TORTICOLLIS DUE TO A COMBINATION OF
STERNOMASTOID CONTRACTURE
AND CONGENITAL VERTEBRAL ANOMALIES

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We report four children with sternomastoid contracture combined with torticollis secondary to congenital vertebral anomalies. Two had features of Klippel-Feil syndrome and a proximal release of the contracted sternomastoid produced good cosmetic correction initially. Progression of the deformity occurred subsequently without recurrence of sternomastoid contracture. One child had such mild deformity that it was merely observed. The fourth child was born with torticollis without sternomastoid tightness and a vertebral anomaly was later recognised. He slowly developed a sternomastoid contracture and his condition was considerably improved by sternomastoid release.

This combination of causes of torticollis has not, as far as we know, been previously reported. The clinician should be aware of it and should also realise that radiographs of the very immature spine may not disclose the bony anomalies.

Muscular torticollis secondary to contracture of the sternomastoid is common, with an incidence estimated to be 0.4% of live births (Coventry and Harris 1959). To our knowledge, the association of congenital muscular torticollis with congenital anomalies of the cervical and upper thoracic spine has not been previously reported; we report four children with this association. In three of these, significant cosmetic improvement followed release of the contracted muscle.

CASE REPORTS

Case 1. A 19-month-old boy presented with a right torticollis that was first noted when he began to sit at seven months of age. There was no history of a sternomastoid tumour. A diagnosis of muscular torticollis was made although the deformity appeared to be greater than expected from the amount of sternomastoid shortening. Radiographs were taken in an endeavour to establish if bony anomalies were present; these were considered to be normal. A proximal release of the sternomastoid was carried out at the age of three years and good correction was obtained. However, the deformity progressed and, three years after operation, radiographs showed a mixed lesion of failure of segmentation and failure of formation of the first and second cervical vertebrae. Five years after surgery, there was a right torticollis with associated facial asymmetry and restricted neck movement (Fig. 1). The neck was short as in Klippel-Feil syndrome and there was no evidence of a residual sternomastoid contracture. A radiograph taken at that time is shown in Figure 2.

Case 2. A boy was born with multiple skeletal anomalies, which included some of the upper cervical and lower thoracic vertebrae. At seven months, he showed a contracture of the right sternomastoid without a preceding history of a sternomastoid tumour. Physiotherapy was unsuccessful and for the persisting contracture a proximal release of the sternomastoid was carried out at the age of one year. Before operation the parents were told that complete correction was unlikely because of the skeletal anomalies. However, there was significant cosmetic improvement.

Since then the deformity has progressed and eight years after operation he had a right torticollis with facial asymmetry (Fig. 3) and a reduced range of movement of the neck. His neck was short and he had a right
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**Case 1.** At eight years of age, five years after operation, there is a right torticollis and a short neck indicating Klippel-Feil syndrome, but no sternomastoid contracture. An anteroposterior radiograph of C1 and C2 shows a mixed lesion of failure of segmentation and formation.

![Fig. 1](image1.png)  ![Fig. 2](image2.png)

**Case 2.** This boy aged nine years has a right torticollis, a short neck, and a right Sprengel shoulder consistent with Klippel-Feil syndrome. A lateral radiograph of the cervical spine shows multiple bony fusions from C1 to C5.

![Fig. 3](image3.png)  ![Fig. 4](image4.png)

Sprengel's shoulder which, with the radiographic abnormalities, indicated Klippel-Feil syndrome. There was no residual sternomastoid contracture. Radiographs eight years after operation show multiple fusions of the first to the fifth cervical vertebrae (Fig. 4).

**Case 3.** A boy was found to have a right torticollis at three months of age, without an associated sternomastoid tumour. Considerable improvement occurred with physiotherapy during the first year of life. He presented to us at five years of age with progression of the deformity, when he had a mild right torticollis with associated mild facial asymmetry. There was a palpably contracted band in the clavicular head of the right sternomastoid (Fig. 5), but no significant restriction of cervical spine movement. Radiographs showed multiple hemivertebrae at the level of the first to third thoracic vertebrae (Fig. 6). In view of the mild nature of the deformity and the presence of bony anomalies, observation has continued; the deformity has not progressed in three years.

**Case 4.** A boy had a right-sided head tilt at birth, but no tightness of the sternomastoid. When seen by us at three months there was no muscle tightness and radiographs suggested that the lower cervical spine was normal but revealed an angular deformity at cervico-occipital level. Over the years, the sternomastoid became tight and there was increasing facial asymmetry with an abnormal posture of the head and poor neck movement. At 10 years (Fig. 7) there was facial asymmetry, a sternomastoid contracture on the right, a right cervicothoracic and a left upper cervical scoliosis with a right-sided head tilt. The
Case 4. At 10 years of age this boy has a short neck, right-sided sternomastoid contracture and facial asymmetry. Two years after a release operation his appearance has improved.

range of rotation of the neck was 20° to the right and 90° to the left. There was full flexion but extension was reduced to 50% of the normal range. Left lateral flexion was about 30° but right lateral flexion was full. The shoulders were slightly asymmetrical with a smaller right scapula which was, however, not elevated. Radiographs showed that the lateral mass of C2 was enlarged on the right, but the deformity made it impossible to produce prints which merit publication.

At the age of 10 years, he had an open sternomastoid release at both proximal and distal ends followed by three weeks in halo traction and a cervicothoracic brace for 12 weeks. Two years later, there is a marked improvement in appearance (Fig. 8) and in the range of neck movement. Rotation to the right is now to 45° and extension is normal. Left lateral flexion remained as restricted as before operation though there was no residual sternomastoid tightness.

DISCUSSION

Congenital muscular torticollis has been reported to occur with other skeletal anomalies. The commonest is hip dysplasia (Hummer and MacEwen 1972) where the association may be due to intra-uterine moulding. More rarely, an association with scoliosis and chest deformities of congenital origin has been noted (Armstrong et al. 1965; Canale, Griffin and Hubbard 1982). Our Case 2 had congenital anomalies of the left rib cage in addition to spinal anomalies. There are no previous reports of the association of congenital muscular torticollis with either
Klippel-Feil syndrome or with any congenital vertebral anomalies (Goldberg 1987).

The late diagnosis of vertebral anomalies in Cases 1 and 4, despite a repeated search, highlights a problem. Where there is doubt as to whether such anomalies are present because of a discrepancy between the deformity and the degree of sternomastoid contracture, a guarded prognosis should be given and search continued as the child matures.

The occurrence of vertebral anomalies in combination with congenital muscular torticollis has considerable clinical importance, since surgery directed to the sternomastoid will not completely correct the deformity and progression is likely as the child matures. On the other hand, division of the contracted muscle should be considered in such cases; operation resulted in improvement in all three patients in whom it was performed. In one, surgery was of considerable long-term benefit; in the other two it is likely that later deterioration was due to progression of the vertebral lesion. There was no evidence of the tight muscle which generally accompanies relapse in sternomastoid torticollis.

In two of our patients, the congenital vertebral anomalies eluded early radiological diagnosis; could it be that subtle congenital anomalies are more commonly associated with sternomastoid torticollis than is now recognised? If so, then the hypothesis that sternomastoid torticollis is of congenital origin should be resurrected.

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REFERENCES


