Identification and Treatment of Congenital Muscular Torticollis in Infants

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Torticollis is a clinical sign or symptom that could be the result of a variety of underlying disorders. Congenital muscular torticollis (CMT) is a postural deformity detected at birth or shortly after birth, primarily resulting from unilateral shortening and fibrosis of the sternocleidomastoid muscle (SCM). [1–3] Infants with CMT display head tilt to one side, which is often combined with rotation of the head to the opposite side (Figure 1). [4,5] CMT is estimated to occur in one infant of every 300 live births. [6] Plagiocephaly is reported as a coexisting impairment in 80% to 90.1% of children with CMT. [1,7] The purposes of this report are to provide an overview of the CMT literature with attention to the relationship between torticollis and plagiocephaly and to describe current management of infants with congenital muscular torticollis with or without plagiocephaly.

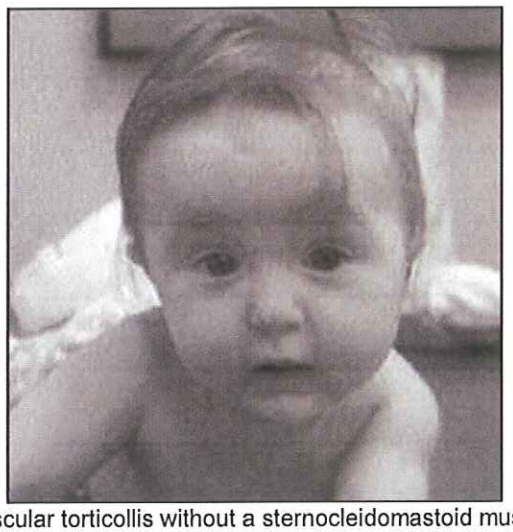

Figure 1. Child with congenital muscular torticollis without a sternocleidomastoid muscle (SCM) tumor viewed prone on elbows. Observe cervical side bending and lateral head tilt to the side of SCM tightness with minimal rotation of the chin to the opposite side. Note compensatory right shoulder elevation and side bending of the trunk.

CMT with impairment of the SCM is the most frequent cause of torticollis in infants, but torticollis could also be the result of other underlying disorders. The congenital and developmental causes of torticollis in children have been classified as osseous, nonosseous, or neurogenic (Table 1). [8,9] The prevalence of nonmuscular causes of torticollis in children could be as high as 18%. [10] Children with ocular torticollis could present with the postural signs of torticollis without restrictions in cervical range of movement. [10,11] The child with impairment of extraocular muscles in one eye could tilt or turn the head to achieve a clear binocular view. Paresis of the superior oblique muscle could result in head tilt away from the side of impairment and is the most common source of ocular torticollis. [11]

Osseous types
Occipitocervical dysfunction
Cervical vertebral dysfunction
Klippel-Feil syndrome
Congenital scoliosis
Hemivertebrae
Nonosseous types
Congenital muscular torticollis
Sandifer syndrome
Neurogenic types
Central nervous system tumors
Arnold Chiari malformation
Ocular torticollis
Paroxysmal torticollis

Table 1. Congenital and developmental disorders with signs or symptoms of torticollis [8,9]

The pathophysiology and etiology of sternocleidomastoid impairment in CMT is still unknown. Prominent theories related to the cause of sternocleidomastoid impairment in CMT include intrauterine crowding, [12–13] muscle trauma during a difficult delivery, [2,4] soft tissue compression leading to compartment syndrome, [14] and congenital abnormalities of soft tissue differentiation within the SCM muscle. [15,16]

Children with CMT can be assigned to one of three clinical subgroups: 1) children with a palpable swelling or pseudotumor of the sternocleidomastoid, 2) children with SCM tightness but no tumor, and 3) children with all the features of muscular torticollis without muscle tightness or tumor. [1,16] In an alternative system of classification, pseudotumor of infancy and CMT are described as separate diagnoses. [17,18]

NATURAL HISTORY AND OUTCOMES OF CONGENITAL MUSCULAR TORTICOLLIS

The diagnosis of torticollis is usually made by the pediatrician in the first 2 or 3 months of life when a pseudotumor of the SCM, abnormal head posture, restricted cervical range of movement, or plagiocephaly is noted during a "well baby" visit. The mean age for diagnosis of torticollis is reported as 24 days, [9] 1.1 months, [19] and 4 months. [17]

