LRI Children's Hospital

Management of Dystonia and Status dystonicus in children

Staff relevant to:	Medical staff caring for Children within UHL Children's Hospital presenting with status epilepticus
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Contents

1. Introduction and Who Guideline applies to

This guideline is intended for children presenting with dystonia including status dystonicus under 16 years of age.
This guideline aims to help UHL clinicians to identify, take a focused history to identify possible cause/s, investigate and manage the child presenting with dystonia and status dystonicus.
Specialised treatment options such as Intrathecal Baclofen (ITB) and Deep brain stimulation (DBS) is beyond the scope of this guideline and hence not discussed.

Related Documents.

Basic Life Support or Choking UHL Childrens Hospital Guideline C2/2016
Vascular Access UHL Policy B13/2010
IV (Intravenous Therapy) UHL Policy B25/2010
Status Epilepticus UHL Childrens Hospital Guideline D1/2022
Pain Agitation Neuromuscular Blockade, Delirium & Early Mobility UHL Paediatric Intensive Care Guideline C10/2009

A) What is Dystonia?

The term dystonia originated in 1911 with Oppenheim's describing 4 individuals who were floppy at rest yet developed stiffness when they tried to move.

The word ***dys-tonia*** literally means ***abnormal tone*** caused by sustained or intermittent involuntary contraction of muscles.

This abnormal tone could either be hyper or hypo or both involving one or different groups of muscles at any single point of time and may change to a different tone at another time.

According to the 2013 international consensus of movement disorder society and the Dystonia Europe society,

Dystonia is a movement disorder characterized by

1. **Sustained or intermittent muscle contractions causing abnormal, often repetitive movements, postures, or both.**
2. **Dystonic movements are typically patterned, twisting, and may be tremulous.**
3. **Dystonia is often initiated or worsened by voluntary action and associated with overflow muscle activation.**

B) What is Status dystonicus?

Status dystonicus is a severe and potentially life-threatening condition of increasingly frequent and severe episodes of generalised dystonia which requires urgent (hospital) management.

Status dystonicus is a clinical diagnosis based on the identification of severe, generalised dystonic movements. It usually occurs in a child who is already known to have dystonia, although new-onset cases are reported.

Status dystonicus is mainly triggered by severe gut dysmotility, medication withdrawal, infection or disruption to deep brain stimulation.

It can cause rhabdomyolysis leading to multi-organ failure and in severe cases death.

Diagnosis should involve identification of associated comorbidity (bulbar and respiratory difficulties, metabolic derangements, exhaustion and pain).

2. Focused history and examination

Take focused history especially if dystonia is the initial presentation in a child.		
History	***Pregnancy/Delivery***	Previous Miscarriages; Infections or bleeding during pregnancy; gestation at delivery
	Neonatal Period	Resuscitation/APGARs/ problems establishing breastfeeding; jaundice; concerns about weight loss; neonatal infection/sepsis; neonatal encephalopathy; neonatal seizures.
	Development and Schooling	Ages milestones achieved. Developmental delay/plateauing/regression; extra support in school. Visual/hearing difficulties
	Movement Disorder/Dystonia	At what age initial concerns raised; body distribution at onset and with progression; cause of dystonia over time; other associated movement problems; fluctuating during day; exacerbating factors such as sudden motion
	Family History	Consanguinity; movement disorders (not just dystonia); psychiatric history
	Complications of Dystonia	Feeding problems; mobility issues; communication issues; pain; gastroenterological issues; musculoskeletal deformities/growth
	Medications	Current medications: previous medications to treat dystonia (and why stopped); medications which have worsened dystonia
Examination	***Growth Parameters***	Height; weight; head circumference
	Motor Disorders	Dystonia- regions of body affected; other hyperkinetic movements; spasticity; rigidity; eye movements (including saccades); weakness; ataxia; selective motor control; dyspraxia
	General examination	Neurocutaneous stigmata; organomegaly; musculoskeletal deformity/ scoliosis; cardiovascular abnormalities; respiratory abnormalities

3. Classification of Dystonia

Dystonia is classified by three main factors:

