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).

### Pre-status dystonicus
- 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.

### Status dystonicus

#### More severe
- Severe generalized dystonia
- As grade 4 will full metabolic decompensation (metabolic, renal) or respiratory-cardiovascular compromise: organ support

#### Less severe
- Sits comfortably Stable on medication
- Irritable and cannot settle Posturing interferes with seating Can only tolerate lying despite baseline medication
- Can’t tolerate lying Sleep disturbed No sign of metabolic (e.g., fever, dehydration) or airway compromise
- Clinically as grade 3, with metabolic disturbance: fever, dehydration, abnormal electrolytes, CK > 1,000 IU/L, myoglobinuria

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](#)