# CHAPTER 38
# Sternomastoid Tumour of Infancy and Congenital Muscular Torticollis

**Lukman O. Abdur-Rahman**
**Brian H. Cameron**

## Introduction

Sternomastoid tumour (SMT) of infancy is usually associated with congenital muscular torticollis (CMT); these will be discussed together in this chapter. The term "tumour" is a misnomer because it is a congenital fibrotic process; it also is referred to as congenital fibromatosis coli. The terms "torticollis" and "wryneck" actually describe the tilting and rotation of the head and neck that results from the contracture of the sternocleidomastoid muscle. The torticollis, when untreated, results in plagiocephaly, hemifacial hypoplasia, and body distortion. The key to preventing deformity is early diagnosis and passive stretching exercises (PSEs) of the affected muscle, with only 5% of cases needing surgical intervention in a large prospective series.[1]

## Demography

The prevalence of SMT and CMT in Africa is unknown.[2] At the University of Ilorin Teaching Hospital in Nigeria over a period of 10 years (1999–2008), only 15 cases presented at the outpatient clinic and only one had surgical intervention at 10 months of age because of severe torticollis. CMT is reported to occur in 0.3–2% of all births, with a slight male preponderance. The right side is affected in 60% of the cases, and 2–8% are bilateral.[3]

## Aetiology

The sternocleidomastoid muscle is enveloped by the deep cervical fascia as it attaches to the clavicle, manubrium, and mastoid process. The aetiology of SMT is unknown, but the current theory is that it results from abnormal intrauterine positioning, leading to intramuscular compartment syndrome and ischaemic muscle injury with subsequent fibrosis and contracture of the sternomastoid muscle.[4] Muscle biopsy shows replacement of muscle bundles with a mass of maturing fibrous tissue—evidence that the pathology may begin prenatally.[3]

Contracture of the sternomastoid muscle on one side causes the head and neck to tilt (flex) to the ipsilateral side and the face to turn toward the contralateral side (Figure 38.1).

If untreated, the sternomastoid tumour naturally resolves completely in 50–70% of the cases by 6 months of age, with muscle shortening persisting in 5–7% after 1 year.[1,5] Hence, late presentations are usually accompanied by fibrotic and shortened sternocleidomastoid muscle from a missed or unrecognised sternomastoid tumour.

## Clinical Presentation

### History

Infants present with either a lump in the neck, or with a head tilt that is not correctable by repositioning. SMT is usually absent at birth and presents between 3 weeks to 3 months of age. There is a high incidence of breech presentation or assisted delivery. It is important to reassure the parents that the obstetric difficulty is thought to be the result rather than cause of the shortened sternomastoid muscle.[4]

Many parents present their children after 3 months of age in the African setting because they presume the abnormal position to be due to poor neck control in infants before this age (Figure 38.2). Some of the cases are first noticed by grandmothers while doing the traditional body massage for the babies.

### Physical Examination

A sternomastoid tumour is defined by the presence of a palpable, hard, spindle-shaped, painless, 1–3 cm diameter swelling within the substance of the sternocleidomastoid muscle, usually located in the lower and middle third of the muscle. The mass may be confused with a lymph node or neoplasm, which is far less likely at this age.

Children presenting with CMT should have the entire length of the sternomastoid muscle palpated to determine whether the swelling or area

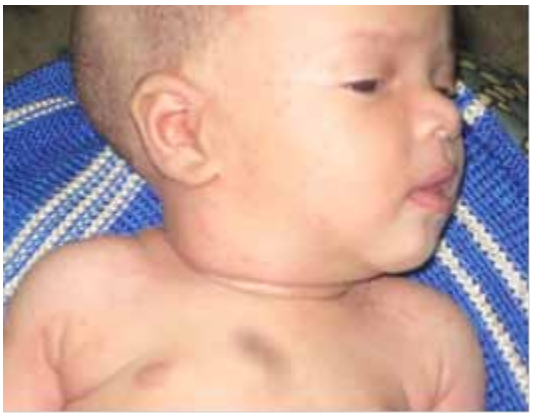

*Figure 38.1: Sternomastoid tumour on the right side, with the child's head turned away from the affected side.*

*Figure 38.2: Note the posture of the older baby with left sternomastoid tumour.*

of fibrosis is present and in what proportion. More than 50% of children with CMT will have SMT at the time of presentation.[1] The anterior border of the sternomastoid muscle may reveal a tight band of muscle, especially in older children. Bilateral sternomastoid tumour with torticollis creates difficulty in confirming the diagnosis from examination.