1. **Age at which symptoms develop,**
2. **Areas of the body affected and,**
3. **Underlying cause.**

Axis	Dimension for Classification	Subgroups
Axis I: Clinical features	***Age at Onset***	• Infancy (birth to 2 years) • Childhood (3–12 years) • Adolescence (13–20 years) • Early adulthood (21–40 years) • Late adulthood (40 years and older)
	Body Distribution	• Focal (one isolated body region) • Segmental (two or more contiguous regions) • Multifocal (two or more non-contiguous regions) • Hemidystonia (half the body) • Generalized (trunk plus two other sites)
	Temporal Pattern	• Disease course (static vs. progressive) • Short-term variation (e.g., persistent, action specific, diurnal, or paroxysmal)
	Associated features	• Isolated (with or without tremor) • Combined (with other neurological or systemic features)
Axis 2: Aetiology	***Nervous system Pathology***	• Degenerative • Structural (e.g., focal static lesions) • No degenerative or structural pathology
	Heritability	• Inherited (e.g., sex linked or autosomal, dominant or recessive, or mitochondrial) • Acquired (e.g., brain injury, drugs/toxins, vascular, or neoplastic)
	Idiopathic	• Sporadic and Familial

4. Grading of Dystonia Severity:

Dystonia is a fluctuating state of tone. Categorisation into different grades helps planning management strategy boundaries between the grades can be very subtle on times.

