

Effectiveness of drug management along with physical and nutritional rehabilitation in children with dyskinetic cerebral palsy

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Abstract

Objective: To assess the improvement in functional activities of children with dyskinetic cerebral palsy after drug management along with physical and nutritional rehabilitation.

Design: Observational analytical study.

Setting: community based rehabilitation centre & a private medical college at Malappuram, Kerala.

Methods: This is a follow up cohort study conducted at the community level. 34 Children with dyskinetic cerebral palsy aged 4-18 years were followed up with medications for dystonia along with physical and nutritional rehabilitation for 9 months. These children were assessed with Barry- Albright dystonia scale prior to medications and at intervals of 3 months prospectively up to 9 months.

Results: Over the 9 months prospective study, 34 children were enrolled and started with medications on different occasions. 6 children were excluded (2 of them developed adverse reactions to drugs and 4 were lost to follow up). Out of 28 cases, 21 were males and 7 were females with an average age of 8 years. During the study period, all of them responded well with a significant improvement after 9 months.

Conclusion: Optimal drug management of dystonia, physical and nutritional rehabilitation along with management of co morbid conditions are associated with significant improvement in functional abilities of children with dyskinetic cerebral palsy.

Keywords: Dyskinetic cerebral palsy, Dystonia, Physiotherapy, Barry-albright dystonia scale.

Introduction

Cerebral palsy (CP) is a disorder of posture, movement and tone due to a static encephalopathy acquired during brain growth in fetal life, infancy or in early childhood.¹ Cerebral palsy is caused by a broad group of developmental, genetic, metabolic, ischemic, infectious and other acquired etiologies that produce a common group of neurologic phenotypes.² Though the brain disorder is unchanging, the effects are dynamic, as the brain matures, and the child's developmental capabilities extend.¹

High survival rates of at-risk neonates with improvement in neonatal care have led to an increased number of children with cerebral palsy in developing countries like India.³ In India, children with disability constitutes 3.8% of the population & nearly 15-20% of this physically disabled children are affected by cerebral palsy. In India, the estimated incidence of CP is around 3/1000 live births.⁴ Cerebral palsy is the most common motor disability in childhood. Some may also have associated problems such as epilepsy, mental sub normality etc. All these add to increased concern among parents and economic burden to healthcare system.

The physiologic classification identifies the major motor abnormality, whereas the topographic taxonomy indicates the involved extremities.

Dyskinetic cerebral palsy makes up approximately 15-20% of all CP.² Hypoxic ischemic injury to the brain & chronic bilirubin encephalopathy (kernicterus) are the major causes of dyskinetic cerebral palsy. They are

characterised by hypotonia with poor head control. They also develop variably increased tone, rigidity and dystonia over several years. Dystonia is physiologically characterised by the involuntary co-contraction of agonist and antagonist muscle groups at a joint and an overflow into muscles or limbs not normally involved with intended movement, resulting in abnormalities in posture and twisting movements. Dyskinetic cerebral palsy has an estimated prevalence of 0.27 per 1000 live births. In dystonia, muscle tone typically fluctuates, varying from normal or low to extreme hypertonia. It can be precipitated or worsened by attempts to move and can vary according to the emotional state. Dystonia typically diminishes or disappears with distraction and sleep. The basal ganglia have long been implicated in the pathophysiology of dystonia.^{5,6} In the European CP study, 76% of patients with this form of CP had lesions in basal ganglia & thalamus.⁷ Unlike spastic diplegia, the upper extremities are generally more affected than the lower extremities in dyskinetic CP. Feeding may be difficult, the tongue thrust and drooling may be prominent. Speech is typically affected because the oropharyngeal muscles are involved.²

Dystonia usually appears as one of an abnormal posture rather than an involuntary movement. Involvement may be of a single body part (focal dystonia), two or more contiguous body parts (segmental dystonia), the arm and leg on one side of the body (hemidystonia) or one or both legs and the contiguous trunk and any other body part (generalized dystonia).

For children who have a diagnosis of dyskinetic CP, a team of physicians including neurodevelopmental paediatricians, paediatric neurologists and physical medicine and rehabilitation specialists as well as occupational and physical therapists, social workers, educators, and developmental psychologist are important to reduce abnormalities of movement and tone and to optimize normal psychomotor development.

Methods

This is a follow up cohort study of quantitative nature using the existing data. Study was conducted at a private medical college and a community based rehabilitation centre in Malappuram district, Kerala, South India. Convenient sampling method was used and the sampling size was around 30. Children diagnosed with dyskinetic cerebral palsy aged 4-18 years with persistent dystonic movements were included in the study. Those with chorea-athetoid movements were excluded. Children with known neuro-metabolic

disorders, genetic syndromes and recurrent refractory seizures not responding to anti-epileptics were excluded. Children with other serious systemic illness were also excluded.

