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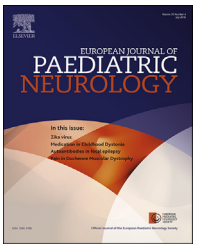

Case Study

Status dystonicus in children: Early recognition and treatment prevent serious complications

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ABSTRACT

This is a retrospective study of all patients presenting to our paediatric unit with status dystonicus (SD) over a period of five years. Anonymous information was collected and a descriptive analysis is made. There were four episodes of SD in three children between 11 and 15 years of age. All children are known to have severe dyskinetic cerebral palsy and presented with an acute or sub-acute deterioration in their symptoms. Symptoms were triggered by infections in three of the four episodes. Early features included frequent and repetitive generalized muscle spasms, poor swallowing, poor sleep, distress and pain. Patients responded to supportive treatment, rehydration, benzodiazepines, baclofen and L-dopa. Intensive care was not necessary in any of the patients and patients made full recovery within 5–14 days. This report shows the value of early recognition and treatment of SD can be successful in preventing serious complications.

1. Introduction

A recent consensus report defined dystonia as, 'a movement disorder characterized by sustained or intermittent muscle contraction causing abnormal, often repetitive, movements, postures or both. These movements are typically patterned, twisting and may be tremulous.[1] Dystonia has numerous causes but cerebral palsy is probably the most common in children, brought on by injury to the basal ganglia, thalamus, brainstem, and cerebellum from hypoxic-ischemia in the perinatal period or during infancy. Other causes include encephalitis, vascular diseases, autoimmune disorders, cerebral malformations, metabolic disease, and neurodegenerative disorders.[2] Primary dystonia in children (mostly genetic) is relatively rare. L-dopa plays an important role in muscle tone management of children and adolescents with primary dystonia and Baclofen is the most commonly used drug in the treatment for children with secondary dystonia. There are no randomised controlled trials and most drugs are used on the basis of open studies.

Status dystonicus (SD) is considered rare, but is a serious and potentially life threatening, condition. SD is defined as 'increasingly frequent and severe episodes of generalized dystonia requiring urgent hospital admission.[3] Complications include respiratory failure, which can be due to one, or a combination, of muscle spasm, exhaustion, aspiration pneumonia or the heavy sedation used in managing the condition.[4] Associated muscle spasms can also lead to rhabdomyolysis, acute renal failure, hyperpyrexia and dehydration.[4]

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This definition of SD may be appropriate for severe forms of the disease, but for many patients, may be imprecise and lead to under-recognition and under-treatment.[5] A recent publication introduced a "dystonia severity action plan" that allows a comprehensive assessment of symptoms and disease progression in order to provide treatment appropriate to different grades of deterioration and to prevent disease progression.[6]

In this report we describe the presentation, recognition and successful early treatment of four episodes of SD in three patients.

2. Patients and methods

This is a retrospective study of all children with SD who were admitted to Forth Valley Royal Hospital over a five-year period between 2010 and 2015. Children were known to one of us (IAA) and cases were confirmed by reviewing hospital records. The diagnosis and grading of SD were based on the clinical criteria in Table 1.[6] The project is registered as a service evaluation within NHS Forth Valley and ethical approval was not necessary.

3. Results

Four episodes of SD were identified in three children over 5 years with an average of 0.8 episodes per year. Mean age at presentation was 13 years (11–15). All children had severe pre-existing dyskinetic cerebral palsy. The details of case presentations are given below and summary of main features are presented in Table 2.

3.1. *Case A*

This 12 year-old boy has a background of dyskinetic athetoid cerebral palsy, hearing impairment, global developmental

Table 1 – Dystonia severity assessment.[6]

Grade	Clinical description
Grade 1	The child sits comfortably and has regular periods of uninterrupted sleep. Child stable on medication
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.
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
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.
Grade 5:	Immediate life-threatening. As above with: full metabolic decompensation, respiratory, cardiovascular or renal compromise, requires intensive care

Adapted from Lumsden et al. (2013).

Table 2 – Summary of the main features of 3 cases (4 episodes) of SD.

Feature	Case A	Case B- 1	Case B- 2	Case C
Gender	M	M	M	F
Age (years)	12	11	15	14
Weight (kg)	36	25.8	31.6	26.5
Dyskinetic cerebral palsy	Yes	Yes	Yes	Yes
GMFCS	5	5	5	5
SD Grade	3	4–5	4–5	3–4
Precipitating factors	Chest infection, constipation	Fever, anorexia, vomiting	Urine infection, gastroenteritis	None
Days of prodrome	3	3	3	21
Medications on admission	Baclofen 0.6 mg/kg/day	L-dopa 9 mg/kg/day, ITB 17 mcg/kg/day, Midazolam 2 mg PRN, Baclofen 5 mg PRN	L-dopa 7 mg/kg/day ITB 27 mcg/kg/day	Chloral hydrate 19 mg/kg/day
Supportive treatment	Rehydration, Pain relief	IV fluids, co-amoxyclav,	NGT Rehydration solution	Oral rehydration fluids
Acute treatment of SD	IV then oral Diazepam 5 mg	Added oral Baclofen 5 –10 mg Oral diazepam 5 mg	IV then oral Diazepam 5 mg, three times/day	L-dopa in increasing doses
Duration of SD (days)	12	14	8	5
Medications on discharge	Diazepam 5mgtwice/day L-dopa 5.2 mg/kg/day	Diazepam 2 mg twice/day Oral Baclofen 1.2 mg/kg/day Midazolam PRN Review ITB	L-dopa 75 mg three times/day, Diazepam 5 mg three times/day, Review of ITB	Chloral hydrate 500 mg/day; 19 mg/kg/day L-dopa 62.5 mg four times/day; 9.4 mg/kg/day

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