Infants with a head tilt will prefer to look away from the affected muscle. The most important part of the physical exam is to determine the presence and severity of limitation of passive neck rotation. Gentle neck rotation with the baby supine and head held over the side of the examining table should normally allow the chin to reach to or past the shoulder, 90–110 degrees from the neutral position (Figures 38.3 and 38.4). With CMT, there is limited rotation towards the affected side, which can be graded as mild (>80 degrees), moderate (45–80 degrees), or severe (<45 degrees) (Figure 38.5).[6]

The head should be examined from the back and top of the baby to document any plagiocephaly, a flattening of the contralateral occiput that results from persistent lying on one side; this is sometimes accompanied by contralateral flattening of the forehead (Figure 38.6).

Mild facial asymmetry may be noted, even at an early presentation; this asymmetry worsens when the torticollis is severe and untreated. The degree of hemifacial hypoplasia can be determined by the angle between the plane of the eyes and the plane of the mouth (Figure 38.7).

Older children with long-standing torticollis may have secondary compensation resulting in musculoskeletal deformities, including elevation of the ipsilateral shoulder to maintain a horizontal plane of vision, twisting of the neck and back to maintain a straight line of sight, and wasting of the neck muscles from disuse atrophy (Figure 38.8).There may be accompanying muscle spasm with cervical and thoracic scoliosis.

Developmental dysplasia of the hip (DDH) is seen in 5–8% of children with CMT, so this should be screened for on the initial physical examination. Clues on inspection are asymmetric thigh folds and apparent leg length discrepancy; the Ortolani and Barlow tests for hip stability should be done. The American Academy of Pediatrics recommends an ultrasound at 6 weeks of age or radiographs of the hips at 4 months of age in children at higher risk, which includes girls having breech presentations.[7] No specific mention is made of torticollis as a risk factor, but DDH may occur in at least 4% of infants with torticollis,[5] so it may be prudent to screen children with hip ultrasound if it is available. Metatarsal adductus and calcaneovalgus may also be associated with abnormal intrauterine positioning.

## Differential Diagnoses

The clinical features of SMT when associated with CMT are pathognomonic and should not be confused with other lateral neck masses, such as cystic hygroma, branchial cyst, or hemangioma. Enlarged cervical nodes are rare in infancy, as are neoplasms.

Congenital torticollis may present without an SMT, but most will still have some palpable thickening and shortening of the muscle. If there is a head tilt without any limitation of rotation of the neck, then causes of postural torticollis should be considered. These would include congenital hemivertebra, Klippel-Feil syndrome (atlanto-axial fusion), strabismus, and Sandifer syndrome caused by chronic gastro-oesophageal reflux (see Table 38.1). Familial and hereditary sternomastoid muscle aplasia have also been reported.[8,9]

## Investigations

Clinical examination confirms the diagnosis of sternomastoid tumour and torticollis in most cases, and no investigations are routinely required. However, imaging studies can occasionally be used to exclude other conditions when the clinical findings are equivocal or atypical.

Plain cervical radiographs are of limited use in nontraumatic infant torticollis due to their low true-positive yield; more false-positives were identified in one retrospective review.[10] In only 1 of 502 cases was there a craniocervical anomaly, and the study concluded that physical examination could safely eliminate the need for routine radiography in infant torticollis.

