Congenital muscular torticollis

General Information

Description

- condition presenting at or soon after birth in which asymmetry of sternocleidomastoid muscle length or strength leads to head tilt toward side of shorter or stronger muscle, typically with contralateral rotation of face and chin

Also called

- neonatal torticollis
- infantile torticollis
- congenital torticollis
- pseudotumor of infancy
- fibromatosis colli
- idiopathic generalized dystonia
- sternocleidomastoid torticollis
- sternomastoid torticollis
- postural torticollis
- wryneck

Types

- general presents as 1 of 3 types
  - torticollis associated with sternocleidomastoid tumor (also called fibromatosis colli)
    - about 43% of congenital muscular torticollis
    - characterized by palpable hard, moveable, fibrotic mass within sternocleidomastoid muscle
    - mass usually regresses during first year of life
  - muscular torticollis
    - about 35% of congenital muscular torticollis
    - muscular tightness, but no tumor
  - postural torticollis
    - about 22% of congenital muscular torticollis
    - no tightness or tumor

Epidemiology

Who is most affected

- infants
- male to female ratio 3:1

Incidence/Prevalence
- reported incidence typically ranges from 0.3% to 3.92%, but 16% incidence has been reported in healthy newborns\(^1\)\(^2\)\(^3\)
- 1.9% incidence of torticollis reported based on 43 cases among 2,160 infants delivered 1978-1981 (Int Orthop 1984;8(3):175)

**Likely risk factors**
- breech presentation
  - based on 1,086 cases of congenital muscular torticollis in infants delivered 1985-1997
    - 13% had breech presentation
    - another 1.6% had presentations other than usual vertex
    - incidence of torticollis was 1.9% overall (43 cases)
    - incidence was 1.2% in cephalic presentations, 4.2% in footling presentations, 6.5% in double breech presentations, and 34% in single (frank) breech presentations
    - Reference - Int Orthop 1984;8(3):175
- difficult birth\(^1\)\(^2\)

**Associated conditions**
- developmental dysplasia of hip \(^1\)
- metatarsus adductus\(^1\)
- plagiocephaly
  - torticollis reported in 5%-67% of infants with plagiocephaly
    - based on study of Texas Birth Defects registry
    - 18 facilities recorded > 20 cases of plagiocephaly
    - rates of co-diagnosis of torticollis in these 18 facilities ranged from 5% to 67%
    - rates of co-diagnosis of torticollis in 3 facilities with most recorded cases were 10% (16 of 159 cases), 29% (108 of 376 cases), and 54% (122 of 228 cases)
    - wide range may be related to inconsistently applied definition rather than true patient variability

**Etiology and Pathogenesis**

**Causes**
- etiology unclear, but theories include trauma to sternocleidomastoid muscle during head descent or due to\(^1\)\(^2\)
  - abnormal fetal positioning
  - difficult delivery
  - venous occlusion caused by persistent lateral flexion and rotation of neck in utero

**Pathogenesis**
asymmetry of sternocleidomastoid muscle length or strength leads to head tilt toward side of shorter or stronger muscle and contralateral rotation of face and chin\textsuperscript{1, 2}

asymmetry is due to replacement of affected sternocleidomastoid muscle by dense fibrous tissue\textsuperscript{1}

theories of pathogenesis include\textsuperscript{1, 2}
  o congenitally shortened sternocleidomastoid muscle tears during delivery, hematoma forms, leading to collagen deposition, fibroblast migration, and atrophy of muscle fibers
  o intrauterine or perinatal flexion, lateral bending, and rotation of head and neck causes ischemia and edema within sternocleidomastoid muscle, resulting in compartment syndrome and subsequent interstitial fibrosis

**History and Physical**

**History**

**Chief concern (CC)**
  - infant with\textsuperscript{1, 2}
    o neck mass or swelling
    o head tilt and rotation

**History of present illness (HPI)**
  - ask about\textsuperscript{1}
    o difficult labor or delivery
    o birth history suggestive for fetal malpositioning or constraint, such as
      - decreased fetal movement
      - oligohydramnios
      - high birth weight
      - breech position
      - multiple births

**Physical**

**General physical**
  - postural abnormalities may be present, including shoulder elevation or lateral bending on side of affected sternocleidomastoid muscle\textsuperscript{1, 2}

