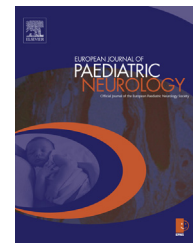




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## Original article

# Interventional studies in childhood dystonia do not address the concerns of children and their carers



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## ABSTRACT

**Aims:** This study aimed to determine the main concerns/priorities of the parents and carers of children with dystonia referred to our service and whether medical interventional studies addressed these concerns.

**Methods:** Records of children assessed by our service from June 2005–December 2012 were reviewed and expressed parental/carer concerns at initial assessment categorized using the International Classification of Functioning (ICF) Framework. Medline, CINAHL and Embase databases were searched for outcome measures of medical and surgical interventional studies in childhood dystonia.

**Results:** Data was collected from 273 children and young people with dystonia. The most commonly expressed concerns were: pain (104/273, 38.1%); difficulties in delivering activities of daily-living (66/273, 24.2%), difficulties with hand-use (59/273, 21.6%) and seating (41/273, 15.0%). Literature review identified 70 interventional studies, 46 neurosurgical and 24 pharmacological. The majority of neurosurgical studies (34/46) used impairment scales to measure change, with pharmacological studies typically reporting more subjective changes in motor symptoms. Only a minority of studies used assessments or scales capable of objectively addressing the concerns reported by our cohort.

**Interpretations:** Existing interventional studies in childhood dystonia poorly address the main concerns of children with dystonia and their carers, limiting the conclusions which may be drawn as to true impact of these interventions in childhood.

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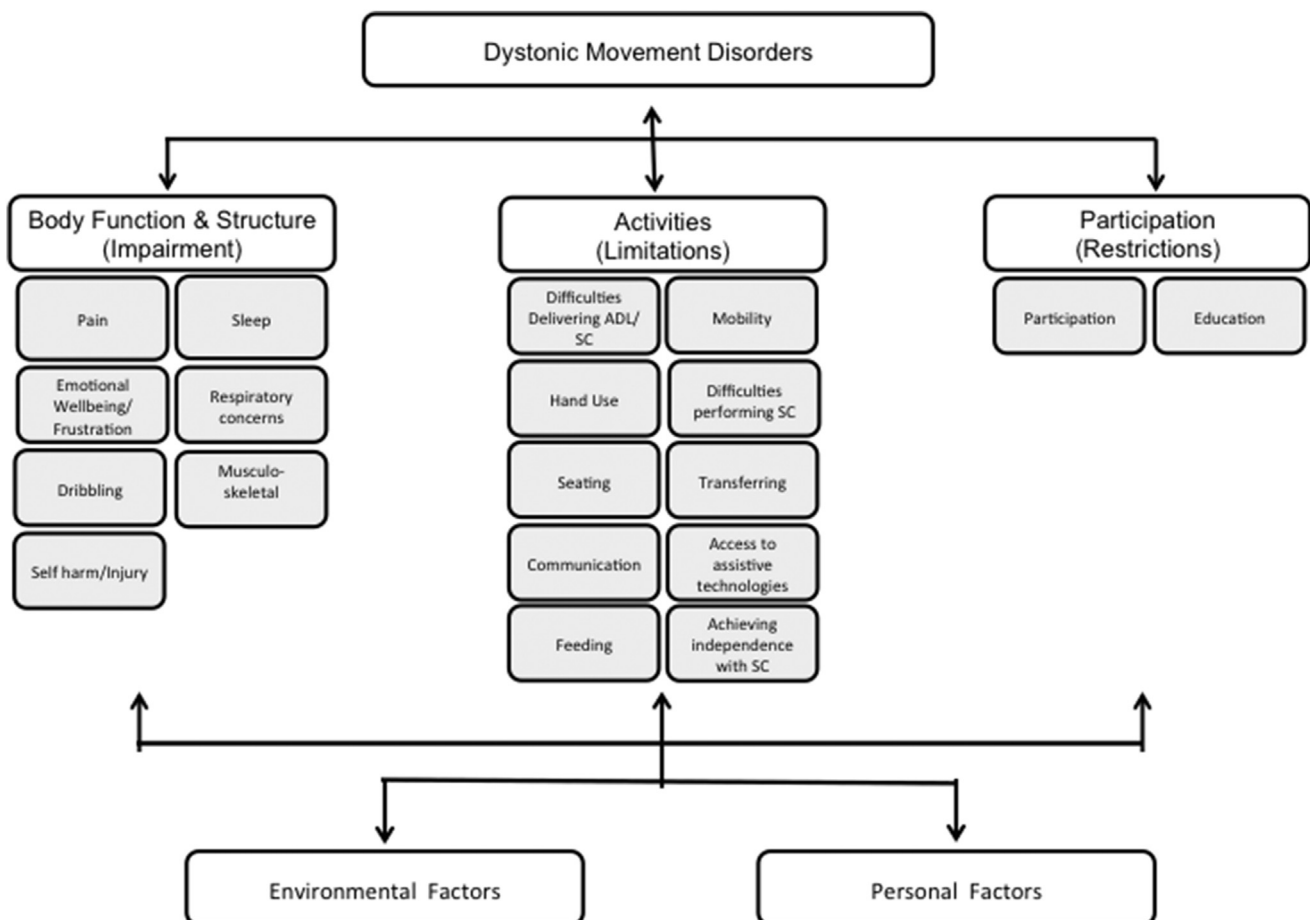
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## 1. Introduction