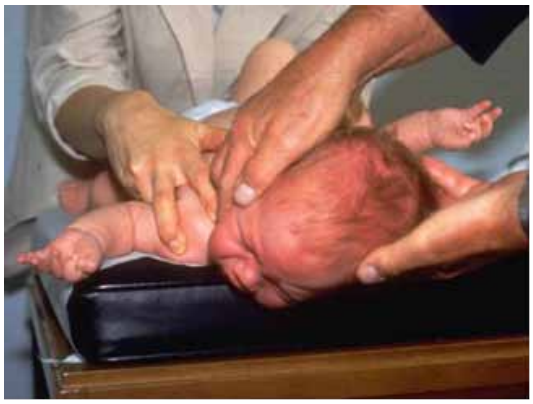

*Figure 38.3: Normal rotation of the neck to the left past the shoulder.*

*Figure 38.4: Limitation of passive neck rotation towards the right (affected) side.*

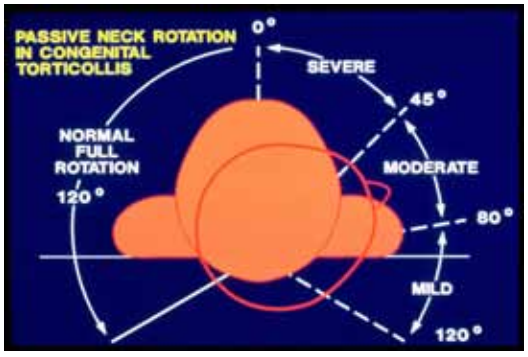


*Figure 38.5: Measurement of passive range of neck motion from the midline.*

*Figure 38.6: Right occipital flattening (plagiocephaly) with left sternomastoid shortening.*

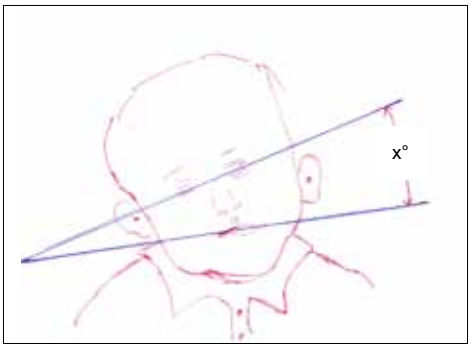


*Figure 38.7: Degree of hemifacial hypoplasia measured by the angle between the plane of the eyes and mouth (x°).*

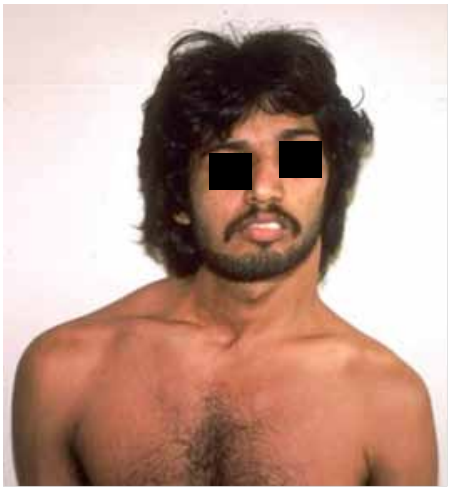

*Figure 38.8: Long-standing right sternomastoid shortening causing facial asymmetry with secondary neck, shoulder, and back deformities.*

*Table 38.1: Differential diagnoses of torticollis.*

| |
|---|
| **Congenital** |
| 1. Sternomastoid tumour |
| 2. Muscular torticollis |
| 3. Congenital vertebral anomalies (cervical hemivertebral) |
| 4. Klippel-Feil syndrome (atlanto-axial fusion) |
| **Trauma** |
| 1. Rotary subluxation of atlantoaxial or atlanto-occipital joints (post ear, nose, throat (ENT) surgery, such as tonsillectomy; retropharygeal abscess drainage) |
| 2. Cervical spine fracture |
| 3. Clavicular fracture |
| **Inflammatory** |
| 1. Grisel syndrome |
| 2. Diskitis |
| 3. Vertebral osteomyelitis |
| 4. Juvenile rheumatoid arthritis |
| 5. Cervical disc calcification |
| 6. Retropharyngeal abscess |
| 7. Cervical lymphadenitis |
| 8. Acute lymphoblastic leukaemia |
| **Neurologic** |
| 1. Posterior fossa tumour |
| 2. Syringomyelia |
| 3. Arnold-Chiari malformation |
| 4. Paroxysmal torticollis of infancy |
| 5. Cerebral palsy |
| 6. Strabismus |
| **Others** |
| 1. Sandifer syndrome in chronic gastro-oesophageal reflux |
| 2. Thymitis |
| 3. Thyroiditis |
| 4. Postural; familial |

