Early recognition of status dystonicus in children: a case-based approach for the general pediatrician

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Purpose of review
This review highlights the importance of promptly recognizing and correctly naming status dystonicus as a neurologic emergency in the outpatient and inpatient settings, and aims to equip general pediatricians with practical guidance to trigger rapid escalation of care through the recently published status dystonicus pathways.

Recent findings
In 2024, Vogt *et al.* introduced two consensus algorithms – the acute dystonia pathway and the refractory status dystonicus pathway – aimed to unify diagnostic criteria, triage steps, and staged pharmacological and supportive interventions for status dystonicus across healthcare environments.

Summary
Effective application of these pathways empowers frontline clinicians to identify status dystonicus early, initiate first-line treatments without delay, and expedite transfer to specialized teams, thereby reducing the morbidity and mortality associated with this life-threatening movement disorder emergency.

Keywords
dystonia, hyperkinetic movement disorders, pediatric deep brain stimulation, status dystonicus

INTRODUCTION

Clinical vignette

A 6 year-old female with spastic-dystonic cerebral palsy (GMFCS level 5) secondary to preterm birth presents in your pediatrics clinic with fever, increased posturing of the limbs and trunk, and associated irritability. Her home rescue dose of clonazepam has been used 3 times in the past 24 hours with only worsening of increased muscle tone, crying, and poor sleep. Similar prior episodes of increased tone are always resolved with clonazepam rescue doses.

Within pediatric movement disorders, dystonia is a common diagnosis, both in the context of secondary causes, i.e. due to hypoxic or ischemic brain injury, or primary genetic forms. Defined by sustained or intermittent muscle contracture resulting in writhing movements and abnormal postures, our understanding of its etiologies, phenotypes, and therapies has expanded considerably over the past century [1,2]. As demonstrated in our case vignette above, chronic dystonia can escalate into a neurologic emergency known as *status dystonicus*. Status dystonicus is defined as: "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 complication(s) of these movements, regardless of the patient's neurological condition at baseline" [3,4▪]. Recognition of status dystonicus as a neurologic emergency is essential to appropriate management. Similar to prior efforts to streamline clinical recognition and proper naming of clinical emergencies such as sepsis, shock, stroke, and status epilepticus to expedite access to appropriate management, experts in dystonia care advocate for education of all providers to ensure rapid recognition of and intervention for status dystonicus. Management of status dystonicus, has been the subject of prior reviews [1,3,4▪,5]. The development

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KEY POINTS

- Status dystonicus is a medical emergency requiring prompt recognition of worsening dystonia, evaluation for underlying triggers, and management with both supportive care and dystonia targeted therapies.
- Treatment of status dystonicus requires inpatient admission, however stabilization and medical therapy can initiated in the outpatient setting, particularly if there may be delays in accessing inpatient care.
- Inpatient management of status dystonicus requires surveillance of dystonia symptoms, evaluation of patient comfort, and monitoring of the patient's clinical stability as they continue to receive escalating therapies targeting dystonia and sedation.
- For patients with certain genetic etiologies of dystonia or for patients with refractory status dystonicus, deep brain stimulation targeting the globus pallidus internus may prove the most effective therapy.

of the acute dystonia pathway and refractory status dystonicus pathway by Vogt *et al*. in 2024 offers a unified, stepwise approach spanning outpatient triage through intensive care [4▪]. This review targets general pediatricians, who are often the first clinicians to witness the early signs of status dystonicus. We summarize the pathways, underscore practical bedside clues, and illustrate their application through case vignettes that capture the heterogeneity of pediatric dystonia.

BACKGROUND

Dystonia in children can arise from a variety of etiologies. Historically considered a basal ganglia disorder, converging evidence from lesional studies and animal models support the conceptualization of dystonia as a network disorder involving multiple brain regions (basal ganglia, cerebellum, thalamus, cortex, brainstem) [6–8]. Injuries within this circuit can cause acquired dystonia, which can be seen in hypoxic ischemic encephalopathy, intracranial hemorrhage, central nervous system (CNS) infections, neurometabolic, and autoimmune conditions [1]. Genetic forms of dystonias constitute a second major category. Some manifest in childhood as pure progressive dystonia (*TOR1A*), or co-occur with additional movement symptoms, such as parkinsonism (*ATP1A3*), myoclonus, and other dyskinetic movements (*GNAO1*, *KCNMA1*, *ADCY5*) [9–13]. Consideration of the underlying etiology of the patient's dystonia is important, as it can guide assessment, treatment, and prognostication.

