

Status Dystonicus

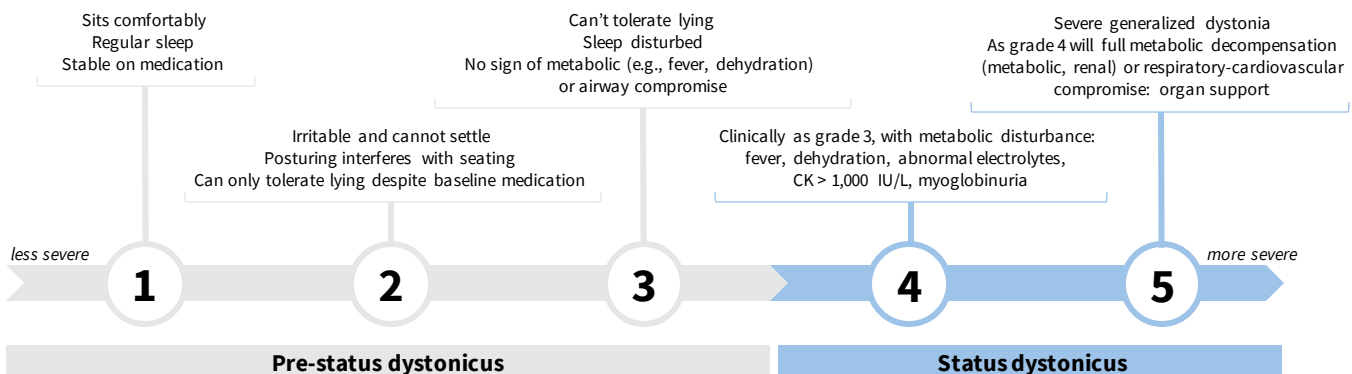
A quick reference guide for recognition and management

What is status dystonicus?

- **Status dystonicus** (or 'dystonic storm') is a medical emergency involving increased frequency or continuous episodes of severe generalized dystonic spasms.¹
- Status dystonicus typically arises in individuals with an underlying dystonia diagnosis, including individuals with **cerebral palsy**.^{1,2}
- Status dystonicus can be difficult to recognize and can lead to life-threatening complications, including respiratory compromise and severe metabolic disturbances (e.g., acute renal failure) requiring **immediate medical attention**.³
- **Management** requires hospital admission and can be both challenging and resource-intensive.³

How to identify & manage status dystonicus

- The **Dystonia Severity Action Plan (DSAP)**^{3,4} is a simple, 5-grade scoring system for grading the medical severity of dystonia, facilitating early recognition of **status dystonicus** and helping to guide management decisions
- The scale is based on **characteristic clinical indicators** of status dystonicus (e.g., irritability, sleep disruption, seating intolerance, metabolic disturbance, respiratory compromise)



- Establish a **personalized plan** for the family to implement at the first sign of worsening dystonia (e.g., pm doses of chloral hydrate, clonidine, benzodiazepines)
- At **grade 2**, assess and consider adjusting medication or dystonia plan
- At **grade 3**, conduct urgent assessment to exclude metabolic decompensation (e.g., CK) and escalate management

Supportive care

- Admit to the hospital and assess respiratory (e.g., O₂ sats/blood gases) and metabolic status (e.g., renal function, electrolyte, CPK, myoglobinuria, acidosis). ICU input is often required and intubation/ventilation needs to be considered.
- Introduce IV hydration, antipyretics
- Assess for 'triggers' (e.g., underlying infection, medication withdrawal, pain)

Temporizing measures

- Encourage sleep without compromising breathing patterns (e.g., chloral hydrate, clonidine, dexmedetomidine)
- Consider increasing or introducing other dystonia medications

Dystonia-specific management

- Consider increasing other dystonia medication(s)
- Consider consultation regarding intrathecal baclofen (ITB) or deep brain stimulation (DBS)

Resources



Learn more and consult the DSAP (Lumsden, Lundy, Fairhurst & Lin, 2013) [here](#)



For a complete practical approach to screening and management, consult a practice guide (Allen, Lin, Lynch & King, 2014) [here](#)