CONGENITAL TORTICOLLIS

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"As knots, by the conflux of meeting sap, 
Infect the sound pine, and divert his grain 
Tortive and errant from his course of growth."

(Troilus and Cressida. Act I, Scene 3)

The pathology of congenital torticollis has been the subject of investigation for more than a hundred years, ever since Stromeyer suggested that congenital sternomastoid tumour and congenital torticollis might be stages in one continuous process. The deformity is so obvious that it must have been known from the time that man first began to observe the traits of his fellows. Alexander the Great was claimed as the hero of torticollis (Colonna 1927, Little 1928), the evidence being in Plutarch's "Lives" where the writer noted "Alexander's manner of holding his neck hanging down towards the left side." But Alexander was renowned for his physical beauty, and the few authentic portraits and busts that are available show no facial deformity (Fig. 1). Moreover it should be remembered that slight lateral inclination of the head has often been favoured by distinguished persons, and that in the representation of mediaeval saints "torticollis" has played no less a role than the halo over their heads.

Antyllus is said to have practised tenotomy for torticollis in the second century but it was only the writings of Oribasius, one century later, that preserved our knowledge of him. Fabricius of Padua, the tutor of William Harvey, devised a brace for correction of the deformity but his results were not recorded. In 1641 a German doctor, Isaac Minnius, made the first known attempt at surgical correction (Hough 1934, Little 1928); and in 1670 the operation was again described by Rouenhuysen in Holland. In mediaeval times, subcutaneous tenotomy was performed by itinerant practitioners in the fairs of England to the accompaniment of clanging cymbals by which to drown the cries of their victims (Power 1933). The Vicar of Stratford-on-Avon, the Reverend John Ward (1663–1681) described the insertion of a lancet beneath the muscle, cutting from within outwards. It appears that these practitioners never injured the underlying veins, which, if true, is a humiliating thought when we recall how often the subcutaneous operation is now condemned because of the danger to great vessels. In 1749 William Cheselden of St Thomas's Hospital published a treatise, "Observations on Le Dan's Operations of Surgery" in which is described and illustrated the operation of open tenotomy of the sternomastoid (Fig. 2). He asserted that after division of the sternal

Fig. 1
Bust of Alexander the Great. The alleged wry-neck is shown, but no facial asymmetry. This is not a deformity: it is a pose. (Reproduction by courtesy of the British Museum.)
attachment the clavicular head of the muscle could be stretched. Dupuytren performed subcutaneous tenotomy on one of his servants in 1828, and demonstrated his technique to Symes who later introduced the operation to Edinburgh.

**PATHOLOGY**

The records of one hundred patients with congenital torticollis seen between 1938 and 1945 at the Hospital for Sick Children, Great Ormond Street, have been studied. Congenital skeletal abnormalities were excluded. The cases fell into two groups according to the age at which the deformity became obvious.

Group A comprised sixty-seven children with torticollis noticed at or soon after birth who were brought to hospital between the ages of three weeks and three months, the average age being six weeks; in no case was operation needed. In Group B there were thirty-three patients with torticollis first noticed long after birth who were brought to hospital between the ages of one and ten years, usually at about the age of four years; and in twenty of these patients operation was necessary. Evidence will be presented to show that there is an essential difference in the pathology and treatment of these two groups. In Group A the condition is postural and temporary; in Group B there is a true congenital deformity associated with pathological changes in the sternomastoid muscle.

Stromeyer said that injury to the foetal neck at birth caused a haematoma of the sternomastoid muscle which then contracted (Hough 1934). His contribution was supported only by the evidence of four cases but it was important because it correlated the sternomastoid tumour with torticollis and emphasized the significance of the obstetric history. The relationship between sternomastoid tumour and torticollis has been debated ever since. Four sternomastoid tumours were recorded in eighteen cases of torticollis by Clutton (1888), twenty tumours in 106 cases by Power (1893), and twenty-three tumours in one hundred cases by Gray (1935). In this series of one hundred patients a sternomastoid tumour was recorded in fourteen. Middleton (1930) reported experimental evidence in favour of the pathological continuity of the two diseases and showed that the “tumour” consisted of interstitial fibrosis which went on to fibrous contracture; but he was unable to explain why in four cases out of five the tumour appeared to resolve and leave a normal muscle.

**The causation of torticollis**—It seems probable that there is a relationship between sternomastoid tumour and congenital torticollis and nearly all hypotheses are based on this assumption. At least six are worthy of consideration.

