

Congenital Sternomastoid Torticollis: aetiology, diagnosis and management*

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INTRODUCTION

Congenital sternomastoid torticollis (CST) was studied during the personal observation of more than 20,000 newborn infants during the 1960s. The findings were first presented to the British Orthopaedic Association at their annual meeting in Bristol in May 1973 and subsequently to the European Society of Pediatric Research (Seville, October 1973) and the British Paediatric Association (York, April 1974).

DEFINITION

Congenital sternomastoid torticollis may be defined as a contracture of the muscle, present at birth, which limits lateral rotation of the head to the affected side and may cause increasing lateral flexion and twisting of the head so that the chin points towards the contralateral side. The condition is almost invariably unilateral. The anatomy of the normal sternomastoid muscle is illustrated in Figure 1. The affected muscle feels increasingly hard to palpation and may develop a sternomastoid 'tumour' one to two weeks after birth.

CLINICAL DIAGNOSIS

Although torticollis may be present at birth, it is often 'latent' because of the shortness of the neck in early infancy, and may only become apparent as the neck progressively lengthens. However CST may be demonstrated and diagnosed at birth by examining for a contracture in this muscle. Normally the head of the newborn infant may be passively rotated laterally until the chin points over the back of the shoulder joint (Fig 2). Inability to rotate the head beyond the front of the joint suggests that the sternomastoid muscle on that side is significantly contracted.

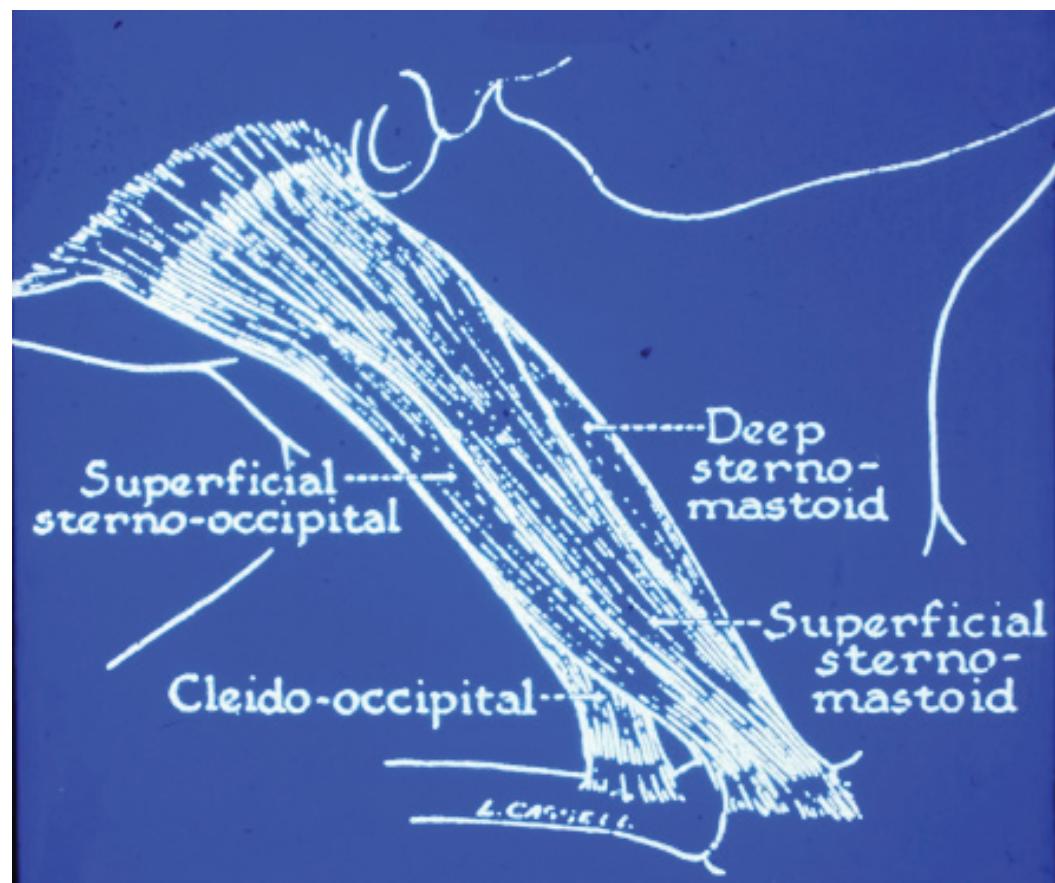
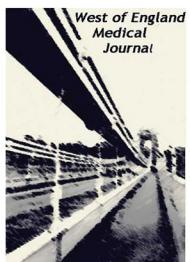


