Congenital Muscular Torticollis: Bridging the Gap Between Research and Clinical Practice

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Congenital muscular torticollis (CMT) is a common postural deformity evident shortly after birth, typically characterized by ipsilateral cervical lateral flexion and contralateral cervical rotation due to unilateral shortening of the sternocleidomastoid muscle. New evidence is emerging on the pathogenesis of CMT, the negative long-term consequences of delaying intervention, and the importance of early identification and early intervention to maximize outcomes. Our purpose in this article is to inform pediatricians and health care providers about new research evidence and share selected recommendations and implementation strategies specifically relevant to pediatric practice to optimize outcomes and health services for infants with CMT.

Congenital muscular torticollis (CMT) is a common postural deformity evident shortly after birth, affecting 3.9%[1,2] to 16%[3] of infants. It is characterized by ipsilateral cervical lateral flexion and contralateral cervical rotation due to unilateral shortening of the sternocleidomastoid muscle, with or without a sternocleidomastoid mass. In Fig 1, an infant with right CMT is depicted. Craniofacial asymmetry is a coexisting impairment in up to 90% of infants with CMT[4] and increases the risk of facial,[5] ear,[5] and mandibular asymmetry.[6] Outcomes are best when infants are diagnosed early and start comprehensive physical therapy management before 3 months of age.[7–9] If untreated or treated after early infancy, CMT can lead to craniofacial deformities,[10] cervical spine dysmorphism,[11,12] and painful limited cervical motion, requiring more invasive interventions such as botulinum neurotoxin injections[13] and surgery.[14,15]

The etiology of CMT is unclear. Historically, it has been attributed to birth trauma,[16] prenatal or perinatal compartment syndrome,[17] and impairment of the developing sternocleidomastoid due to intrauterine constraint.[18–20] Recent immunohistochemical and gene expression studies provide stronger support for an intrauterine impairment of the developing sternocleidomastoid.[18,19] The primary pathologic features of the involved sternocleidomastoid include excessive endomysial and perimysial fibrosis, adipocyte hyperplasia, and muscle fiber atrophy,[19] leading to tightness of the involved sternocleidomastoid and limited cervical motion. The degree of fibrosis is proportionate to the age of the child when untreated; the older the child, the more fibrosis is present.[19] Ultrasonography has been used to quantify the initial severity of muscle fibrosis and its resolution with intervention.[21] Emerging immunohistochemical research supports deposition of type III collagen as a key factor in generating sternocleidomastoid fibrosis, and its

abstract

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Drs Sargent, Kaplan, and Coulter conceptualized the need for the manuscript and drafted the initial manuscript; Dr Baker conceptualized the need for the manuscript; and all authors reviewed and revised the manuscript, approved the final manuscript as submitted, and agree to be accountable for all aspects of the work.

DOI: https://doi.org/10.1542/peds.2019-0582

Accepted for publication Apr 2, 2019

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PEDIATRICS (ISSN Numbers: Print, 0031-4005; Online, 1098-4275).

FINANCIAL DISCLOSURE: The authors have indicated they have no financial relationships relevant to this article to disclose.

FUNDING: Supported by grants from the American Physical Therapy Association and the Academy of Pediatric Physical Therapy. In addition, Dr Sargent's salary was supported by National Institutes of Health grant K12-HD055929. The content is solely the responsibility of the authors and does not necessarily represent the official views of the National Institutes of Health. Funded by the National Institutes of Health (NIH).

To cite: Sargent B, Kaplan SL, Coulter C, et al. Congenital Muscular Torticollis: Bridging the Gap Between Research and Clinical Practice. *Pediatrics.* 2019;144(2):e20190582

hyperplasia is associated with accelerated apoptosis and transforming growth factor beta 1 over-expression.[22] Gene expression studies further support this by proposing that 1 of the critical pathways to the pathogenesis of CMT is fibrosis with collagen and elastin fibrillogenesis, with evidence of DNA repair and cytoskeletal rearrangement possibly related to mechanical strain.[23] Secondary changes of the cervical spine in CMT have also been reported.[11,12,24] A recent study supports that the onset of deformable changes in the cervical spine starts as early as 8 months of age, and the severity of the deformity increases with age and with the severity of the sternocleidomastoid tightness.[11] To minimize these secondary impairments, it is imperative that infants with CMT participate in early intervention.

