Case Report

Management of a Focal Dystonic Crisis in Cerebral Palsy: Use of Botulinum Toxin

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Introduction

Dystonic Cerebral Palsy (CP) is the smallest classified group of CP, coming within the sub-group of athetosis, which results from a lesion primarily in the basal ganglia. Although the mechanism behind observed involuntary movements is unclear we do know that the basal ganglia are involved in the refinement of information being transferred to the cortex and motor areas, the scale and amplitude of movements as well as the control of automatic background activity. The reported incidence of dystonic CP varies from 2-15% of the CP population (Reid et al, 2010).

This case report focuses on a 5 year old boy, ‘E’, with a primary diagnosis of CP secondary to prematurity. He presents with a mixed tonal pattern, predominantly those of a child with dystonic CP. The management of a growing number of such children is increasingly complex, putting additional demands on therapy departments.

A child presenting with dystonic CP secondary to prematurity requires careful assessment and management. The child often presents with central hypotonia, poor dissociation between the pelvic and shoulder girdles, fluctuating tone and dystonic posturing, resulting in ever-increasing stresses on the musculoskeletal system.

Botulinum Toxin has been used to treat spasticity since the early 1990s. Its efficacy in reducing spasticity in children with CP has been validated in studies (Keewon et al 2011; Love, Graham et al 2010).

Signed written consent for publication was obtained from the child’s parents prior to writing this case report.

History

‘E’ was born at 24 weeks and required 9 months of respiratory support. He was diagnosed with 4 limb CP (GMFCS level 5). All areas of his development have been and continue to be affected. Tone fluctuates throughout his body from hypo to severe hypertonia, exacerbated by effort, emotion and pain. Support is required to maintain symmetry in all lying sitting and standing positions. He had a past history of left shoulder posturing and possible subluxation. ‘E’ was already known to the tertiary movement disorder clinic prior to this episode. The clinic that had advised a trial of Trihexyphenidyl with positive effect.

Acute intervention

On initial examination at home, ‘E’ was unable to be placed in any position and was being held by his mother at all times. When placed on the bed, his whole body arched into extension with rotation to the left. His left upper limb was fixed in pronation and extension at the shoulder and elbow, with his hand fisted and thumb adducted.

On palpation his left humerus appeared subluxed and at risk of further injury. When held by his mother, with his left arm against her body, he settled for approximately 15 minutes at a time. Different positions were trialled including cradling him in a more flexed position. The left arm could be brought into neutral flexion/extension in this position, but remained in pronation and the child had difficulty settling.

Discussion between his mother, physiotherapist and paediatrician concluded that ‘E’s’ pain was worsening and he was becoming exhausted secondary to lack of sleep and possible infection. Admission to the local hospital was arranged via A&E.

Inpatient Local Hospital Care

Following Morphine, two doses of Diazepam and Chloral Hydrate, ‘E’ slept and his shoulder position improved. Discharged the next day, within hours his symptoms reappeared. His overall posture was improved, the left arm had reverted to the same extreme position and he was in pain on any movement.
The physiotherapist trialled various positioning techniques, specifically for the left upper limb. Triceps was palpably rigid and attempts to massage it were clearly painful. By abducting the thumb, the physiotherapist was able to position and strap his left arm across the body in more flexion. ‘E’ could then be placed in a Tumbleform chair for short periods of time. Liaison with the local hospital ensured that the child could return to the ward for more medication if required.

Referral to a Tertiary Centre and Care
An urgent review was requested at the tertiary movement disorder clinic. This review highlighted ‘E’s’ significant pain levels, for which three analgesics and an increased dose of Trihexyphenidyl had already been prescribed. The treatment plan, following assessment by a neurologist and physiotherapist, was to inject Latissimus Dorsi and Triceps (both palpably rigid) with 50 units of Botulinum Toxin A (Dysport) (Btx A). ‘E’ was also provided with Diazepam and Chloral Hydrate to use at home if necessary and commenced on a small dose of Baclofen.

Goals for Btx A intervention
Short Term
1. To reduce pain levels, measured through monitoring analgesia levels
2. To increase length of time that ‘E’ was able to sleep at night
3. To allow ‘E’ to be placed in his postural management equipment (seating and lying support)

Long term
1. To allow a window of opportunity for ‘E’ to develop a preferred switch position to improve communication
2. To help determine suitable treatment strategy to manage ‘E’s’ left shoulder posturing and help reduce the risk of further dystonic crises.

