THE ASSOCIATION BETWEEN CONGENITAL ELEVATION OF THE SCAPULA AND DIASTEMATOMYELIA

A PRELIMINARY REPORT

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Of forty-six patients with congenital elevation of the scapula, diastematomyelia was confirmed in four and was probably present in five others. Possible reasons for the previously unsuspected association are presented.

Congenital elevation of the scapula was first described by Eulenburg in 1863 and there have been many descriptions and reviews of the literature since then (Horwitz 1908; Cavendish 1972; Ross and Cruess 1977). The condition is usually known as Sprengel's shoulder (Sprengel 1891), and because it is clinically recognisable and partially impairs function it is more familiar to orthopaedic surgeons than is diastematomyelia.

Diastematomyelia consists of complete separation of the spinal cord into two, not necessarily equal, parts which are often contained in separate dural sheaths and lie together in an expanded spinal canal over a distance of several segments. A septum, which is frequently bony, separates the two parts in an approximately sagittal plane, and is attached ventrally to the rear of a vertebral body and dorsally to the often malformed lamina. In most cases the onset of progressive neurological deficiency in the lower limbs or disturbance of control of the bladder lead to the discovery of the spinal abnormality. Several authors have stressed the danger to the spinal cord during corrective operations for congenital scoliosis that can arise from diastematomyelia (Faithfull 1973; Gillespie et al. 1973; Keim and Greene 1973; Winter 1973; Winter et al. 1974; Leonard 1975; Banniza von Bazan et al. 1976). Recently we have reported several patients with diastematomyelia and abnormal caudal fixation of the spinal cord in whom there was no neurological deficiency (Banniza von Bazan, Krastel and Lohkamp 1978). We suggested that diastematomyelia is not as rare as had previously been thought and that the abnormal anatomical findings frequently give rise to no symptoms and remain undetected, so that in many instances the abnormality itself may have no orthopaedic or neurological significance.

Neither of these uncommon deformities has yet been fully explained, but we have seen them occurring together in the same individual on several occasions and believe they may have a common aetiology. This is the basis of the present paper.

MATERIAL AND METHODS

Since 1950 sixty-two children have been treated at the University Orthopaedic Hospital in Heidelberg for congenital elevation of the scapula, fifteen of whom had undergone operation. An additional girl who was operated upon in 1935 is also included. The clinical records of all sixty-three patients were available together with the radiographs of sixty-one of the children, so that the diagnosis of "congenital elevation of the scapula" was confirmed according to the following three criteria. There was limitation of abduction at the shoulder on clinical examination. There were radiographic signs of a difference in height between the two scapulae of at least one spinal segment in unilateral cases; of dysplasia of the scapula indicated by small size, anterior deformation of the superior angle or an alteration in the scapular index (width to height ratio); and of malrotation of the scapula. There was radiological evidence or discovery at the operation of a bony or fibrous omovertbral band.

A diagnosis of diastematomyelia may be suspected on the basis of certain clinical symptoms (Hamby 1936; Matson et al. 1950; James and Lassman 1958, 1964), and the suspicion is strengthened by characteristic radiographic findings which are usually present (Neuhauser, Wittenborg and Dehlinger 1950). Unfortunately inadequate radiographs were only available for forty-six of our sixty-three patients; however, in nine of those with congenital elevation of the scapula we felt justified in diagnosing that diastematomyelia was also present. Of these nine patients some were no longer available for review while others refused further radiographic examination so that confirmation of the diagnosis by myelography or by computer tomography was limited.

Axial computer tomography (Weinstein et al. 1975; Claussen, Lohkamp and Banniza von Bazan 1977) confirmed the diagnosis of a bony spur in four patients, one of whom also underwent myelography because the bony spur transfixcd the conus medullaris unusually far caudally.

RESULTS

Table I lists four patients with Sprengel's shoulder in whom we were able positively to indentify an intraspinal bone spur by axial computer tomography. A myelogram was also performed for Case 1. The findings are illustrated in Figures 1 to 4.