Infants diagnosed with CMT are not expected to spontaneously resolve. They require a comprehensive physical therapy program of cervical stretching, cervical and trunk strengthening, activities to promote symmetrical movement, environmental adaptations, and parent or caregiver education and support to provide a daily, intensive home program.[25] There is strong evidence that earlier physical therapy intervention is more effective than intervention started later.[26] If started before 1 month of age, 98% of infants with CMT achieve normal cervical range of motion within 1.5 months.[7] Waiting until after 1 month of age prolongs the physical therapy episode of care up to 6 months, and waiting until after 6 months to begin physical therapy may require 9 to 10 months of physical therapy intervention, with progressively fewer infants achieving normal range.[7] Clearly, the costs of service and time to families could be reduced when a comprehensive physical therapy program is started earlier. However, infants in the United States are not being referred to physical therapy early enough. Physical therapists report that two-thirds of infants with CMT are referred at ages 3 to 4 months and one-third at 5 to 6 months[27]; these ages are much later than is ideal.

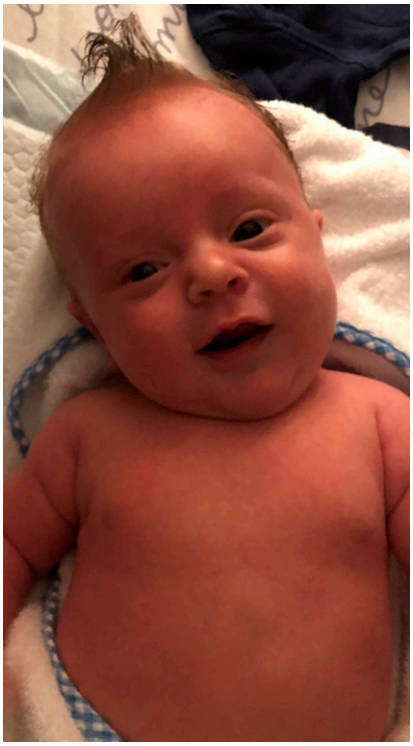

FIGURE 1
Infant with right CMT.

An emerging approach to increasing cervical range with a shorter episode of care is microcurrent delivered below the sensory threshold. Two small randomized clinical trials support that single-frequency microcurrent added to conventional care, versus conventional care alone, results in greater reductions in head tilt,[28] increases in passive cervical rotation,[28,29] greater reductions in sternocleidomastoid thickness,[29] and shortened average treatment duration (2.6 vs 6.3 months).[29] This approach may be especially useful for older infants or toddlers with CMT based on a recent case study of a 19 month old with untreated left CMT.[30] In this case, the CMT resolved after only 10 weeks of conventional care, dual current, frequency-specific microcurrent, and massage.[30] Although these studies are promising, more research is needed before microcurrent becomes part of routine care.

The incidence of CMT and craniofacial asymmetries has risen in recent years, requiring increased collaboration between physicians and physical therapists to appropriately manage the conditions. The increased incidence has been attributed to cultural factors, including the Safe to Sleep campaign,[31–34] decreased prone positioning for play, and the use of infant positioning equipment[35] (eg, infant swings, car seats and strollers) that has increased the time an infant's head rests against a firm surface. The American Academy of Pediatrics endorsed guidelines for the management of infants with positional plagiocephaly that are freely available at www.cns.org. In addition, the Academy of Pediatric Physical Therapy has updated clinical practice guidelines (CPGs) for the physical therapy management of CMT (2018 CMT CPG)[25] freely available at www.pedpt.com. The purpose of the 2018 CMT CPG[25] is to provide recommendations to physical therapists to improve clinical outcomes and health services for children with CMT. Adherence to the physical therapy recommendations, originally outlined in the 2013 version of the CMT CPG,[36] can improve clinical outcomes by shortening the physical therapy episode of care required to meet discontinuation criteria.[37]

To bridge the gap between research and clinical practice, this article focuses on 5 themes from the 2018 CMT CPG[25] that require collaboration between pediatricians and physical therapists to facilitate prevention of asymmetries and earlier referral for physical therapy. The ultimate goal is to optimize outcomes for infants with CMT with a shorter treatment duration, reduce the burden on

families, and decrease medical costs associated with CMT.