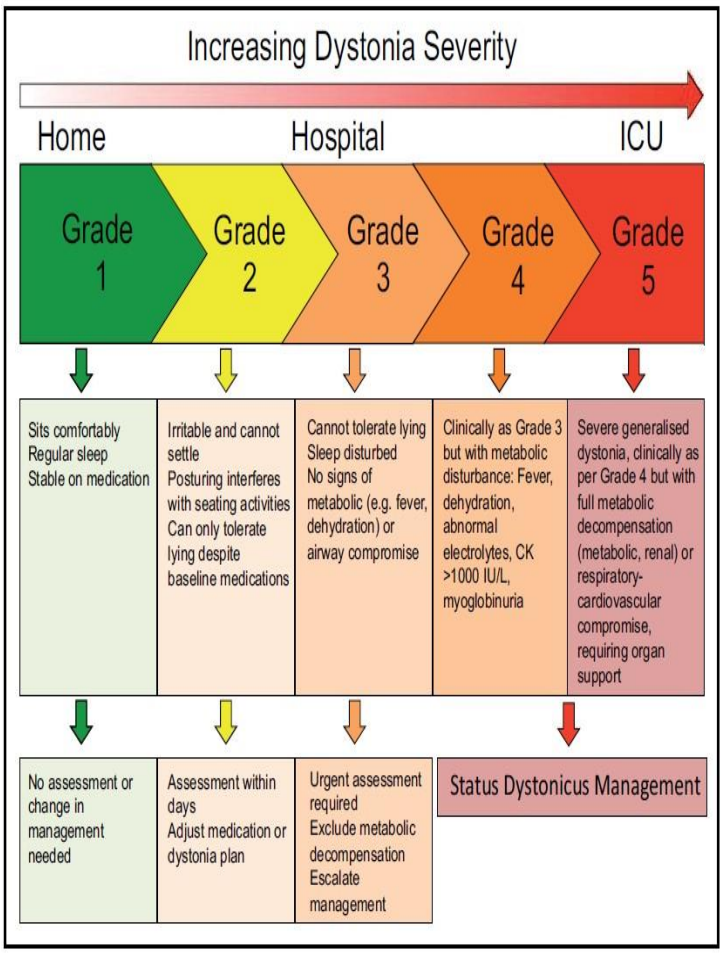
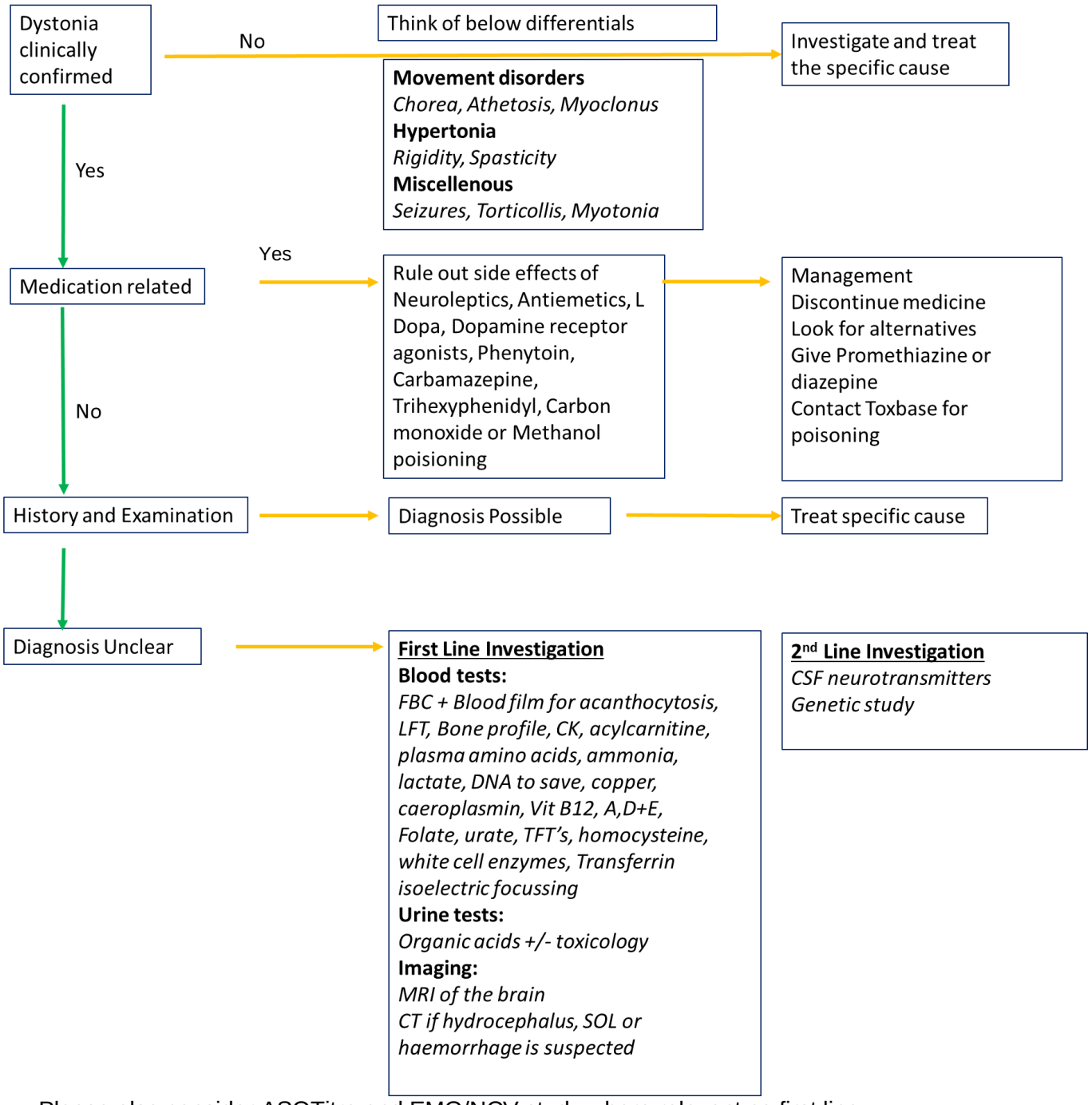


FIGURE 1. Screening for dystonia severity (grade) and action plan. Dystonia severity action plan (DSAP) (for established dystonia patients); Modified with permission from Lumsden *et al.* [10].

