New data on diastematomyelia

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Five cases of diastematomyelia are described and illustrated. These and a further 60 cases from the literature are analyzed. The data corroborate the opinion that early diagnosis and treatment result in a better prognosis.

KEY WORDS • diastematomyelia • diplomyelia • lumbosacral bands • computerized tomography • early treatment

DIASTEMATOMYELIA as defined by Matson,[28] is a rare congenital division of the spinal cord or its intraspinal derivatives. The split may extend in a sagittal plane for a variable number of segments. There may be a septum of bone or fibrocartilage within the split. James and Lassman[18] emphasized that diastematomyelia refers to the split neural tissue, not to the septum, since 20 of their 41 cases had no septum. They note that “since Mat- son[26] does not refer to diastematomyelia without a septum, readers have erroneously assumed that the septum is diastematomyelia.”

The term “diplomyelia” refers to duplication of the spinal cord. Since it can be differentiated only by examination of the transected cord, James and Lassman[18] and Matson[26] preferred the term “diastematomyelia” for these clinically indistinguishable conditions.

The first description of congenital back conditions was made by Professor Nicolai Tulp in 1624.[28,85] In 1761, congenital lesions of the back were further described by Giovanni Battista Morgagni.[28] It is Ollivier[80] who is credited by Perret[81] as being the first to refer to the split cord as a diastematomyelia. He coined the term in 1837 from the Greek “diastema” two, and “myelon” marrow. In 1842, Cruveilhier described the septum.[7] Experimental diastematomyelia was produced by Lereboulet in 1868,[24] Roux in 1888,[28] and Hertwig in 1892.[14] In 1886, Humphrey[16] described six specimens of spina bifida with extensions of bone from the bodies of the vertebrae projecting into the spinal canal. In 1920, Lipshutz[22] described a patient with a double spinal cord.

In 1936, Hamby[12] reported a case with multiple anomalies of the cord, and reviewed the literature. In 1940, Lichtenstein[29] classified diastematomyelia as a dysraphic myelodysplasia and considered it a pseudoduplication. In the same year, Herren and Edwards[13] described 43 cases from the literature, 15 of which had an “osseo-chondromatous process.” In 1949, Neushauser, et al.,[29] stated for the first time the radiological criteria for diagnosis, and since then Cowie,[9] Scatliff, et al.,[34] and Hilal, et al.,[15] have made significant radiological contributions.

A milestone was marked by Matson in 1969,[25] when he published his series of 31 operated cases. He stressed the importance of early diagnosis, and others have since agreed.[9,15,17,18,21,23,24,27,31,32,34,40,41]

Clinical Material and Methods

A search of the literature produced almost 200 cases of diastematomyelia. However, only 60 of these patients[1,8-12,19,21,23,24,27,31,32,35-37] were reported in sufficient detail for analysis. These cases were reviewed along with five new cases.

In this analysis the frequency of occurrence of the presenting features and the clinical findings in each case were compared with additional details on age, sex, level of the lesion, and outcome. The cases were divided into age groups: infants up to 1 year, children 1 through 9 years old, adolescents aged 10 through 19 years, and adults aged 20 years or more.

Case Reports

Case 1

This 17-year-old adolescent was referred in 1975 for assessment before fusion of a severe and progressive scoliosis. His only complaint was inability to stand up straight. When he tried to do so, he felt a tingling in his lower limbs. Neurological examination was nor-
mal. He said three sisters had died with “spina bifida.” X-ray examination showed an abnormal midline calcification at T3-5. The diagnosis of diastematomyelia was confirmed at myelography (Fig. 1), which also showed a low conus medullaris. At operation, a septum of bone, unusually high in the thoracic spine, was removed (Fig. 2). At a second operation, the lumbosacral region was explored, and a thickened and taut filum terminale was found; this was considered the cause of the low conus medullaris. On division of the filum, the ends promptly moved apart by 3 cm or more.

Postoperatively, the patient said he felt “much straighter” and no longer had the tingling in his legs. On account of his improved stance and gait, orthopedic correction was deferred. He remained well. In 1978, his back was finally fused, and the scoliosis improved from 77° to 40°. Four months later he continues to do well and is markedly taller.

Case 2

This 9-year-old boy was referred in 1975 with incontinence for many years. During radiological investigation of his genitourinary tract he was noted to have the three classic signs of diastematomyelia at the L-4 spinal level, namely, an abnormal midline calcification, spina bifida occulta, and a widened interpedicular distance. Clinical examination on referral showed that his left leg was shorter than the right by 1.5 cm (Fig. 3). Both ankle jerks were absent, his feet were hyperesthetic, he had mild hammer toes, and lumbar hypertrichosis. Air myelography outlined the diastematomyelia, and also indicated traction on the conus medullaris. Two operations were performed, the first to remove the septum from within the split cord, the second to divide a fibrous band adhering the cauda equina to the dura. Three years later he is asymptomatic and plays football.