Data was analyzed by comparing the pre and post intervention status using paired T test or Wilcoxon signed rank test depending on the distribution of data.

Results

Over the 9 months of prospective study, 34 children diagnosed with dyskinetic cerebral palsy were started with medications on different occasions. Six children were excluded from the analysis as two of them developed adverse reactions to the medication and four of them were lost to follow up. The majority were males (21), and females were 7 with an average age of 8 years. During the study period, 28 of them responded well. (Table 1)

Table 1: Functional improvement in children with Dyskinetic cerebral palsy (n=28)

Functional improvement	Mean	SD	T value	P value
Pre –Medication	20.57	7.36	7.75	<0.001
Post –Medication	13.25	6.26		

Table 1: shows premedication mean improvement from 20.57 to 13.25 after nine months

Changes in Score

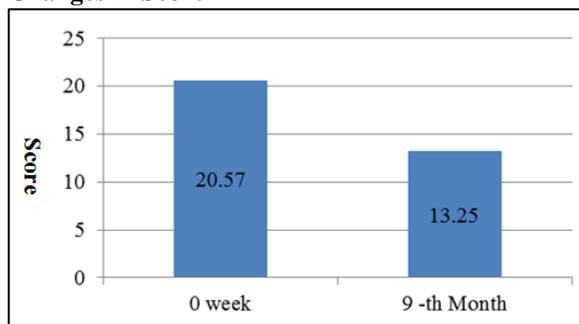


Fig. 1: Shows there is a significant functional improvement after 9 months

The mean change in score was 7.32 (SD 0.94) and it was found to be significant ($p < 0.001$).

Discussion

We have followed more than 500 children diagnosed with cerebral palsy in a community based rehabilitation centre. Out of these, 34 children were diagnosed with dyskinetic cerebral palsy in between 4-18 years. These children were started with trihexyphenidyl and levetiracetam and they were also given anti-spasmodic medications (Baclofen and/ or benzodiazepines) based on the severity for 9 months. They were assessed with Barry- Albright Dystonia Scale prior to the medications and at intervals of three months prospectively up to nine months of giving medications and physiotherapy.

Detailed nutritional assessment was carried out in all children. Out of 28 children, 14 had mild to moderate undernutrition and appropriate dietary modification was advised. 12 children had nutritional anemia and were treated with iron supplements, folic acid and Vit B12 depending on the underlying problem. 2 children with clinical, biochemical and radiological features of rickets were treated with 6 lakhs IU vitamin D followed by maintenance dose of Vitamin D and calcium for 4 months. All other children received Vit D3 (60,000 IU weekly X 4 weeks), calcium and phosphorus supplementation for 4 months. Anti-epileptics were continued to those who had been receiving them. Intercurrent infections were treated accordingly. Physical and occupational therapy were continued as per the usual protocol.

Various treatment modalities for cerebral palsy include medical management, physiotherapy, injection of Botulinum toxin, conventional multistage surgeries and surgical procedures like single event multi-level surgery. Numerous medications have been used in the management of dystonia, including anti-cholinergic medications (e.g. trihexyphenidyl), tetrabenazine, benzodiazepines (e.g. diazepam) and baclofen. Several drugs have been used to treat spasticity including oral dantrolene sodium, benzodiazepines and baclofen.⁸ Baclofen may be used for both oral and intrathecal administration.⁹ Patients with rigidity, dystonia, and spastic quadriplegia sometimes respond to levodopa, and children with dystonia may benefit from trihexyphenidyl or Levetiracetam.¹⁰ Intrathecal

baclofen can affect specific aspects of functioning in dyskinetic cerebral palsy. Sitting, communication, and fine motor functions improved.¹¹ Dachy et al evaluated the effect of intrathecal baclofen in a group of dystonic children by electrophysiological procedures previously validated in spastic children. They found that H reflex and area of flexor reflex significantly decreased after baclofen, though no significant clinical improvement of the Barry-Albright Dystonia Scale (BADs) was observed.¹²

In the present study significant improvement was noted after 9 months compared to the pre medication level. The long term follow up of these children with dystonic medications is required for knowing the complete outcome since there is no consensus regarding the duration of anti-dystonic medications to be used in dyskinetic cerebral palsy.

Conclusion

Optimal drug management of dystonia, physical and nutritional rehabilitation along with management of co morbid conditions are associated with significant improvement in functional ability of children with dyskinetic cerebral palsy.

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