EDUCATE EXPECTANT PARENTS AND PARENTS OF NEWBORNS TO PREVENT ASYMMETRIES AND/OR CMT

Evidence strongly supports that early identification of postural preference and CMT results in shorter durations of physical therapy and full resolution of asymmetries.[7,26] Therefore, it is recommended that all expectant parents and parents of newborns be educated on the importance of supervised prone and tummy play when awake for 3 or more times daily, full active movement in all developmental positions, prevention of postural preferences, and the role of a pediatric physical therapist if asymmetries or motor delays are concerns. This information can be provided by prenatal educators. Pediatricians can support this education by distributing and reviewing the Academy of Pediatric Physical Therapy 2018 CMT CPG resources with parents of newborns (https://pediatricapta.org/clinical-practice-guidelines/). The goals of this education are twofold. First, to prevent asymmetries from developing by strongly encouraging supervised "tummy time while awake" in addition to safe sleep practices because many parents do not regularly place awake infants on their stomach.[38] Time in the prone position encourages activation of the cervical muscles and minimizes the potential for cranial deformation.[39] Second, if asymmetries or CMT is evident, education empowers parents to report their concerns to their pediatrician quickly so that early physical therapy intervention is initiated in a timely manner.

ASSESS NEWBORN INFANTS FOR ASYMMETRIES AND/OR CMT AND REFER FOR PHYSICAL THERAPY AS SOON AS THE ASYMMETRY IS NOTED

During the first postnatal examination, newborns (up to the first 3 days of life) can be easily screened for CMT by checking for full passive cervical rotation range of motion in both directions (chin turns past shoulder to 100°)[3,40] and lateral flexion range of motion (ear approximates shoulder)[3,40] while the infant is positioned in supine. If cervical asymmetries in range are noted, prompt referral to physical therapy is warranted to provide a comprehensive and supportive intervention program. In the past, it was common for pediatricians to instruct parents in neck-stretching exercises and only refer infants to physical therapy if the asymmetries did not resolve within a few months.[41] Recent studies support that early physical therapy significantly reduces the time to resolution as compared with parent-only stretching[42] or later referral to physical therapy,[26] that infants become more difficult to stretch as they age and develop neck control,[8] and that earlier intervention can negate the need for more invasive interventions such as surgery.[43] Therefore, the evidence supports immediate referral to a pediatric physical therapist for a comprehensive evaluation and management program, including parent and caregiver education, to reduce the costs of service and time to families and prevent secondary sequelae.

Pediatric physical therapists provide a comprehensive examination and evaluation for infants with CMT as detailed in the 2018 CMT CPG.[25] This comprehensive physical therapy examination and evaluation includes assessment of the infant's entire musculoskeletal system, classification of the severity of CMT, a standardized motor assessment, and assessment of parental routines for feeding, positioning, and equipment use, providing a thorough understanding of the potential contributing factors to asymmetrical postures. In addition, the physical therapist screens for nonmuscular causes of asymmetry and conditions associated with CMT, as detailed in the next section.

Physical therapy management of CMT is also comprehensive, focusing on 5 components as the first-choice intervention: (1) neck passive range of motion, (2) neck and trunk active range of motion, (3) development of active symmetrical movement, (4) environmental adaptations, and (5) parent or caregiver education to integrate interventions into the family's routine.[25] Thus, physical therapists do not focus only on direct or appointment-based stretching of the tight neck muscles and strengthening of the weak neck muscles but also address a broad range of developmental, environmental, and educational factors that support parents and influence resolution. These include family-tailored strategies to alternate the infant's position to encourage head turning toward the desired direction,[40] use positions for breast or bottle feeding that encourage cervical symmetry,[40] progress infant tolerance and endurance for "tummy time" or prone play,[40,44] encourage symmetry through positioning and handling,[40,45,46] minimize time in car seats and infant carriers,[40] identify and address potential motor delays,[46] and educate and support parents and caregivers.