Evaluation of dystonia severity can be challenging. Symptoms can fluctuate from focal and relatively mild dystonia to generalized posturing and movements resulting in extreme discomfort and metabolic decompensation [1,4▪]. The development of the Dystonia Severity Scale (DSS) by Lumsden *et al*. in 2013 with further refinement by Vogt *et al*. in 2024 has proven a useful clinical framework to standardize assessment [3,4▪]. In its mildest form (DSS Grade 1), this represents the patient's baseline: Dystonia is generally well controlled and the patient remains comfortable [3,4▪]. As severity progresses, patients can show signs of discomfort and irritability (DSS Grades 2–3) [3,4▪]. In developmentally delayed or medically complex children, an acute worsening of dystonia can be hard to recognize; their discomfort and abnormal posturing are often attributed solely to pain or infection, obscuring the fact that these very stressors may be precipitating their movement disorder exacerbation. Failure to recognize and treat dystonia at this stage can allow escalation to status dystonicus (DSS Grade 4), a state in which continued uncontrolled dystonia has now resulted in end-organ decompensation requiring emergent management of worsening dystonia and metabolic abnormalities [3,4▪]. If initial interventions prove ineffective, refractory status dystonicus (DSS Grade 5) follows, characterized by multiorgan decompensation driven by unremitting dystonia [3,4▪]. Status dystonicus is life-threatening and requires emergent treatment.

Recognition of status dystonicus, identification and management of underlying triggers, and prompt treatment as detailed below are cornerstones to successful management. While the general pediatrician may not be directly involved in ICU level management of status dystonicus, they have a critical role in rapid and accurate recognition of status dystonicus presenting in the outpatient setting (Fig. 1a) and escalation of care (Fig. 2) for affected children to appropriate inpatient facilities and teams. The following cases are meant as illustrative guides for the recognition and management of status dystonicus.

CASE 1

A 10-year-old male with cerebral palsy due to extreme prematurity complicated by severe generalized dystonia (GMFCSV) presents with fever, vomiting, and diarrhea. At baseline, his dystonia is managed with trihexyphenidyl, baclofen, clonidine, and diazepam; for acute exacerbations, he receives as-needed clonidine and diazepam. His parents report that discomfort is usually signaled by vocalizations and grimacing.

Two days prior to admission, he appeared increasingly uncomfortable. The following day, he