A subgroup of children with CMT develop a pseudotumor or swelling in the body of the SCM, which can be palpated as early as 2 to 3 weeks of age. [14,18] When a pseudotumor is present in the SCM within the first few weeks of life, it is usually a soft, nontender enlargement of the muscle belly. [16] The most frequent site of the pseudotumor is the middle to lower third of the sternal portion of the SCM (Figure 2). [1,10] A pseudotumor is palpated in 28.2% [1] to 47.2% [17] of infants with CMT. The pseudotumor usually becomes larger after it is first noted and then slowly resolves over a period of 5 to 21 months. [19]

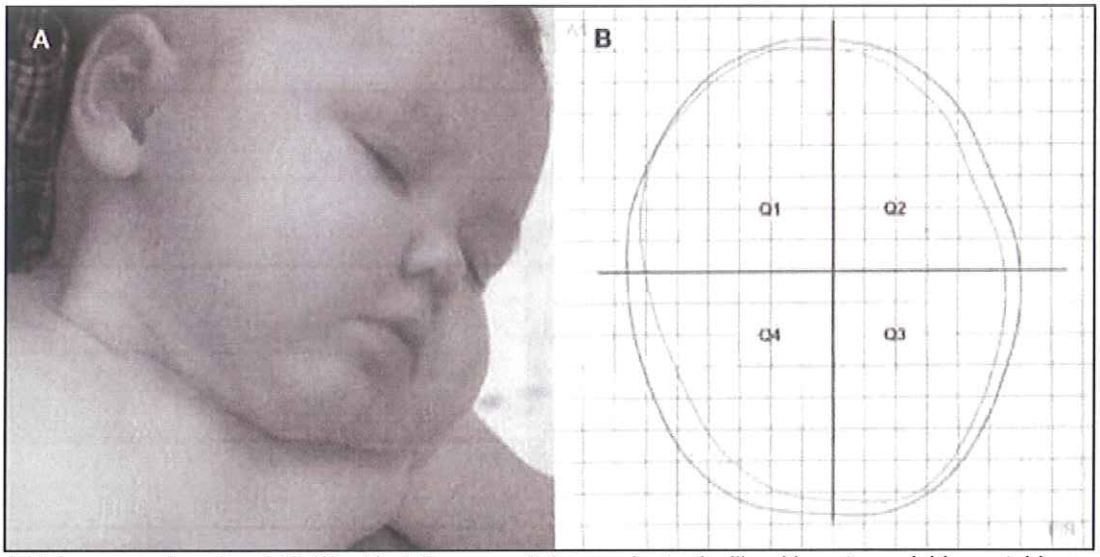

Figure 2. Initial presentation of a child (A) with right congenital muscular torticollis with a sternocleidomastoid muscle tumor and left plagiocephaly. (B) Laser scan image of infant skull at initial presentation is seen as the interior line. Final scan (outer line) shows marked increase of symmetry in the left posterior and right anterior quadrants. Conservative treatment involved 5 months of physical therapy and use of a cranial remolding orthosis. Surgery for muscle release preceded cessation of the cranial remolding orthosis.

Pediatricians are advised to refer infants with plagiocephaly or torticollis to physical therapy by 2 to 3 months of age if neck movement does not improve after intervention with parent instructions in the physician's office. [20] Children with less than 10° difference in range of cervical rotation between sides could comprise a group that is easily managed with parental advisement from their pediatricians. [1] Conservative treatment is not different for children with and without tumor.

Outcomes at 12 months of age are described as "good" for 69.3% of American children who receive conservative treatment of CMT. [21] Cheng et al. [1] have developed a useful rating scale for determining the outcomes of treatment for children with CMT (Table 2). Outcomes of conservative treatment were rated "good" for 91.1% of the 1086 Chinese children with CMT using the Cheng scale. [1] Tight bands of residual fibrosis resulting from contracture of the SCM muscle could be present after resolution of the pseudotumor in children for whom conservative intervention does not yield satisfactory results. Surgical intervention could be required to release the fibrotic tissues (Figure 3).