5. Approach to Dystonia

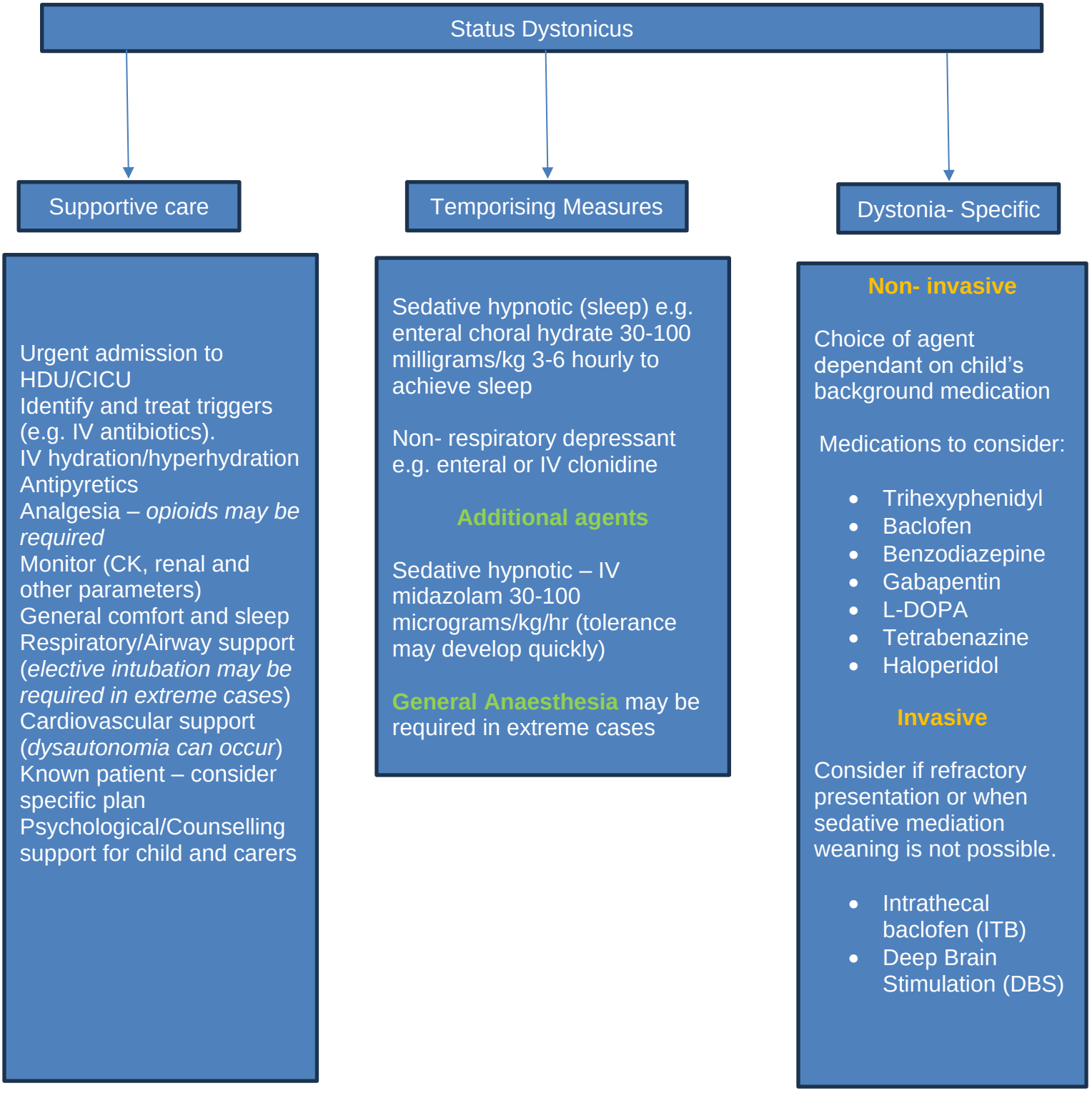


Please also consider ASOTitre and EMG/NCV study where relevant as first line

See Dystonia severity grading for guidance about step wise medication use for different grades of dystonia.