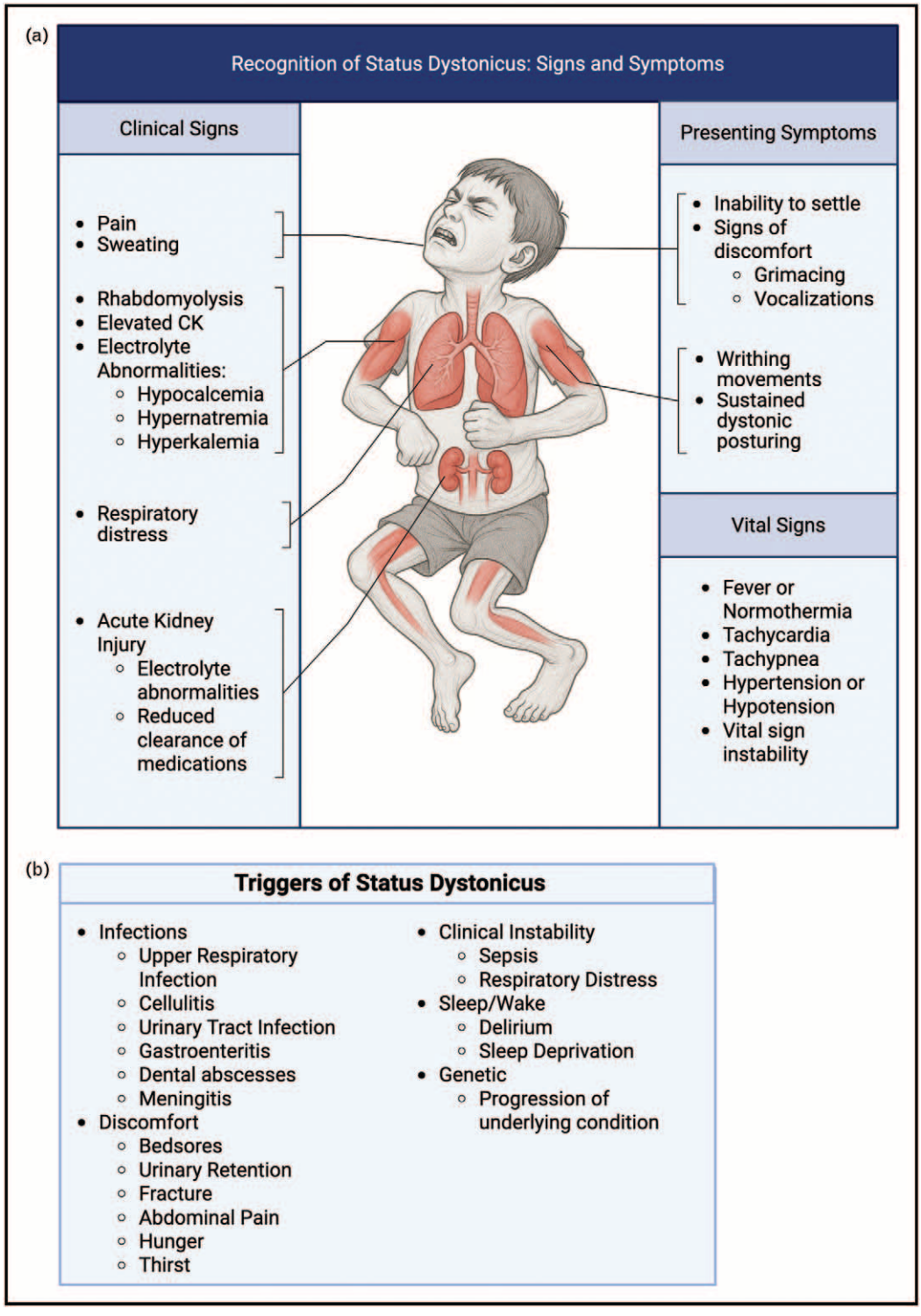

FIGURE 1. (a) Clinical signs and symptoms of status dystonicus. Early recognition of status dystonicus is vital for prompt treatment. (b) Common triggers of status dystonicus. Figures created in BioRender. O'Malley, J. (2025) https://BioRender.com/tdfaxqn. (a) Image generated OpenAI's DALL·E image generation tool for illustrative purposes with prompt "status dystonicus, school aged child, anatomical sketch, medical journal, and overlay muscles, lungs and kidneys".

developed fever, emesis, and profuse diarrhea. At his pediatrician's office, he was found to be febrile, tachycardic, and tachypneic with an oxygen saturation of 95% on room air. On exam, he exhibited clear signs of distress, including leftward head deviation, flexion posturing of the limbs, intermittent opisthotonic posturing, and increased vocalizations, particularly during bowel movements. Labs are notable for