	Scoring criteria				
Category	3 Points	2 Points	1 Point	0 Points	Points awarded
Rotation deficit (degrees)*	<5	6–10	11–15	>15	
Lateral bending deficit (degrees)*	<5	6–10	11–15	>15	
Craniofacial asymmetry	None	Mild	Moderate	Severe	
Residual band	None	Lateral	Lateral/cleidal	Cleidal/sternal	
Head tilt	None	Mild	Moderate	Severe	
Subjective assessment by parents (cosmetic and functional)	Excellent	Good	Fair	Poor	
				Total score	

Modified from Cheng et al.[1] Adapted with permission from *J Bone Joint Surg.*
Outcome rating determined by total score: excellent = 16–18 pts; good = 12–15 pts; fair = 6–11 pts; poor = <6 pts.
*Measured with arthrodial goniometer; deficit determined by comparison to contralateral side.

Table 2. Scoring system for assessment of treatment outcomes in children with congenital muscular torticollis

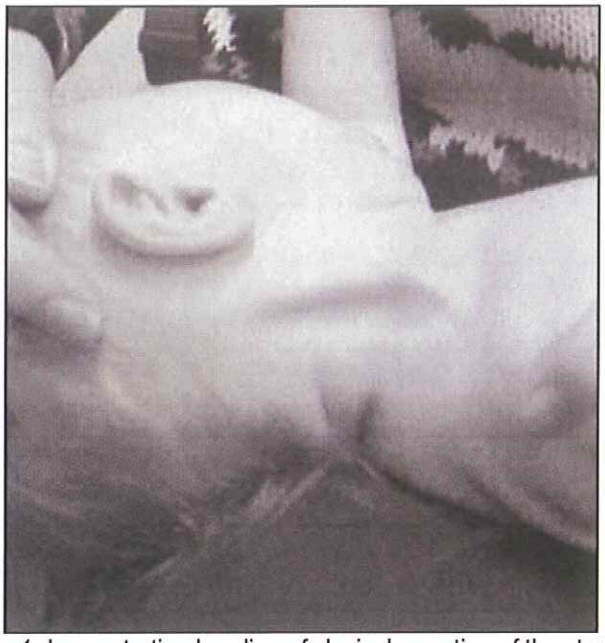

Figure 3. Child pictured in Figure 1 demonstrating banding of clavicular portion of the sternocleidomastoid muscle, lateral and posterior cervical skin folds. Cheng outcome score was "poor" after 7 months of conservative treatment. The child was referred to surgery with good outcome.

Surgical release of the SCM is required more frequently when children have a history of SCM tumor or deficits of greater than 30° in cervical rotation at the time of diagnosis. Children who are younger at presentation and initiation of physical therapy are less likely to require surgery to lengthen their sternocleidomastoid muscles. [4,21] No prospective studies were found comparing conservative treatment with physical therapy with untreated control groups. Several studies describe the postural and craniofacial deformities present among individuals who received no intervention or who began treatment after 1 year of age.

PLAGIOCEPHALY AND CONGENITAL MUSCULAR TORTICOLLIS

Plagiocephaly is reported in up to 90% of children with CMT. [1] Torticollis could result in deformities of the developing skull base, cranium, or face. When plagiocephaly and torticollis coexist in a neonate, they could both be the result of a limitation of the intrauterine space that caused persistent asymmetric compression of the cranium and unilateral shortening of the SCM in the final weeks of gestation. [22]

CMT could cause plagiocephaly in infants who do not present with plagiocephaly at birth. [4,16,20] In cultures where young infants sleep in the supine position, unilateral compression of the skull base will occur in the child with torticollis if there is rotation of the chin away from the shortened muscle. [22] Unilateral shortening of the SCM causes the young infant to consistently position the head on the occiput contralateral to the tight SCM while unloading the occiput on the ipsilateral side. With continued unilateral weight bearing, the skull base and cranium will deform so that the vertex view reveals a parallelogram-shaped head (Figure 2B). [23]

