Combined diastematomyelia and hemivertebra
A REVIEW OF THE MANAGEMENT AT A SINGLE CENTRE

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A combination of hemivertebrae and diastematomyelia is rare. We have identified 12 such patients seen during a period of 11 years in the orthopaedic, spinal and neurosurgical units in Nottingham and analysed their treatment and outcome.

Diastematomyelia, the congenital splitting of the spinal cord at one or more levels is a rare condition often associated with a bony, cartilaginous or fibrous spur extending into the median plane across the spinal canal. Between 50% and 60% of these patients have an associated scoliosis secondary to vertebral anomalies such as a butterfly vertebra or hemivertebra.2

A hemivertebra lacks half of the body and neural arch on one side, due to the absence of one of the centres of chondrification.3 The condition is found in five to ten per 10 000 births, occurring more commonly in girls.4 Patients with hemivertebrae often present with congenital scoliosis. Various methods exist for correction of scoliosis secondary to hemivertebrae including hemivertebrectomy5 and/or epiphysiodesis.6,7 These are axial shortening procedures.

Patients with a combination of diastematomyelia and hemivertebrae are rare. There is very limited information on the outcome of patients with both diastematomyelia and hemivertebrae, particularly following surgery. A search of recent literature yielded no reported outcome studies. We have analysed the outcome of patients with the two abnormalities treated at a single centre to determine how they were managed and whether surgery influenced the result.

Patients and Methods
A prospectively-acquired database containing information of cases seen jointly by the neurosurgical and orthopaedic spinal units at the Queen’s Medical Centre, Nottingham was analysed. Data regarding the diagnosis, the number of hemivertebrae, the type and location of the spinal deformity, the type of surgery undertaken, the duration of follow-up and the outcome were collected. All patients had previously been referred to the units from paediatricians, general practitioners, and other sources.

Results
Between 1990 and 2001, 12 patients with both hemivertebrae and diastematomyelia were seen. There were 11 women and girls and one boy; the age at diagnosis ranged from 4 months to 41 years and the median period of follow-up for ten patients was 9.5 years (2 to 26). Two were followed up elsewhere. The number of hemivertebrae ranged from one to four, with a mean per patient of 2.2. Spinal deformity secondary to hemivertebrae and diastematomyelia was seen in ten patients (Table I). One patient had diastematomyelia and hemivertebra without spinal deformity as the hemivertebra was at L5. In another patient the spinal deformity was not classified according to region. Before operation, all but two patients were independently mobile. One of

<p>| Table I. Location of the spinal deformity associated with diastematomyelia and hemivertebra |</p>
<table>
<thead>
<tr>
<th>Curve</th>
<th>Location</th>
<th>Number of patients*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Scoliosis</td>
<td>Cervicothoracic</td>
<td>2</td>
</tr>
<tr>
<td>Thoracic</td>
<td>4</td>
<td></td>
</tr>
<tr>
<td>Thoracolumbar</td>
<td>3</td>
<td></td>
</tr>
<tr>
<td>Kyphosis</td>
<td>Thoracolumbar</td>
<td>1</td>
</tr>
</tbody>
</table>

* in one patient no deformity was identified and in another a deformity was not specified

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these had an associated spina bifida while a further four had spina bifida occulta.

Associated abnormalities of the limbs were found in three patients. One had a small left leg, another a Sprengel shoulder deformity and the third a progressive sensory deficit in the lower limbs. A further patient had an associated horseshoe kidney.

All patients had either an orthopaedic spinal or neurosurgical operation, or both (Tables II and III). Cord monitoring was used in all cases where spinal surgery was performed. Four patients had an operation on the spine for progressive deformity. Convex anterior/posterior epiphysiodesis was carried out where there was a complex arrangement of multiple hemivertebrae; while spinal fusion using Harrington instrumentation, or hemivertebrectomy was used when the deformity was caused by an isolated hemivertebra. Where the cord was deemed vulnerable or where surgical access was likely to be difficult if intervention were needed should there be neurological deterioration, a prophylactic neurosurgical procedure was performed. This occurred in three patients. In two, untethering of the cord was required and in the other mobilisation of the diastematomyelia. Where the cord was deemed vulnerable or where surgical access was likely to be difficult if intervention were needed should there be neurological deterioration, a prophylactic neurosurgical procedure was performed. This occurred in three patients. In two, untethering of the cord was required and in the other mobilisation of the diastematomyelia. The mean length of time between the neurosurgical and orthopaedic operations was three years. In two, further patients concurrent procedures were performed. One had an anterior wedge resection, osteotomy and instrumentation with removal of a diastematomyelia and mobilisation of the cord and the second had a posterior fusion and excision of diastematomyelia. Spinal fusion was carried out because both patients had reached skeletal maturity. A decision to remove the diastematomyelia was taken as it was felt that should it become symptomatic surgery would be difficult in the presence of a fused spine. One patient remained unhappy with the cosmetic result and the spinal fusion was revised with the subsequent development of paraparesis of the left leg. Other complications following spinal procedures included post-operative paraparesis due to intra-operative hypotension in one patient, and the position of the rod in two others which required its removal. Superficial wound sepsis occurred in a further patient.