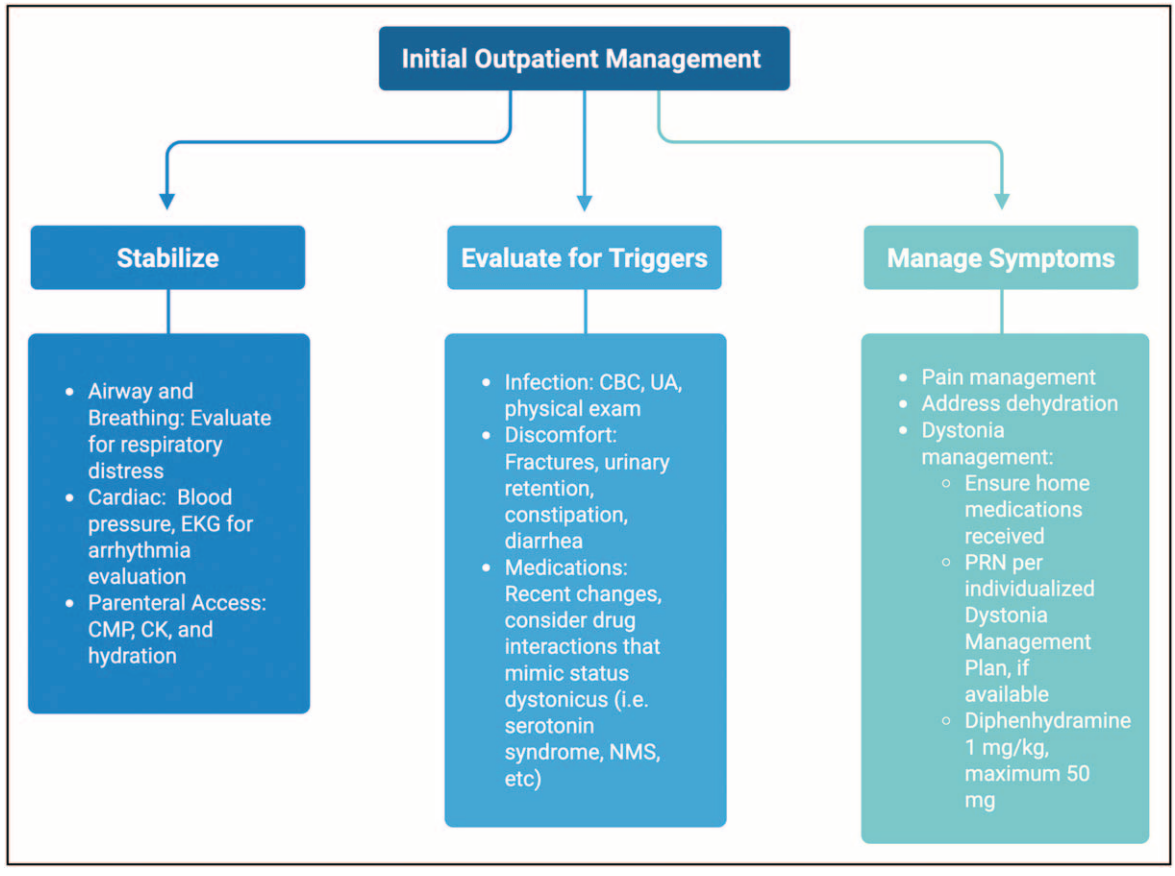

FIGURE 2. Initial outpatient management of status dystonicus. If there is concern for status dystonicus in the outpatient setting, the focus should be on stabilization and transfer to inpatient management. Initial evaluation for cardio-respiratory stability then prioritizing access and dystonia management. If the patient has an individualized dystonia management plan, administration of recommended PRN medications is advised. If no individualized plan is available, diphenhydramine is a reasonable first line PRN that is often accessible in the outpatient setting. Management of pain and dehydration should also be addressed.

leukocytosis, mild hypernatremia, elevated creatinine, and a positive norovirus test. He is admitted to the acute care unit for management of presumed viral gastroenteritis with dehydration and started on intravenous fluids while continuing his home medications.

Despite supportive care and as-needed clonidine and diazepam doses for dystonia, his discomfort persists over the next 48 h, with severely disrupted sleep. Repeat labs reveal worsening leukocytosis and increasing creatinine. Urinalysis shows pyuria and positive nitrites, raising concern for a urinary tract infection, and antibiotics are initiated. However, over the following days, his symptoms progress: He remains unable to sleep, with persistent dystonic posturing of the trunk and limbs, frequent writhing arm movements, tachycardia, diaphoresis and distress. A serum creatine kinase level, drawn for the first time, is elevated to 2,500 IU/l. He is diagnosed with status dystonicus.

CASE 1 DISCUSSION

Status dystonicus is frequently precipitated by identifiable triggers such as infection, pain, or medication changes. In this case, the patient's progression from intermittent dystonia to status dystonicus was triggered by a combination of viral gastroenteritis, dehydration, and a urinary tract infection. However, the contribution of dystonia itself to the patient's worsening renal function and electrolyte disturbances – hallmarks of end-organ involvement – was not initially recognized, resulting in a delay in the escalation of dystonia-directed management.

This case highlights the clinical progression from prestatus dystonicus to status dystonicus: a state of sustained, severe dystonia associated with systemic complications such as rhabdomyolysis and acute kidney injury. Recognition of status dystonicus can be challenging, particularly when the physical exam is confounded by mixed motor features – spasticity,

rigidity, or other hyperkinetic movements. The differential diagnosis for status dystonicus should also include other hypertonic emergencies, such as neuroleptic malignant syndrome, serotonin syndrome, malignant hyperthermia, and medication-induced dyskinesias, depending on the clinical context and medication history.

Prompt recognition of worsening dystonia can occur in both outpatient and inpatient settings. Figure 1a and 1b outlines common symptoms and triggers that warrant evaluation for status dystonicus. Although hospitalization is warranted for patients in status dystonicus, there are critical diagnostic and therapeutic steps that can and should be initiated by outpatient providers when encountering patients in prestatus dystonicus or status dystonicus (see Fig. 2). The first step in any acute evaluation is stabilization using the ABC framework: assessment of airway, breathing, and circulation [14]. Additional early workup should include establishment of IV access, EKG, and laboratory studies including serum electrolytes, renal and hepatic panels, creatine kinase (CK), and infectious markers. CK is of particular use as a biomarker; a recent retrospective study of patients with dystonia showed elevation in CK levels when diagnosed with status dystonicus as compared to their prestatus dystonicus baseline [15▪].