Craniofacial asymmetry of varying degrees was noted in 80% of 624 Chinese infants referred for treatment of torticollis between 1985 and 1991. [7] Mild to moderate facial asymmetries usually resolve with conservative treatment, including orthotics if necessary. The classic facial deformities associated with muscular torticollis that is severe, discovered late, or untreated have been described as "a suborbital torsional deformity of the face toward the affected side" (p. 7). [24] The craniofacial deformities associated with CMT were described in a clinical series of 16 patients referred to the Variety Center for Craniofacial Rehabilitation at the New York Medical Center between 1992 and 1997. [4] Deformities observed in clinical photographs included posterior displacement of the ipsilateral ear (94%), posterior regression of the ipsilateral zygoma (87%) and forehead (81%), mandibular deviation toward the affected side (44%), inferior positioning of the affected eye (31%), and deviation of the nasal tip to the affected side (19%). [4] Skull base and cranial deformation toward a rhomboidal shape occurs before onset of facial deformities. [4,23–25]

Deformational plagiocephaly has increased in frequency since initiation of the "back to sleep" program. Secondary contractures of the SCM could develop in infants with deformational plagiocephaly resulting from consistent positioning with the head turned to one side. [20] In the authors' experience, infants with deformational plagiocephaly often present with limited extensibility of

cervical muscles. However, the flattened occiput is not always opposite to the side of SCM tightness, and examination often reveals that the SCM is not the muscle causing the limited extensibility.

EXAMINATION OF THE CHILD WITH CONGENITAL MUSCULAR TORTICOLLIS

The examination of the child with torticollis should begin with a careful history to reveal the presence of coexisting conditions and to determine if the history is compatible with a diagnosis of CMT. Recommendations for use of diagnostic procedures, including x-ray and imaging, vary among authors. Clinical practice guidelines for management of positional skull deformities in infants are available in the July 2003 issue of *Pediatrics* . [20] No clinical practice guidelines were found for pediatricians to use in the diagnosis of CMT. Ballock and Song published a useful decision tree for differential diagnosis of young children with torticollis (Figure 4). [10]

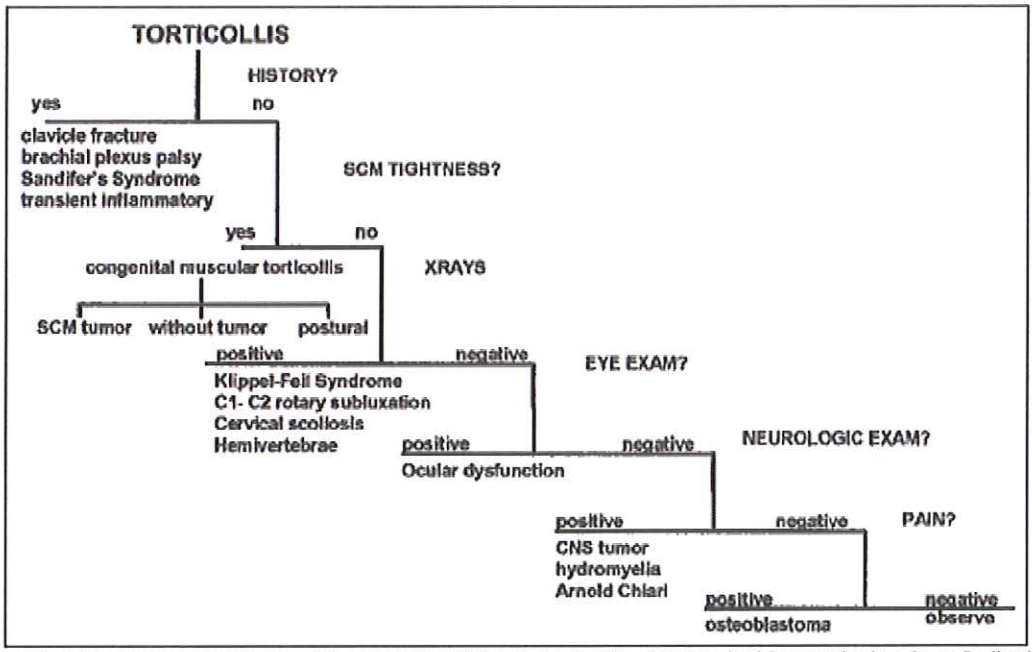

Figure 4. Decision tree for assessment of the young child with torticollis. Adapted with permission from Ballock RT, Song KM. The prevalence of nonmuscular causes of torticollis in children. *J Pediatr Orthop* 1996;16:500–504.

