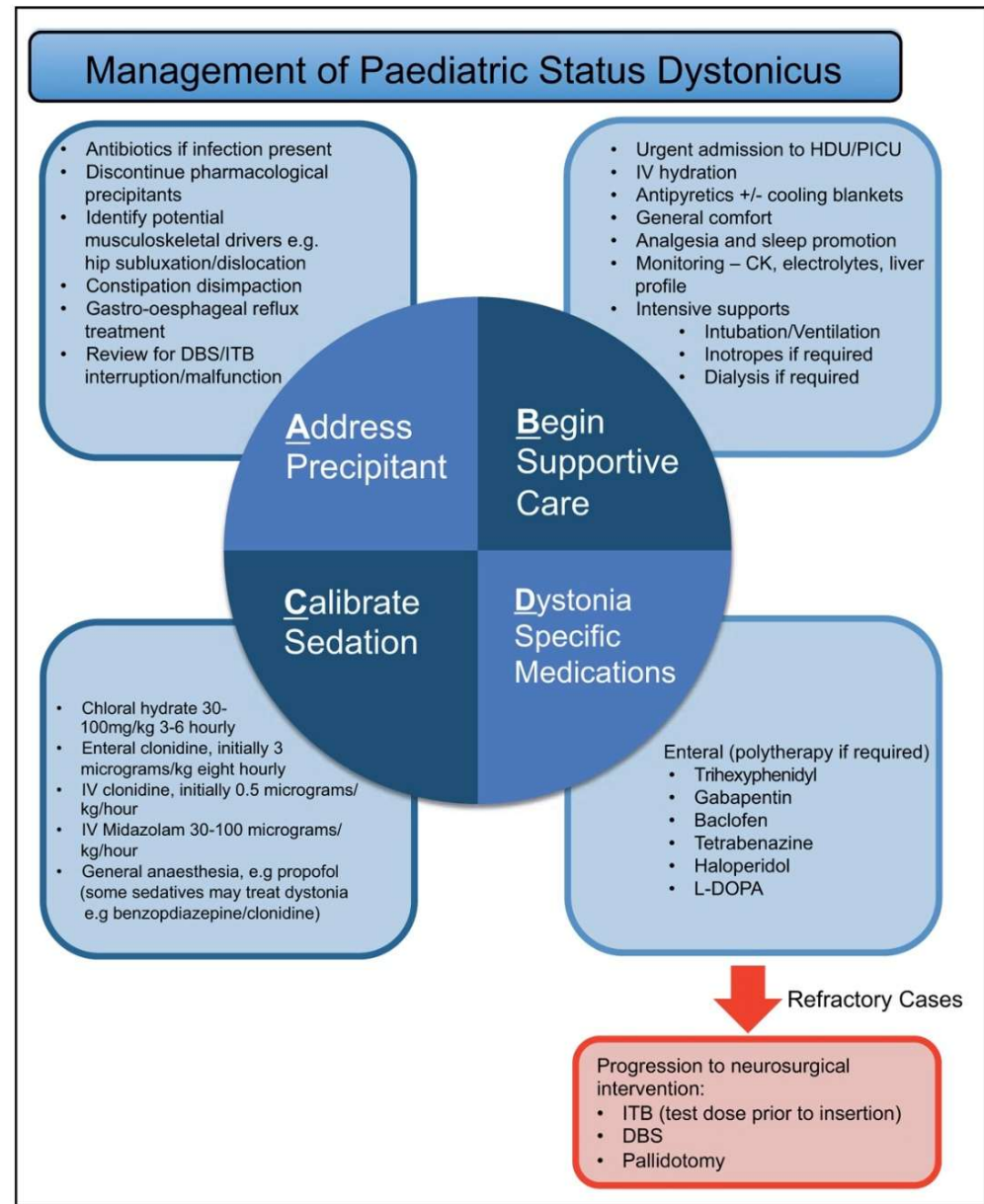


Lumsden et al
2017

COMPLICATIONS OF SD

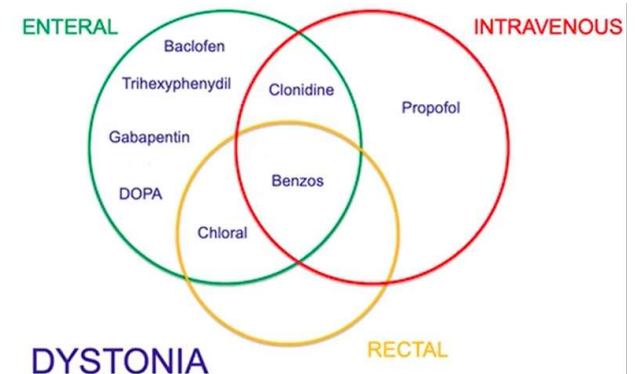
- Respiratory
 - Bulbar dysfunction/Spasm, truncal/respiratory muscle spasm, diaphragmatic spasm, exhaustion, aspiration pneumonia
- Cardiovascular
 - Dysautonomia, dehydration
- Metabolic compromise
 - Rhabdomyolysis, myoglobinaemia/myoglobinuria, electrolyte imbalances, acid-base derangement
- Hyperpyrexia
- Mental Health and Wellbeing