Common precipitants of status dystonicus include infection, discomfort, dehydration, or recent changes in medications. Initial management should address these potential triggers, including empiric treatment of pain, constipation, urinary retention, or other sources of distress identified on physical exam. Dystonia-targeted treatment should include both resumption of home maintenance medications and administration of PRN medications, ideally guided by a personalized dystonia action plan. When such a plan is unavailable, empiric treatment can include diphenhydramine 1 mg/kg (max 50 mg), clonidine 0.1–0.2 mcg/kg (max 100 mcg), or diazepam 0.1 mg/kg (max 10 mg), followed by immediate transfer to a pediatric emergency department.

In the inpatient setting, recognition of status dystonicus should prompt activation of a structured management algorithm such as the Acute Dystonia Pathway (Fig. 3a). In our patient, there were no signs of respiratory compromise. Initial management included IV diphenhydramine 1 mg/kg; when dystonia persisted, he received enteral clonidine 1 mcg/kg, followed by diazepam 0.1 mg/kg. This resulted in modest improvement, though writhing movements and grimacing persisted. A repeat dose of clonidine (0.2 mcg/kg via G-tube) was given, leading to further improvement and restoration of sleep. Given limited availability of chloral hydrate in the United States, it was not administered to our patient [16].

Supportive measures were implemented in parallel with pharmacologic treatment. Interventions included minimizing stimulation, consolidating care activities to promote uninterrupted rest, initiating dextrose-containing IV fluids to mitigate catabolic stress, and administering scheduled acetaminophen for presumed pain due to gastroenteritis and UTI.

Following stabilization, his maintenance regimen was modified: standing doses of clonidine and diazepam were increased, while trihexyphenidyl and baclofen were continued at baseline doses due to concerns about gastrointestinal side effects. Over the subsequent days, his gastrointestinal symptoms resolved, and the UTI was effectively treated. He was discharged with a plan to gradually taper back to his baseline home regimen.

CASE 2

A 10-year-old female with GNAO1-related disorder – characterized by intellectual disability, epilepsy, and generalized chorea – presented to her pediatrician with a 1-week history of worsening generalized chorea and dystonic episodes. At baseline, she is ambulatory with assistance and communicates using an augmentative and alternative communication (AAC) device. Her dyskinesias and dystonia have shown partial response to trihexyphenidyl. She also has well controlled epilepsy on levetiracetam.

Over the week prior to presentation, her parents observed a marked increase in her usual dyskinetic movements, progressing to near-constant generalized chorea and intermittent dystonic postures of the neck, trunk and arms. For the preceding three days, she had been unable to sleep or tolerate solid foods. On evaluation, she is febrile, tachycardic, tachypneic, and mildly hypertensive, with an oxygen saturation of 98% on room air. Examination reveals continuous involuntary movements of all extremities and facial muscles, with signs of discomfort. Laboratory studies showed modest leukocytosis and a creatine kinase of 1900 IU/l.

She is diagnosed with status dystonicus and urgently transferred to a pediatric emergency department, where the Acute Dystonia Pathway was initiated. She receives diphenhydramine, diazepam, and chloral hydrate with partial improvement, though she remained unable to rest. A second cycle of the pathway is administered with similar results. Given persistent symptoms and emerging end-organ involvement, she is transferred to the pediatric intensive care unit for escalation to the refractory status dystonicus pathway.

In the ICU, she is started on a dexmedetomidine infusion, titrated to the maximum dose of 2 mcg/kg/h, and her maintenance dystonia regimen is