The primary method for estimating active range of motion in young infants is through active head turning during visual tracking. [20] A reliable and valid method for assessment of passive cervical range of motion in infants using an arthrodial protractor is described in Cheng et al. [1] Valid studies determining age-related norms for cervical range of motion in infants are lacking. Inspection of the skin of the neck for asymmetry of skin folds and skin condition, as well as palpation of all superficial cervical muscles, is essential to discovery of the sources of abnormal head and neck posture. Postural assessment should include the entire spine as a result of the frequency of development of cervical and/or thoracolumbar scoliosis as the child attempts to achieve vertical orientation of the head in the presence of SCM contracture. [19] The lower extremities should be screened for hip dislocation and other associated problems. Examination and evaluation of head and face shape are important components of assessment for every child with torticollis. [18]

Observation of the infant's abilities to perform ageappropriate motor skills is an important component in the assessment of the infant with torticollis or plagiocephaly. Underlying impairments of muscle force could be revealed in the observation of motor skills. The infant could be positioned to vary the resistance of gravity to reveal weakness. Feeding problems can require special positioning and complicate care. Gastroesophageal reflux and any other problems with feedings should be identified, and positioning for feeding should be specifically assessed. Intolerance of prone positioning is a common problem in infants with torticollis and/or plagiocephaly. [26] Assessment of family routines and positions of the baby throughout the day is essential before planning a therapeutic positioning program.

CLINICAL MANAGEMENT OF THE CHILD WITH MUSCULAR TORTICOLLIS

Treatment of the infant with torticollis is guided by the age of the infant, the severity of the torticollis, the abilities of the parents to perform the exercises and repositioning procedures, the diagnosis of plagiocephaly, and the presence of associated

neuromuscular or orthopedic impairments. [27–30] The first line of treatment for young infants with either deformational plagiocephaly or torticollis is aggressive repositioning. American Academy of Pediatrics guidelines [20] instruct physicians to counsel parents in repositioning their infants in the first weeks of life.

PHYSICAL THERAPY

Conservative management of infants with torticollis consists of positioning, gentle range of motion, and strengthening through activation of head and trunk muscles as the infant gains control of upright postures. [28–30] Overall goals of the physical therapist in the management of infants with CMT could include: 1) age-appropriate active and passive range of motion of cervical and trunk movements if possible; 2) prevention of contractures or further loss of motion in infants with SCM nodules or significant fibrosis; 3) symmetry of shape in the face, head, and neck; 4) development of postural reactions in all directions; 5) centered upright posture of the head and neck without persistent tilt to the involved side; and 6) symmetry of gross motor patterns throughout development. [28–31]

The physical therapist's responsibilities include assessment of the infant and family to determine the causes of the child's movement disorder followed by implementation of a management program. The home program should be incorporated into the family's routines and include: ways to handle, feed, carry, and position the baby; activities to encourage midline head and trunk postures; and gentle active and/or passive cervical range-of-motion exercises opposite to the torticollis posture and away from the plagiocephalic, flattened side. [28–31] Parental compliance is mandatory for successful outcomes. [20]

Manual stretching is the most common form of treatment for CMT. [1,5,16,13,30,32,33] Proper stabilization and hand placement is vital for the success of each stretch; however, all child/parent pairs will not be comfortable with the same method of stretching or the same stretch positions. The severity of the torticollis, the age of the child, the tolerance of the child for handling, and the parent's ability to carry out the exercise program will determine the method of stretching. When performing stretching exercises, the position of the head and neck in flexion versus extension will impact the effectiveness of the stretch. [28] One point of agreement among authors who discuss stretching methods is that the stretches should not be painful and should be carried out by the parents and caregivers whenever possible. [27,29,34] Reducing the intensity of the stretch slightly could avoid pain and muscle guarding. The tight muscles of infants with SCM tumors should be gently stretched toward end range using active and passive exercises with similar intensity to the stretch imposed on tight muscles without SCM nodules. [29]

