

In-Hospital Management of Acute Dystonia Exacerbations: Initial and Refractory Status Dystonicus

Version: 2

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1.0 Introduction

Dystonia is a hyperkinetic movement disorder characterized by involuntary muscle contractions. These muscle contractions result in twisting and posturing movements (stiffening, bending, arching, clenching, or recurrent patterned movements). Dystonia can be sustained or intermittent.¹ Dystonia can be focal (one body area), multifocal (multiple body areas), or generalized (involving the majority of the body). Dystonia can be caused by genetic or metabolic disorders, or it can be acquired by damage through brain injury or stroke.² We commonly see dystonia in children with severe neurological impairment, who may also have other neurological comorbidities such as spasticity, seizures, or neuro-irritability.

Dystonia is state dependent. This means that dystonia often worsens with voluntary movement, pain, illness, or strong emotions. Dystonia remits with sleep. Therefore, it is common for children to experience worsening dystonia in hospital, where triggers like pain, illness, and emotion are present. Dystonia can escalate and become severe. Status dystonicus (also known as dystonic storm or dystonic crisis) is a life-threatening emergency that requires urgent treatment. Status dystonicus is the most severe manifestation along a spectrum of worsening dystonia (Figure 1).³ Status dystonicus is characterized by increasingly frequent episodes of severe dystonia. It can lead to extreme pain and discomfort with complications including respiratory compromise, and metabolic derangements, including death.⁴⁻⁷

A trigger for worsening dystonia is often identified. It is imperative to address and treat the trigger to improve dystonia. Since dystonia abates with sleep, we also treat concurrently with sedatives.⁶ These urgent treatments take the same approach regardless of the underlying cause or etiology of the dystonia.

This clinical pathway has been created to standardize the approach to treatment of worsening dystonia at SickKids and to minimize potential complications.^{8,9} The target population includes: 1) Patients presenting to the Emergency Department or admitted to hospital with dystonia and 2) Patients requiring PICU care for refractory status dystonicus.

2.0 Purpose of guideline

- Reduce delays to treatment of dystonia
- Limit variation in the acute management of patients with severe dystonia
- Provide pharmacological dosing that is efficacious while minimizing risk of potential side effects
- Decrease incidence of progression to status dystonicus/refractory status dystonicus and prevent its secondary complications

3.0 Definitions⁹

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Term	Definition
Pre-Status Dystonicus	A child demonstrating worsening dystonia but without end-organ involvement or airway compromise. They may be able to achieve intermittent sleep; however, it could be fragmented or easily disrupted by dystonia. Generally, grade 2-3 on the DSS.
Status Dystonicus	Worsening dystonia over >15 minutes, characterized by discomfort, tachycardia, and diaphoresis, with the presence of one or more end-organ metabolic decompensation (hyperthermia, major electrolyte abnormalities, renal failure, myoglobinuria or elevated serum CK level). Generally, grade 4 on the DSS.
Refractory Status Dystonicus	Status dystonicus that persists despite attempted drug therapy, and displays one or more life-threatening complications (bulbar weakness, compromised upper airway patency, exhaustion/pain, metabolic imbalances, renal or respiratory failure). Generally, DSS grade 4-5.
Resolved Status Dystonicus	Dystonia that has improved to DSS grade 1 or 2 for > 24 hours.

Major electrolyte abnormalities are defined as hyperkalemia > 5.5 mEq/L.

Renal failure is defined as serum creatinine > 1.5x baseline and urine output < 0.5 mL/kg/hr for 6-12 hours.

Respiratory compromise is defined as the need for respiratory support in the form of CPAP/BiPAP/intubation.