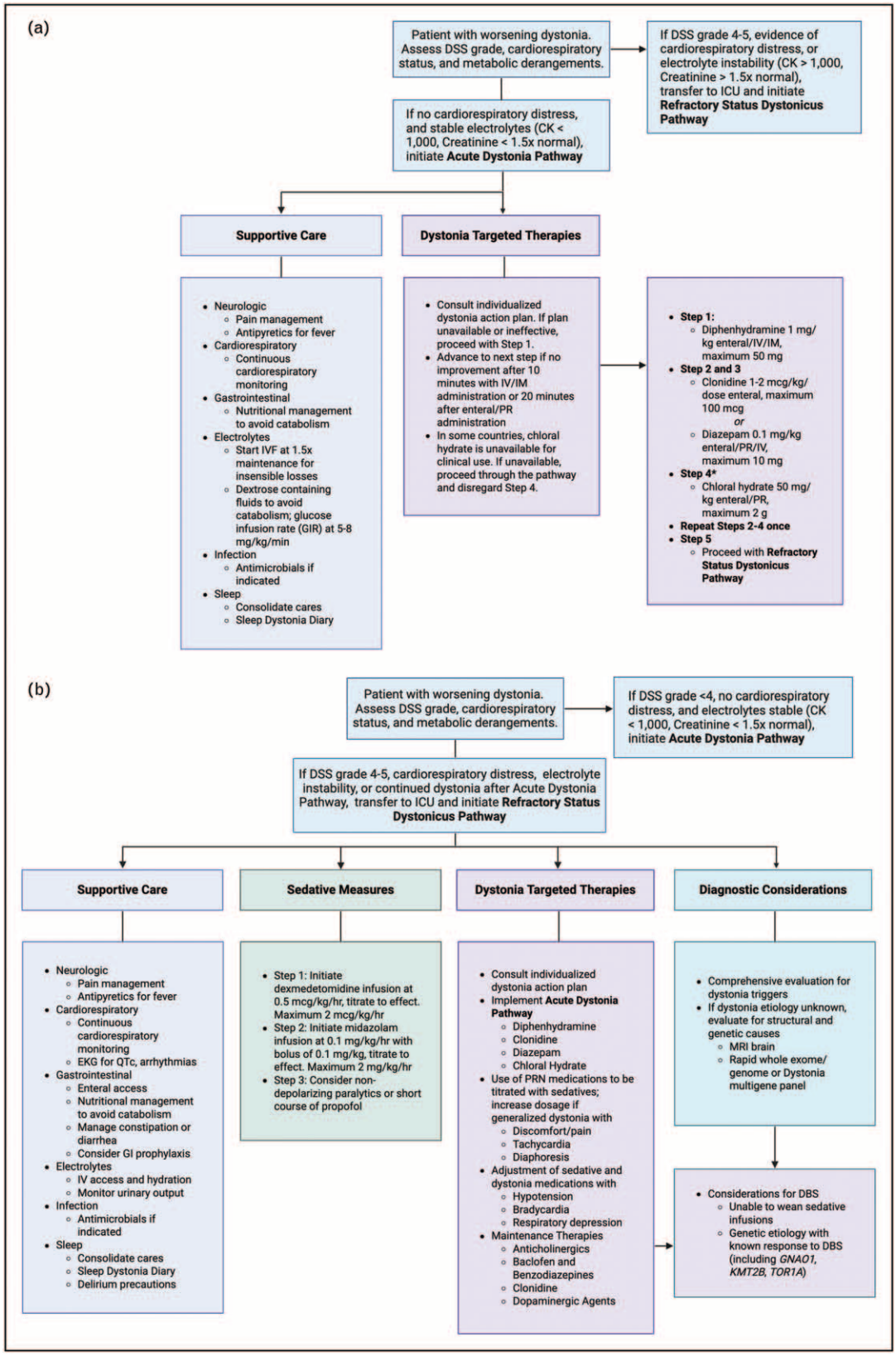

FIGURE 3. (a) Acute dystonia pathway. This figure is reproduced from Vogt *et al.* with modifications to supportive care for emphasis of general management [4▪]. *Chloral hydrate is no longer commercially available in the United States, although there may be limited availability via compounding pharmacies [16]. (b) Refractory status dystonicus pathway, adapted from Vogt *et al.* with modifications to supportive care for emphasis of general management and sedation [4▪].

intensified. Trihexyphenidyl is up-titrated, and with enteral access established, baclofen and clonazepam are added. Supportive measures included continuous cardiorespiratory monitoring, initiation of slow nasogastric feeds alongside parenteral nutrition, and implementation of a Sleep–Wake Dystonia Diary to monitor rest cycles and guide sedation.

Despite these interventions, her dystonia and chorea persisted over the following week, with ongoing elevation of CK. She is started on scheduled gabapentin and tetrabenazine, and later transitioned to a midazolam infusion with subsequent intubation for airway protection. Sedation resulted in clinical comfort and a downward trend in CK. However, attempts to taper sedative infusions led to immediate recurrence of dystonic and choreiform movements.

A comprehensive re-evaluation for triggers - including infection, metabolic derangements, and occult pain - is unrevealing. With no reversible cause identified and inability to wean from continuous sedation, the decision is made, in consensus with the family, to proceed with bilateral globus pallidus interna deep brain stimulation implantation.

CASE 2 DISCUSSION

Refractory status dystonicus (SD) is a medical emergency that often requires a combination of deep sedation and escalation of dystonia-targeted therapies. Alpha-2 adrenergic agonists (e.g., clonidine, dexmedetomidine) and benzodiazepines (e.g., diazepam, midazolam) are foundational agents for both dystonia management and sedation (Fig. 3b). When administered as continuous infusions, these agents can be titrated to achieve clinical comfort and suppress dystonic movements. In more severe cases, escalation to neuromuscular blockade or short-term propofol infusions may be necessary to achieve cessation of dystonia and rhabdomyolysis. As always a safe airway, respiratory support and appropriate monitoring of cardiorespiratory function is essential.

