Scoliosis is a common complication of Duchenne muscular dystrophy, developing in almost all affected children once they become confined to a wheelchair. Individuals with a stable, hyperextended spine are usually spared. Once curves develop, patients are treated by instrumented fusion of the spine, carried out before cardiopulmonary function deteriorates to the point at which surgical intervention is dangerous.

The caudal extent of spinal fusion and instrumentation is a matter of debate. Historically, the recommendation was from the upper thoracic spine to the sacrum. More recently, stopping at the fifth lumbar vertebra has been advocated in selected patients because of a shorter operating time, reduced blood loss and fewer complications due to anaesthesia.

Patients with Duchenne muscular dystrophy have a progressive deterioration in pulmonary function of approximately 4% per year. This decline makes general anaesthesia dangerous in the older individual so that extension of the fusion from L5 to the sacrum later in life may not be possible. It is therefore important to determine whether ending the fusion and instrumentation at the fifth lumbar vertebrae caudally will control spinal alignment in the long term.

Fusion to the fifth lumbar vertebra has been recommended in cases in which pelvic obliquity, measured as the angle between a line tangential to the superior margins of both iliac crests and a line perpendicular to one through the spinous processes of L4 and L5, is less than 10°, and the Cobb angle is less than 40°. This is based on the results of ten cases followed for a mean of 34 months. Our study was undertaken to determine if instrumentation and fusion to L5, based on these recommendations, would control spinal alignment over a longer period of time.

Patients and Methods

We reviewed all patients undergoing spinal surgery for Duchenne muscular dystrophy between 1988 and 1995 at two institutions where selected cases were fused caudally to L5. A diagnosis of Duchenne muscular dystrophy was based on clinical findings and confirmed by muscle biopsy, DNA testing, or both. Discussion about operative intervention for scoliosis began when boys became full-time wheelchair users, and surgery was recommended when curves developed and began to progress. The fusion was carried out cranially to the upper thoracic spine (T1-4). Patients with curves measuring less than 40° and with less than 10° of angulation between the sacrum and the caudal two lumbar vertebrae, were instrumented and fused, stopping caudally at the fifth lumbar vertebra. The procedure was carried out using multiple-level sublaminar wires.
(Luque) with either a modified unit rod with Galveston extensions to the pelvis cut off, a modified unit rod with a cross-link placed at the caudal end, or two Luque rods. The pelvis was included when the curve had a Cobb angle greater than 40° or pelvic obliquity was greater than 10°, using the Galveston technique, with either the unit rod or two Luque rods. All fusions included facetectomy, decortication and allograft bone.

Radiographs of the spine in the sitting position were obtained before operation, shortly after, either while in hospital or at follow-up at six weeks, and at each further review, usually annually. The Cobb angle, end vertebra, apical vertebra, and the angle between a line tangential to the iliac crests and one joining the spinous processes of L4 and L5 were determined on the preoperative radiographs. Pelvic obliquity and thoracic decompensation were measured using several previously reported techniques: 1) torso decompensation was the distance between the spinous process of T1 and a line through the centre of the sacrum, perpendicular to the iliac crests;10 2) sitting obliquity was the angle between a line tangential to the iliac crests, and one parallel to the floor;10 and 3) spinal obliquity was the angle between a line drawn from the spinous process of the first thoracic vertebra and the middle of the top of the sacrum, and one perpendicular to a line tangential to the iliac crests.11 The angle between the iliac crests and a perpendicular to the spinous processes of L4 and L5 was not measured after operation since the spinous processes had usually been removed. We determined intraclass correlation coefficients for these three measurements using those made on ten radiographs by two paediatric orthopaedic surgeons.12 Postoperative radiographs were also used to measure the angle between the endplates at each extreme of the fused segment. We calculated the means and standard deviations for the clinical and radiological parameters and determined the 95% confidence interval (CI) for the difference between the two means. An interval including the value zero is not significant.13

The records were reviewed to identify intraoperative or postoperative complications which might have affected the ultimate result. The use of bracing after surgery was recorded. We reviewed notes made in the outpatient and seating clinic to identify problems related to spinal alignment. Some patients died before the completion of this study; in these cases the last radiograph and clinic notes were used as their latest follow-up.