Two patients underwent excision of the diastematomyelia and division of the filum, for progressive lower limb paraparesis. An expectant policy was adopted for these patients with respect to their spinal deformity. The mean age of nine of the patients at neurosurgery was 10.8 years (3 months to 25 years). A single separate patient underwent division of a diastematomyelia eight years after spinal fusion.

At follow-up, two patients had persistent paraparesis due to intra-operative hypotension from excessive blood loss and from traction of the cord in a straight spine. Despite this all were independently mobile. The two patients who had paraparetic lower limbs initially had a resolution of their symptoms following neurosurgery.
Discussion

The embryology of diastematomyelia. Diastematomyelia is the congenital splitting of the spinal cord due to a fibrous, bony or cartilaginous spur. How this occurs is unclear, though various theories exist. One suggests an aberrant event such as the application of a teratogen on the primitive streak and node which form the notochord during the third week of embryogenesis, resulting in failure of separation of the neuroectodermal and endodermal layers. Cephalad growth of the notochord leads to a divergence at the level of the neuroectodermal adhesion, forming a split cord. A study in 1992 coined the term split-cord malformation which encompassed conditions involving double spinal cords including diastematomyelia. It proposed a common embryogenetic mechanism leading to the formation of split-cord malfunctions. Adhesions formed between the ectoderm and the endoderm result in the appearance of an accessory neuroenteric canal. Around this condenses an endomesenchymal tract which divides the developing notochord, resulting in the formation of two hemineural plates leading to two cords. Two types have been classified. Type I consists of two hemicords each with its own dual tube and separated by a rigid, dural-sheathed osseocartilaginous median septum (diastematomyelia). This is the most common type of cord malformation. Figure 1 shows the MRI image of a 14-month-old girl with multiple segmental vertebral anomalies, detected on antenatal screening, and a twin sac diastematomyelia with a bony spur at T11/12. Type II has the two hemicords contained within a single dural tube and separated by a non-rigid fibrous median septum (diplomyelia). Figure 2 illustrates this in an MRI of a five-month-old boy.

Embryology of hemivertebrae. Gastrulation occurring in the embryonic period converts the bilaminar embryonic disc to a trilaminar structure containing ecto-, endo- and mesoderm. Spinal development involving the formation of mesodermal-derived somites then culminates in the formation of primitive vertebrae. By day 36, chondrification, which begins in the mesenchymal centrum on either side of the notochord, leads to a rapid increase in vertebral size. By day 63, small vessels have invaded the centrum and ossification begins. Failure of chondrification of the vertebral precursor due to unilateral lack of vascularisation, decreased nutrition or sclerotomal deficiency results in the formation of hemivertebrae.

Another proposed mechanism for hemivertebral formation involves the independent but synchronous development of each somite pair. Under normal circumstances, these somites position themselves opposite to one another prior to fusion in the midline to form a normal vertebra. If however, the paired somite derivatives are not in precisely the same development phase, the slow side moves one segment caudally, leading to the formation of a solitary hemivertebra. The patients in this study exhibited a variety
of vertebral anomalies. These ranged from a failure of formation (true hemivertebrae), and failure of segmentation (block vertebra, unsegmented bar) to a combination of both (Table IV\textsuperscript{11}). Seven of the patients had complex vertebral abnormalities involving an overlap of hemivertebra types. It was not possible to classify the different types accurately since the clinical management did not justify the use of computed tomography to assess the structure and the complexity of the multiple hemivertebrae precluded accurate assessment using plain radiographs.