Ultrasonography has been used to classify, monitor progress, and predict outcome of congenital muscular torticollis (Table 38.2).[11] However, it is not used routinely in most centres, and treatment decisions should be based on clinical findings. Fine needle aspiration (FNA) cytology and open muscle biopsy are not necessary and may be misleading because the histology may look similar to a fibrous neoplasm. These procedures should be reserved for atypical cases or when the muscle tumour does not resolve as expected (Figure 38.9).

## Management

Most cases of SMT and CMT are managed nonoperatively. Babies are encouraged to actively look towards the affected side. In the University of Ilorin Teaching Hospital, mothers are encouraged to be turning the face of the child in the ipsilateral direction while "backing" their babies in the traditional way (Figure 38.10). This position also helps in keeping the hip flexed and abducted, preventing or reducing the chance of a hip dislocation; this could serve as treatment for associated hip dysplasia. This management replaces the Pavlik harness in the African setting. For kangaroo bag (frontal child carrier bags) users, this affords the mother the opportunity to do the massage of the neck and manual stretching even when in transit.

Minor cases will resolve spontaneously with or without treatment, but there is evidence that early PSEs are effective in almost all cases when initiated prior to 3 months of age.[5,6,12,13]

### Passive Stretching Exercises

The PSE technique is as follows:[6] With the baby supine and head suspended over the side of the examining table, one adult holds the baby's shoulders while the other rotates the neck to the same side as the affected muscle. Gentle but firm pressure and some flexion are applied at the limit of rotation to maximally stretch the muscle for 10–15 seconds (Figure 38.11). This procedure is repeated 10 times, twice daily.

The keys to success are to explain the diagnosis and prognosis to the parents, demonstrate the PSE, watch them do the PSE in the clinic, and then follow-up in the clinic at 2 and 6 weeks to ensure progress. Parents are motivated by telling them that PSE prevents the need for surgery in most cases. Reassure the parents that babies get used to the exercises and do not remember the discomfort when they are older. With proper instruction, PSE will not harm the child. The sternomastoid scar will occasionally "snap", which may lead to some temporary swelling, but this often results in an improved range of motion.[14]

The PSE should be consistent and continuous until the muscle tumour and contracture resolve, generally within 6–8 months.[12] PSE treatment is successful in more than 90% of cases when commenced within the first 3 months of life.[1,6,13] Emery[15] followed 101 children who started treatment at a mean age of 4 months and found that the average PSE treatment duration was 4.7 months, with all but one child achieving a full passive range of motion.

A child older than 6–8 months of age is less cooperative with the PSE, and surgery is more likely to be necessary because the contracture is tighter and more fibrotic.

A physiotherapist can be engaged to do the initial demonstration and follow-up of progress, especially when mothers are afraid to do the stretching or there is no response to this treatment as expected. Physiotherapy would need to be done at least three times a week to be effective;[5] a professional physiotherapist service is an additional cost that many families would want to avoid and may lead to hospital default. To prevent this, the confidence of the parents is gained by providing adequate information and instruction on the pathology and the treatment options.

A neck brace has been used in older children as an adjunct to PSE or after surgical treatment.[16] If used, the neck brace should be carefully fitted and not worn at night.

Plagiocephaly can be prevented or treated by positioning the head to avoid persistent lying on the same side of the occiput. Measures tried

*Table 38.2: Ultrasound prediction of outcome of congenital muscular torticollis.*

| | | |
|---|---|---|
| Type I, 15% | Fibrotic mass | Spontaneous resolution |
| Type II, 77% | Diffuse fibrosis mixing with normal muscle | Spontaneous resolution |
| Type III, 5% | Without normal muscle in the involved muscle | Surgical intervention |
| Type IV, 3% | Fibrotic cord in the involved muscle | Surgical intervention (odds ratio = 31.54, p = .0196) |

Source: Adapted from Hsu TC et al. Correlation of clinical and ultrasonographic features of congenital muscular torticolis. Arch Phys Med Rehabil 1999; 80:637–641.