1) The hypothesis of birth injury was formulated by Stromeyer and presented fully by Witzel (1891) who believed that there might be abnormal intra-uterine development of the sternomastoid producing a shortened muscle which was torn at birth with the formation of a haematoma which underwent fibrous contracture. There is no doubt that sternomastoid
TABLE I
CLASSIFICATION OF CONGENITAL TORTICOLLIS

<table>
<thead>
<tr>
<th></th>
<th>Group A</th>
<th>Group B</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of cases</td>
<td>67</td>
<td>33</td>
</tr>
<tr>
<td>Average age</td>
<td>6 weeks</td>
<td>4 years</td>
</tr>
<tr>
<td>Number requiring operation</td>
<td>Nil</td>
<td>20</td>
</tr>
</tbody>
</table>

TABLE II
REPORTED INCIDENCE OF STERNOMASTOID TUMOUR IN CONGENITAL TORTICOLLIS

<table>
<thead>
<tr>
<th>Author</th>
<th>Year</th>
<th>Total cases</th>
<th>Sternomastoid tumour</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td>Cases</td>
</tr>
<tr>
<td>Clutton</td>
<td>1888</td>
<td>18</td>
<td>4</td>
</tr>
<tr>
<td>Power</td>
<td>1894</td>
<td>106</td>
<td>21</td>
</tr>
<tr>
<td>Gray</td>
<td>1934</td>
<td>100</td>
<td>23</td>
</tr>
<tr>
<td>Hospital for Sick Children</td>
<td>1947</td>
<td>100</td>
<td>14</td>
</tr>
</tbody>
</table>

Tumours occur with frequency after difficult labour; and in this series there was a history of difficult labour in more than half the patients. But the evidence against this interpretation is overwhelming. The tumour is hard and discrete, and there is no discolouration of the skin such as might occur after a haematoma. Moreover, ruptured muscles elongate; they do not contract. There is no haemosiderin in the tissues in old cases. Even early cases, examined within a few weeks, show a mass of glistening white fibrous tissue with no evidence at all of blood. The tumour is certainly not caused by a haematoma.

2) The ischaemic hypothesis, first suggested by Mikulicz, was brought into prominence by Nové-Josserand and Viannay (1906) who compared the muscular fibrosis of torticollis with that of Volkmann's contracture. They showed that the blood supply of the sternomastoid was precarious. The sternal head and middle fibres of the muscle are supplied by the sternomastoid branch of the superior thyroid artery which has been proved to be an end-artery. The circulation of this part of the muscle may be obstructed by placing the head in certain positions. Necropsies of twenty newly born infants showed that lateral flexion of the head with elongation and torsion of the neck prevented material injected into the carotid artery from reaching the muscle. The results of such experiments in the cadaver should not of course be applied without reserve to the arterial circulation of the living; and it is to be noted that those who have excised the sternomastoid tumour have noted the vascularity of the tissues. Moreover, Brooks (1922) pointed out that ligation of an artery does not cause atrophy and massive fibrosis.

Middleton (1930) postulated that venous occlusion produced ischaemia in muscle. He believed that venous drainage of the sternomastoid was separated into three fields—the mastoid, clavicular and sternal parts. The clavicular and sternal veins might be obstructed by torsion of the foetal head in the second stage of labour, thus producing ischaemia of the muscle. This view has been criticised by recent observers who claim that the muscle is drained by a common venous bed which cannot be obstructed so easily (Chandler and Altenberg 1944). Middleton went further and suggested that birth injury caused venous
thrombosis in the muscle; but this again has not been confirmed by later investigations (Chandler and Altenberg 1944).

The supposition that torticollis might be due to ischaemic necrosis is open to even more serious criticism. The susceptibility of various tissues to ischaemia differs greatly. Nerve cells are damaged irreparably by deprivation of their blood supply for a quarter of an hour, and muscle dies after six or seven hours (Watson-Jones 1946). If ischaemic necrosis is produced in the sternomastoid muscle by stretching or torsion of the head during the second stage of labour it must necessarily be presumed that the foetal head has been retained in this position for several hours with consequent interference with the cerebral circulation. In these circumstances the breech delivery that is characteristic of most patients with this condition must surely have proved fatal. It might be argued that birth injury produced arterial spasm

![Image](image-url)  

**Fig. 3**  
The histological appearances of ischaemic necrosis. There is a massive necrosis of muscle tissue, with a peripheral cellular reaction.  
(Reproduced by permission of Mr D. Ll. Griffiths and the British Journal of Surgery.)

but this is ruled out on histological grounds. Griffiths (1940) showed that the histological appearances of ischaemic muscles in Volkmann's contracture of the forearm differed from those of the sternomastoid tumour, and from the experimental lesions produced by Middleton. The histological appearance in Volkmann's ischaemia is that of a muscle sequestrum: the general contour is preserved but the tissue is dead, with no living nuclei in the muscle fibres; the necrotic area is surrounded by a zone of phagocytic activity and eventually there is replacement by fibrous tissue which invades the slough from the periphery inwards (Fig. 3). In contradistinction, the sternomastoid tumour contains islands of muscle tissue surrounded by masses of cellular fibrous tissue and the characteristic muscle slough of Volkmann's ischaemia is not seen (Fig. 4).

