THE LUMBAR THECO-PERITONEAL SHUNT SYNDROME AND ITS SURGICAL MANAGEMENT

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Seven patients treated in infancy by a lumbar theco-peritoneal shunt for idiopathic communicating hydrocephalus presented later in childhood after developing a characteristic orthopaedic syndrome. This included a severe, rigid and progressive lumbar hyperlordosis, severe bilateral restriction of straight leg raising and abnormalities of stance and gait.

Four of the patients, who had severe hyperlordotic curves of over 90°, required operations to correct their extreme deformity. The recommended method of correction is a three-stage procedure: first, anterior wedge resection osteotomies at several levels in the lumbar spine, then a period of “90-90” femoral traction, and finally a posterior fusion and stabilisation using Harrington instrumentation. The results were good, with few complications.

This paper describes the presentation and treatment of an orthopaedic syndrome which can develop many years after the insertion of a theco-peritoneal shunt for the treatment of idiopathic hydrocephalus in otherwise normal children. This shunt procedure drains cerebrospinal fluid from the lumbar subarachnoid space to the peritoneal cavity by means of a small polyethylene tube (Matson 1949). More recently, because of recurrent blockages, this shunt has been largely superseded by the more successful ventriculo-atrial shunt. However, some children who had theco-peritoneal shunts inserted many years previously have presented to the Edinburgh Scoliosis Clinic having developed a severe spinal deformity. These children were found to have a characteristic syndrome of extreme lumbar hyperlordosis (Fig. 1), bilateral limitation of straight leg raising and an abnormality of stance and gait. The incidence of this syndrome is not known. In the only reported long-term review, Kushner et al. (1971) examined 28 children several years after the theco-peritoneal shunting procedure and found that 13 had developed a scoliosis and that six of these had a lumbar hyperlordosis.

CLINICAL MATERIAL

Between 1973 and 1979, seven patients treated in infancy by lumbar theco-peritoneal shunt for idiopathic communicating hydrocephalus have presented at the Edinburgh Scoliosis Clinic with a severe lumbar hyperlordosis. There were four girls and three boys, aged between 8 and 16 years (mean 11 years) when they presented.

Theco-peritoneal shunt had been carried out on them between the ages of two months and 18 months (mean seven months). During the four years after the shunt, all the patients had required revision operations to renew the tube, which had become blocked, from once to four times in each patient (on average twice). In three patients the lumbar theco-peritoneal shunt was eventually abandoned and replaced by a Pudenz ventriculo-atrial shunt.

Symptoms. The earliest onset of symptoms was at a mean age of nine years, the most common initial complaint being increasing difficulty in walking. Three patients stated that they were able to walk only on their toes with their knees bent, having previously walked normally. The other four patients complained that they had a shuffling gait and could walk less far than formerly. Four patients complained of an increasing hollowing of their lower backs. Low backache was present in four patients and two had used a spinal support without improvement. Clinical signs. All but one of the patients were of normal intelligence and attended normal schools. All seven had two major clinical abnormalities: an increased lumbar lordosis, and severe bilateral restriction of straight leg raising. Six patients also had some scoliosis but increased lumbar lordosis was the major deformity. Lumbar hyperlordosis. The increase in lumbar lordosis was mild in two children, moderate in one and very severe in four. Clinical details of these severe cases are given in Table I, while photographs and radiographs of two of them are shown in Figures 1 to 12. The patients with severe hyperlordosis stood with marked protrusion of the abdomen and shortening of the trunk (Figs 1 and 7). The pelvis was rotated forward to such a degree that the anus faced posteriorly. The sacrum was hori-
Case 2. Photographs and radiographs before treatment. Figure 1—A girl aged 10 years with severe lumbar hyperlordosis. She can stand only on tiptoe with hips and knees flexed, because of the tightness of her hamstrings. Figure 2—A lateral radiograph, taken standing, shows a 109° lumbar lordosis with a horizontal sacrum (inclination 90°). Figure 3—A radiograph, taken standing, shows a 62° left lumbar scoliosis, causing the upper trunk to list to the right.

After treatment. Figure 4—One year after anterior wedge osteotomies and fusion, the lumbar lordosis and posture are normal. The patient has gained 7.6 cm in height. Figure 5—At the age of 14 years, there is solid anterior spinal fusion from L3 to the sacrum. The lumbar lordosis and sacral inclination have remained normal. Figure 6—A radiograph at 14 years shows that the lumbar scoliosis which was only 14°, after her operations, is now 34°.
Table 1. Details of the four patients with severe lumbar hyperlordosis who had operative treatment