This comprehensive physical therapy program enhances outcomes because the therapist partners with the family to determine the best ways to integrate the exercises and positioning activities throughout the infant's and family's daily routines. Physical therapy appointments are typically scheduled weekly or biweekly to strongly reinforce parent or caregiver confidence and adherence to the daily intensive home program required to resolve CMT. Although this initial visit intensity is ideal, the frequency is negotiated with the families to facilitate optimal outcomes, taking into account access

TABLE 1 These Screens by Physical Therapists Prompt a Referral to the Primary Physician for Nonmuscular Causes of Asymmetry and Conditions Associated With CMT

Screens	Description
Musculoskeletal system	Asymmetrical shape of the face, skull, and spine; asymmetrical alignment of the shoulder and hip girdles, with particular attention to cervical vertebral anomalies, rib cage symmetry, and DDH; asymmetrical PROM of the neck; and sternocleidomastoid masses or restricted movement.
Neurologic system	Abnormal or asymmetrical tone, retention of primitive reflexes, resistance to movement, atypical cranial nerve integrity, and BPI; temperament (irritability, decreased alertness); inability to perform age-appropriate developmental milestones inclusive of cognitive and social integration within the family setting.
Visual system	Asymmetrical eye tracking, noting visual field defects and nystagmus as potential ocular causes of asymmetrical postures.
Integumentary system	Skinfold asymmetry of the hips and cervical regions; atypical color and condition of the skin, with special attention to signs of pressure and trauma that might cause asymmetrical posturing.
Cardiopulmonary system	Asymmetrical coloration, rib cage expansion, and clavicle movement to rule out conditions that might cause asymmetrical posturing (eg, BPI, Grisel syndrome); check for acute upper respiratory tract distress.
Gastrointestinal history	History of reflux, constipation, preferred feeding from 1 side or difficulty with latching on to 1 side.

BPI, brachial plexus injury; DDH, developmental dysplasia of the hip; PROM, passive range of motion.

to physical therapists, travel, and cost burdens. Weekly or biweekly visits are especially important when parents are first learning to implement the home program, the infant is progressing rapidly and parents need to update the foci of the home program, or when infants are less tolerant of the home program and parents need alternative strategies to maximize the program's impact. A comprehensive management program, especially when started as soon as asymmetry is noted, can resolve the CMT in as little as 4 to 8 weeks when infants are younger than 2 months; time to resolution increases sharply when intervention is delayed until the infant is 4 to 6 months of age or older.

SCREEN INFANTS FOR NONMUSCULAR CAUSES OF ASYMMETRY AND CONDITIONS ASSOCIATED WITH CMT AND CONSULT WITH SPECIALISTS IF INDICATED

Many states allow for direct access of patients to physical therapists. In these cases, physical therapists conduct a comprehensive screen and refer infants to the primary physician if indicated by the screen. The physical therapist assesses the musculoskeletal, neurologic, visual, integumentary, gastrointestinal, and cardiopulmonary systems for nonmuscular causes of asymmetry (eg, congenital scoliosis, abnormal muscle tone, visual impairments, reflux) and conditions associated with CMT (eg, cranial deformation, brachial plexus injury, developmental dysplasia of the hip), as listed in Table 1. Up to 18% of cases with asymmetrical head and neck posturing may be due to nonmuscular causes,[47] including cervical vertebral dysfunction such as congenital scoliosis, hemivertebra, and Klippel-Feil syndrome[47,48]; neurologic