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Case 1. A girl aged seventeen years. Figure 1—Ten years after operation for a left Sprengel shoulder. Figure 2—Myelogram showing a long cleft in the spinal cord.

Table I. Diastematomyelia proven

<table>
<thead>
<tr>
<th>Case number</th>
<th>Sex</th>
<th>Sprengel shoulder</th>
<th>Operation</th>
<th>Myelogram</th>
<th>Computer tomogram</th>
<th>Level</th>
<th>Type*</th>
<th>Spina bifida</th>
<th>Neurological signs</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>F</td>
<td>Left</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>L3–4</td>
<td>1</td>
<td>L4–S1</td>
<td>—</td>
</tr>
<tr>
<td>2</td>
<td>F</td>
<td>Right</td>
<td>+</td>
<td></td>
<td>+</td>
<td>L1–2</td>
<td>2</td>
<td>L3–S1</td>
<td>—</td>
</tr>
<tr>
<td>3</td>
<td>F</td>
<td>Right</td>
<td>–</td>
<td></td>
<td>+</td>
<td>L2–3</td>
<td>3–4</td>
<td>C7–T1</td>
<td>L2–S1</td>
</tr>
<tr>
<td>4</td>
<td>F</td>
<td>Bilateral</td>
<td>–</td>
<td></td>
<td>+</td>
<td>T12</td>
<td>1</td>
<td>T12–S1</td>
<td>Paraplegia T12</td>
</tr>
</tbody>
</table>

*Type: 1, massive bone spur; 2, spur with central “pore”; 3, “atypical spinous process”; 4, haze.

Table II lists a further five patients where diastematomyelia was strongly suspected together with elevation of the scapula. The suspicion was based upon the criteria proposed by Neuhauser et al. (1950). In Cases 5 to 7 a “haze” was present in place of the normal spinous process (Figs. 5 and 6). It has been shown by myelography and also by computer tomography that the intramedullary bone spicule of diastematomyelia can assume this hazy appearance (Banniza von Bazan et al. 1976; Kühner et al. 1976). In Case 8 (Figs. 7 to 9) initial radiographs revealed the typical fusiform widening of the spinal canal in the upper thoracic region while later fusion of the vertebrae C7 to T6 occurred. No bony spur was present and the diagnosis of diastematomyelia rested on the clinical findings. The girl showed a tuft of hair between the scapulae (Fig. 9), the left leg was shorter by one centimetre and the left foot by two centimetres. The muscle bulk was reduced and the left
Case 7. A girl aged ten and a half years. Figure 5—Left Sprengel shoulder with an omovertebral bone. Figure 6—Haze replacing the spinous processes of L1 and 2.

Table II. Diastematomyelia suspected

<table>
<thead>
<tr>
<th>Case number</th>
<th>Sex</th>
<th>Sprengel shoulder</th>
<th>Operation</th>
<th>Elevation of vertebral segments</th>
<th>Malrotation</th>
<th>Omovertebral bone</th>
<th>Wide spinal canal</th>
<th>Spur type*</th>
<th>Hair patch</th>
<th>Level</th>
<th>Spina bifida</th>
<th>Neurological signs</th>
<th>Clinical signs</th>
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<tbody>
<tr>
<td>5</td>
<td>M</td>
<td>Left</td>
<td>-</td>
<td>2</td>
<td>+</td>
<td>-</td>
<td>+</td>
<td>4</td>
<td>-</td>
<td>L2–4</td>
<td>L2–S1</td>
<td>Left ankle reflex absent</td>
<td>2 cm leg difference</td>
</tr>
<tr>
<td>6</td>
<td>F</td>
<td>Right</td>
<td>-</td>
<td>2</td>
<td>+</td>
<td>-</td>
<td>+</td>
<td>3–4</td>
<td>+</td>
<td>L1–2</td>
<td>T3–S1</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>7</td>
<td>F</td>
<td>Left</td>
<td>+</td>
<td>2–3</td>
<td>(+)</td>
<td>+</td>
<td>4</td>
<td>+</td>
<td>+</td>
<td>L2–3</td>
<td>L1–L4</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>8</td>
<td>F</td>
<td>Left</td>
<td>-</td>
<td>2</td>
<td>+</td>
<td>-</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>T4</td>
<td>C7–T3</td>
<td>Asymmetric reflexes</td>
<td>—</td>
</tr>
<tr>
<td>9</td>
<td>F</td>
<td>Left</td>
<td>+</td>
<td>2</td>
<td>(+)</td>
<td>+</td>
<td>1</td>
<td>+</td>
<td>L3–4</td>
<td>T5–6</td>
<td>L3–L3</td>
<td>—</td>
<td>—</td>
</tr>
</tbody>
</table>