In some patients, sedation cannot be safely weaned without recurrence of severe dystonia, necessitating consideration of surgical intervention. Deep brain stimulation (DBS) has emerged as a life-saving therapy for children with refractory SD. First reported in 1999 in an 8-year-old girl with sustained benefit, DBS has since been used in dozens of pediatric cases with various underlying etiologies, including genetic, neurodegenerative, metabolic, and acquired causes [17▪▪,18,19]. The globus pallidus internus (GPi) is the most common target, though the subthalamic nucleus has also been utilized in select cases. Among monogenic causes of pediatric dystonia, *GNAO1*-related disorder is one of the most frequently reported etiologies associated with positive response to DBS for refractory SD [17▪▪]. In our patient, following bilateral GPi DBS implantation, stimulation was initiated intraoperatively. Over the ensuing weeks, she was successfully weaned from continuous sedative infusions and extubated. At discharge, she remained on a regimen of trihexyphenidyl, baclofen, clonidine, clonazepam, and gabapentin. With subsequent DBS programming adjustments, these medications were gradually tapered, and her motor symptoms remained stable.

GENERAL CONSIDERATIONS ABOUT THE MANAGEMENT OF STATUS DYSTONICUS

The cornerstone of effective treatment for status dystonicus is *timely recognition and clear communication* of the diagnosis. All subsequent management hinges on shared situational awareness among providers. Figure 4 provides a sample communication script using the "SBAR" format (Situation, Background, Assessment, Recommendation), which can be used to convey concerns about a patient's worsening motor status, outline relevant background (including dystonia etiology and baseline function), summarize the clinical assessment, and recommend urgent next steps.

Management of status dystonicus requires a multimodal, interdisciplinary approach that accounts for the underlying etiology, precipitating factors, and medication side effects. Standardization of care through structured pathways enhances quality and safety. We recommend using the "ABCD" mnemonic as a guiding framework: Address triggers, Begin supportive care, Calibrate sedation, and Deliver dystonia-specific therapies [1,4▪,20▪]. While algorithmic, these pathways must be individualized and adapted to each patient's clinical scenario.

Initial steps across care settings should include rapid assessment of dystonia severity using tools like the DSS, alongside evaluation for acute complications such as respiratory distress, rhabdomyolysis, or metabolic derangements. Identifying triggers is critical: infection, pain, medication changes, and discomfort are among the most common precipitants, identified in over 50% of pediatric cases [21–23]. Many children with status dystonicus are nonverbal or medically complex, making a thorough and systematic evaluation essential.

Supportive care should follow immediately. This includes consolidating nursing care to promote rest, initiating hydration and nutrition (via enteral or parenteral means), controlling hyperthermia, and addressing pain. These interventions are not unique to status dystonicus, but their implementation is particularly important, as untreated stressors can perpetuate the dystonia-pain cycle and contribute to clinical deterioration.