6. Management of Status Dystonicus

(adapted from Allen N, et al - Status dystonicus: A Practice Guide. Dev Med Child Neurol 2014:56:105-112)



See next page for Dystonia specific medication and their uses

Dystonia Specific Medication**:**

Medications	**Dosages**	**Comments**
1.Trihexyphenidyl (Enteral) BNFC	Initially 1-2mg/day in 1-2 doses (all ages >1 month) Doses should be increased by 0.5-1 mg per dose per week up to maximum dose as listed below Maximum recommended doses (equates to 2mg/kg/day) 1month – 2 years – 3mgTDS 2-12 years – 10mg TDS >12 years – 30mg TDS	Anticholinergic agent. Side effects include dry mouth, blurred vision, constipation and urinary retention. May be better tolerated in younger children and with slower dose escalation. Depression may also be seen. Once maximum dose reached, maintain for 3 months and review response.
2. Baclofen (Enteral) BNFC	Initial dose (all ages) 75microgram/kg QDS Increase by 0.25mg/kg/dose each week Maximum recommended dosage (equates to 2mg/kg/day) Under 9 years – 40mg/day Over 8 years – 60mg/day Review if no benefit seen after 6 weeks	GABAminergic agent. Not likely to be beneficial below 1 year of age Side effects commonly include sedation and nausea. Wean over 2 weeks Poorly crosses blood brain barrier, and so higher doses may be required. Bulbar function may also be adversely affected by baclofen.
3.Benzodiazepine (Enteral) BNFC/ Evelina Childrens Hospital	**Diazepam - preferred** 4 wks–1 year 0.25 mg/kg BD 1-4 years 2.5mg BD 5-12 years 5mg BD >13 years 10mg BD - QDS Doses given short term up to 4 hourly in status dystonicus **Nitrazepam** <1 year 0.25mg-0.5mg/kg BD 1-4 years 2.5mg BD 5-12 years 2.5-5mg BD >12 years 2.5-15mg BD	Acute side effects include respiratory suppression and increased drooling. Dependency develops with regular use, and so slow wean over weeks required to avoid symptoms of withdrawal. Tolerance to dosage also builds over time.
4.Clonidine (Enteral/Intravenous/ Patches) Evelina Children's	Initially: 3micrograms/kg **(maximum 50 micrograms**) at night. Dose and frequency may be	Oral and intravenous doses interchangeable. Role in acute dystonia as

Hospital	increased weekly, according to response. Doses may be non-evenly distributed throughout the day for individual symptom control. Inpatient setting (with appropriate BP and respiratory monitoring) doses may be escalated up to the equivalence of 2microgram/kg/hour IV e.g. 12micrograms/kg four times daily. Higher doses of IV clonidine may be required in some cases and has to be discussed with Paediatric neurology team for an individualised plan. Continuous IV infusions or patches may be considered (at 1:1 dose conversions) for children unable to take enterally. (Please mention the total enteral dose in microgram per day for conversion to a patch format when submitting request to pharmacy)	benzodiazepine sparing sedative agent. Bradycardia may occur with higher doses. Doses >48microgram/kg/day may be used, but only following discussion with clinicians with experience with such dosage regimes. If used for >2 weeks, wean over at least 6 days
5.Gabapentin (Enteral) Evelina Children's Hospital	Day 1: 5 mg/kg OD Day 2: 5 mg/kg BD Day 3: 5 mg/kg TDS Can increase to 10mg/kg TDS or 3.6g daily Reduced dose required in renal Impairment – discuss with pharmacist	Potentially most useful when pain is the significant feature of dystonia.
6.Chloral Hydrate (Enteral) Evelina Childrens Hospital	30-60mg/kg (max 1g) 3 - 6 hourly – 3 hourly dosing under the supervision of a paediatric neurologist	Acute sedative agent
7.L-Dopa (Enteral) BNFC Use Co-careldopa (Sinemet) – each 62.5mg tablet contains 50mg levodopa and 12.5 mg carbidopa – contains 1:4 ratio of carbidopa:levodopa	Doses expressed as LevoDopa >3months initially 250mcg/kg BD - TDS Can be increased every 2-3 days to total 1mg/kg TDS	Significant side effects include nausea, which may limit dosage. Must be stopped for a minimum of 72 hours prior to CSF neurotransmitter metabolite analysis, unless analysis aimed at monitoring efficacy of treatment in children, e.g. with a diagnosis of tyrosine hydroxylase deficiency.