4.0 Dystonia Severity Scale (DSS)^{3, 9,10} ([printable version](#))

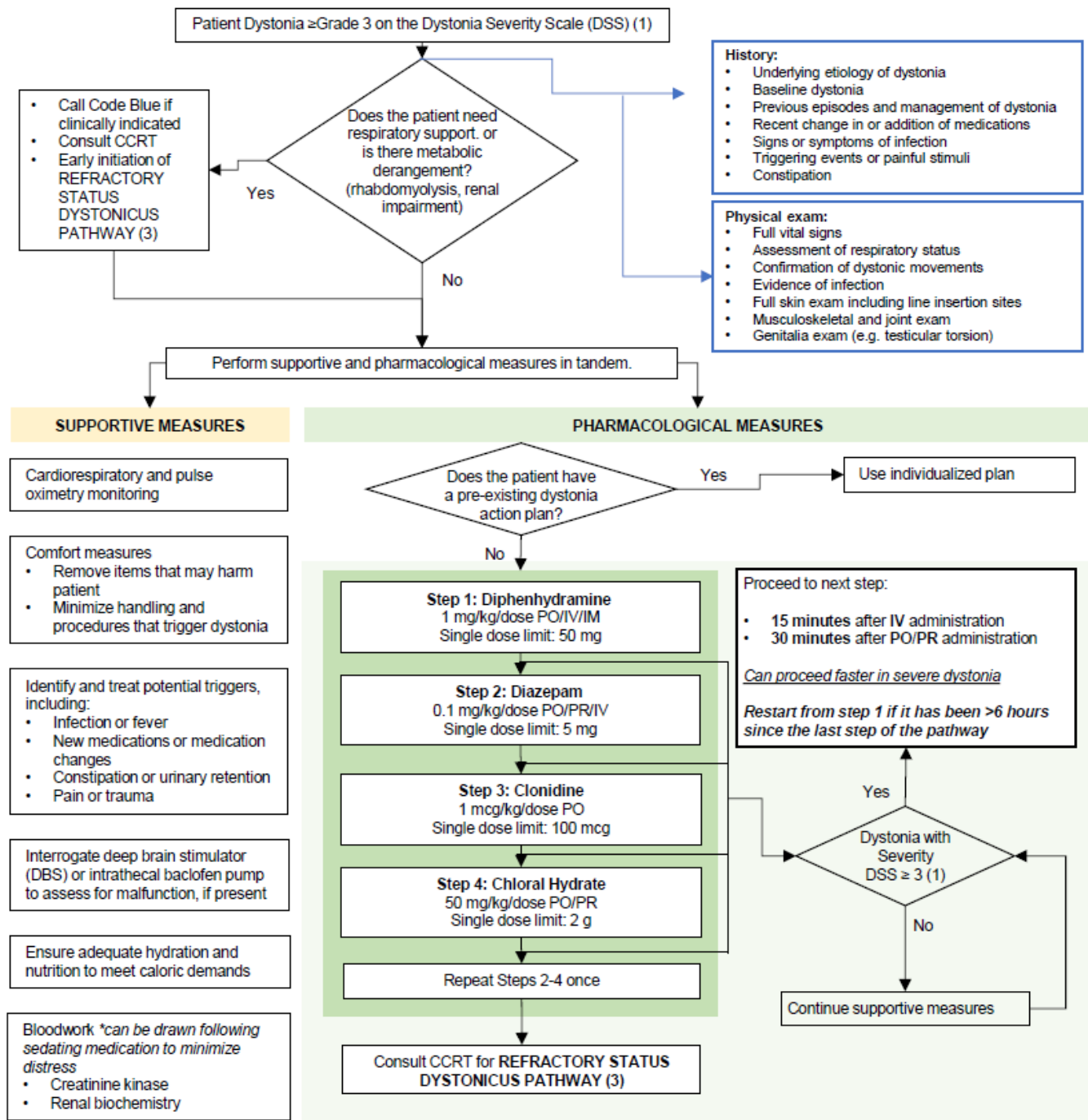
Dystonia Severity Scale (DSS)		Grade	Grade 1	Grade 2	Grade 3	Grade 4	Grade 5
		Features		Patient sits comfortably Regular periods of uninterrupted sleep	Irritable and cannot settle Dystonic posturing interferes with sitting activities or sleep Patient can only tolerate lying still	Not able to tolerate lying still and/or unable to sleep No evidence of metabolic decompensation	Not able to tolerate lying still and/or unable to sleep Early end-organ and metabolic effects Fever Metabolic compromise Myoglobinuria Elevated creatinine kinase > 1000 IU/L
Terms	Controlled Dystonia		Intermittent Dystonia	Pre- Status Dystonicus	Status Dystonicus	Refractory Status Dystonicus	
Urgency of Assessment			PROMPT	URGENT	IMMEDIATE		
Treatment Suggestions	Acute Dystonia Pathway		Consideration of acute dystonia pathway	Implementation of acute dystonia pathway	Transition to Refractory Pathway		
	Refractory Status Dystonicus Pathway			Consideration & preparation for escalation of care	Refractory Status Dystonicus pathway		
	Adjustment of background dystonia medications	Cornerstone of management alongside supportive and temporizing measures					

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5.0 Clinical Practice Recommendations^{9,11} ([printable version](#))

2) ACUTE DYSTONIA PATHWAY

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COMMON QUESTIONS/CONCERNS

What if the PRN medications coincide with regular maintenance medications?

If the patient requires PRN dystonia medications within 1 hour that their maintenance medications are due:

