CLINICAL RESEARCH

IM botulinum toxin-A improves ambulation in severe haemophilia and cerebral palsy

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Introduction: The use of BtA in spasticity management is well documented in the literature for the management of hypertonicity in children with cerebral palsy.
Methods: We report a case of a 2-year-old boy with severe haemophilia and cerebral palsy who received intramuscular injections of botulinum toxin-A (BtA) to reduce his spastic diplegia lower limb hypertonicity.
Results: Following treatment, clinical and formal assessments demonstrated positive changes in ambulation, tolerance of orthotics, functional abilities and musculoskeletal range of movement and no adverse bleeding side effects.
Conclusion: The use of BtA should be considered for the management of hypertonicity in children with haemophilia and cerebral palsy.

Key words: haemophilia, cerebral palsy, botulinum toxin-A

Intramuscular botulinum toxin-A (BtA) to produce neuromuscular blockade has been effective in treating spasticity in children with cerebral palsy (CP) [1-3]. The use of BtA in children with a combination of CP and haemophilia has not previously been reported. We report a case of a 2-year-old boy with severe haemophilia and CP who received intramuscular injections of BtA to reduce his spastic diplegia lower limb hypertonicity.

Intramuscular BtA injections are indicated to relieve dynamic spasticity affecting function in the absence of a fixed deformity, or pain, or to facilitate the use of orthotics [4-7]. The literature also supports the use of BtA in children 4 years old or younger in the hope of preventing or delaying deformity [6, 8].

Materials and methods

Presentation

The 2-year-old boy (for the purpose of this report named Joshua) was examined in February 2008, and assessment included physical examination, video analysis and the Gross Motor Function Measure (GMFM-66). The modified Tardieu Scale was used to estimate the contributions of spasticity and contracture [9, 10]. For his haemophilia, Joshua received prophylaxis three-times a week with no reports of bleeds.

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Spasticity in cerebral palsy may be treated with neuromuscular blockade

He presented with hypertonicity in his lower limbs, with loss of passive extensibility and dynamic overactivity in hip adductors, hamstrings and gastrocnemii. On physical examination Joshua had bilateral hot swollen anterior ankle joints as a result of the supramalleolar splints, which had been provided for ankle stability. This was worse on the left, reflecting the fact that the left ankle was tighter. This was compounded by the dynamic element of his gastrocnemii.

Functionally, Joshua was mobile indoors with the assistance of walking aids, albeit slowly and with adducted and internally rotated hips, semi-flexed knees and a toe-toe gait pattern. On observation, his bare foot gait using his walking aids was slow and effortful, with a small base of support. Joshua had a bilateral equinus gait and hyper-extended his knees during stance phase. There was some hip excursion, but he maintained hip flexion throughout his walking pattern and had poor pelvic stability with little co-activation of his trunk. Utilising his supramalleolar splints there was an improvement in foot clearance during swing and foot stability in stance. All other joint positions remained the same.

The GMFM-66 is a criterion-referenced observational measure designed and validated to measure change in

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TABLE 1: Joint ranges before and after (BtA)

Joint	Movement	Passive range of movement (degrees)					
		Right			Left		
		Pre	8wk	3 Months	Pre	8wk	3 Months
Supine hip	Flexion (0-125)	Full	Full	Full	Full	Full	Full
	Extension (Thomas test)	Negative	Negative	Negative	Negative	Negative	Negative
	Abduction in flexion	40	45	45	40	45	45
	Dynamic catch	25	No	25	25	No	25
	Abduction in extension (0-45)	20	30	30	20	40	30
	Dynamic catch	15	no	20	15	no	20
Knee	Flexion (0-140)	Full	Full	Full	Full	Full	Full
	Extension	0	0	+5	0	+5	+5
	Popliteal angle unilateral	35	35	30	35	35	40
	Dynamic catch	60	No	No	60	No	No
Ankle	Dorsiflexion knee flexed (0-20)	15	25	20	15	20	20
	Dorsiflexion knee extended(0-20)	0	20	20	0	20	20
	Dynamic catch	-30	-5	90	-40	-10	90
Spine			Straight				
Clonus			Not apparent				

gross motor function over time in children with cerebral palsy [11]. Joshua scored 83% in the crawling and kneeling dimensions and 26% in the standing dimension. Furthermore he was classified as 'Level ll' using the Gross Motor Function Classification System (GMFCS): "Children floor sit, but may have difficulty with balance when both hands are free to manipulate objects. Movements in and out of sitting are performed without adult assistance. Children pull to stand on a stable surface. Children crawl on hands and knees with a reciprocal pattern, cruise holding on to furniture and walk using an assistive mobility device as preferred methods of mobility," [11].

