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CASE SERIES

Botulinum Toxin Type A in the Treatment of Children with Congenital Muscular Torticollis

ABSTRACT

Oleszek JL, Chang N, Apkon SD, Wilson PE: Botulinum toxin type A in the treatment of children with congenital muscular torticollis. Am J Phys Med Rehabil 2005;84:813–816.

This is a retrospective case series describing the use of botulinum toxin type A in the treatment of children with congenital muscular torticollis who fail to progress with conservative management. A total of 27 children with congenital muscular torticollis, 6–18 mos of age, received 30 botulinum toxin type A injections into their sternocleidomastoid or upper trapezius muscle, or both, at a pediatric tertiary care center between 1995 and 2001. Three children received repeat injections. Twenty of 27 children (74%) had improved cervical rotation or head tilt after the injections, and 2 of 27 (7%) experienced transient adverse events, specifically, mild dysphagia and neck weakness. This series suggests that botulinum toxin type A may be a safe and effective treatment option for children with congenital muscular torticollis who are unresponsive to a traditional regimen of physical therapy and a home program. A prospective, randomized controlled trial is necessary to definitively assess the role of botulinum toxin type A in this population.

Key Words: Congenital Muscular Torticollis, Botulinum Toxin Type A, Children, Physical Therapy

Congenital muscular torticollis (CMT) is a neck deformity that involves shortening of the sternocleidomastoid (SCM) muscle. Infants with torticollis typically tilt their head toward the side of the affected muscle and rotate toward the contralateral side. We have found that the upper trapezius muscle can also be involved and contribute to the neck deformity. The prevalence of CMT varies from 0.3% to 2.0%.[1] Although many theories have been proposed, the true cause of CMT remains unknown. If torticollis persists, craniofacial deformities or plagiocephaly can occur.

Treatment of CMT may include observation, formal physical therapy program, home exercise program, or use of an orthosis. The majority of published studies use a combination of physical therapy and a home program completed by the parents.[1–4] Surgery is often considered for resistant cases of CMT. Binder et al.[2] reported full recovery in 70% of subjects receiving physical therapy by 12 mos of age, with 7% requiring surgery. In a study by Demirbilek and Atayurt[5] of 57 children with CMT who were involved in a therapy program, all children

under the age of 3 mos had excellent results, without the need for surgery. However, surgery was required in 25% of the 3- to 6-mo-old infants and 70% of the 6- to 18-mo-old infants.

In our experience, an aggressive stretching program can become challenging because of the strength and tightness of the child's affected SCM and because of the child's resistance to therapy with increasing age. For these reasons, and to avoid the sequelae of persistent CMT and surgery, we began using a combination of botulinum toxin type A (BTX-A) and therapy as a treatment option when therapy alone was ineffective. BTX-A, a neurotoxin derived from the bacteria *Clostridium botulinum*, produces a protein that inhibits release of acetylcholine and results in localized reduction in muscle activity. The goal of BTX-A is to temporarily weaken the affected SCM or upper trapezius muscle, resulting in an easier and more successful stretching program and an improved ability to strengthen the opposing neck musculature. BTX-A has been shown to be safe and effective in the treatment of cervical dystonia in adults, and it has been used safely in children with limb spasticity. To our knowledge, there have been no studies examining the use of BTX-A in children with CMT. The purpose of this report is to describe our experience using BTX-A in the treatment of children with CMT who fail to progress with conservative management.

METHODS

Children 6–18 mos of age with CMT who received BTX-A injections between 1995 and 2001 in the rehabilitation clinic at a tertiary care children's hospital were included in this series. Children with torticollis as a result of neurologic abnormalities, vertebral anomalies, or ocular disturbances were excluded.

Retrospective data recorded from the subjects' records included: age at injection, side of involvement, sex, weight, therapy program, documentation of plagiocephaly, muscle injected, units of BTX-A injected, range of motion preinjection and postinjection, caregiver report of adverse events, and subjective assessment of outcome by the caregiver or physician.