<table>
<thead>
<tr>
<th>Case number</th>
<th>Age years</th>
<th>months</th>
<th>Sex</th>
<th>Lordosis (°)</th>
<th>Sacral inclination (°)</th>
<th>Scoliosis</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>16</td>
<td>3</td>
<td>M</td>
<td>T11 to S1</td>
<td>165</td>
<td>120</td>
<td>Pre-operative halo traction</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>T7 to L5</td>
<td>52</td>
<td>right lumbar</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>List to right</td>
<td></td>
<td></td>
<td>Anterior lumbar osteotomies L2 3, L3-4 and L4/5</td>
</tr>
<tr>
<td>2</td>
<td>10</td>
<td>0</td>
<td>F</td>
<td>L1 to S1</td>
<td>109</td>
<td>90</td>
<td>Anterior lumbar osteotomies L3 4, L4 5 and L5/S1</td>
</tr>
<tr>
<td></td>
<td>(Figs 1 to 3)</td>
<td></td>
<td></td>
<td>T10 to S1</td>
<td>62</td>
<td>left lumbar</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>List to right</td>
<td></td>
<td></td>
<td>90-90 femoral traction</td>
</tr>
<tr>
<td>3</td>
<td>13</td>
<td>7</td>
<td>F</td>
<td>T11 to S1</td>
<td>150</td>
<td>98</td>
<td>Anterior lumbar osteotomies L3 4 and L4 5</td>
</tr>
<tr>
<td></td>
<td>(Figs 7 to 9)</td>
<td></td>
<td></td>
<td>T5 to T12</td>
<td></td>
<td>right thoracic</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Slight list to right</td>
<td></td>
<td></td>
<td>90-90 femoral traction</td>
</tr>
<tr>
<td>4</td>
<td>12</td>
<td>9</td>
<td>M</td>
<td>T11 to S1</td>
<td>92</td>
<td>88</td>
<td>Anterior lumbar osteotomies L3 4, L4 5 and L5/S1</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>T2 to T12</td>
<td>56</td>
<td>left thoracic</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>List to left</td>
<td></td>
<td></td>
<td>90-90 femoral traction</td>
</tr>
</tbody>
</table>

Horizontal and the apex of the lumbar lordosis was so deep that the posterior elements of the spine could be palpated only with difficulty. In compensation for the lumbar hyperlordosis all patients had a smooth thoracic kyphosis. The hyperlordosis was completely rigid and neither suspension nor forward bending produced any correction. The thoracic kyphosis was not as rigid and could be partly corrected by suspension. In five patients there was a tendency for the upper trunk to list to one side.

Scoliosis. Four patients also had a mild to moderate left lumbar scoliosis and two patients had a moderate right thoracic scoliosis. In four patients the upper trunk listed towards the concave side of the scoliosis. The list was most pronounced in those patients with a lumbar scoliosis and became more severe when they attempted to bend forward at the hips.

Straight leg raising. All seven patients had severe limitation of straight leg raising due to tightness of their hamstrings. In the four patients with severe lumbar lordosis (over 90°) straight leg raising was zero, and in the other three patients it was 40° or less. Because of the tightness of their hamstrings, two patients were able to stand only on their toes with hips and knees bent (Fig. 1), and two other patients had to walk in this manner. In these four patients ankle dorsiflexion could only be achieved when the knee was flexed more than 90°. All the patients had a waddling gait, swinging the pelvis forward on the same side as the leading leg.
Table II. The results of operative treatment of the four patients who had severe lumbar hyperlordosis

<table>
<thead>
<tr>
<th>Case number</th>
<th>Lordosis</th>
<th>Sacral inclination</th>
<th>Scoliosis</th>
<th>Complications</th>
<th>Age</th>
<th>Lordosis</th>
<th>Sacral inclination</th>
<th>Scoliosis</th>
<th>At final review</th>
</tr>
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<tbody>
<tr>
<td>1</td>
<td>T11 to S1</td>
<td>42</td>
<td>T7 to L5</td>
<td>30</td>
<td>No list</td>
<td>T11 to S1</td>
<td>55</td>
<td>56</td>
<td>T7 to L5</td>
</tr>
<tr>
<td>2</td>
<td>L1 to S1</td>
<td>47</td>
<td>T10 to S1</td>
<td>14</td>
<td>No list</td>
<td>L1 to S1</td>
<td>50</td>
<td>45</td>
<td>T10 to S1</td>
</tr>
<tr>
<td>3</td>
<td>T11 to S1</td>
<td>43</td>
<td>T5 to T12</td>
<td>39</td>
<td>No list</td>
<td>T11 to S1</td>
<td>40</td>
<td>48</td>
<td>T5 to T12</td>
</tr>
<tr>
<td>4</td>
<td>T11 to S1</td>
<td>54</td>
<td>T2 to T12</td>
<td>32</td>
<td>No list</td>
<td>T11 to S1</td>
<td>49</td>
<td>40</td>
<td>T2 to T12</td>
</tr>
</tbody>
</table>

There were no motor or sensory disturbances and the only abnormal neurological finding was a moderate increase in the ankle and knee reflexes of one patient.