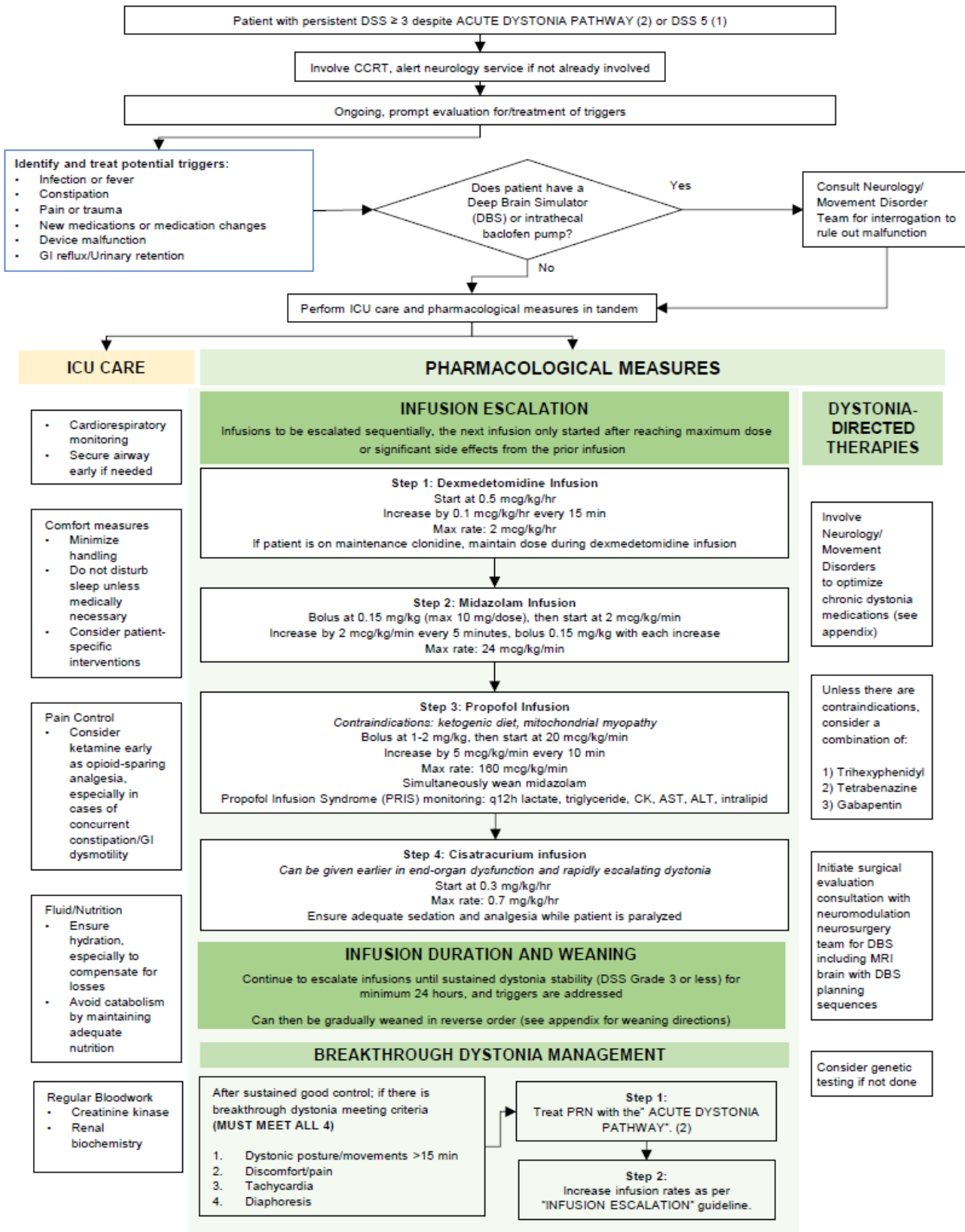
1. Maintenance medications should still be given and can be given up to 1 hour early
2. If the two medications are the same, then please give the higher dose medication first
3. Following this, the other dose can be given if:
 1. The patient continues to be dystonic after 15 minutes, or if they develop dystonia again within 1 hour
 2. After 1 hour regardless of whether the patient has active dystonia
 3. If the patient is excessively sedated after 1 hour, the dose can be held for discussion with MD

When to involve neurology or movement disorders?

- If a patient is requiring the pathway for >2 days in a row
- If a patient proceeds through more than 4 steps of the pathway in 1 day

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3) REFRACTORY STATUS DYSTONICUS PATHWAY



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6.0 Appendix ([printable version](#))^{9,12}

Infusion weaning instructions:

Note: infusion weaning in the setting of status dystonicus is typically slower than weans for other conditions to prevent rebound worsening of dystonia

1. **Midazolam:** Wean infusion by 2mcg/kg/min every 8-12 hours.
2. **Propofol:** Wean infusion by 5-10 mcg/kg/min every 2 hours.
 - Urgent discontinuation for suspected propofol infusion syndrome (bradycardia, metabolic acidosis, rhabdomyolysis, triglycerides greater than 500 mg/dl, or new liver or renal injury)
3. **Dexmedetomidine:** Wean infusion by 0.1-0.2 mcg/kg/hr every 8-12 hours.
 - Consider adding/increasing maintenance clonidine during wean
 - Neurology/Movement Disorders to advise on further clonidine management

Dystonia-directed therapies

To be optimized or initiated in conjunction with Neurology/Movement Disorders (refer to SickKids Formulary for dosing recommendations):

- Baclofen
- Carbidopa/levodopa (4:1 levodopa/carbidopa ratio)
- Clonazepam
- Clonidine
- Diazepam
- Gabapentin
- Tetrabenazine
- Trihexyphenidyl

7.0 Related Documents

AboutKidsHealth Patient Information Sheet: [Dystonia AboutKidsHealth Article](#)

Dystonia Action Plan: Provided by the Movement Disorders Team (Patient Specific Dystonia Action Plan for Providing to families)

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