The fact that the sternomastoid tumour which appears towards the end of the second week of extra-uterine life contains cellular fibrous tissue should be noted. It seems unlikely that a pathological process occurring at the time of birth could produce mature fibrous
tissue within two weeks, and this observation casts doubt upon any theory that is based on causative factors operating at the time of birth.

3) The hypothesis of intra-uterine malposition—The conception that congenital deformity is the consequence of abnormal intra-uterine position dates from the time of Hippocrates and has long been considered a possible explanation of torticollis (Browne 1936). In 1944 Chandler and Altenberg described a case in which ante-partum radiographs showed breech presentation with the foetal head in right lateral flexion. The baby was delivered by Caesarian section; and right-sided torticollis which was present at birth became increasingly pronounced. A sternomastoid tumour was noticed after fourteen days and it was excised. Rossi (1928) reported a similar case of torticollis found at Caesarian section in which the tumour was present at birth. Joachimstal and Volcher (1904) described torticollis in an extra-uterine pregnancy. These reports suggest that torticollis is present at or before birth and that the sternomastoid tumour may be the result rather than the cause of the torticollis.

The high incidence of obstetric abnormalities, especially breech presentation, has been explained in various ways. Sippel (quoted by Chandler and Altenberg 1944) suggested that inability of the wry-necked foetus to engage normally in the pelvis caused breech presentation. Browne (1936) pointed out that postural torticollis was associated with flattening of the opposite side of the head, as if it had been moulded by the uterine wall; and he described a number of infants with moulding of the lobe of the ear which suggested pressure against the shoulder while the head was flexed to that side. One case with such moulding also showed facial paralysis from compression of the facial nerve. Of sixty-seven cases of postural torticollis in the series now reported, four showed the triad of torticollis, crumpled ear and facial paralysis. The frequency with which these signs are associated is enough to justify the assumption that they may have a common cause. Moulding
of the head was seen in nearly all children with postural torticollis, and in these cases the clinical evidence in favour of intra-uterine malposition seems to be convincing. Nevertheless postural torticollis was never associated with a tumour and never gave rise to structural contracture. It seems clear therefore that some other factor must be responsible for the muscular type of torticollis with sternomastoid tumour.  

4) *The hereditary hypothesis* which was advanced in the last century has never found favour (Tubby 1912). In this series there was only one example of the deformity occurring in mother and child. No series published in the literature has shown any higher incidence.  

5) *The hypothesis of infective myositis*—Since the sternomastoid tumour is characterised by interstitial fibrosis it has been suggested, though unconvincingly, that it is the end-result of an infective myositis. MacLennan (1943) described the appearances of infective myositis; they are quite different from those of the sternomastoid tumour. Moreover no infective agent, and no evidence of syphilis, has ever been discovered.  

6) *The nervous hypothesis*—Atrophy of the anterior horn cells has been reported in an adult case of torticollis but this is comparable to the similar atrophy that occurs after amputation of a limb and there is no evidence whatever that it is a significant factor in causation.  

**Summary of hypotheses**—The *postural* type of torticollis can be attributed to intra-uterine malposition on the evidence of associated moulding of the head with occasional deformity of the ear and facial palsy. The *muscular* type of torticollis with its associated sternomastoid tumour is usually attributed to ischaemic necrosis. That this view should be reconsidered is quite clear from the evidence here presented. The histology of sternomastoid tumours is not that of any known type of ischaemic necrosis. Interstitial fibrosis of this type cannot be produced experimentally; and it has been shown that the tumour cannot be produced at birth. It must be acknowledged that the real cause of this peculiar fibrous tumour remains unknown.  

**CLINICAL FINDINGS**  

The accepted clinical picture of congenital torticollis—About ten days after birth a tumour appears in the substance of the sternomastoid which may or may not be associated with transient spasmodic torticollis lasting a few days or weeks. The tumour occupies most of the muscle but is circumscribed. Both heads of the muscle are usually involved but sometimes the swelling is confined to one head. It persists for two or three months and then disappears gradually in from four to six months. The development of torticollis is delayed until the age of three or four years when the short neck of the infant lengthens into that of the young child. As growth proceeds the torticollis leads to facial asymmetry which becomes permanent if the deformity is allowed to persist. The mastoid process on the affected side becomes elongated and ossification may occur in the clavicular origin of the muscle. In neglected cases there is cervical scoliosis and sometimes contraction of the field of vision.  