Radiographs. The angles of lordosis and scoliosis were measured, using the Cobb method (1948), on standard lateral and anteroposterior radiographs, taken with the patient standing. Lumbar hyperlordosis extended from the thoracolumbar junction to the sacrum and measured from 65° to 165° with a mean of 100°. Normal lumbar lordosis ranges from 40° to 60° (Moe et al. 1978).

Sacral inclination was measured on the lateral spinal radiograph using the method described by Wiltse and Winter (1983). Normal sacral inclination is approximately 45°; a horizontal sacrum would have 90° inclination. The inclination in our patients ranged from 72° to 120° with a mean of 85°. In three of the four patients with very severe lordosis, sacral inclination was over 90° and the upper sacral segments were angled downwards towards the floor (Fig. 8).

Lumbar scoliosis, present in four patients, extended

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Case 3. After treatment. Figure 10—One year after anterior and posterior correction and fusion, lumbar lordosis and sacral inclination are both normal. The patient has gained 15 cm in height. Figure 11—At the age of 16 years a lateral radiograph shows solid anterior and posterior fusion. Lumbar lordosis and sacral inclination have remained normal. Figure 12—The radiograph shows that the spontaneous correction of thoracic scoliosis has remained stable at 35°.
from the lower thoracic region to the sacrum (Fig. 3). The mean Cobb angle was 28° (range 20° to 62°). These curves were moderately rotated, and all were associated with a list towards the concave side. Two other patients had thoracic scoliotic curves (Fig. 9) of 46° and 65° with mild rotation.

Pelvic obliquity due to asymmetrical hip flexion was seen on the erect anteroposterior radiographs of two patients. This disappeared completely when the radiographs were repeated with the patients supine. Myelography. Myelograms were performed on the four patients with severe hyperlordosis. Lumbar puncture was impossible because of the severity of the lordosis and Myodil had to be injected by cisternal puncture. In one patient a complete block was seen at the level of the tenth thoracic vertebra and was thought to be due to adhesions. In two there was a block at the apex of the lumbar lordosis which was partial in the supine position, but became complete when the patient was prone. Three patients showed loculation of the Myodil suggestive of arachnoiditis.

**TREATMENT**

The four patients with very severe hyperlordosis required operations in several stages to obtain complete correction of their extreme deformity. Three of these patients were operated upon by MJM.

**Anterior spinal osteotomies.** The lower lumbar spine was exposed anteriorly through a transperitoneal approach and the great vessels mobilised to either side. The lower three lumbar discs and parts of the adjacent vertebrae were excised as anteriorly based wedges which extended back to the posterior longitudinal ligament. Partial correction of the lordosis was obtained by flexion of the hips, with firm downward pressure on the spine.

**Femoral traction.** Complete closure of the anterior open wedges left by resection was obtained slowly over the first few days after operation, by applying increasing femoral traction with both hips and knees flexed to 90° (Fig. 13). If neurological abnormalities developed they could be detected early and the traction released.

**Posterior spinal fusion and stabilisation.** Posterior fusion and stabilisation of the spine by means of Harrington instrumentation was done about two weeks after the anterior operation. No further correction was attempted and the Harrington rods were used only as splints from the thoracolumbar junction to the ala of the sacrum on each side (Figs 11 and 12).

After this operation the patient was returned to an ordinary bed without traction. A well-fitting underarm lightweight jacket was applied when the wound had healed. The patient was then encouraged to walk and was allowed home when confident. The jacket was removed nine months later when the spine was solidly fused.

**RESULTS**

The details of the operative treatment and results of the four patients are shown in Tables I and II. Follow-up was for a mean of four years (range 2 years 8 months to 5 years 6 months), by which time three patients had reached skeletal maturity.

**Lumbar hyperlordosis.** The mean lumbar hyperlordosis in the four patients before treatment was 129° (range 92° to 165°); soon after treatment this was reduced to 46° (range 42° to 54°). At final follow-up good correction had been maintained (Figs 5 and 11). The mean lordosis was 45°, and the range of 42° to 54° is within normal limits.

**Scoliosis.** Two patients (Cases 1 and 2) also had a lumbar scoliosis which was associated with a marked list of the upper trunk towards the concave side of the scoliosis. Both patients were treated by anterior wedge resection osteotomy and "90-90°" traction. Neither had a posterior spinal fusion. The lumbar scoliosis and the list of the trunk were corrected significantly after these procedures, but during follow-up the scoliosis tended to recur (Table II).