Case 3

This 12-year-old girl was referred in 1975 for assessment of an area of calcification seen radiologically at T9-10 many years before. Her left leg was shorter than the right by 2.5 cm, with a cavus deformity of her left foot. She also had muscle weakness of her left leg. She walked with a limp and was scoliotic. Myelography showed a midline filling defect at T9-10, and suggested the presence of an internal meningocele in the lumbar region. At the first operation the septum of bone was removed, and, at the second, a fibrous band adhering the cauda equina to the dura and supporting a pocket of cerebrospinal fluid was divided, releasing the meningocele. She made a satisfactory recovery. Three years later she is fully active and enjoys cycling. Her back, leg, and foot deformities are no worse.
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Case 4

This 10-year-old girl was referred in 1975 with a 9-month history of a limp. She was scoliotic, and had hip pain and lumbar hypertrichosis. Her left leg was shorter than the right by 4 cm. A pes cavus deformity had been corrected in infancy. Her brother had died with a "spina bifida" condition. Plain x-ray films showed abnormal midline calcification at L2-3, and myelography showed the bands securing the filling defect. In two stages, the septum was removed and the cauda equina was separated from the dura. Her pain decreased rapidly, although it persisted. Two years later her scoliosis was treated with traction for 1 month but has remained at 65°. Three years postoperatively, she still complains of occasional pain in the left side of her back, but is active at netball and swimming.

Case 5

This case was discovered at operation in 1977 for closure of a draining myelomeningocele. At birth, this baby girl had normal leg movements, normal muscle tone, normal withdrawal to pinprick, and normal anal sphincter tone. At operation when she was 10 days of age, diastematomyelia was found in addition to the myelomeningocele. The cauda equina was split by a septum of bone. The cutaneous defect was closed without removal of the septum. She recovered well, and the wound healed without complication. The septum will be removed only if symptoms or signs develop. A computerized tomographic (CT) scan showed hydrocephalus, and ventriculography showed an Arnold-Chiari malformation. Accordingly, a ventriculoperitoneal shunt was inserted. She continues to do well.

Discussion

In the following discussion are summarized the results of the analysis of the 60 cases fully reported in the literature and the five new cases. Only new data revealed by the analysis of the total 65 cases are presented.

Sex

There was a female to male ratio of 2.4:1 among the 65 cases. This is different from other reported ratios, but is derived from a larger number of patients.

Age

The age at presentation ranged from 10 days to 59 years in life, and 64 years post mortem. Figure 4 shows the age of patients at presentation. The average is 4.3 years.

Genetic Predisposition

Although most authors deny a genetic influence, Kapsalakis reported the condition in two sisters. Dale, Hamby, and Perret mention a family history, and two of our patients had a history of dysraphism in parents or siblings.

Presenting Features

Table 1 shows that the two most common presenting features differ radically from group to group, a fact not previously recognized. Matson and James and Lassman stated that foot deformities, limp, and lumbosacral hypertrichosis were the most common presenting features in all cases, but this has not been borne out in this analysis.

Clinical Findings

Table 2 shows that the two most common clinical findings differ markedly from group to group, a fact not previously recognized.
FIG. 4. Graph showing the age of 65 patients at the time of presentation.

TABLE 1

<table>
<thead>
<tr>
<th>Feature</th>
<th>Total Cases</th>
<th>Adults</th>
<th>Adolescents</th>
<th>Children</th>
<th>Infants</th>
<th>Total</th>
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<td>4*</td>
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<td>3</td>
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<td>8</td>
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<td>3</td>
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<td>6</td>
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<td>5*</td>
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<td>8</td>
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<td>1</td>
<td>4</td>
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<td>11</td>
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<td>2</td>
<td>1</td>
<td>1</td>
<td>3</td>
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<td>1</td>
<td>1</td>
<td>1</td>
<td>2</td>
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<td>1</td>
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<td>1</td>
<td>1</td>
<td>1</td>
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</tr>
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</table>

*The two most common presenting features in each group.

TABLE 2

<table>
<thead>
<tr>
<th>Findings</th>
<th>Total</th>
<th>Adults</th>
<th>Adolescents</th>
<th>Children</th>
<th>Infants</th>
<th>Total</th>
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<tr>
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<tr>
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<td>40</td>
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<td>46</td>
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<td>22</td>
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<td>8</td>
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<td>22</td>
<td>40</td>
<td>50</td>
<td>8</td>
<td>37</td>
</tr>
<tr>
<td>unequal legs</td>
<td>29</td>
<td>33</td>
<td>40</td>
<td>30</td>
<td>8</td>
<td>29</td>
</tr>
<tr>
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<td>20</td>
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<td>8</td>
<td>21</td>
</tr>
<tr>
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<td>12</td>
<td>6</td>
<td>10</td>
</tr>
</tbody>
</table>

*The two most common clinical findings in each group.
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Matson\textsuperscript{25} emphasized the importance of hypertrichosis, having found it in 45\% of his series of 31 infants and children. However, in this analysis of 46 such cases, hypertrichosis was found in only 26\%. Furthermore, although it was the most common feature in his series, it was only the sixth most common in this series. In view of this, its importance is in doubt. As Table 2 shows, stretch reflex changes are prominent findings.

