# Congenital Muscular Torticollis in Older Children: Treatment With Z-Plasty Technique

Nur Yucel Ekici, MD,\* Ahmet Kizilay, MD,† Mustafa Akarcay, MD,† and Yezdan Firat, MD‡

Abstract: Congenital muscular torticollis (CMT) is a common congenital disorder of the musculoskeletal system in neonates and infants. The aim of this study was to evaluate the results of inferior Z-plasty in older children with CMT. They had mean age of 10 years (range, 5-14 years) and were followed up for 1 to 6 years. Postoperative protocol included a neck exercise program composed of active and passive movements in all cases and immobilization with a cervical collar in only 4 patients. This study concluded that surgical management of older children with CMT using Z-lengthening gives excellent clinical and functional results. The procedure is much more effective than other techniques and relatively complication-free and safe. Postoperative cervical collar and a well-planed physiotherapy protocol go a long way toward ensuring good to excellent results. Early diagnosis and treatment are necessary for good results.

Key Words: Congenital muscular torticollis, children, Z-plasty

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ongenital muscular torticollis (CMT) is an idiopathic condition that begins in infancy as a rotation and flexion deformity of the neck caused by sternocleidomastoid (SCM) muscle shortening. It is the third most common congenital musculoskeletal anomaly after congenital hip dysplasia and clubfoot. Its frequency in the newborn ranges from 0.3% to 2%. It involves contracture and shortening of the SCM muscle, causing the head to tilt toward the affected side and the chin to the opposite side.<sup>2-4</sup> Birth trauma, intrauterine malposition, ischemia-venous occlusion, infection, constitutional and growth arrest causes, hereditary predisposition, and neurogenesis have all been implicated. But its pathogenesis and treatment protocol remain controversial.<sup>2-5</sup> Skull and facial asymmetry or plagiocephaly may occur in the presence of prolonged uncorrected head tilt.<sup>6</sup>

The main reason for consultation is cosmetic deformity of the head and face. Ultrasound (US) is advocated for the diagnosis and follow-up of CMT because it noninvasively provides reliable and dynamic information without sedation. The diagnosis of CMT is based on the clinical palpation of a firm mass, fibrous band within

From the \*Department of Otorhinolaryngology, Kozan State Hospital, Adana; †Department of Otorhinolaryngology, Inonu University Faculty of Medicine, Malatya; and §Department of Otorhinolaryngology, Memorial Hospital, Istanbul, Turkey.

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Address correspondence and reprint requests to Mustafa Akarcay, MD, Department of Otorhinolaryngology, Inonu University Faculty of Medicine, Elazig yolu 7th km 44067, Malatya, Turkey; E-mail: makarcay@inonu.edu.tr

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SCM muscle, or contracture of the SCM.4 Ultrasound is the imaging modality of choice for the evaluation of CMT, which must be clearly differentiated from other congenital and acquired types of torticollis, such as congenital cervical vertebral anomalies, posttraumatic conditions, infections and inflammation of adjacent structures, tumors, ocular torticollis, hearing deficit, and miscellaneous neurological structural and functional causes.

When diagnosed early, CMT can be managed conservatively with good or excellent results. Spontaneous resolution is expected in most patients. The optimal time for surgical treatment is contentious. In patients older than 1 year, corrective surgery has both cosmetic and functional benefits, with the best outcomes being obtained between the ages of 1 and 4 years. Some authors have stated that surgical treatment is of little value after age 5 years, and the results are even worse when the operation is done after puberty and may lead to more complications.<sup>2,4,7</sup> Nonsurgical treatment includes stretching exercises and physiotherapy (especially in early age) and botulinum toxin injections. Surgical options comprising subcutaneous and open tenotomies, resection, and Z-lengthening of the SCM muscle. 4,7-11 The aim of our study was to evaluate the results of inferior Z-lengthening procedure prospectively in older children.

## PATIENTS AND METHODS

Between 2002 and 2007, 6 children with neglected CMT presented to our clinic and were followed up prospectively (1–6 years). Six patients (4 girls, 2 boys) whose ages ranged from 5 to 14 years were included in the study. The right side was involved in 4 patients and the left side in 2 patients.

After the first examination, cervical US was performed to clarify the diagnosis of CMT and the severity of fibrosis. According to US findings, all of the cases had diffuse hyperechogenicity along the entire muscle with hypoechogenic background muscle, without any palpable mass, but they had severe fibrosis. Following a standardized initial assessment, parents were taught our intensive home treatment protocol, consisting of passive stretching exercises repeated every 3 hours. After a mean follow-up of 5.5 months (5–6 months) with physiotherapy, no patients were treated by use of our intensive protocol of passive stretching exercises. Therefore, surgery was necessary. The indications for surgery were a persistent head tilt and deficits of passive rotation. There were no other congenital deformities in our patients.

Inferior Z-plasty technique was performed in all patients. Under general anesthesia, the patients lay supine with the head turned away from the affected side. The involved side was placed under tension by hyperextending the neck and rotating the head toward the shoulder on the unaffected side. Horizontal incision was done 1 to 2 cm above the medial third of the clavicle. The platysma, adjacent fascia, and other soft tissues were also resected. The sternal and clavicular attachments were dissected and divided by electrocautery. The clavicular head was released while the sternal head was lengthened by Z-plasty. The tendon ends were approximated with 3-0 monofilament absorbable sutures in the stretched position to preserve the cosmetic V-shape of the neck. Subcuticular closure was



**FIGURE 1.** In patient 1 (an 8-year-old boy), soft cervical collar was used for 6 weeks postoperatively.

done in layers to reduce scar formation, taking due precaution not to injure any neurovascular structure.