**HEENT**
  - head position\textsuperscript{1}
    o head typically tilted toward side of affected muscle, and rotated to opposite side
    o ipsilateral tilt and rotation possible but less common, and other causes should be considered
  - skull and facial asymmetry may be present, including\textsuperscript{1}
    o frontal flattening ipsilateral to affected sternocleidomastoid muscle, and contralateral parietal occipital flattening (plagiocephaly)
- jaw asymmetry with mandibular hypoplasia
- tilting of lower jaw and gum line
- smaller ear ipsilateral to affected sternocleidomastoid muscle, forward displacement of contralateral ear
- smaller eye ipsilateral to affected sternocleidomastoid muscle
- visual gaze may be oriented in direction of head rotation
- if presenting later in childhood or adulthood, deviation and decreased vertical height of orbit and maxilla on affected side
- infants with bilateral torticollis (rare) may have brachycephaly (flattening and widening of back of head)

**Neck**

- assess for limited range of motion
  - full cervical passive range of motion in rotation
    - 100-110 degrees in children < 3 years old (chin 10-20 degrees past shoulder)
    - 90 degrees in older children and adults (chin to shoulder)
  - full cervical passive range of motion in lateral flexion
    - 65-75 degrees in children < 3 years old
    - 45 degrees in older children and adults
- mass common
  - hard, moveable mass may be palpable within sternocleidomastoid muscle
  - may be tender
  - usually presents by age 3 weeks, reaches maximum size by age 1 month, and regresses during first year of life

**Back**

- older children and adults with uncorrected congenital muscular torticollis may have compensatory scoliosis

**Extremities**

- check for common associated conditions such as
  - developmental dysplasia of hip
  - metatarsus adductus

**Diagnosis**

**Making the diagnosis**

- diagnosis usually based on clinical findings and response to treatment in infant with abnormal posturing of head and neck
- imaging and other testing may be indicated if torticollis fails to respond to physical therapy

**Differential diagnosis**
- other causes of abnormal posturing of head and neck\(^1\)
  - vertebral malformation, as in Klippel-Feil syndrome
  - intracranial tumor
  - ocular abnormality, for example weakness of superior oblique or lateral rectus muscles
  - posttraumatic infection and inflammation
  - Sandifer syndrome - hiatal hernia and abnormal posture of head and neck (posturing attributed to pain associated with gastroesophageal reflux)
  - paroxysmal torticollis
- dystonic drug reaction

**Testing overview**
- testing to consider if little or no improvement after 4-6 weeks of physical therapy
  - neck ultrasound\(^3\)
  - plain x-rays\(^1\)
    - anteroposterior and lateral cervical spine
    - lateral skull
  - magnetic resonance imaging (MRI) of cervical spine\(^1\)
  - ophthalmologic exam\(^1\)

**Imaging studies**
- radiographs appear to have very low yield in congenital muscular torticollis in infants with typical clinical findings
  - based on retrospective study of 502 infants aged 0-12 months who had plain x-ray for torticollis or "heat tilt"
  - only 1 infant (who also had abnormal cervical kyphosis on exam) had unstable abnormality that might contraindicate physical therapy
  - only 4 infants had true bony vertebral abnormalities
  - 6 others had false-positive x-rays which were normal on subsequent studies
  - 25 additional infants also had spine computed tomography (CT) or magnetic resonance imaging (MRI) which were all normal
  - authors suggest that imaging be reserved for physical therapy failures or for atypical physical findings
  - Reference - *Pediatrics 2006 Dec;118(6):e1779*
- ultrasound
  - affected sternocleidomastoid muscle appears larger and hyperechoic compared to contralateral muscle\(^3\)
  - ultrasound classification of severity of fibrosis in sternocleidomastoid muscle correlated with duration of treatment for congenital muscular torticollis
    - based on cohort study
    - 50 infants < 3 months old with palpable neck mass and initial deficit of passive neck rotation > 10 degrees were evaluated by ultrasound and treated with physical therapy
ultrasound classification of severity of sternocleidomastoid fibrosis correlated initial deficit of passive neck rotation (p for trend = 0.003) and mean treatment duration (p for trend < 0.001)