**Results**

We identified 48 patients with Duchenne muscular dystrophy treated by spinal fusion between 1988 and 1995; 38 underwent fusion to L5 and the remaining ten to the sacrum. Preoperative radiographs showed that every patient fused to L5 had an angle between the sacrum and L4 and L5 measuring less than 10°, and had a Cobb angle of less than 40°. Table I gives the clinical details before operation. The group fused to the sacrum had a greater initial curve, than the remainder of the baseline characteristics did not vary between the two groups.

The intraclass correlation coefficients for pelvic obliquity showed acceptable reliability for spinal and for sitting obliquity, but an unacceptable value for torso decompensation (values 0.74, 0.68, and 0.38, respectively). Therefore, the latter measurement was felt to be unreliable for use in this study and was abandoned.

There were two intraoperative complications recorded in the medical records, both being fractures of the L5 lamina. In one patient the fusion had been extended to the sacrum using the Galveston technique, while in the other sublaminar hooks had been used in a ‘claw’ configuration at L4 and L5.

The mean follow-up was for four years (2 to 8); ten patients had died, and one had moved away and could not be traced. All patients had at least a two-year follow-up. The mean age at the time of fusion was 13 years (10 to 18).

There was an increase in spinal or sitting obliquity between the first postoperative and latest follow-up radiograph in 32 of the 38 patients fused to L5; the mean

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**Table I.** Details of the 48 patients undergoing spinal fusion

<table>
<thead>
<tr>
<th></th>
<th>Fused to L5</th>
<th>Fused to sacrum</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of patients</td>
<td>38</td>
<td>10</td>
</tr>
<tr>
<td>Mean age at surgery (years ± sd)</td>
<td>12.8 ± 2.5</td>
<td>13.1 ± 2.8</td>
</tr>
<tr>
<td>Mean Cobb angle (degrees ± sd)</td>
<td>24 ± 11</td>
<td>48 ± 10</td>
</tr>
<tr>
<td>Mean preoperative obliquity (L4 to S1) (degrees ± sd)</td>
<td>5 ± 4</td>
<td>13 ± 6</td>
</tr>
<tr>
<td>Mean FVC (% predicted ± sd)</td>
<td>56 ± 15</td>
<td>52 ± 14</td>
</tr>
</tbody>
</table>

**Table II.** Radiological outcome for the 38 patients fused to L5 and the ten fused to the sacrum

<table>
<thead>
<tr>
<th></th>
<th>All patients</th>
<th>Preoperative apex above L1</th>
<th>Preoperative apex below L1</th>
<th>Fused to sacrum</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of patients</td>
<td>38</td>
<td>28</td>
<td>10</td>
<td>10</td>
</tr>
<tr>
<td>Mean follow-up (years ± sd)</td>
<td>4.6 ± 1.8</td>
<td>4.2 ± 2.0</td>
<td>4.8 ± 1.7</td>
<td>4.4 ± 1.6</td>
</tr>
<tr>
<td>Mean increase in (degrees ± sd)</td>
<td>6 ± 5</td>
<td>3 ± 3</td>
<td>14 ± 8</td>
<td>2 ± 2</td>
</tr>
<tr>
<td>Spinal obliquity</td>
<td>8 ± 6</td>
<td>4 ± 4</td>
<td>16 ± 8</td>
<td>3 ± 2</td>
</tr>
<tr>
<td>Cobb angle</td>
<td>5 ± 3</td>
<td>2 ± 2</td>
<td>15 ± 7</td>
<td>2 ± 2</td>
</tr>
<tr>
<td>Number of patients with increase in obliquity &gt;20°</td>
<td>3</td>
<td>0</td>
<td>3</td>
<td>0</td>
</tr>
</tbody>
</table>
increase in spinal obliquity was 6° and for sitting obliquity 8°. There was no change in either of these measurements for the patients fused to the sacrum and instrumented to the pelvis. Table II summarises the radiological data.

Of the patients fused to L5, three had an increase in spinal obliquity of more than 20° and all had an increase in sitting obliquity of at least 10°. Of the three boys who developed an increase in obliquity of at least 20°, two required revision surgery, which was performed when their pulmonary function was below 25% of predicted values. The outpatient records of the third patient, who died during this study, made no reference to any problems of seating or sitting balance. Another patient required removal of prominent metalwork and one developed a late infection which needed removal of this material, but neither of these patients complained about sitting balance or seating difficulty.