Treatment approach

In consultation with the Movement Disorder Team, it was decided that intramuscular BtA was indicated to Joshua's hip adductors, bilateral hamstrings and gastrocnemii, because of dynamic contractures that were interfering with walking and wearing splints.

Collaborative goals of easing stretching, reducing scissoring and equinus during gait and increasing tolerance to orthotics were agreed. He received a review of his splints and was cast for hinged ankle foot orthoses. These were chosen as it was felt that a longer lever splint would help to improve stability at his ankle, reduce knee hyperextension and control his dynamic equinus in conjunction with the BtA. These changes would be expected to result in a more even pressure exerted by the orthotic and as such reduce his swollen ankle joints.

One day prior to Joshua's injections he commenced 250 mg (15mg/kg) tranexamic acid three-times-daily and was advised to continue taking it for 7 days post procedure. Dosage of BtA injected was calculated as international consensus guidelines [3]. He received BtA (Dysport) under general anaesthetic with 1000 IU factor VIII and 250mg tranexamic acid cover pre and post operatively for one day. The following day Joshua returned to his usual prophylaxis regime.

On discharge one day later, Joshua had no pain or bruising and the family were advised to continue with his stretching programme and local physiotherapy.

Treatment outcome

On reassessment after 8 weeks, the parents reported improvements in walking-speed, general movement and in providing for Joshua's personal care. On examination the dynamic element of his hypertonicity had either reduced or was absent, thereby assisting his fluidity and range of movement (Table 1).

On observation, his bare foot gait using his walking aids had improved in both speed and fluidity with a wider base of support. He had a bilateral toe heel gait, with improved heel contact more on the right than the left. He retained some hyperextension at his knees.

When using hinged ankle foot orthoses Joshua's walking pattern had improved, with a notable reduction in knee hyperextension and the achievement of bilateral heel strike. Functionally, the ability to stand holding on to furniture with only one hand was observed.

The GMFM-66 was not completed at this assessment, because significant functional changes would not be expected at this point.

At the 3 month follow up, Joshua's parents reported he was able to high-kneel, walk forward on his knees and had managed to stand independently for 18 seconds and on occasions had taken 5-8 steps independently. He was able to climb the stairs utilising alternate feet on each stair and his walking speed was quicker. Functionally Joshua was able to sit in the bath holding on to the rails. On the GMFM-66 he scored 95.2% on the crawling and kneeling dimension and 33% on the standing dimension: demonstrating an overall clinical improvement in function. His GMFCS remained at level II.

On observation, Joshua's bare foot gait was flat-footed with early heel rise when walking slowly and a bilateral equinus gait as he increased his speed. It was evident that

he had retained knee hyperextension during stance with some hip flexion. When walking in orthoses he was able to achieve a bilateral heel strike, but often chose to walk with a flat footed gait, with knees in slight flexion. Pelvic stability and active hip extension during gait remained poor, but his balance and stability had improved. This was evidenced by standing and stepping and holding on to only one pod. Joshua's gait pattern remained fluid with increased speed from his initial assessment. Limited independent walking was observed. Overall, Joshua's confidence while walking bare foot or using his splints and pods had improved significantly.

Conclusion

This is the first report to our knowledge to demonstrate the benefits of using BtA in a child with severe haemophilia A and a co-existing diagnosis of CP. Our clinical and formal assessments demonstrated positive changes in ambulation, tolerance of orthotics, functional abilities and musculoskeletal range of movement and no adverse bleeding side effects. The use of BtA in spasticity management is well documented in the literature for the management of hypertonicity in children with CP and should be considered in children with haemophilia and CP.

Acknowledgements

The authors stated that they had no interests, which might be perceived as posing a conflict or bias.

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