Five days postoperatively, a neck exercise program including active and passive movements was started in all patients and immobilization with a cervical collar in 4 patients. Cervical collar was applied to patients for 5 to 8 hours a day for a mean of 2 to 6 weeks (Fig. 1). All patients were educated about performing stretching exercises for at least 10 minutes, 4 to 6 times daily.

The neck movements were compared with the uninvolved side. The head tilt and operative scar were evaluated by clinical observation and patient satisfaction. The described operative technique, combined with intensive postoperative physical therapy and application of a soft cervical collar, provides good functional and cosmetic results.

### **RESULTS**

Mean age of the patients was 10 years (5–14 years). Four of the patients were girls, whereas 2 were boys. Four of the patients had involvement of the right side, whereas 3 had involvement of the left side. The mean follow-up for the patients was around 3.5 years (range, 1–6 years). Excellent and good results were noted in all patients. No patients experienced a complication or required revision surgery for residual bands. There was not a bad scar formation in any patient. Cosmetic improvement as reduction in head tilt and chin deviation was present in all patients. The head tilt and operative scar were evaluated by clinical observation and patient satisfaction.

#### DISCUSSION

Most of cases with CMT resolve completely, either spontaneously within months after birth or following the early initiation of conservative measures such as passive manual stretching exercises on the affected side. Conservative management is usually tried before the age of 1 year. Sönmez et al<sup>12</sup> found that 95% of the patients diagnosed and treated effectively before the age of 1 year did not need surgical treatment. Celayir<sup>5</sup> reported that there is no need for surgical treatment in early-treated patients with CMT, and a successful outcome depends on good cooperation with the parents. Nonoperative therapy after the age of 1 year is rarely successful. The primary goals of surgery for patients with CMT are improved movement of the neck and an improvement in head tilt. Cosmetic improvement is also an important consideration. Surgical correction after 1 year of age has been advised because of the need for compliance with postoperative rehabilitation.<sup>13</sup> Chen and Ko<sup>14</sup> and Minamitani et al<sup>15</sup> reported that late release of SCM muscle for the patients older than 6 years could yield acceptable results. We

also agree with these authors, and according to these criteria, we were successful in managing all our patients, despite the long-standing nature of the deformity. Satisfactory results will be obtained regardless of the patient's age when a suitable operation and rehabilitation are performed.

Patients with CMT with severe fibrosis in the entire SCM muscle on US did not improve after conservative treatment and were more likely to require surgical intervention than those with mild fibrosis. However, for clinical outcome, it is well known that the success rate of conservative treatment depends mainly not only on the degree of SCM muscle damage but also on the patient's age at initiation of treatment. 1,2,4 Furthermore, patients with severe fibrosis on US were likely to be older than those with mild fibrosis. The severity of restriction of neck rotation at the beginning of treatment was a predictor of treatment duration and that severity of fibrosis on US was intimately associated with clinical outcome. 16,17 Ultrasound features of the SCM muscle correlate with the clinical severity of CMT in young infants.<sup>18</sup> Thus, to elucidate the true relationship between clinical features and severity of fibrosis on US, the effect of patient age should be considered. All our patients had severe fibrosis on US at the first examination, and they were older children. Thus, we did not perform ultrasonographic classification.

There are various surgical procedures for CMT. Omidi-Kashani et al,<sup>3</sup> in a prospective study of 14 cases, concluded that bipolar electrocautery SCM division, even in patients with irreversible facial asymmetry, improves quality of life by improving neck movement and head tilt. All our patients had an excellent outcome. Bipolar electrocautery release has been suggested as the operation choice for patients with CMT.<sup>8,9,14,16</sup> If bipolar electrocautery release is performed without Z-lengthening, loss of the normal contour of the neck may affect the cosmetic appearance. We also believe in that Z-plasty preserves the normal V contour of the SCM and ensures a better cosmetic outcome. In our patients, we performed Z-plasty technique in all cases. And all of them reported improved movement and head tilt and good cosmetic results.

The postoperative immobilization protocol for CMT is also controversial. In the initial days following surgery, the patient has a tendency to keep the head in its former position to reduce pain, and compliance with the prescribed exercises is poor. If the head remains in this position, the released structures will regain their former tightness. Immobilization in an overcorrected position has been claimed to give better results<sup>9,13</sup>; others have advocated the long-term use of braces to prevent recurrence<sup>17</sup> or intermittent cervical traction in the early postoperative period followed by application of a cervical collar and exercises.<sup>9</sup> We combined the operative technique with intensive postoperative physical therapy and application of a soft cervical collar; we think that it provided us good functional and cosmetic results.

Surgical scarring and loss of the SCM column are reported as generally more common in older patients. <sup>18</sup> Transaxillary subcutaneous endoscopy for the release of the SCM muscle procedure provides direct access to the fibrous bands, enables us to release without risk of damage to other tissues, and leaves no visible neck scars. <sup>19,20</sup> We made incision away from the clavicle and used subcutaneous sutures and were thus able to produce less scar tissue. However, Z-plasty preserves the normal V contour of the SCM muscle and ensures a better cosmetic outcome.

Our study has limitations. The sample size is small, but CMT presenting in older children is rare. We did not specifically measure facial asymmetry, head tilt, neck range of motion, scar, and so on. The head tilt and operative scar were evaluated by clinical observation and patient satisfaction. However, our patients reported improved self-perception of facial asymmetry, and most had good cosmetic results.

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In conclusion, surgical management of older children with CMT using Z-lengthening gives excellent clinical and functional results. The procedure is much more effective than other techniques and relatively complication-free and safe. Postoperative cervical collar and a well planed physiotherapy protocol go a long way toward ensuring good to excellent results.

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