Clinical features in this series of cases—Onset of the tumour—The tumour was usually noted within the first month of life, and four were seen within the first few days. Sternomastoid tumours have been seen at birth by Rossi (1928) and Chandler and Altenberg (1944).  

Clinical nature of the tumour—Our cases corresponded closely with the classical description. The fibrous process involved nearly the whole sternomastoid muscle but was confined to it. The patient was a male in 54 per cent. of cases; and the lesion was on the right side in 52 per cent.  

Disappearance of the tumour—The tumour usually disappeared by about the seventh month. The earliest disappearance was at two months and the latest at eleven months.  

Onset of torticollis—In Group A cases the postural deformity was present at birth, was transient, and left no structural deformity. In Group B cases the deformity was sometimes, though not always, associated with a history of antecedent sternomastoid tumour. Several patients were first brought for advice at the age of six or seven years, and in one the deformity was not noticed until the age of eight years. On the other hand deformity was noticed in one patient
soon after birth, in two at four weeks, and in one at six weeks. These findings are in general agreement with those of Middleton (1930) who found that the deformity seldom became obvious until the neck began to lengthen at about the age of three years. *Facial asymmetry* was observed at an earlier age than is usually described. Asymmetry of slight degree can be detected by examination of the head and neck from behind—the "scoliosis capit" of Middleton. In the same way that dorsal scoliosis with vertebral rotation causes flattening on one side of the chest and bulging on the other, rotation of the cervico-cranial vertebrae gives rise to asymmetry of the skull. This may even be present in new-born babies when it is probably due to intra-uterine torsion. Facial asymmetry may of course complicate any longstanding cervical deformity whether from dislocation, tuberculosis disease or scar contracture. Walter (quoted by Hough 1894) produced reversal of the facial asymmetry by over-correction maintained for seventeen weeks; and Tubby (1912) believed that facial asymmetry always disappeared within three years of operation.

*The obstetric history—*In our series of patients with sternomastoid tumour the incidence of obstetric complications was: breech delivery, 40 per cent.; forceps delivery, 20 per cent.; difficult labour, 9 per cent.; total incidence, 69 per cent. These figures are compared with those of other authors in Table III.

**TABLE III**

<table>
<thead>
<tr>
<th></th>
<th>Breech</th>
<th>Forceps</th>
<th>Difficult labour</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Witzel (1891)</td>
<td>44</td>
<td>19</td>
<td>9</td>
<td>72</td>
</tr>
<tr>
<td>Middleton (1930)</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>72</td>
</tr>
<tr>
<td>Chandler and Altenberg (1944)</td>
<td>30</td>
<td>23</td>
<td>8</td>
<td>61</td>
</tr>
<tr>
<td>Hospital for Sick Children (1947)</td>
<td>40</td>
<td>20</td>
<td>9</td>
<td>69</td>
</tr>
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</table>

Breech presentation occurs in about 3 per cent. of all pregnancies, and the greater incidence in cases of sternomastoid tumour is significant. Breech delivery seldom causes injury and it seems probable that it is the position in utero, rather than the nature of delivery, that is the factor of etiological importance. This relationship was stressed by Chandler and Altenberg who quoted the radiographic evidence of Isigkeit and Sippel. The high incidence of difficult delivery has long been considered significant in the causation of the tumour but doubt is cast upon this view by the occurrence of the condition in babies delivered by Caesarian section.

**TREATMENT**

*Non-operative measures*—Most patients with sternomastoid tumour, and all with postural torticollis, were treated successfully by early manipulation by the physiotherapist and the mother. There has been some misunderstanding as to the method of manipulation. The combined action of both sternomastoid muscles is to extend the head on the neck and draw forward the cervical spine. The insertion of the muscle into the mastoid process is behind the occipital condyles, so that it is an extensor of the atlanto-occipital joint (double torticollis, which is very rare, is characterised by extension of the head and flexion of the neck—the "sniffing the air" position). The neck should of course be flexed laterally to the opposite side, and the head rotated to the same side (Fig. 5); but flexion of the neck is obviously a fundamental part of the manipulative treatment (Fig. 6). Lorenz advocated subcutaneous rupture, or myorrhesis, after the manner of his treatment of the contracted adductors in congenital dislocation of the hip; but Reiner (1896) and Brackett (1897) recorded sudden and dangerous collapse during this procedure.
Operative treatment—Subcutaneous tenotomy is condemned by most writers but defended vigorously by others (Elmslie 1943). It is practised to-day by many surgeons of repute. The operation is said to endanger the great vessels, particularly the external jugular and anterior jugular veins, and the communicating channel between them. A further objection is that division of the contracted tissues may be incomplete. The operation for torticollis is largely cosmetic and the subcutaneous operation has the advantage that it produces no scar. Injury to the vessels can be prevented by performing the tenotomy low down and cutting on to the clavicle, and if the operation is performed through a quarter-inch incision the risk to the vessels is further reduced. This is a good method which should be practised more widely.