TABLE 2 Implementation Strategies for Pediatricians

1. Download the following free resources for distribution to parents and other health professionals
   a. The full 2018 CMT CPG[25] at https://journals.lww.com/pedpt/Fulltext/2018/10000/Physical_Therapy_Management_of_Congenital_Muscular.2.aspx
      - A Summary of the 2018 CMT CPG[25] Action Statements at http://links.lww.com/PPT/A223
      - Figure 1 from the 2018 CMT CPG,[25] an algorithm for early identification of CMT and referral to physical therapy at http://links.lww.com/PPT/A221
      - Figure 2 from the 2018 CMT CPG,[25] an updated 8-grade CMT severity classification system and decision tree for CMT management at http://links.lww.com/PPT/A222
   b. Academy of Pediatric Physical Therapy 2018 CMT CPG[25] resources at https://pediatricapta.org/clinical-practice-guidelines/
      - Newborn Tips for Positioning and Play
      - 2018 CMT CPG Resource Packet for Parents and Caregivers
      - 2018 CMT CPG Discharge Guide
      - 2018 CMT CPG Resource Packet for Healthcare Providers
2. Identify pediatric physical therapists and medical specialists in your community to establish collaborative relationships for referral and comanagement of infants with CMT. Physical therapists can be found at http://aptaapps.apta.org/findapt
   a. In communities with limited access to pediatric physical therapists, it may be necessary to establish consultative relationships between pediatric physical therapists and community physical therapists to provide optimal care for infants with CMT.
   b. In some states, early start programs provide pediatric physical therapy services for infants with CMT.
3. Discuss implementing the 2018 CMT CPG[25] with colleagues to improve clinical outcomes, shorten episodes of care, reduce burden on families, and decrease cost of care for infants with CMT.
   a. Collaborate with pediatric physical therapists to provide training to relevant health care providers (eg, midwives and lactation specialists) on ways to educate all parents of newborns, screen for CMT, and use the CMT CPG resources.
   b. Establish clinical pathways and electronic health record adaptations to facilitate implementation of the 2018 CMT CPG[25] recommendations, including educating all parents of newborns to prevent CMT, early identification of CMT, and earlier referral to physical therapy.
4. Provide feedback at torticolliscpg@gmail.com to inform updates to the 2018 CMT CPG[25] and the 2018 CMT CPG resources.

disorders such as central nervous system tumors, brainstem malformations, and paroxysmal torticollis that alternates sides[47,48]; ocular disorders[47–50]; and Sandifer syndrome.[47] The intent of the screen is twofold: to ensure comprehensive care regardless of whether an infant is referred from a pediatrician or the infant is first brought to a physical therapist through direct access, and to facilitate collaboration with the pediatrician and other appropriate specialists (eg, ophthalmologists, pediatric neurologists, and pediatric orthopedic surgeons).

IF THE INFANT IS OLDER AT INITIAL REFERRAL OR IS NOT PROGRESSING AS ANTICIPATED WITH PHYSICAL THERAPY MANAGEMENT, CONSULT WITH SPECIALISTS IF INDICATED

Physical therapists are encouraged to collaborate with infants' pediatricians when children present with more severe cases of CMT or when the rate of change is slower than anticipated.[25] The family may benefit from shared decision-making to determine the most appropriate plan of care for their infant when the following conditions are present:

- The infant is older than 12 months and either facial asymmetry and/or 10 to 15° of difference between the left and right sides exists in passive or active cervical rotation or lateral flexion.
- The infant is 7 months or older with a sternocleidomastoid mass.
- The infant presents with asymmetries of the head, neck, and trunk that are not starting to resolve after 4 to 6 weeks of initial intense physical therapy intervention.
- The infant demonstrates a plateau in resolution after 6 months of physical therapy intervention.

In these cases, physical therapy may be initiated or continued; however, collaboration with the pediatrician and possible specialists may be appropriate for a more in-depth examination of the musculoskeletal system, including radiograph and/or ultrasound imaging, and to determine the appropriateness of nonconservative interventions such as botulinum neurotoxin therapy or surgery.

IMPORTANCE OF A PHYSICAL THERAPY REASSESSMENT 3 TO 12 MONTHS AFTER DISCONTINUATION OF DIRECT SERVICES, THEN DISCHARGE PHYSICAL THERAPY IF APPROPRIATE

Physical therapists are encouraged to discontinue direct or appointment-based physical therapy services when infants meet these 5 criteria: (1) cervical passive range of motion is within 5° of the nonaffected side; (2) symmetrical active movement patterns are used; (3) age-appropriate motor development is achieved; (4) no head tilt during static or active postures is observed; and (5) the parents or caregivers understand what to monitor as the child grows.[25] Additionally, it is recommended that the discontinuation status be communicated to the infant's physician along with the importance of a single physical therapy assessment 3 to 12 months later. Although the short-term outcomes of conservative management are well documented, there is little evidence of