*Type: 1, massive bone spur; 3, “atypical spinous process”; 4, haze.
ankle reflex was absent. Despite these findings the girl was good at sports and complained of no symptoms.

In Case 9 there was a strong suspicion of diastematomyelia, with spina bifida occulta below L3 and a massive bone spur at the L4 level at the site of a noticeable widening of the spinal canal. Externally a large hairy patch was present at the lumbosacral level.

In summary, all nine children had elevation of the scapula, four had a definitely proven intraspinal bone spicule associated with diastematomyelia and in five other patients, four girls and one boy, there were strong grounds to suspect that diastematomyelia was also present. With the exception of one patient (Case 8) the abnormality was found or presumed to exist at the thoracolumbar or lumbosacral level. Only three patients (Cases 4, 6 and 8) demonstrated neurological abnormality, one of whom had a lumbosacral myelomeningocele with complete paralysis below the level of L1. In this child the elevation of the scapula was bilateral while in the other patients the proportion of left-sided to right-sided elevation was five to three. This agrees with previously reported statistics.

**DISCUSSION**

The literature reveals occasional reports of findings similar to those occurring in our nine patients. Greig (1911) published a picture of a young woman with elevation of the scapula and a large lumbosacral hair patch, similar reports were made by Flotow (1929) and Junge (1942), while Schwarzweller (1937) illustrated a possible diastematomyelia at the L3 level.

There have been other occasional reports of patients with congenital elevation of the scapula and neurological disturbance including urinary and faecal incontinence (Hensinger and MacEwen 1975).

In a review of ninety cases of diastematomyelia, Burrows (1968) mentioned one child with elevation of the scapula. Cavendish (1972) reported 112 children with elevation of the scapula of whom three had diastematomyelia. Keim and Greene (1973) found 153 cases of diastematomyelia in the literature and added a further twenty new patients, one of whom, a woman, also had elevation of the scapula. None of these authors attribute any special significance to the coincidental finding of the two relatively rare deformities.

We believe that the combination of these developmental defects may be rather frequent, and suggest it may be due to a common embryological maldevelopment. One explanation could be the hydromyelic theory of Gardner and Collis (1969), in conjunction with the “bleb” theory of Engel (1940, 1943). The alternative theory, which we believe is more likely, is that of the “split notochord syndrome” (Feller and Sternberg 1929, 1932; Bremer 1952; Bentley and Smith 1960), which relates diastematomyelia to other known malformations of the spinal column. If one assumes that the notochord induces the development of the anlage of the spinal cord and of the vertebral bodies, while the spinal cord induces the formation of the laminar arches as a secondary stage (Tondury 1958), it is possible to imagine that both these developmental organisers could have an inductive influence on the formation of the upper limb buds. Such a disturbance of the development of the notochord at this time might explain the malposition of the scapula.

I am grateful to Professor Dr H. Cotta, director of the University Orthopaedic Hospital, Heidelberg, for allowing me to review his patients and make this report; and I am indebted to Dr C. W. Claussen, Deutsches Krebsforschungszentrum Heidelberg, for the helpful confirmation of Case 3 by computerised tomography. I wish to express my thanks to Mr Hugh McKim Thomas, Nottingham, for his help in preparing this paper.

**REFERENCES**


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