### Treatment

#### Treatment overview

- usual first-line therapy is **neck-stretching exercises**
  - aggressive stretching exercises appear more likely to be successful if started before age 3-6 months ([level 2 [mid-level] evidence](#))
  - stretching treatment by physical therapist may lead to resolution of torticollis about 2 months earlier than home stretching by parents in infants with congenital muscular torticollis ([level 2 [mid-level] evidence](#))
- advise handling and positioning of infant that encourages active neck stretching and strengthening
  - addition of strength exercises (and further addition of physical therapy training) may not improve muscle function more than handling strategies alone in infants with congenital muscular torticollis ([level 2 [mid-level] evidence](#))
- consider early referral for physical therapy
- botulinum toxin A injections may be considered as adjunct to stretching exercises
  - botulinum toxin type A reported to be effective for congenital muscular torticollis unresponsive to conservative management in children aged 6-18 months ([level 3 [lacking direct] evidence](#))
  - botulinum toxin injection reported to have inconsistent efficacy for congenital muscular torticollis in older children and adults ([level 3 [lacking direct] evidence](#))
- other adjunctive therapies, such as orthotics or microcurrent therapy, may also be considered
- surgery may be indicated for congenital muscular torticollis refractory to 6-12 months of conservative treatment

#### Activity

- neck stretching exercises
  - passive neck stretching
    - exercises should stretch involved sternocleidomastoid muscle, not both sides of neck
    - hold each stretch for 30-50 seconds
    - perform 3 repetitions of each stretch
    - perform series of exercises 6-8 times/day
  - aggressive stretching exercises appear more likely to be successful if started before age 3-6 months ([level 2 [mid-level] evidence](#))
    - based on retrospective cohort study
    - retrospective cohort of 57 consecutive children treated for congenital muscular torticollis, children < 18 months old treated with passive and active stretching exercises
    - 100% of 28 children < 3 months old had excellent outcome without surgery
4 (25%) of 16 infants aged 3-6 months had persistent symptoms despite exercises and underwent surgery

5 (71%) of 7 infants aged 6-18 months had persistent symptoms despite exercises and underwent surgery

100% of children > 18 months old had surgery


- passive stretching exercises every 3 hours reported to have 100% success rate (level 3 [lacking direct] evidence)
  - based on prospective series of 45 infants with congenital muscular torticollis starting treatment at age 15-120 days

- stretching treatment by physical therapist may lead to resolution of torticollis about 2 months earlier than home stretching by parents in infants with congenital muscular torticollis (level 2 [mid-level] evidence)
  - based on small randomized trial
  - 20 infants with congenital muscular torticollis randomized to stretching treatment by physical therapist vs. stretching treatment by parents
  - in physical therapist group stretching exercises in both rotation and lateral flexion done for about 15 minutes 3 times/week without any stretching at home
  - in parent stretching group stretching exercises done for up to 15 minutes twice daily 7 days/week
  - 2 of 10 infants in parent stretching group were changed to physical therapist treatment but analyzed by intention-to-treat
  - all 20 infants achieved good range of motion (in rotation and lateral flexion)
  - comparing physical therapist stretching vs. parent stretching
    - mean time to achieve good range of motion 0.9 months vs. 3.1 months (p < 0.001)
    - median time to achieve good range of motion 0.7 months vs. 3 months
    - mean time to symmetric head posture 2.5 months vs. 4.5 months (p = 0.03)
    - median time to symmetric head posture 2.1 months vs. 4.1 months
  - Reference - PM R 2010 Dec;2(12):1073

- infant handling and positioning
  - advise caregivers on how to increase active neck stretching
  - encourage infant to look at objects on less preferred side
  - position infant in crib so that door is on less preferred side
  - hold infant with less preferred side facing outward
  - encourage "tummy time" while awake to strengthen neck muscles
  - use car seats only while in moving vehicles and not as infant carrier to avoid prolonged periods in 1 position

- addition of strength exercises (and further addition of physical therapy training) may not improve muscle function more than handling strategies alone in infants with congenital muscular torticollis (level 2 [mid-level] evidence)
  - based on small randomized trial
37 infants aged 1-10.5 months (mean 4.5 months) with congenital muscular torticollis randomized to 1 of 3 groups
  o handling strategies
  o handling strategies plus specific strength exercises
  o handling strategies plus specific strength exercises plus weekly training by physical therapist
4 dropped out
infants assigned to handling strategies alone were given specific strength exercises if no improvement after 2 months
31 infants achieved symmetric head posture by age 12 months
mean treatment time 3.5 months (range 1-5.5 months)
no significant differences in mean treatment time or achievement of symmetric head posture among groups
Reference - Physiother Theory Pract 2011 Oct;27(7):463 full-text