Fig 1. Anatomy of the sternomastoid muscle



Fig 2 The head of a newborn infant may normally be rotated until the chin points over the back of the shoulder joint. (With parental permission)

*Based on a presentation to the
British Orthopaedic Association, April, 1973



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Fig 3.

Infant delivered by Caesarean section because of breech presentation. CST noted at birth. Sternomastoid 'tumour' observed at 12 days. Note the upturned ear. (With parental permission)

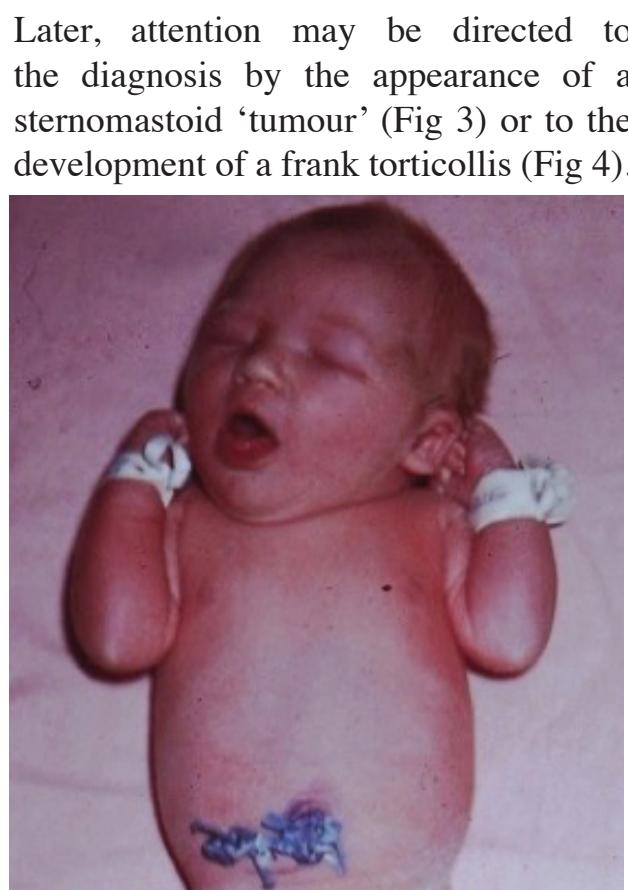


Fig 5. Position of comfort of an infant with left-sided congenital sternomastoid torticollis on day 1 (with parental permission)



Fig 4.

Right-sided CST at the age of 6 years. Sternomastoid tumour and plagiocephaly were noted soon after birth. (With parental permission)

Later, attention may be directed to the diagnosis by the appearance of a sternomastoid 'tumour' (Fig 3) or to the development of a frank torticollis (Fig 4).

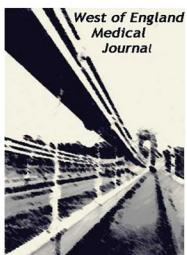
CLINICAL ASSOCIATIONS

Characteristically the infant lies with the head flexed laterally onto the shoulder on the affected side with the head rotated so that the chin points away to the other side (Fig 5). Often the lower lobe of the ear on the same side is upturned while the mandible is tilted away. This alters the outline of the chin giving a hollow on the affected side and a prominence on the other (Fig 6). Occasionally there is a lower facial nerve palsy (neuropraxia) on the same side. Plagiocephaly is another common association with flattening of the contralateral temporal region. Epidemiological studies in the early 1960s (Dunn, 1969, 1974, 1976) also showed a highly significant association between CST and other congenital postural deformities including those of the face, the spine, hip and feet. A similar pattern of maternal pregnancy characteristics

was also observed as were encountered in association with other congenital postural deformities. Thus, 53% of infants with CST were firstborn (controls 36%) and 20% presented by the breech (controls 5%). Maternal oligohydramnios was a frequent accompaniment and CST was noted to occur in association with bilateral renal anomalies and other malformations giving rise to fetal anuria.