**Natural history of diastematomyelia/hemivertebra without surgical intervention.** The prognosis of patients with diastematomyelia and hemivertebrae without surgery is not clear. Some studies have suggested that if a diastematomyelia is left alone no significant problems occur. A retrospective study involving 12 children\textsuperscript{12} found that even though patients had a neurological deficit no new symptoms appeared with no worsening of the neurological state in a follow-up of two to ten years following conservative management. This suggests that in patients with asymptomatic or non-progressive diastematomyelia an expectant policy should be adopted.\textsuperscript{1,13} However, in some patients with pre-existing neurological abnormality, unresected diastematomyelia caused progressive neurological deterioration.\textsuperscript{13} Whether excision of hemivertebrae is needed depends on the type. Semi-segmented and incarcerated hemivertebrae usually do not require treatment,\textsuperscript{14} but fully segmented non-incarcerated hemivertebrae may require prophylactic management to prevent significant spinal deformity,\textsuperscript{14} as was the case in our study. In eight of the cases, the Cobb angles were > 45° and were caused by a complex combination of vertebral anomalies.

**Advantages and disadvantages of surgery.** Surgery in patients with diastematomyelia and hemivertebrae aims to prevent progressive deformity and neurological deficit.\textsuperscript{1,15} A variety of strategies is available to correct the spinal deformity due to hemivertebrae. These include convex epiphysiodectomy,\textsuperscript{9} hemivertebrectomy via a posterior approach,\textsuperscript{5} convex fusion and concave distraction\textsuperscript{16} and anterior release with posterior stabilisation.\textsuperscript{17} These techniques are thought to be reliable and safe.

It has been suggested that treatment of congenital scoliosis in the presence of diastematomyelia should be based on the size of the curve and its progression. In cases where a neurological deficit is present resection of the diastematomyelia should be performed prior to spinal correction.\textsuperscript{14} However, significant neurological damage has been recorded during the prophylactic removal of bony spurs.\textsuperscript{18}**

**Prophylactic surgical options when both conditions co-exist.** Where both diastematomyelia and hemivertebra exist, there remains controversy regarding management. Some studies have suggested anecdotally that the presence of a spur in diastematomyelia in the absence of progressive neurological, is not a sufficient indication for its primary removal, even if a concomitant deformity needs correction.\textsuperscript{13,18} However, others\textsuperscript{1,15} have recommended that prior prophylactic excision of any tethering lesion, including diastematomyelia, be undertaken before spinal correction in order to prevent the risk of neurological deficit developing from cord traction. Pang et al\textsuperscript{18} recommended that split-cord malformations are cord-tethering lesions which are likely to cause neurological deficit and should be treated, since there was subsequently a demonstratable improvement or stabilisation of the neurological status of the patient. From our study it appears that prophylactic treatment of diastematomyelia is not necessary in the presence of significant spinal deformity, as surgery for the latter will shorten the neuro-axis relieving the tethering lesion. However, concurrent correction of the spinal deformity and mobilisation or excision of the diastematomyelia should be undertaken if there is a potential neurological problem which would be technically difficult to resolve after spinal fusion.

**Antenatal diagnosis.** Diastematomyelias appear as echogenic foci located in the posterior aspect of the spine and are associated with widened interpedicular spaces seen on sonography during the antenatal period.\textsuperscript{7,19} This technique has also been used to identify anomalies in the fetal vertebral bodies such as hemivertebrae,\textsuperscript{20-22} allowing prenatal counselling and early, even prophylactic, surgical management if necessary.

Patients with concurrent diastematomyelia and hemivertebrae are rare. The justification for surgery is based on the degree of neurological impairment and spinal deformity. Good surgical and functional outcomes can be achieved in selected groups of patients. Diastematomyelia and hemivertebrae are abnormalities of development. For a satisfactory outcome management begins prior to conception. A multidisciplinary approach involving geneticists, obstetricians, paediatricians, neurosurgeons, spinal surgeons, physiotherapists and psychologists is necessary. Individuals with a positive family history of spinal abnormalities should be counselled and advised prior to conception. Facilities such as genetic testing and antenatal ultrasonography are available to diagnose problems following conception. Once identified, surgical management can be planned and performed and treatment made available to ensure that patients remain mobile and independent.

\textsuperscript{11} Table IV. Classification of hemivertebra\textsuperscript{11}

<table>
<thead>
<tr>
<th>Classification</th>
<th>Description</th>
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<tbody>
<tr>
<td>A</td>
<td>Failure of formation&lt;br&gt;Hemivertebra&lt;br&gt;Semi-segmented&lt;br&gt;Fully segmented&lt;br&gt;Wedge vertebra</td>
</tr>
<tr>
<td>B</td>
<td>Failure of segmentation&lt;br&gt;Block vertebra&lt;br&gt;Unsegmented bar&lt;br&gt;Combination</td>
</tr>
<tr>
<td>C</td>
<td>Unsegmented bar with hemivertebrae</td>
</tr>
</tbody>
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No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.
References