A thoracic scoliosis was present in the other two patients with severe hyperlordosis (Cases 3 and 4). These curves corrected spontaneously after anterior osteotomies of the lumbar spine; because of this it was decided that neither curve required treatment. During follow-up one of these thoracic curves remained stable (Case 3, Fig. 12), but the other (Case 4), increased from 32° to 62° and after one year required correction by means of Harrington instrumentation and a posterior spinal fusion. The thoracic scoliosis was corrected to 30° and then remained stable.

**Stance and gait.** Before treatment all four patients tended to walk on tiptoe with knees flexed and a rolling gait. One patient could stand only with hips and knees flexed and heels off the ground (Case 2, Fig. 1). After correction of the lumbar hyperlordosis all four patients could stand.
normally (Case 2, Fig. 4 and Case 3, Fig. 10), and walk with a heel-and-toe gait. However, straight leg raising was minimally improved and as a result they all still tended to walk with a rolling gait, swinging the pelvis forward on the same side as the leading leg.

The correction of the lumbar hyperlordosis in all the patients produced a marked gain in height, ranging from 5 to 15.3 cm. Three patients who had pain before treatment gained complete relief of their low backache after spinal surgery.

**Complications.** There were no deaths. Complete unilateral paralysis of the quadriceps occurred in one patient (Case 1), who had been treated with halo traction before operation. Traction was immediately released but the nerve palsy did not recover. After this experience, traction was not applied to any patient before operation. This was the only permanent and severe complication in the series.

Another patient (Case 2), developed bilateral weakness of dorsiflexion and eversion of the ankles with paraesthesia in the feet while in “90-90” traction after anterior spinal osteotomy. Traction was immediately reduced and no further correction was attempted. Second-stage posterior fusion was not performed for fear of increasing the neurological deficit. A complete neurological recovery took place over the next three months.

**DISCUSSION**

Infants who have had a lumbar theco-peritoneal shunt inserted for communicating hydrocephalus may present later in childhood having developed a characteristic syndrome which is not seen in any other condition.

The main deformity in all our patients was a severe lumbar hyperlordosis. This was first noted at a mean age of nine years and was progressive, giving an extremely ugly appearance. Normal lumbar lordosis ranges from 40° to 60° (Moe et al. 1978); in our severely affected patients the mean lumbar lordosis was 100° and the sacrum was rotated into a horizontal position. The cause of this very rigid deformity is unknown. It is possible that the insertion of the shunt, and the many revisions which were often necessary, could cause irritative arachnoiditis and tethering of the cauda equina to the bony structures. Then, as the child grows, a reflex protective spasm may develop in the posterior spinal muscles to produce an increasing lumbar lordosis which later becomes fixed. Of the other possible causes of hyperlordosis we found no evidence of interlaminar fusion or of significant scars in the paraspinous muscles.

The second component of the syndrome was very severe bilateral limitation of straight leg raising caused by tightness of the hamstrings. This prevented the child from striding forward normally by flexion of the hip and extension of the knee. The patients compensated for this restriction by swinging the ipsilateral side of the pelvis forward with the leading leg and limiting extension of the knee by walking on tiptoe. In our opinion this tightness of the hamstrings was due to protective spasm resulting from arachnoiditis and limitation of movement of the cauda equina. After surgical correction of the hyperlordosis, both stance and gait were improved because of the relative lengthening of the hamstrings which occurred when the pelvis was rotated back into a more normal position.

The few published papers on the surgical correction of hyperlordosis in the theco-peritoneal shunt syndrome have reported a high incidence of serious neurological complications (Hall and King 1971; Steel and Adams 1972; Moe et al. 1978). In our series the anterior closing wedge osteotomies shortened the vertebral column, and so minimised the risk of stretching the tethered cauda equina. Complete correction was obtained postoperatively by slowly increasing femoral traction with both hips and knees in 90° flexion. This technique allows neurological abnormalities to be detected early and reversed by releasing the traction. Pre-operative traction or first-stage correction by means of Harrington distraction rods are absolutely contra-indicated because of the very high incidence of serious neurological complications. We found that the anterior fusion stabilised the corrected hyperlordosis, but failed to prevent recurrence of lumbar scoliosis. To control the lumbar scoliosis a posterior spinal fusion was needed as a second stage. Harrington distraction rods were then used to hold the correction obtained by the anterior operation and post-operative traction, but no further correction was attempted for fear of stretching the tethered cauda equina. Thoracic scoliosis, when present, usually corrected spontaneously after the anterior operation and, because of this, the thoracic curve was not included in the posterior fusion. Any progressive thoracic scoliosis could be treated separately at a later date.

The methods reported here make possible the safe and satisfactory correction of the very severe spinal deformity which can occur as a result of a theco-peritoneal shunting procedure. In our opinion this neurosurgical procedure is now contra-indicated because of these serious orthopaedic complications.

**REFERENCES**


VOL. 67-B, No. 2, MARCH 1985