7. Education and Training

Ensure healthcare professionals managing children with Status Dystonicus are APLS trained and is up to date.

8. Monitoring Compliance

What will be measured to monitor compliance	How will compliance be monitored	Monitoring Lead	Frequency	Reporting arrangements
Admission to CICU with status dystonicus	Audit	Consultant Paediatric Neurologist	2 Yearly	Paediatric Neurology group

8. Supporting References

1. Advanced Paediatric Life Support (APLS) Advanced Life Support Group (ALSG), Blackwell Publishing.
2. Lumsden D. The Child With Dystonia. Paediatrics and Child Health 2018 28(10): 459-467
3. Allen M N, Pierre J, Lunch T, King D M. Status Dystonicus: A Practice Guide. Developmental Medicine and Child Neurology 2014;56: 105-112
4. Lin JP. Advances in pharmacotherapies for movement disorders in children: current limitations and future progress Curr Opin Pediatr. 2017 Dec;29(6):652-664
5. Albanese A, Bhatia K, Bressman SB, Delong MR, Fahn S, Fung VS, Hallett M, Jankovic J, Jinnah HA, Klein C, Lang AE, Mink JW, Teller JK. Phenomenology and classification of dystonia: a consensus update. Mov Disord. 2013 Jun;28(7):863-73
6. Koy A, Lin JP, Sanger T, Marks W, Mink J, Timmerman L. Advances in management of movement disorders in children. Neurology 2016 15:719-735
7. Lumsden D, Lundy C, Fairhurst C, Lin JP, Dystonia Severity Action Plan: a simple grading system for medical severity of status dystonicus andlife threatening dystonia. Dev Med Child Neurology 2013; 55:671-672
8. Lumsden DE, King MD, Allen NM, Status Dystonicus in Childhood. Current Opinion in Paediatrics 2017 29(6): 674-682
9. Lumsden DE, King MD, Allen NM, Status Dystonicus in Childhood. Current Opinion in Paediatrics 2017 29(6): 674-682
10. Tomlin S (ed.). (2015) Evelina London Paediatric Formulary. Evelina London Childrens Hospital, St Thomas' NHS Foundation Trust. 1st Electronic Edition adapted from 8th printed edition http://cms.ubqo.com/public/d2595446-ce3c-47ff-9dcc-63167d9f4b80
11. Kliegman RM, Stanton BMD, St. Geme J, Schor NF, eds. Nelson' s Textbook of Pediatrics. 20th ed. Philadelphia, PA: Saunders Elsevier; 2016.

12. Koy A, Lin JP, Sanger TD, Marks WA, Mink JW, Timmermann. Advances in management of movement disorders in children. Lancet Neurol 2016; 15: 719-35.
13. Joint Formulary Committee (2018-2019) British National Formulary for Children.September 2018-19. London: British Medical Association and Royal Pharmaceutical Society of Great Britain.
14. Jassal,SS,et al The Association of Paediatric Palliative Medicine Master Formulary 4th Edition, 2017
15. Egmond ME, Kuiper A, Eggink H, et al. Dystonia in children and Adolescents; a systematic review and a new diagnostic algorithm, Neurol Neurosurg Psychiatry 2015;86:774-781

9. Key Words

Abnormal tone,Dystonia, Status dystonicus

The Trust recognises the diversity of the local community it serves. Our aim therefore is to provide a safe environment free from discrimination and treat all individuals fairly with dignity and appropriately according to their needs.
As part of its development, this policy and its impact on equality have been reviewed and no detriment was identified.

Contact and review details	
Guideline Lead (Name and Title) Dr B Rai, Consultant	**Executive Lead** Chief Medical Officer
Changes made during review: Introduction updated Pg. 6 Added 'Please also consider ASOTitre and EMG/NCV study where relevant as first line' to the bottom of Approach to Dystonia flow chart New Section 6 – Management of Status Dystonicus	