Neck range of motion was measured by one of four physiatrists. Measurements recorded included cervical rotation (passive or active) and active head tilt (defined as lateral deviation from midline), which were measured either by goniometer or observation. The same physician performed each individual subject's pre- and post-BTX-A injection measurements. The results were divided into quantitative and qualitative measures. In determining quantitative improvement, changes in rotation and tilt were recorded. Qualitative comments by the physician or caregiver were recorded, such as "marked improvement," "no asymmetry," or "no head tilt," when formal measurements were not obtained.

BTX-A injections were performed by one of four physiatrists. The muscles selected for injection (SCM or upper trapezius muscle, or both) were based on the clinical judgment of the treating physician. The upper trapezius injections correlated with documented muscle tightness or shoulder elevation on examination. Subjects received injections in the clinic setting without sedation or EMG guidance. One to two assistants were needed to secure the child in a supine or side-lying position. The SCM was isolated by turning the child's head toward the contralateral, thus allowing the muscle to be easily grasped between two fingers during the injection. The upper trapezius was isolated by palpation. Other cervical muscles, such as scalenes or splenius, which are often injected in adults with cervical dystonia, are more difficult to isolate in an awake, young child and are not injected in our clinic. BTX-A was reconstituted with normal saline to a concentration of 100 units in 0.5 ml for all injections. A 27-gauge, 30-mm needle with a 1-ml syringe was used. The dose of BTX-A used was based on the child's age and muscle size, not weight. When injecting the SCM and upper trapezius muscles, one to two injection sites per muscle were often used.

RESULTS

A total of 27 children (59% male and 41% female) with CMT received 30 BTX-A injections between 1995 and 2001. Three of these children received repeat injections. Thirteen of 27 (48%) had left-sided involvement, and 14 of 27 (52%) had right-sided involvement. The age range at the time of injection was 6–18 mos, with a mean age of 10.1 mos. The three children who received repeat injections were initially injected between 7.5 and 8.5 mos of age. The weight range at the time of injection was 6.5–12 kg, with a mean of 8.8 kg.

Plagiocephaly was observed in 52% of children. All children were involved in a home program, and 23 of 25 (93%) received formal physical therapy.

BTX-A injections were performed as follows: 18 injections into the SCM muscle alone, using a mean of 29.7 units (range, 20–50 units); three injections into the upper trapezius muscle alone, using a mean of 28.3 units (range, 25–35 units); and nine injections into both the SCM and upper trapezius muscles, using a combined mean of 55 units (range, 25–80 units).

Of the 27 children who received BTX-A injections, 20 (74%) had documented improvement in neck rotation or active head tilt. Of these, two had

only qualitative assessments. Lack of improvement was seen in two (7%), but only subjective assessments were documented in these children. Five of the 27 children (19%) were lost to follow-up. Fifteen of 20 had improvement in rotation between 10 and 50 degrees, with a mean of 30.7 degrees, and 14 of 20 had improvement in active head tilt between 15 and 45 degrees, with a mean of 29.6 degrees. Ten of these subjects had improvement in both rotation and head tilt. Tables 1 and 2 depict the preinjection and postinjection measurements and the degree of change for rotation and head tilt, respectively. Only the 18 subjects with measurable data were included. Some data represent repeat treatments. Twelve children had full rotation of ≥90 degrees, whereas ten children had a neutral head position after the injections. Of the two children with only qualitative documentation of change, one had "no head tilt" postinjection and the other had "no asymmetry" postinjection, but neither had preinjection measurements recorded. One of the children without improvement underwent an SCM surgical release. This child received the injection at 17 mos of age. Of the three children who received repeat injections, one had improved rotation after the first injection but no follow-up after the second, one had no improvement after the first injection but improved rotation after the second, and the last child had improved head tilt and rotation after both injections.