*Figure 38.9: Residual left sternomastoid swelling in a five-month-old infant at 8 weeks of physiotherapy. The tumour progressively reduced over 4 months.*

*Figure 38.10: A woman backing an infant with the head turned to the ipsilateral side of the sternomastoid tumour.*

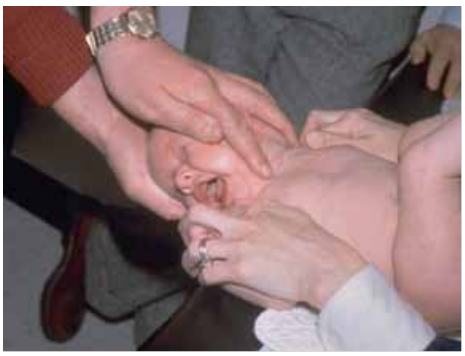

*Figure 38.11: The baby's mother does PSE assisted by a friend, while the surgeon watches.*

have included placing toys and desirable objects on the ipsilateral side of the lesion so that the child turns towards it. The child could also be put to sleep with the head facing the ipsilateral side. Helmet treatment has been described in some children with severe plagiocephaly,[17] but it is usually impractical.

## Operative Treatment

Surgical treatment is required in only about 5% of patients seen early, but is necessary in half of those presenting after 6 months of age.[6] The indications for surgery are:

1. late diagnosis, after 12 months of age; and
2. failure after at least 6 months of PSE with a significant head tilt, persistent deficit of passive neck rotation, and a tight band in the sternomastoid muscle, often with hemifacial hypoplasia.

There is evidence that surgical release of the sternomastoid contracture between 12 and 18 months of age maximises spontaneous correction of plagiocephaly. Surgical correction also results in adequate mobility and acceptable cosmetics in more than 90% of cases, with some benefit even in older children.[18] Other authors recommend delaying operative treatment until after 5 years of age, when the patient can comply with postoperative bracing and physiotherapy.[19]

### Operative Procedure—Open Technique

1. The patient is placed under general anaesthesia with endotracheal intubation or the use of a laryngeal mask airway. Anaesthetic intubation difficulty may arise from abnormal tilt in the trachea, so a prior cervical x-ray will guide the anaesthetist. Where available, fibre-optic guided endotracheal intubation is the best.
2. The patient is positioned supine with the shoulder raised and neck rotated to the contralateral side.
3. A 3–4cm transverse skin incision is made 1 cm above the sternal and clavicular origin of the sternomastoid muscle.
4. The platysma is carefully divided along the line of incision to avoid injury to the external jugular vein and accessory nerve.
5. The two heads of the sternomastoid muscle are dissected free from the anterior and posterior layers of the investing fascia and are subsequently divided using diathermy to prevent bleeding; some advocate excision of a 1-cm segment of the muscle.[18]
6. Tight deep cervical fascia should be released, testing lateral and rotational movement carefully under anaesthetic.
7. In severe cases of contracture, division of the upper end of the sternomastoid may be necessary.
8. The platysma is then sutured with 4-0 interrupted absorbable suture, and the skin is closed with 5-0 absorbable suture. There is no need for a drain provided haemostasis is well secured.

Postoperatively, physiotherapy is resumed after wound healing to maintain a full range of motion; some authors recommend using a neck brace for several months.[18]

Alternative surgical approaches that have been described are a sternomastoid lengthening technique using a Z-plasty,[20] and endoscopic tenotomy of the sternomastoid contracture.[21]

### Postoperative Complications

1. A hematoma usually will resolve, but sometimes requires aspiration or drainage.
2. Residual contracture from incomplete division of both heads of sternomastoid or cervical fascia over the posterior triangle of the neck would need a reoperation.
3. Recurrent CMT is rare.