The most recent consensus agreement on the definition of dystonia states that: 'Dystonia is defined as a movement disorder characterized by sustained or intermittent muscle contractions causing abnormal, often repetitive, movements, postures, or both. Dystonic movements are typically patterned and twisting, and may be tremulous. Dystonia is often initiated or worsened by voluntary action and associated with overflow muscle activation'.<sup>1</sup> This adds further clarity to an earlier definition of dystonia in children as 'a movement disorder in which involuntary sustained or intermittent muscle contractions cause twisting and repetitive movements, abnormal postures, or both'.<sup>2</sup> Dystonia in childhood is a heterogeneous disorder, with a wide number of causes, varying severity and response to medical managements. On an aetiological basis dystonia in childhood is most commonly classified as secondary, arising as a consequence of an identified disease process, e.g. hypoxic-ischaemic encephalopathy.<sup>3</sup> Dystonia may interfere with functional activities and participation. Pharmacological and neuro-surgical management choices for childhood dystonia are complicated by the lack of high quality evidence, and current recommendations are made on a largely pragmatic basis.<sup>4</sup>

In recent years there has been considerable focus on emergent neurosurgical interventions for the management of dystonia. The advent of Deep Brain Stimulation (DBS) has directed new interest in dystonia which may be under-recognised in children with cerebral palsy or mistakenly classified as spasticity owing to a failure to implement rigorous operational definitions.<sup>5</sup> For the management of primary dystonia randomized control trial data has established the efficacy of DBS<sup>6</sup> in dystonia reduction. The role for DBS in secondary dystonia, and how the efficacy of this contrasts to primary dystonia, is currently the focus of much interest.<sup>7,8</sup> Evidence from adult trials of dystonia interventions cannot be easily applied to the paediatric population, due to differences in the most common aetiologies of childhood dystonia,<sup>3</sup> the possible co-morbid associations of dystonia, spasticity and other dyskinesias<sup>9,10</sup> and the influence of ongoing brain maturation during childhood.<sup>4</sup> The adverse influence of dystonia on motor development in children further supports the need for robustly designed interventional trials within the paediatric age group. Establishing the functional priorities of children and young people with dystonia is essential if the efficacy of an intervention for those child is to be understood.<sup>11</sup> This is consistent with recommendations of the International Classification of Functioning, Disability, and Health Framework (ICF) (Fig. 1).<sup>12</sup>



**Fig. 1** – Concerns expressed by children with dystonia and their carers mapped against the domains of the Internal Classification of Functioning (ICF) framework.

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