postural control may not reduce sternocleidomastoid tumor thickness compared to manual stretching in infants < 6 months old ([level 3 [lacking direct] evidence])
  o based on randomized trial without clinical outcomes
  o 76 infants < 6 months old with congenital muscular torticollis were randomized to postural control vs. manual stretching twice weekly
  o goal of postural control was to strengthen unaffected side of sternocleidomastoid muscle and elongate posterior fibers of affected side
  o 8% withdrew (all in postural control group) and were excluded from analyses
  o comparing postural control vs. manual stretching
    ▪ mean reduction in sternocleidomastoid tumor thickness by ultrasound 6.9 mm vs. 6.1 mm (not significant)
    ▪ mean treatment duration 93 days vs. 88 days (not significant)

addition of manual therapy to physical therapy not associated with improved symptoms in infantile torticollis ([level 2 [mid-level] evidence])
  o based on small randomized trial
  o 32 infants aged 3-6 months with torticollis randomized to manual therapy plus child physical therapy vs. child physical therapy alone
  o addition of manual therapy to child physical therapy associated with
    ▪ nonsignificantly greater improvement in lateral flexion (p = 0.092)
    ▪ no significant difference in improvement in head righting reaction
  o Reference - Acta Paediatr 2011 May;100(5):687

Medications
  • botulinum toxin A injections may be considered to[3]
    o enhance effectiveness of stretching exercises
    o allow strengthening of overstretched, weakened contralateral neck muscles
- Botulinum toxin type A reported to be effective for congenital muscular torticollis unresponsive to conservative management in children aged 6-18 months (level 3 [lacking direct evidence])
  - Based on 2 case series
  - 27 children ages 6-18 months with congenital muscular torticollis and failure to progress with conservative management received botulinum toxin type A injections into sternocleidomastoid and/or upper trapezius muscle
    - 3 children had repeat injections
    - 20 children (74%) had improved cervical rotation or head tilt
    - 2 children (7%) had transient adverse events, including dysphagia and neck weakness
- Botulinum toxin type A reported to be effective in 14 of 15 children with congenital muscular torticollis poorly responsive to conservative therapy (*J Craniofac Surg* 2005 Mar;16(2):321)
- Botulinum toxin injection reported to have inconsistent efficacy for congenital muscular torticollis in older children and adults (level 3 [lacking direct evidence])
  - Based on 2 case series with inconsistent results
  - Case series with 7 cases of congenital muscular torticollis presenting in late childhood or adults
    - Only 2 patients had modest benefit
  - Reference - *Neurology* 2006 Sep 26;67(6):1083
  - Case series of 3 adult patients with persistent congenital muscular torticollis
    - All 3 treated with botulinum toxin injections with long lasting benefit

**Surgery and procedures**
- Surgery to lengthen sternocleidomastoid muscle may be indicated for
  - Persistent symptoms after 1 year of conservative therapy
  - > 15 degree deficit in rotation range persisting after 6 months of stretching exercises
- Procedure options include
  - Unipolar release of sternal and/or clavicular heads of sternocleidomastoid muscle
  - Bipolar release at muscle insertion at mastoid process with or without Z-plasty to preserve normal contour of sternocleidomastoid muscle in neckline
- Endoscopic release may be possible
- Postoperative intensive physical therapy including passive range of motion and active strengthening exercises indicated for ≥ 3 months
- Complications may include
  - Hematoma
  - Adverse cosmetic outcomes, such as
    - Hollowing at base of neck
    - Bony prominence of sternal head of clavicle
Consultation and referral
- consider early referral for physical therapy\(^1\)
  - to instruct caregivers on positioning and how to perform stretching exercises
  - to address associated motor delays
- referral to orthopedic surgeon if congenital muscular torticollis identified after early infancy, or if refractory to conservative management\(^1\)

Other management
- adjunctive therapy tools\(^1\)
  - may be considered in infants with lateral head tilt refractory to exercise
  - devices provide sensory feedback when head is tilted in preferred direction, prompting neuromuscular response to correct head tilt
  - infant must show signs of motor readiness to respond to feedback
  - options include tubular orthosis for torticollis collar and elastic therapeutic tape
- in infants with associated plagiocephaly, cranial remodeling orthosis may be considered\(^1\) (see Positional head deformity for additional information)