The cosmetic appearance may be disfiguring, especially in the older child with severe contracture. This is usually due to anomalous reattachment of the clavicular head of sternomastoid or loss of the sternomastoid column of the neck.

## Prognosis and Outcome

The prognosis for sternomastoid tumour is generally excellent if treated early. The child is able to achieve full range of head movement, and the swelling resolves within 6 months. The majority of those who have surgery also do well.

Follow-up should continue until the sternomastoid muscle resolves and feels normal, and full neck rotation is achieved. Older children should be monitored for development of scoliosis.

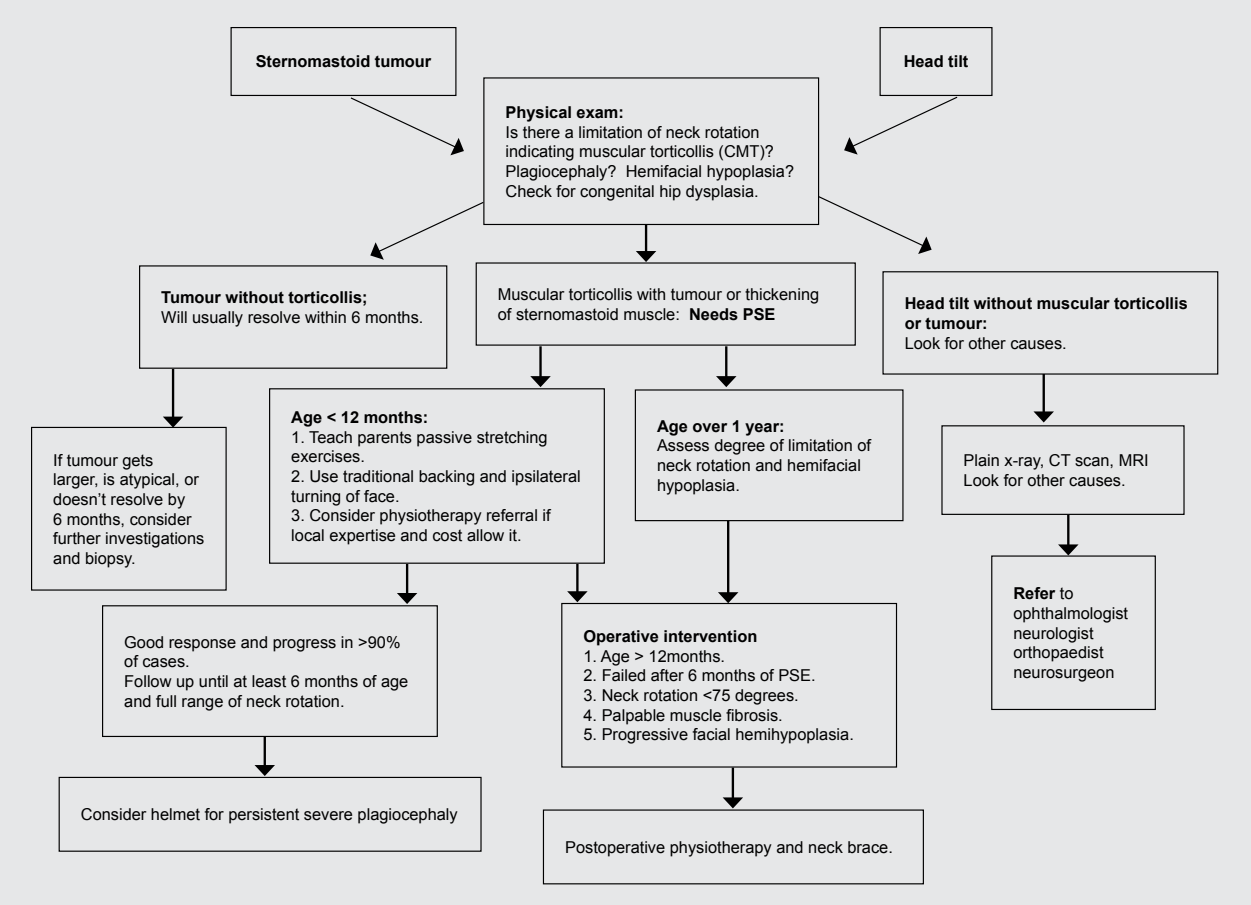


*Figure 38.12: Flow chart.*

## Prevention

There is no primary prevention because the aetiology of sternomastoid tumour is unknown. Secondary and tertiary prevention strategies will identify early cases of CMT in children with SMT or breech presentation, initiate early PSE to prevent further deformity, and avoid the need for surgery. The contracture and torticollis resulting from the sternomastoid tumour should be prevented by early identification, adequate physiotherapy, and good follow-up.