Stretching and strengthening exercises can be carried out through holding, carrying, and playing with the baby in postures and positions selected to achieve the desired active and passive movements. Strengthening overstretched muscles on the side opposite to the torticollis could be accomplished through postural reactions as the infant matures and gains better control of the head and trunk. [35] Torticollis influences acquisition of motor skills. Restrictions and imbalances in the cervical muscles can lead to asymmetry of motor development. [10] Since the introduction of the "back to sleep" program, there has been an increase in the incidence of delayed gross motor development. [32,36–38] Torticollis can augment the impact of supine sleeping on motor development by further diminishing head control in antigravity positions leading to decreased weight bearing on the arms or by causing uneven weight shifts through the trunk. Trunk shortening on the torticollis side and asymmetrical transitional movements into sitting and standing are common. The adaptive compensations of the musculoskeletal system in the trunk and upper extremities can contribute to abnormal movements and posture. [28,29]

Massage of tight neck muscles and subcutaneous tissues is commonly used in the management of infants with CMT to aid in increasing pain-free range of motion. [28,34,39] Joint mobilization, myofascial release, and craniosacral therapies are emerging areas of practice in infants with torticollis. [28] Therapeutic taping is sometimes used to support weak and overstretched muscles. [31]

Guidelines for discharge from physical therapy and for monitoring after discharge are not well defined. [5,34] Torticollis posture could reappear during periods of growth. The SCM on the involved side may not grow at the same rate as on the uninvolved side, creating a risk of return of contracture. [13] During periods of illness, teething, and acquisition of new motor functions, regression to the torticollis posture could occur. [28]

ORTHOSES

Many infants with torticollis have coexisting deformational plagiocephaly severe enough to warrant use of a cranial remolding orthosis. The duration of treatment with a cranial remolding orthosis could be longer for children with torticollis and plagiocephaly who have residual limitations in cervical range of motion. Criteria and guidelines for cranial remolding are found throughout this consensus report.

Cervical orthoses have been used as treatment adjuncts for those children whose lateral head tilt does not resolve with

exercises. The most commonly used collar in children with torticollis is the TOT collar, Tubular Orthosis for Torticollis. [30,40–42] The TOT collar is adjusted to support the neck on the impaired side in the neutral position. The TOT collar is worn only during waking hours and is not used for children younger than 4 months of age. [28,30,41]

BOTULINUM TOXIN INJECTIONS AND SURGICAL INTERVENTIONS

Botulinum toxin (Botox) could enhance the effectiveness of stretching on the side of the contracture and allow strengthening of overstretched and weakened muscles on the opposite side of the neck. [34] Surgery is indicated if symptoms persist after 1 year of age despite conservative treatment. [17,29,32,33,42,43] An alternate criterion reported for surgical intervention is the presence of residual deficits in rotation range of greater than 15° of rotation after at least 6 months of controlled manual exercises. [1,5] Children who enter conservative treatment at older ages are more likely to require surgical release of the SCM. [4] Surgical techniques to lengthen tight SCMs include unipolar release, [1,5] bipolar release, [8] endoscopic release, [44] and subperiosteal lengthening. [43] Postoperative physical therapy consisting of range-of-motion exercises is recommended after surgical release of the SCM. [1,5,8,28,29,43] A cervical collar could also be included in the postoperative management. [8,28,29]

CONCLUSION

Contemporary management of infants and children with congenital muscular torticollis must address the needs of a changing population. Since the introduction of the "back to sleep" program, there has been an increase in the incidence of infants with plagiocephaly with and without torticollis. Early detection and initiation of physical therapy is related to improved outcomes and less need for surgical lengthening of the SCM. Repositioning is a required element of early management of both torticollis and plagiocephaly. Cranial remolding orthoses and cervical collars could be necessary as adjuncts to treatment. The literature upholds active and passive cervical and trunk range of motion as an effective treatment for children with torticollis, but no randomized, controlled trials exist using alternative treatments.

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