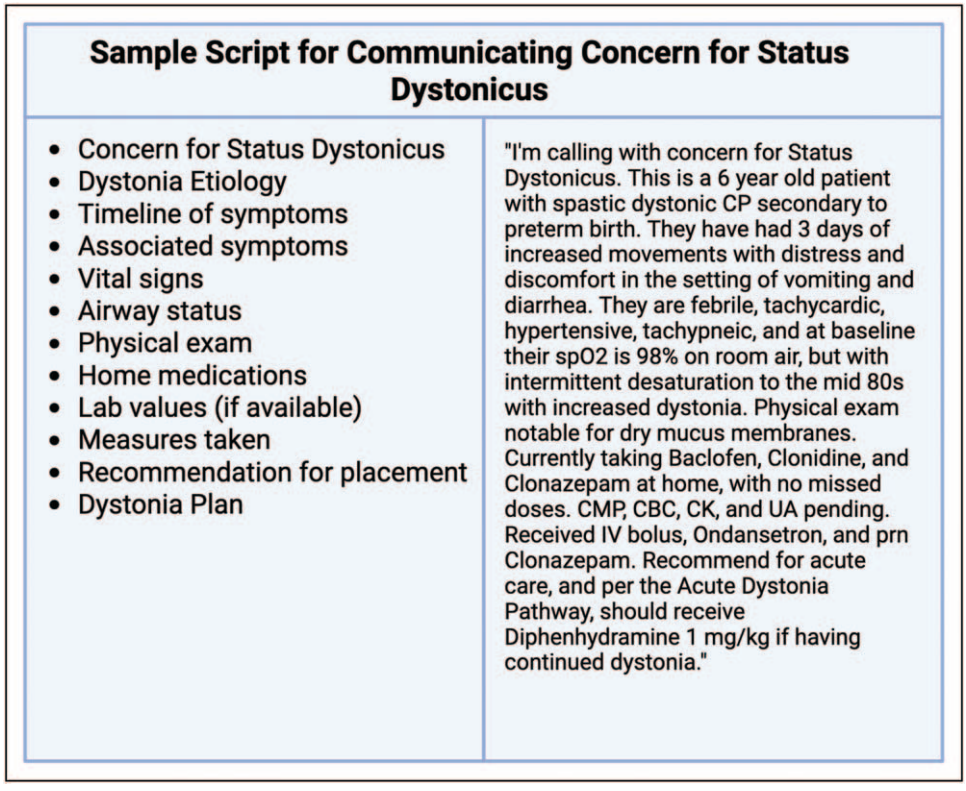

FIGURE 4. Sample script for communicating concern for status dystonicus. Utilizing an SBAR (situation, background, assessment, recommendation) format, this script emphasizes patient status, evaluation of dystonia etiology and triggers, and next steps for management.

Next, targeted pharmacologic treatment should be initiated. As outlined in the Acute dystonia pathway proposed by Vogt *et al.* [4▪], escalation typically follows a stepwise approach beginning with diphenhydramine and progressing to clonidine, diazepam, and, where available, chloral hydrate. The sequence is designed to achieve symptom control while minimizing sedation. Medication selection should be informed by patient-specific factors such as hemodynamic stability, enteral access, and prior response.

Patients with baseline dystonia often have individualized medication regimens that can be remembered using a second "ABCD" mnemonic: Anticholinergics (e.g., trihexyphenidyl), Baclofen and benzodiazepines, Clonidine and clonazepam, and Dopamine-targeted therapies (including both dopaminergic and dopamine-depleting agents) [4▪,5]. Optimizing these maintenance therapies in parallel with acute interventions may improve outcomes.

Persistent DSS Grade 4 or 5 despite acute management signals refractory status dystonicus, necessitating escalation to intensive care. ICU-level supportive measures should include continuous monitoring, enteral access, nutritional support, and structured assessments of dystonia severity. Sleep–wake patterns should be tracked using a Sleep–Wake Dystonia Diary, which facilitates correlation with medication timing and vital signs and can be integrated into the electronic health record [4▪].

Management of refractory status dystonicus may require deep sedation, including dexmedetomidine, midazolam, and, when necessary, short-term propofol infusions or neuromuscular blockade. Escalation of standing dystonia medications should occur in parallel to facilitate eventual weaning of sedatives. Adjunct therapies such as botulinum toxin should also be considered, especially if focal dystonia contributes to pain or functional impairment [4▪,24]. Hospital admission may disrupt outpatient botulinum injection schedules, so re-administration should be prioritized when appropriate.

If patients remain dependent on continuous sedation and no reversible trigger is identified, surgical interventions should be considered. Deep brain stimulation for refractory status dystonicus requires presurgical imaging to assess suitability for implantation, typically targeting the globus pallidus internus or, less commonly, the subthalamic nucleus. Contraindications include active CNS infection, unstable vital signs, or contraindications to anesthesia [17▪▪]. For patients who may benefit from DBS but are

contraindicated from implanted hardware, magnetic resonance guided focused ultrasound (MRgFUS) pallidotomy may be an option [25].

CONCLUSION

Dystonia exists along a continuum of severity, with status dystonicus representing the most severe and life-threatening manifestation. If left untreated, status dystonicus can result in multiorgan failure and death. Prompt recognition and diagnosis are therefore paramount. Recent consensus guidelines – including the acute dystonia pathway and the refractory status dystonicus pathway – offer structured frameworks to guide clinicians. However, these should be applied with careful attention to the individual patient's etiology, baseline function, comorbidities, and treatment history.

Effective use of these pathways depends on early diagnosis and collaborative care, beginning with outpatient providers and pediatricians. These clinicians play a vital role in identifying early warning signs, initiating rescue treatments, and activating appropriate referral pathways. A standardized, team-based approach is essential to improving outcomes for children with this complex and often under-recognized neurologic emergency.

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Conflicts of interest

There are no conflicts of interest.

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- ■ of special interest
- ■■ of outstanding interest

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