Journal Watch 2017

Dyskinetic Cerebral Palsy (CP) is challenging to manage. Many studies have evaluated spastic cerebral palsy however there are fewer studies identifying the diagnostic difficulties associated with dyskinetic CP. An overview of the latest cerebral palsy guidelines, functional assessment of patient with dyskinetic CP and two articles aimed at managing dyskinetic CP are attached.

Managing dyskinetic CP remains challenging because many of the studies are retrospective and comprise of small patient numbers. These articles highlight the challenges with choosing the appropriate functional assessment tool and correlating it with the degree of severity of these symptoms. The next two articles highlights the management challenges and evaluates newer management techniques aimed at improving this condition. We hope that you would find reading these articles stimulating and that it may assist with the further management of these patients.

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Cerebral palsy in under 25s: assessment and management

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nice.org.uk/guidance/ng62

Review of the current NICE Guidelines in Cerebral Palsy

This is the most recent comprehensive guideline outlining the assessment and management of individuals with CP below 25 years of age.

The new guidelines include:

Risk factors

Causes of cerebral palsy

Looking for signs of cerebral palsy

Red flags for other neurological disorders

Multidisciplinary care

The principles of management are directed toward, both the individual and the family. The goals of management aim to optimise individual outcomes by maximising functional ability and promoting choice and independence. The guidelines recommend ongoing support to the individual with cerebral palsy, the family and all persons who care for these individuals.

The guidelines focus on the management of co –morbidities including:

- Eating, drinking and swallowing difficulties
- Speech, language and communication
- Optimising nutritional status
- Managing saliva control
- Low bone mineral density
- Pain, discomfort and distress
- Sleep disturbances
- Mental health problems
- Registering and processing sensory information

The guidelines also highlight interventions that are not recommended in the initial assessment and management of children and young adults with CP. It addresses the challenges of transition of care which is becoming more relevant in our setting with increased survival.

Functional Outcomes in Children and Young People with Dyskinetic Cerebral Palsy

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Developmental Medicine & Child Neurology

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Dyskinetic cerebral palsy (CP) is the most common subtype after spastic cerebral palsy. The functional impairment associated with dyskinetic CP is difficult to quantify due to the prevalence of choreoathetoid and dystonic movements. This study was aimed at advancing our insights in the functional profile of individuals with dyskinetic CP. This study aimed to investigate the interrelationships between the functional classification systems and the relationship of functional abilities with dystonia and choreoathetosis severity.

This study included 55 participants and were divided in 2 groups, comprising of children (<15 years) and young people (>15-22 years). The Gross Motor Function Classification System (GMFCS), Manual Ability Classification System (MACS), Communication Function Classification System (CFCS), Eating and Drinking Ability Classification System (EDACS), and Viking Speech Scale (VSS), Dyskinesia Impairment Scale (DIS) were evaluated.

Study findings: Dystonia is significantly related with all classifications, except the Communication Function Classification System (CFCS). Eating and Drinking Ability Classification System (EDACS) and Viking Speech Scale are significantly related with Gross Motor Function Classification System (GMFCS), Manual Ability Classification System (MACS), and CFCS. No significant relationship between choreoathetosis and classification scales at young age. Higher relationship between choreoathetosis and MACS and EDACS at older age.

Efficacy of oral pharmacological treatments in dyskinetic cerebral palsy: a systematic review

Riccardo Masson, Emanuela Pagliano, Giovanni Baranello

Developmental Medicine & Child Neurology

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The present systematic review aims to examine the actual level of evidence on the efficacy of oral medications in the management of dyskinetic CP, considering treatments for both dystonia and choreoathetosis.

Sixteen articles met the eligibility criteria. Eight studies on trihexyphenidyl and two on levodopa showed contradictory results. Low efficacy was reported for diazepam, dantrolene sodium, perphenazine, and etybenzatropine. Tetrabenazine, gabapentin and levetiracetam are considered as potential therapeutic options that should be investigated in more detail.

The updated available evidence does not support any therapeutic algorithm for the management of dyskinetic CP. It emphasizes the lack of standardized outcome measures and challenges with classification in dyskinetic CP. Recommendations are for the consideration of other therapeutic options, including intrathecal baclofen and deep brain stimulation in future research.

Intrathecal Baclofen in Dyskinetic Cerebral Palsy: Effects on Function and Activity

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Dyskinetic cerebral palsy (CP) is characterized by involuntary, uncontrolled, recurring, occasionally stereotyped movements. Primitive reflex patterns predominate and muscle tone varies. Oral and intra-thecal (IT) baclofen has been used to reduce spasticity and dystonia in children with cerebral palsy. Studies have been directed at evaluating the role of IT baclofen in spasticity. This study aimed to investigate the effect of intrathecal baclofen (ITB) on function and activity in dyskinetic CP.

This was a retrospective cohort study of records of 25 children with dyskinetic CP who received IT baclofen between 2004 to 2013.

This study identified statistically significant improvements in activities of daily life, sitting, and communication, sleep, pain. Dystonia was measured using the Barry–Albright Dystonia Scale. A statistically significant improvement with dystonia was noted. The modified Ashworth scale was used to measure muscle spasticity. A statistically significant decrease in muscle tone was demonstrated. This study did not demonstrate changes in gross motor function, fine motor function, range of motion or spinal alignment post IT baclofen.

This study supports the use of IT baclofen for the management of dyskinetic CP. Due to the small study population and lack of a control population it is difficult to generalise these findings to larger populations. IT baclofen holds much potential as a treatment modality and further studies in this area are required.

Deep brain stimulation in cerebral palsy: Challenges and opportunities

Anne Koy, Lars Timmermann

European Journal of Paediatric Neurology

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This review considered the role of Deep brain stimulation (DBS) in Dystonic CP. DBS has been increasingly recognized as a safe and effective treatment option for patients with pharmacological-refractory inborn, monogenetic dystonia. An increasing number of patients with acquired forms of dystonia such as dyskinetic CP have been treated with DBS over the last decade.

There are few studies demonstrating the response to DBS in dyskinetic CP. The response to DCS appears to be more variable and often delayed in individuals with CP. This is thought to be partly explained by the altered neuronal network activity due to lesions in several functionally connected elements of the cortex and basal ganglia network.

Outcome assessment is however challenging due to lack of validated tools to measure dyskinesia.

The current problems associated with DBS in the paediatric population is the higher risk for infection and other adverse outcomes post-surgery. The increasing head circumference especially during puberty makes electrode placement difficult. Lack of established eligibility criteria due to the heterogeneity in the outcomes of these patients, timing of surgery, and utilisation of age-appropriate assessment functional classification scales to objectively identify improvement in outcome.

DBS could improve quality of life and potentially function in children with dystonic CP but more research is indicated.