Transient adverse events were reported in two cases (7%), specifically, mild dysphagia in one child (age of 12 mos) and neck weakness in the other (age of 10 mos). No medical treatment was necessary in either case, and both events were self-limited.

TABLE 1 Quantitative cervical rotation measurements before and after botulinum toxin type A injection and degree of change

Subject	Cervical Rotation, Degrees		Change, Degrees
	Before	After	
2	75	90	15
5	50	75	25
6	45	90	45
7	45	90	45
8	30	70	40
9	60	90	30
11 (injection 2)	45	70	25
13	40	90	50
14	70	90	20
15 (injection 1)	45	90	45
19	70	90	20
23 (injection 1)	50	80	30
2 (injection 2)	80	90	10
26	45	90	45
27	75	90	15

TABLE 2 Quantitative active head tilt measurements before and after botulinum toxin type A injection and degree of change

Subject	Head Tilt, Degrees		Change, Degrees
	Before	After	
1	20	0	20
2	20	5	15
3	45	10	35
5	45	0	45
6	30	5	25
7	30	0	30
9	20	0	20
10	45	0	45
13	30	10	20
14	45	10	35
19	25	0	25
23 (injection 2)	30	5	25
24	30	0	30
26	45	0	45

DISCUSSION

The use of BTX-A as a treatment option for CMT began in our clinic in the early 1990s. The concerns regarding refractory CMT, plagiocephaly, and potential surgery warranted a more aggressive therapy. All children with CMT in our clinic are involved in physical therapy and a home program, and this is particularly important after the BTX-A injections. Families are taught proper techniques of stretching the involved neck muscles and strengthening the opposing muscles, and methods of incorporating these exercises into daily routines of carrying, positioning, and feeding. In addition, ways of avoiding undesirable postures that perpetuate the torticollis and plagiocephaly are emphasized.

Surgery has been recommended for children with persistent range-of-motion deficits when there is no improvement after 6 mos of manual stretching or when they are older than 1 yr of age.[1,6] Use of BTX-A can be an additional option when the traditional regimen is ineffective. In our small series, only 1 of 27 children (age of 17 mos) required surgery. There were six other children >1 yr of age at the time of injection. Four of these had improved rotation and head tilt, and two were lost to follow-up.

We have found that tightness in the upper trapezius muscle can also occur with CMT. Both the SCM and upper trapezius muscles rotate, extend, and laterally flex the neck. To our knowledge, there has been only one report discussing the effect

of the upper trapezius muscle in CMT, and the child in this report was particularly resistant to therapeutic efforts, including surgery.[6] In our series, 12 children received upper trapezius injections, with nine of these children receiving concomitant SCM injections. Ten of these children had improved rotation, two were lost to follow-up, and none required surgery.

A significant limitation of this series is the lack of a standardized measurement paradigm. In addition, four physiatrists performed nonstandardized range-of-motion measurements and the BTX-A injections, and this could certainly affect the reliability of our results.

BTX-A has been used to treat cervical dystonia in adults and has been shown to be a safe intervention. However, significantly larger doses of BTX-A in multiple locations are often used when compared with the injections performed in children with CMT. Jankovic and Schwartz[7] followed 205 patients with intractable cervical dystonia who received 1074 injections in 505 visits. Of these patients, 28% had adverse events, primarily mild dysphagia (35 patients) or neck weakness (17 patients). Similarly, transient dysphagia and transient neck weakness were documented in 2 of 27 children in our series.

CONCLUSION

In this small case series, BTX-A seems to be a safe and effective treatment option for children with CMT who are unresponsive to traditional management of a home program and physical therapy. A prospective, randomized, controlled trial is necessary to definitively assess the role of BTX-A in this population. Other variables that may affect the outcome of BTX-A injections, such as duration of physical therapy, age at injection, and severity of range-of-motion restriction, need to be explored further.

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