TABLE 3 CMT Quick Reference Guide

	Description
CMT	Atypical head posture due to unilateral shortening or fibrosis of the sternocleidomastoid. Named by side of the shortened or fibrotic sternocleidomastoid.
Signs and symptoms	Asymmetrical head posture into ipsilateral cervical lateral flexion and contralateral cervical rotation. Decreased PROM and AROM into contralateral cervical lateral flexion and ipsilateral rotation.
Prevent asymmetries and/or CMT	Educate all parents on the importance of supervised prone and tummy play when awake for ≥3 times daily, full active movement in all developmental positions, and prevention of postural preferences.
Screen for CMT and cranial deformation	Screen for CMT by checking for full passive cervical rotation in both directions (chin turns past shoulder to 100°) and lateral flexion range (ear approximates shoulder) while the infant is positioned supine. Screen for cranial deformation through visual observation of head shape and facial symmetry, especially the ears, eyes, and jaw line.
Screen for nonmuscular causes of asymmetry and conditions associated with CMT	Screen the following systems: musculoskeletal, neurological, visual, integumentary, cardiopulmonary, and gastrointestinal history.
Prognosis	Better prognosis if referred to physical therapy when <3 months of age.
Physical therapy management	First-choice interventions: neck PROM, neck and trunk AROM, symmetrical movement, environmental adaptations, and parent or caregiver education to integrate intervention into the family's daily routines.
Importance of a physical therapy reassessment 3–12 months after discontinuation of direct physical therapy services	Discontinue physical therapy when the infant displays full PROM within 5° of nonaffected side, symmetrical active movement, age-appropriate motor skills, no visible head tilt, and parents understand what to monitor. Physical therapy reassessment should be done 3–12 months later to assess for persistent head tilt or developmental delay; then discharge from the physical therapy episode of care if appropriate.

AROM, active range of motion; PROM, passive range of motion.

the long-term effectiveness of early physical therapy intervention such as whether the last 5° difference in cervical range of motion will resolve on its own or the CMT will reoccur after discharge from physical therapy. This may be particularly important for infants treated before crawling and walking because residual asymmetries may become more evident as infants begin to move against gravity. Öhman[51] found that 7% of preschool aged children with a history of CMT exhibited a head tilt of 5 to 10° and 26% had some degree of asymmetry in passive cervical range of motion. In addition, although the early motor delay of most infants undergoing physical therapy for CMT resolves by 8 to 15 months, similar to the general population, some children will continue to demonstrate a developmental delay.[52]

The potential for persistent head tilt or cervical asymmetry[51] or developmental delay[52] supports a single physical therapy reassessment after discontinuation of direct intervention to determine if the CMT resolution achieved at an earlier age is maintained at an older age. This comprehensive reassessment evaluates static and dynamic sitting and standing postures and movements, position transitions, neck range of motion and strength, and a standardized developmental test. The purpose is to screen for residual cervical asymmetries or delays and to determine if a new episode of physical therapy is warranted. If a physical therapy reassessment is not possible because of the family's location or health care coverage, pediatricians should be aware of the risk for asymmetries and/or developmental delays during routine physical examinations as children with a history of CMT are managed through their school years.[53]

IMPLEMENTATION RECOMMENDATIONS

Table 2 includes selected implementation strategies that may be most readily integrated into pediatric practice with the intent to enhance communication and collaboration between pediatricians and physical therapists on the management of infants with CMT. Free educational resources available at https://pediatricapta.org/clinical-practice-guidelines/ may be particularly useful for sharing with new parents or other health care providers. Table 3 includes a CMT quick reference guide.

CONCLUSIONS

CMT is a common postural condition that requires early education, identification, and intervention for optimal outcomes and reduced health care usage. Pediatric practitioners can provide parent or caregiver education to minimize or prevent asymmetries from developing and, when asymmetries develop, identify infants with CMT early and refer immediately for physical therapy intervention to improve clinical outcomes, shorten episodes of care, reduce burden on families, and decrease cost of care for infants with CMT.

ABBREVIATIONS

CMT: congenital muscular torticollis
CPG: clinical practice guideline

POTENTIAL CONFLICT OF INTEREST: The authors have indicated they have no potential conflicts of interest to disclose.

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DOI: 10.1542/peds.2019-0582 originally published online July 26, 2019;

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