Open tenotomy—The results of open tenotomy in twelve patients, examined fifteen to twenty years afterwards, were cosmetically imperfect. The scar was usually placed horizontally just above the clavicle. It tended to stretch and become unsightly, and there was sometimes keloid formation. Complete division of both heads of the muscle is usually all that is necessary,

but sometimes the cervical fascia, omohyoid and scalenus anterior may have to be divided. This procedure is criticised by many because the pathology is confined primarily to the sternomastoid muscle. Tubby (1912) used an oblique incision along the lower part of the anterior edge of the sternomastoid and reported good cosmetic results in a few cases. But it is now recognised that almost any incision in the neck is unsightly and even a collar incision if placed too low may leave an ugly scar.

Division of the upper end of the sternomastoid was introduced by Tillaux and Lange. It is said that if the muscle is allowed to slide too far—more than about one inch—traction on the spinal accessory nerve may cause paralysis. Apart from this possible complication, detachment of the occipital and mastoid insertions of the muscle through an incision within the hair line gives good correction and a satisfactory cosmetic result. Hellstadius (1927) practised the combined operation of open division of the upper end and subcutaneous division of the lower end, and Denis Browne has employed a similar operation for many years.
Z-shaped lengthening of the muscle—Jones and Lovett (1929) described a method of lengthening the muscle by an oblique or Z-shaped incision. Another method is that of elongation of the sternal head with oblique or Z-shaped incision (Elmslie 1943).

Resection of the muscle—Resection of the sternomastoid was first advocated by Mikulicz. Recently Chandler and Altenberg (1944) advised removal of the sternomastoid tumour during the first few weeks of life. The operation cannot be recommended because four out of every five tumours resolve completely and leave no deformity; and moreover when torticollis does develop it can be corrected by a much simpler procedure at about the age of three or four years.

Post-operative treatment—There are many ways of maintaining correction after operation (Fig. 7). Mediaeval practitioners used a cap which was fastened by fillets beneath the axilla (Power 1893). Plaster caps with similar bands are used to this day at the Hospital for Sick Children in London. Others rely on physiotherapy started as early as possible and pursued conscientiously over a prolonged period. Treatment should be continued for at least six months and every patient should be kept under observation for several years.

**SUMMARY AND CONCLUSIONS**

1. One hundred cases of torticollis and 117 cases of sternomastoid tumour have been reviewed.

2. Congenital torticollis can be sub-divided into two groups: postural and muscular.

3. Congenital postural torticollis is present at birth; it is not associated with a sternomastoid tumour; it is transient in nature; and it does not require operation for its relief.

4. Congenital muscular torticollis is preceded by a sternomastoid tumour which is clinically evident in one-fifth of all cases.

5. The ischaemic theory of the causation of sternomastoid tumours is not supported by recent histological investigations. Some other cause, which probably is operative before birth, must be sought.

6. Four-fifths of all cases of sternomastoid tumours resolve spontaneously and leave no deformity. Excision of the tumour in infancy is therefore unjustifiable.

7. Open division of the muscle and of the cervical fascia in congenital muscular torticollis cures the deformity but leaves an unsightly scar.

8. Subcutaneous tenotomy can be relied upon to cure the deformity if post-operative treatment is carried out skillfully and assiduously over a prolonged period.

9. If complete correction is not gained at the time of subcutaneous tenotomy a better result can be assured by open division of the upper end of the muscle through an incision within the hair line.

I wish to express my thanks to the surgical staff of the Hospital for Sick Children, and especially to Mr Eric Lloyd and Mr Denis Browne, and to the Department of Clinical Photography at the West Hill Hospital, Dartford.

**Fig. 7**

Trethowan’s collar used in the after-treatment of torticollis. It is not so much a splint as an active reminder that the head and neck must be held away from the flange.
CONGENITAL TORTICOLLIS

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