Community Care
Two days later the local physiotherapist reviewed ‘E’ and advised the mother on:
1. The use of positions to maintain postural alignment
2. The maintenance of a flexed posture or a ‘nested’ position (hips and knees flexed) thus reducing tone and allowing the head to remain in midline.
3. The facilitation of shoulder protraction using pillows.
4. Encouraging hands to midline to touch his face and other activities
5. The use of a small soft object placed in his left hand to help maintain thumb abduction

Because of concern regarding possible ‘neglect’ of his left upper limb, his Mother was encouraged to slowly increase his awareness of that limb through massage, placing objects in that hand, looking at it, etc.

Outcomes
Improvement in symptoms following Btx A injection was observed a few days post-injection, with ongoing improvement over time. His left upper limb position improved dramatically and, when supported in his seating system, he was able to rest both hands on his knees comfortably. Physiotherapy focussed on facilitating the left upper limb into midline and flexion by reading books and playing with toys in various positions. Hydrotherapy in the early post-injection period facilitated rehabilitation goals, decreasing the influence of gravity and providing a gentler environment in which to exercise (Kelly et al 2005).

Orthotic Care
‘E’ was assessed a month later for a shoulder stability lycra garment to be used as tolerated. On further review it was decided to add sleeve length and paneling to facilitate external rotation and a thumb loop to maintain thumb abduction.

Discussion
This case report may be of interest to paediatric physiotherapists as it demonstrates the effective use of Btx A in the management of an acute focal dystonic crisis.

Dystonic CP can be difficult to manage. Fluctuating tone causing extreme posturing through range seems to lead to lower use of Btx A in the early years. Instead therapists and paediatricians cooperate to ensure that the child receives other medication and an integrated postural management/therapy approach to manage tone as efficiently as possible. Therapists’ main concern is often to create proximal stability. They focus on postural management systems to provide this and therapeutic handling to facilitate improved postural tone and the grading and control of movement (Bobath course notes 2009).

Use of medication in the management of tone is widespread. ‘E’ was already prescribed Trihexyphenidyl, an antimuscarinic acting on synapses, blocking receptor sites. He was subsequently prescribed Baclofen, a skeletal muscle
relaxant which acts principally on the central nervous system, is widely used (BNF 2011).

Lycra garments are increasingly used for children with CP. Some evidence suggests there is measurable improvement associated with their use (Angilley 2008, Knox 2003). They provide stability, both centrally and in larger limb joints. Shoulder stability garments provide specific support when the posture of the shoulder is affecting overall posture.

Multi-disciplinary teams regularly refer children with CP for Btx A assessment when there is stiffness and reduced movement related to spasticity. Btx A can give the team a window of opportunity to try to maximize length in the injected muscles whilst strengthening the opposing muscle group. As a result of ‘E’s’ fluctuating tone and previous management, he had relatively good muscle length throughout and had no lower limb pathology. He had not therefore previously been considered a good candidate for Btx A treatment.

This case report highlights that children experiencing a focal dystonic crisis can benefit from Btx A injections. Medication worked initially to sedate him but this was not found to be the most functional management strategy. By directly targeting the muscles causing the posturing, subluxation and pain, a positive effect was achieved. Not only did this give fairly rapid relief but there was also an opportunity for rehabilitation post-injection.

The positive effects of the Btx A lasted approximately 2 months, after which the tone in his left upper limb began to increase and the posturing returned, albeit less severe. This was obviously frustrating for him and his Mother as well as the team working with him. It is considered that repeat injections may be beneficial.

Once again the benefits of multi-disciplinary working with this client group is demonstrated in providing support and care at home, school, local hospital and in a tertiary centre. Through joint working and good relationships with acute partners the local team was able to support ‘E’ and his family through a focal dystonic crisis using the most appropriate intervention. With an increasing number of complex children to manage in primary care, it is vital to have good relationships with tertiary partners. This supports effective communication and a sharing of knowledge and skills between agencies. It also facilitates involvement of the whole team and family in the decision-making process and most importantly, helps to achieve the best possible outcomes for the child.

**Conclusion**

This case study describes one way to manage a child experiencing a focal dystonic crisis. Early identification of the main problems and working in partnership with the tertiary centre meant that this child had access to Btx A as an acute intervention to manage his dystonic posturing subluxation and pain.

The use of Btx A in this way is not widely publicised and may not be at the forefront of community therapists’ minds when they are clinically reasoning through a patient with this presentation. As anticipated, the effects of the Btx A wore off after 2 of months and the symptoms began to return. The question requiring further investigation is whether regular Btx A could be a long term solution to the management of a dystonic upper limb in order to prevent crises occurring.

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**References**


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