INCIDENCE

Approximately one in every 300 infants has a significant contracture of the sternomastoid muscle at birth. However about 80% resolve spontaneously during the first year of life. The remainder either still have significant limitation of lateral rotation to the affected side and/or develop a full torticollis.



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Figures 6a and 6b

Tilted jaw and mandibular asymmetry at birth due to persistent lateral flexion of the fetal head onto the left shoulder.
Left CST present at birth (with parental permission)

SCREENING

All newborn infants should be examined at birth for CST using the clinical diagnostic test described above. In practice, it is worth remembering that CST is invariably accompanied by mandibular asymmetry. It has been the author's practice to examine the mouth first and only test for contracture of the sternomastoid when the jaw is found to be tilted.

PATHOLOGY

Necropsy examination of the sternomastoid muscles was undertaken in the case of five infants diagnosed as having CST at birth and dying later on the first day. In every case the affected muscle was purple instead of pink and significantly shorter than the contralateral muscle (Figure 7).

Histological examination revealed vascular congestion, haemorrhage and the infiltration of fibrous tissue among the muscle fibres. The latter showed widespread fragmentation and necrosis (Figure 8).

MANAGEMENT

At birth and for some weeks thereafter the damaged muscle is very painful and may be further damaged by passive stretching. Therefore management should consist of explaining the situation and prognosis to the parents and requesting them to attend the follow-up clinic when the infant is six weeks old.



Fig 7

The sternomastoid muscles at post-mortem of a baby dying on the first day of life. CST had been diagnosed at birth. Note the crimson appearance and shortening of the left muscle.

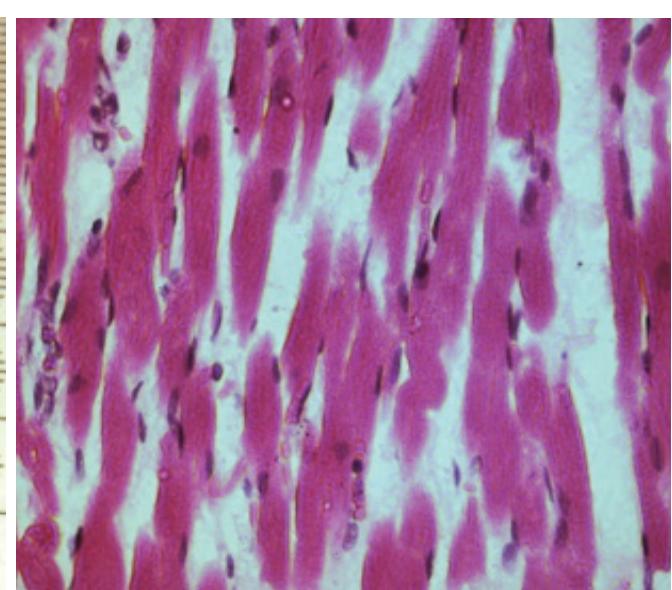


Figure 8a

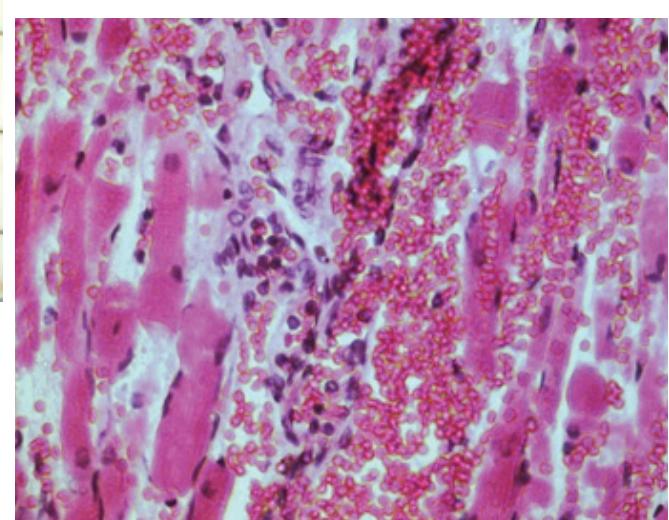
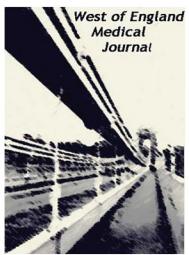


Figure 8b

Figures 8a and 8b

Histology of the normal right and damaged left sternomastoid muscles seen in Fig 7. Note the haemorrhagic necrosis, fragmented muscle fibres, and fibrosis in the damaged left muscle.