Asymptomatic cases have been described throughout the age range. Matson\textsuperscript{25} reported no neurological deficit in six of 31 cases. Three further cases are summarized here. The first was in a 64-year-old woman, who was found at autopsy to have diastematomyelia, although she had been described ante mortem as neurologically normal.\textsuperscript{18} The second case was in a 26-year-old woman who presented for cosmetic surgery to a lower thoracic lipoma, and was found by CT to have a diastematomyelia at the T2-3 level.\textsuperscript{43} Finally, Case 5 in this series appeared to have no neurological signs. These cases emphasize that prophylactic surgery is indicated only for those patients with neurological signs or symptoms, except in young patients as discussed below.

Diagnosis

The diagnosis in the 65 cases was established by clinical findings, plain x-ray findings (Table 3), and CT scanning, along with contrast myelography.\textsuperscript{39,42}

Pathology

The mechanism of neural damage whereby the septum and its dural attachments lead to damage within the split cord have been much discussed, but poorly understood. The etiology of the dermal sinus tract, dermoid cyst, or teratoma in the lumbar region has received less attention. The least is known about the bands in this region, considered by James and Lassman\textsuperscript{18} to be aberrant and atrophied nerve roots, and to be a cause of neurological damage by traction on the spinal cord, conus medullaris, filum terminale, or cauda equina. Bands are detected at myelography when displacement of the conus, filum, or cauda is seen. Table 4 shows that in 15 patients bands were confirmed at operation, whereas in 20 cases they were sought at operation or myelography but not found. Of the remaining cases no information is forthcoming. The data in Table 4 suggest that lumbosacral bands may be more common than is recognized, and should always be sought.

The level of the split, analyzed from 145 patients (the 65 cases plus three further series\textsuperscript{15,18,20}) is shown in Fig. 5. The septum is found more frequently at the level of the L-2 vertebra, whereas in Matson's 31 cases\textsuperscript{25} it was most common at L-4.

\begin{table}
\centering
\caption{Diagnostic plain x-ray findings (at the same level) in 65 cases*}
\begin{tabular}{ll}
\hline
X-Ray Finding & No. of Cases \\
\hline
spina bifida occulta & 49 \\
abnormal midline density & 48 \\
widened interpedicular distance without erosion & 41 \\
\hline
\end{tabular}
\footnotesize*Other features found included hemivertebrae, fused vertebrae, hemilaminae, absent vertebrae, and scoliosis.
\end{table}

\begin{table}
\centering
\caption{Associated pathology among 65 cases}
\begin{tabular}{ll}
\hline
Feature & No. of Cases \\
\hline
lumbosacral bands found & 15 \\
lumbosacral bands not found & 20 \\
cauda equina bands & 5 \\
cauda equina septum & 2 \\
low conus medullaris & 4 \\
filum terminale divided surgically & 4 \\
\hline
\end{tabular}
\end{table}
TABLE 5
Operative outcome in 59 cases

<table>
<thead>
<tr>
<th>Result</th>
<th>No. of Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>neurological improvement</td>
<td>44</td>
</tr>
<tr>
<td>no improvement</td>
<td>9</td>
</tr>
<tr>
<td>&quot;poor&quot; result</td>
<td>2</td>
</tr>
<tr>
<td>death (from other pathology)</td>
<td>1</td>
</tr>
<tr>
<td>result not mentioned</td>
<td>3</td>
</tr>
</tbody>
</table>

Therapy

The treatment of clinically apparent diastematomyelia is primarily surgical. The aim of surgery is the prevention of progressive neurological damage. The timing of surgery is controversial. If, as in the majority of cases, there is a history of recent deterioration, then surgery is indicated. When there is no such history, but there are signs or symptoms, surgery is probably indicated prophylactically, but this point has not been decided. With no history, no signs or symptoms, but an incidentally diagnosed lesion, it would seem prudent to withhold surgery until signs or symptoms develop, reviewing frequently, except in infants or children. For these young patients, Matson and Till have recommended prophylactic surgery.

Surgery is directed at removal of the septum and its dural sleeve, and division of the dural attachments to the neural tissue at that level. Any lumbosacral pathology is treated at a second operation.

In patients with bladder dysfunction, Matson believed that operation would be helpful, and could completely correct the dysfunction if it were of recent onset. In scoliotic patients, removal of the septum and other pathology is obviously necessary before correction of the scoliosis. James and Lassman believe that operation would be helpful, and could completely correct the dysfunction if it were of recent onset. In scoliotic patients, removal of the septum and other pathology is obviously necessary before correction of the scoliosis.

To prevent the irreversible and progressive neurological damage of diastematomyelia, early diagnosis and treatment is of paramount importance.

Acknowledgments

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