MANAGEMENT OF SD



DYSTONIA SPECIFIC MEDICATIONS

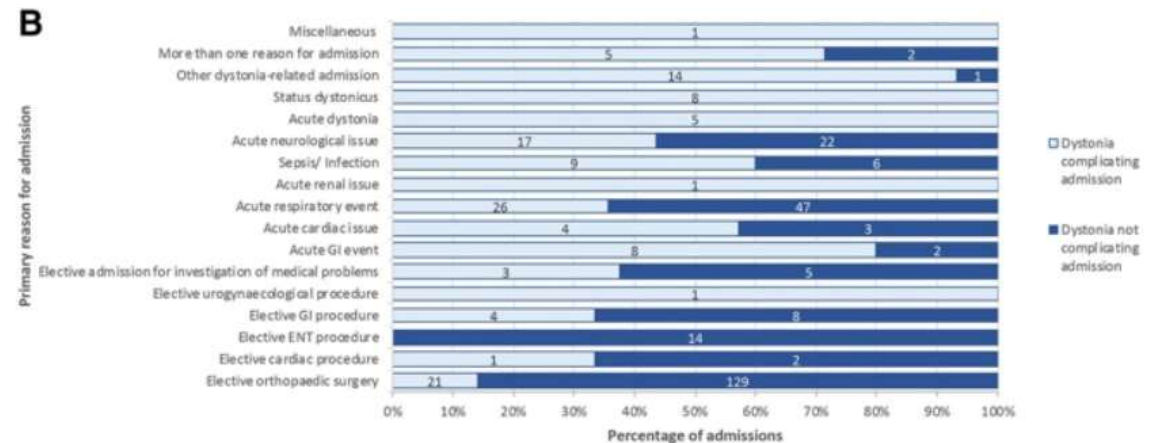
- Gabapentin – 5mg/kg dly -> bd -> tds, then 10mg/kg tds
- Baclofen – good choice if co-independent spasticity
- Clonidine 3mcg/kg tds
- Trihexyphenidyl – anticholinergic agent - start low and go slow
- If marked focal component – botulism toxicity



IMPACT IN GENERAL PAEDS

10-year retrospective study at
Evelina Hospital PICU

- Dystonia was the primary cause of admission in 20 patients – 5%
- **BUT** Dystonia complicated 128 of 369 admissions = 35%
- Median length of stay was longer (21 vs 7 days) in PICU admissions complicated by dystonia



Rumsha Ahmed et al. Arch Dis Child 2020;105:912-914

READING

REVIEW



Status dystonicus in childhood

Daniel E. Lumsden^a, Mary D. King^{b,c}, and Nicholas M. Allen^{d,e}

Purpose of review

Dystonia is a common paediatric neurological condition. At its most severe, dystonia may lead to life-threatening complications, a state termed status dystonicus. This review provides an update on the definition, causes, management and outcome of childhood status dystonicus.

Recent findings

High-quality studies in childhood status dystonicus are lacking, though an increasing number of case series have been published. Status dystonicus appears to occur more frequently in children compared with adults, with a clear precipitant identified in around two-thirds of cases. Although febrile illness remains the commonest trigger for status dystonicus, unplanned interruption to deep brain stimulation (DBS) is increasingly reported as a precipitant. In parallel with this, neurosurgical intervention for status dystonicus appears to have become more widely used, though optimum timing and patient selection remains unclear. In most cases, a multistaged approach is required; we propose an 'ABCD' approach – Addressing precipitants, Beginning supportive measures, Calibrating sedation and Dystonia specific medications. Outcomes following status dystonicus appear to have slightly improved in recent years, potentially as a consequence of increasing use of DBS, though mortality has remained around 10%.

Summary

Future work is needed to inform evidence-based guidelines for the management of status dystonicus. One of many pressing questions is the precise indication, and timing of interventions such as DBS.

Keywords

dystonia, dystonic storm, life-threatening dystonia, rhabdomyolysis, status dystonicus

INTRODUCTION

on providing an update on the more severe presenta-

DOI:10.1097/MPO.0000000000000556

Since then, understanding of dystonia has evolved, with changes in how the disorder is classified, categorized and conceptualized. Originally considered a disorder of basal ganglia function, it is now recognized that dystonia may arise because of disturbed function across much more widely distributed net-

Dystonia is usually a fluctuating state, where clinically the intensity varies over minutes, hours or days and there is paucity of readily available biomarkers for detection. At its most extreme, periods of 'severe dystonia' may be life-threatening but precisely defining when this state is entered remains

Best practice

Fifteen-minute consultation: Management of acute dystonia exacerbation and status dystonicus

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ABSTRACT

Dystonia is a common disorder of movement and tone, characterised by sustained or intermittent muscle contractions causing abnormal movements, postures or both. Children and young people with dystonia can experience episodes of acute worsening tone, which require prompt treatment. When most severe, dystonia may become life-threatening—a state called 'status dystonicus'. This guide aims to provide a framework for how to approach the child with acutely worsening dystonia, following an 'ABCD' approach: Addressing the precipitant, Beginning supportive care, Calibrating sedation and Dystonia-specific medications.

INTRODUCTION

Dystonia is defined as 'a movement disorder characterised by sustained or

very likely significantly underestimated. Around one in four CAYP experiencing SD fail to return to baseline following the episode, with a reported mortality of ~5%.⁵ The risk of developing SD is present for all children with dystonia, though has been most frequently reported in CAYP with dyskinetic CR, pantothene kinase-associated neurodegeneration and in GNAO1-related dystonia.⁶ In the absence of a robust evidence base, this guide will focus on a pragmatic approach to the management of acute dystonia exacerbation.⁷ For a more general approach to the diagnosis and management of dystonia in childhood, the reader is directed to the review by Forman and colleagues.⁸

GETTING THE RIGHT STATUS: DYSTONIA OR DBS PCV?

has many causes, the most common being cerebral palsy (CP).² Dystonic movements may be painful and can interfere with function and the delivery of daily care. Dystonia may occur in isolation, or as part of a more complex motor disorder

that they are not experiencing an epileptic seizure. Prolonged generalised tonic-clonic seizures may be directly harmful to the brain if they continue after 30 min,⁹ and in this situation, the aim of treatment is to turn the seizure off. In contrast

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