## Ethical Issues

Full involvement and education of the parents in the PSE programme requires time and compassionate commitment on the part of the surgeon. Giving information that the mass and head tilt will resolve spontaneously in many of the children may make mothers default from regular visits due to the constraint of cost, distance to the hospital, and long waits in the outpatient clinic. Referral to a costly supervised stretching programme by the physiotherapist should be avoided if it will discourage the parents and lead them to default further outpatient visits. The accessibility and affordability of care make follow-up challenging in the African setting.

## Summary

The flow chart presented in Figure 38.12 summarises the recommendations of this chapter.

## Evidence-Based Research

Many prospective studies have been conducted to predict the outcome of congenital muscular torticollis, but none were randomised. The study in Table 38.3, however, involves a standardised programme of manual stretching.

*Table 38.3: Evidence-based research.*

| | |
|---|---|
| **Title** | Clinical determinants of the outcome of manual stretching in the treatment of congenital muscular torticollis in infants. A prospective study of 821 cases |
| **Authors** | Cheng JCY, Wong MWN, Tang SP, Chen TMK, Shum SLF, Wong EMC |
| **Institution** | The Chinese University of Hong Kong and the Prince of Wales Hospital, Hong Kong, China |
| **Reference** | J Bone Joint Surg Am 2001; 83:679–687 |
| **Problem** | The natural history of congenital muscular torticollis and the outcome of different treatment modalities have been poorly investigated, and the results of treatment have varied considerably. |
| **Intervention** | Standardised program of manual stretching. |
| **Comparison/ control (quality of evidence)** | The study involved three groups:<br>1. Palpable sternomastoid tumour group<br>2. Muscular torticollis group (thickening and tightness of the sternocleidomastoid muscle)<br>3. Postural torticollis group (torticollis but no tightness or tumour).<br>Level 2 evidence. |
| **Outcome/ effect** | Controlled manual stretching is safe and effective in the treatment of congenital muscular torticollis when a patient is seen before the age of 1 year. The most important factors that predict the outcome of manual stretching are the clinical group, the initial deficit in rotation of the neck, and the age of the patient at presentation.<br>Surgical treatment is indicated when a patient has undergone at least 6 months of controlled manual stretching and has residual head tilt, deficits of passive rotation, lateral bending of the neck of >15°, a tight muscular band or tumour, and a poor outcome according to a special assessment chart. |
| **Historical significance/ comments** | A sternomastoid tumour was involved in 55% of the patients in the study.<br>Eight percent of the sternomastoid tumour group needed surgical intervention, compared to 3% and 0% in the muscular and postural torticollis groups, respectively.<br>The worse the neck rotator deficit, the higher the need for surgical intervention. |

## Key Summary Points

1. Sternomastoid tumour is usually associated with congenital muscular torticollis.
2. All children with sternomastoid tumour should be thoroughly examined for associated conditions, such as torticollis, facial and cranial asymmetry, and hip dysplasia.
3. The degree of limitation of rotation of the neck towards the affected side should be assessed. Congenital muscular torticollis is a clinical diagnosis that does not require other investigations.
4. Differentials should be considered and investigated in the absence of sternomastoid mass or nonresolving torticollis with nonoperative treatment.
5. With early diagnosis and treatment, more than 90% of the cases resolve with an adequately supervised passive stretching exercise programme.
6. Surgery is indicated in late presentation (>1 year), persistent limitation of neck rotation with head tilt, and progressive facial asymmetry.
7. Follow-up should continue until the tumour resolves and neck rotation normalises.