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by rotating the head to that side. Later on it may be necessary to use a corrective neck collar or to undertake a myectomy or tenotomy of the muscle. The latter is rarely indicated in the first year. As has been stated previously 80% of cases of CST resolve without the need for such treatment.

DISCUSSION ON AETIOLOGY

In 1938 Stromeier noted the association between a sternomastoid tumour and torticollis and suggested the former was a haematoma caused by birth trauma which on healing led to a contraction of the muscle. This view was supported by Henoch in 1889 who drew attention to the very high incidence of breech delivery among infants developing a sternomastoid tumour. This theory, though incorrect, has received support right up to recent times (Sanerkin and Edwards, 1966). It is true that a shortened muscle may be further damaged during delivery but all the evidence supports the view that CST is one of the congenital postural deformities arising in late fetal life because of persistent lateral flexion of the head onto one shoulder. Pressure on the venous drainage of the sternomastoid muscle may then lead to haemorrhagic necrosis and fibrosis which in turn leads to a contracture of the muscle. The evidence collected in support of this view is overwhelming and may be listed as follows:

- The contracture and other pathological changes in the muscle are present at birth.
- CST may occur in infants born without trauma, as, for instance, by Caesarean delivery.
- Other postural deformities may be mutually related as in the case of the tilted mandible and the upturned ear.
- A coincidence can often be demonstrated between the prenatal posture, the postnatal moulded appearance and the 'position of comfort' of the infant.
- The lesion occurs in association with characteristic prenatal 'deforming' environmental factors such as primiparity, breech presentation and oligohydramnios.
- The deformity has no special association with any teratological malformation except bilateral renal anomalies, which, of course, are

known to produce severe oligohydramnios because of fetal anuria.

- CST is almost invariably unilateral. This would not be expected if the damage was indeed caused by birth trauma. On the other hand it would be expected if it resulted from lateral flexion of the head against one or other shoulder.
- The incidence of CST – about 1 in 300 deliveries – is highest at birth, with a high subsequent spontaneous rate of resolution. This characteristic of all the congenital postural deformities, suggesting release from an adverse environmental situation following delivery.

Heller in 1898 showed experimentally that it was not possible to produce the pathological changes found in the sternomastoid muscle using trauma, and his work has been confirmed since by others. Then in 1922, following earlier studies by Nove-Josserand and Viannay (1906), Brooks of Chicago showed that the characteristic changes could be readily produced by occluding the venous drainage of the muscle. However, such damage did not occur if the artery was occluded either on its own or together with the vein. In 1926 Jepson showed that venous occlusion for as little as 15 hours was sufficient to initiate continuing pathological changes in the muscle. Then in 1930 Middleton confirmed this work with extensive experiments of his own on dogs, and suggested that the damage was caused by ischaemia secondary to acute venous occlusion, consequent on lateral flexion and rotation of the neck during labour. I would like to suggest that Middleton was right except that the venous occlusion probably usually takes place in most cases during late fetal life before labour, producing a damaged, shortened muscle at the time of birth. Recovery of the circulation to the muscle following delivery permits reparatory fibrosis to produce, usually after an interval of 2 or more weeks, a palpable sternomastoid tumour, or at least a hardening of the muscle.

SUMMARY

Congenital sternomastoid torticollis is a congenital postural deformity arising from obstruction to the venous drainage from that muscle by persistent pressure on the shoulder during late fetal life. It

occurs in 1 in 300 births but with a high (80%) resolution rate following delivery. It may and should be diagnosed at birth rather than later. Gentle stretching of the shortened muscle should not commence for at least six weeks after birth.

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