- microcurrent therapy reported to improve head tilting angle and neck rotation range of motion more than ultrasound therapy plus stretching exercises in infants with congenital muscular torticollis \(^{level 3 [lacking direct] evidence}\)
  - based on small nonrandomized trial limited to 14-day follow-up
  - 15 infants with congenital muscular torticollis treated with 1 of 2 approaches
    - microcurrent therapy for 30 minutes then stretching exercises for 2 minutes
    - ultrasound therapy then stretching exercises for 30 minutes
  - patients received 3 treatments weekly and evaluated on day 14
  - comparing microcurrent therapy vs. ultrasound plus stretching exercises at baseline
    - mean age 10 months vs. 7.1 months
    - male gender in 6 of 7 infants vs. 3 of 8 infants
  - comparing microcurrent therapy vs. ultrasound plus stretching exercises at 14 days
    - mean head tilting angle at supine 6.7 degrees vs. 13.9 degrees \(p < 0.01\)
    - mean neck rotation range of motion 80.7 degrees vs. 66.3 degrees \(p < 0.05\) after adjusting for 5-degree difference at baseline

  Reference - PM R 2009 Aug;1(8):736

Complications and Prognosis

Complications
- positional head deformity common\(^1\)
• complications in older children and adults with uncorrected congenital muscular torticollis may include:  
  - craniofacial asymmetry  
  - orbital and maxillary deformity and deviation  
  - mandibular and occlusal abnormalities  
  - scoliosis  
  - progressive limitation of head movement

Prognosis
• > 90% of infants have good-to-excellent outcomes with nonsurgical management.
• early diagnosis and treatment, less severe range of motion limitation, and absence of sternomastoid tumor associated with improved prognosis and reduced treatment duration.
• prognosis by type  
  - postural torticollis - usually resolves with short-term conservative therapy  
  - muscular torticollis - about 3% require invasive therapy if not treated early  
  - torticollis associated with sternomastoid tumor - about 8% require invasive therapy if not treated early

• most infants with congenital muscular torticollis have good outcome  
  - prospective study of 1,086 infants presenting with congenital muscular torticollis in first year of life and followed at 1 center in Hong Kong  
  - 24.5% had initial passive rotation deficit < 10 degrees and had excellent or good outcome with active home positioning and stimulation  
  - of infants with rotation deficits > 10 degrees treated with manual stretching program, 91.1% had excellent or good outcomes and 5.1% had surgery  
  - most important prognostic factors for need of surgery were severe limitation of passive neck rotation and late age of presentation

Prevention and Screening
• screening for congenital muscular torticollis recommended as early as 1-2 days after birth. (see also Initial newborn assessment)

Guidelines and Resources

Guidelines
• Cincinnati Children's Hospital Medical Center (CCHMC) Best evidence statement (BEST) on prognosis of infant development with plagiocephaly, torticollis can be found at CCHMC or at National Guideline Clearinghouse 2012 Jan 2:34044
• CCHMC evidence-based guideline on therapy management of congenital muscular torticollis in children ages 0 to 36 months can be found at CCHMC
Section on Pediatrics of the American Physical Therapy Association clinical practice guideline on physical therapy management of congenital muscular torticollis can be found at Pediatr Phys Ther 2013 Winter;25(4):348

Review articles
- review can be found in Curr Opin Pediatr 2006 Feb;18(1):26
- review can be found in ORL J Otorhinolaryngol Relat Spec 2005;67(6):344
- review of torticollis in infants and children can be found in Instr Course Lect 2006;55:647
- case presentation can be found in Am Fam Physician 2007 Oct 15;76(8):1197 full-text

MEDLINE search
- to search MEDLINE for (Congenital muscular torticollis) with targeted search (Clinical Queries), click therapy, diagnosis, or prognosis

Patient Information
- handout from Parents' Common Sense Encyclopedia
- handout from Texas Pediatric Surgical Associates and in Spanish

ICD-9/ICD-10 Codes

ICD-9 codes
- 754.1 congenital musculoskeletal deformities of sternocleidomastoid muscle
- 723.5 torticollis, unspecified

ICD-10 codes
- Q68.0 congenital deformity of sternocleidomastoid muscle
- P15.2 sternomastoid injury due to birth injury
- M43.6 torticollis

References

General references used

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