Dystonia Severity Action Plan: a simple grading system for medical severity of status dystonicus and life-threatening dystonia

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SIR–Dystonia is the most common movement disorder seen in childhood[1] and may be defined as 'a movement disorder in which involuntary sustained or intermittent muscle contractions cause twisting and repetitive movements, abnormal postures, or both.'[2] Dystonia may arise from a range of pathologies, and impairs quality of life, participation, and function, in addition to causing pain and progressive deformity.

Children and young people with dystonia may experience acute deterioration in their dyskinesia, variously termed 'status dystonicus' or 'dystonic storm', which has been broadly defined as 'generalized, intense and potentially fatal exacerbation of muscle contractions.'[3] This definition is not universally used though, and the precise delineation of the point at which status dystonicus is reached is far less objective than, for example, in status epilepticus. It is important to remember that these acute episodes actually represent the severest end of a continuum of worsening dystonia. Early recognition may potentially facilitate intervention and prevent progression. In a recently reported large retrospective case series of 89 episodes of status dystonicus[4] (58.8% occurring in children <15y) first-line pharmacological treatments of established status dystonicus (most commonly anticholinergics and tetrabenazine) were effective in ~10% of cases. Treatment in this case series frequently necessitated deep sedation in intensive care and there carried a mortality rate of 10.3%.

Numerous scales exist to measure the severity of dystonia (e.g. the Burke Fahn Marsden Dystonia Rating Scale, Barry-Albright Dystonia scale). The limitation of these scales include their time-consuming nature, need for considerable training for use, a plateau affect which limits their utility in the most severely affected cases, and also concerns about reliability and smallest detectable difference, particularly in the secondary dystonias which are most commonly seen in childhood.[5] The recently described Dyskinesia Impairment Scale holds great promise, but the length of time taken to record and score video assessments may restrict its use to the research setting.[6]

The need exists for a pragmatic criterion-referenced clinical scoring system which can be rapidly and reliably applied and which may be understood by parents and all healthcare providers, not just those professionals with an expertise in movement disorders. It is important that such a scale includes graduations up to the point at which a child or young person can be said to have entered status dystonicus, to alert clinicians to a gradual worsening of dystonia and the need for effective intervention. A common agreed nomenclature when describing exacerbations of dystonia is also necessary if clinical trials are to be developed to best guide treatment methodologies.

Between our centres we have devised a simple scoring system (shown in Table I) in an effort to meet this need. The system has been devised to provide simple objective clinical criteria against which the severity of a child's dystonia may be rated and action taken. This system has been specifically designed to be used as a tool by all health care professionals involved in the clinical care of children with dystonia, and not just the movement disorder specialist. To determine the utility of our scoring system, 10 clinical vignettes (Appendix S1; online supporting information) based on real cases managed at our centre, were provided to a mixed group of 30 health care professionals, including nursing staff, allied health professionals, and paediatricians of different grades and specialities. Each participant could score all 10 cases, with average intraclass correlation scores of 0.993 ($p<0.001$) indicating a high level of intrarater reliability. Furthermore, all 30 participants thought that the system would be of use during their routine clinical practice.

Carers, health professionals, paediatricians, neurologists, and neurodisability consultants need to communicate effectively when managing children with worsening dystonia. This simple scale based on commonly seen clinical indicators could help plan treatment and guide where care should be delivered, for example in the ward, high-dependency, or intensive care setting. It may help focus targeted intervention and develop a picture of the child's overall disease trajectory over time while allowing measurement of the health costs of brittle, unstable dystonia, and status dystonicus.

SUPPORTING INFORMATION

The following additional material may be found online:

Appendix S1: Dystonia Severity Action Plan (DSAP) clinical scenarios.

Table I: Dystonia Severity Action Plan

Severity	Assessment	Plan
Grade 1: The child sits comfortably and has regular periods of uninterrupted sleep. Child stable on medication.	No assessment needed	Continue at home or going to school as usual Discharge home if recovering from acute decompensation.
Grade 2: The child is irritable and cannot settle. Dystonic posturing interferes with sitting activities The child can only tolerate lying despite usual baseline medication.	Seek advice from local team including fresh assessment within next few days.	Adjust medication as pre-planned or initiate new plan.
Grade 3: Not able to tolerate lying and/ or unable to get to sleep or sleep disturbed No evidence of metabolic decompensation, with creatinine kinase <1000 IU/L.	No response to adjusted medication? Urgent assessment/review required	Further adjustments to medication Metabolic screen required to look for signs of decompensation; Observe in hospital if indicated; be prepared to escalate management.
Grade 4: Early multi-organ failure: Clinically as above with: Pyrexia (in absence of infection) Evidence of metabolic compromise (e.g. acidosis, elevated potassium, low calcium, evidence of rising creatinine and/or urea) Evidence of myoglobinuria, creatinine kinase >1000 IU/L.	This is an emergency, urgent hospital admission required for multisystem support and attempt to prevent frank renal failure and disseminated intravascular coagulation	Measure creatinine kinase levels monitored regularly along with urea and electrolytes, renal and liver function Nasogastric or gastrostomy or rectal medication if tolerated and working IV. Medication.
Grade 5: Immediate life-threatening: As above with: Full metabolic decompensation Respiratory, cardiovascular or renal compromise Requires intensive care.	Child needs HDU/ PICU	Consider need for : IV Infusion of clonidine[a] and/or midazolam Dialysis/haemofiltration ventilation – specifically if spasms causing airway compromise, bulbar dysfunction with secretions compromising airway, evidence of respiratory failure or impending exhaustion Liver support Intrathecal baclofen infusion Deep brain stimulation.

[a]Non-respiratory depressant.

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