## References


1. Cheng JC, Wong MW, Tang SP, Chen TMK, Shum SLF, Wong EMC. Clinical determinants of the outcome of manual stretching in the treatment of congenital muscular torticollis in infants. A prospective study of 821 cases. J Bone Joint Surg Am 2001; 83:679–687.
2. Nwako FA. Torticollis. In: Nwako FA, ed. A Textbook of Pediatric Surgery in the Tropics. MacMillan Press Limited, 1980, Pp 277–278.
3. Beasley S. Torticollis. In: Grosfeld JL, O'Neill JA Jr, Fonkalsrud EW, Coran AG. Pediatric Surgery, 6th ed. Mosby, 2006, Pp 875–881.
4. Davids JR, Wenger DR, Mubarak SJ. Congenital muscular torticollis: Sequela of intrauterine or perinatal compartment syndrome. J Pediatr Orthop 1993; 13:141.
5. Cheng JC, Tang SP, Chen TM, Wong MWN, Wong EMC. The clinical presentation and outcome of treatment of congenital muscular torticollis in infants—a study of 1086 cases. J Pediatr Surg 2000; 35:1091–1096.
6. Cameron BH, Langer JC, Cameron GS. Success of nonoperative treatment for congenital muscular torticollis is dependent on early therapy. Pediatr Surg Intl 1994; 9:391–393.
7. Goldberg MJ. Early detection of developmental hip dysplasia: synopsis of the AAP Clinical Practice Guideline. Pediatr Rev 2001; 22:131–134.
8. Adams SB Jr, Flynn JM, Hosalkar HS. Torticollis in an infant caused by hereditary muscle aplasia. Am J Orthop 2003; 32: 556–558.
9. Hosalkar HS, Gill IS, Guyar P, Shaw BA. Familial torticollis with polydactyly: manifestation in three generations. Am J Orthop 2001; 30:656–658.
10. Snyder EM, Coley BD. Limited value of plain radiographs in infant torticollis. Pediatr 2006; 118:e1779–e1784.
11. Hsu TC, Wang CL, Wang MK, Hsu KH, Tang FT, Chen HT. Correlation of clinical and ultrasonographic features of congenital muscular torticollis. Arch Phys Med Rehabil 1999; 80:637–641.
12. Celayir AC. Congenital muscular torticollis: early and intensive treatment is critical. A prospective study. Pediatr Int 2000; 42:504–507.
13. Demirbilek S, Atayurt HF. Congenital muscular torticollis and strenomastoid tumour. Result of non operative treatment. J Pediatr Surg 1999; 34:549–551.
14. Cheng JC, Chen TM, Tang SP, Shum SL, Wong MW, Metreweli C. Snapping during manual stretching in congenital muscular torticollis. Clin Orthop 2001; 384:237.
15. Emery C. The determinants of treatment duration for congenital muscular torticollis. Phys Ther 1994; 74(10):921–929.
16. Symmetric Designs: The TOT Collar for Torticollis Treatment. Available at: www.symmetric-designs.com/tot-collar-.html (accessed 30 July 2009).
17. Losee JE, Mason AC, Dudas J, Hua LB, Mooney MP. Nonsynostotic occipital plagiocephaly: factors impacting onset, treatment, and outcomes. Plast Reconstr Surg 2007; 119(6):1866–1873.
18. ChengJC, Tang SP: Outcome of surgical treatment of congenital muscular torticollis. Clin Orthop 1999; 362:190.
19. Azizkhan RG, DeColl JM. Head and neck lesions. In: Ziegler MM, Azizkhan RG, Weber TR, eds. Operative Pediatric Surgery. McGraw-Hill Professional, 2003, Pp 221–240.
20. Shim JS, Jang HP. Operative treatment of congenital torticollis. J Bone Joint Surg Br 2008; 90(7):934–939.
21. Sasaki S, Yamamoto Y, Sugihara T, Kawashima K, Nohira K. Endoscopic tenotomy of the sternocleidomastoid muscle: new method for surgical correction of muscular torticollis. Plast Reconstr Surg 2000; 105:1764–1767.