The preoperative radiographs of these three patients with increasing spinal obliquity were reviewed to determine if they had any characteristics in common. All had an apex of their curve below L1. We studied the radiographs of all patients fused to L5, comparing those with a curve apex below L1 with those with an apex at L1 or higher. The ten curves with an apex below L1 had a mean progression of spinal obliquity of 14°, while the 28 with an apex at L1 or above had a mean increase of only 3°. The 95% CI for the difference did not include the value zero which was statistically significant.

The angle across the fusion masses was measured and four patients showed an increase of at least 5°. The patient with the greatest increase (46°) also had an increase in spinal obliquity of 52° and an increase in sitting obliquity of 29°. There was clear evidence of pseudarthrosis in addition to the worsening pelvic obliquity and a revision procedure was necessary (Fig. 1). None of the patients who had instrumentation to the sacrum required revision or removal of the internal fixation.

Discussion

Most of the individuals fused caudally to the fifth lumbar vertebra had some increase in pelvic obliquity; the degree was relatively small, probably of little clinical significance, and did not require revision during the period of this study. Some patients, particularly those with curves with an apex below L1, did develop greater degrees of obliquity, requiring revision in two cases. None of the patients fused to the sacrum or instrumented to the pelvis showed any increase in their pelvic obliquity. Since most of the patients in this study were healthy at the time of operation, they are expected to have a long life span. Patients fused and instrumented to L5 who have not yet developed obliquity will be continually monitored.

The number of boys who were instrumented to the pelvis is small and complications may have been missed in these few cases. Other series examining fusion to the sacrum in
similar patients show complications related to instrumentation in 0% to 16%. The series with higher complication rates have, however, used non-segmental implants such as Harrington distraction rods, which few surgeons would use today. Most series describing segmental instrumentation to the pelvis for other neuromuscular disorders show relatively low rates of complication.4,7,14,16

Pelvic obliquity is notoriously difficult to measure. This may be even more so after patients undergo spinal fusion and instrumentation, when the landmarks are obscured by the fusion mass and implants. Some of the measurements, such as sitting obliquity, are dependent on the position of the patient at the time at which the radiograph is obtained. We compared three previously reported methods of measuring pelvic obliquity for interobserver reliability, and found that spinal obliquity and sitting obliquity were the best. These measures may be more reliable because they use landmarks at the top of the thoracic spine, the top of the pelvis, or the bottom of the x-ray film, which are relatively easy to identify. Despite their reliability, however, they are likely to have an error, and small changes in the measurement may not represent true changes in alignment, similar to findings for the measurement of the Cobb angle. Patients who developed greater changes in obliquity had a large enough change in alignment for the error in measurement to be unlikely to play a significant role.

Worsening alignment may be due to tilt at the L5-S1 disc space or pseudarthrosis of the spinal fusion. Four of the patients fused and instrumented to L5 showed a significant increase in Cobb angle over the fused segment of the spine, suggesting pseudarthrosis, while none of the patients fused to the sacrum showed any change. While it is possible that increased tilt due to pseudarthrosis is attributable to surgical technique, this is not likely to be the case, since the worsening obliquity was not related to one surgeon and most of each of the surgeons’ cases did not show worsening of obliquity or evidence of pseudarthrosis. The type of instrumentation could be responsible and perhaps utilising a system with screw or hook fixation into the lumbar spine may avoid this complication. The risk of revision surgery in an older individual with Duchenne muscular dystrophy may be too great to advocate trying other instrumentation systems.

One reason for fusing a spine short of the sacrum is that function may be adversely affected; patients with spina bifida have great difficulty in catheterising themselves after fusion to the sacrum. There are no studies comparing functional differences between fusion to the sacrum and fusion to L5 in muscular dystrophy. Studies examining patients with this condition who were fused to the sacrum did not identify significant functional problems.14,15

Decreased blood loss and fewer complications associated with a shorter operating time are potential benefits of fusion to L5. Both blood loss and perioperative complication rates have declined in recent years because of advances in techniques of anaesthesia and blood preservation. Therefore, to compare the perioperative rates of complication of recent cases, when most of the fusions to L5 were carried out, with those of older cases when all patients were fused to the sacrum, is not appropriate.

Patients with Duchenne muscular dystrophy develop a progressive decline in cardiopulmonary function with time, eventually making it dangerous to carry out spinal surgery. It is therefore important to fuse the appropriate levels at the initial operation. Fusion to L5 may decrease the operating time, but it may allow a subsequent increase in pelvic obliquity. This may require a revision operation which may be life-threatening. We recommend fusion to the sacrum for scoliosis in Duchenne muscular dystrophy, particularly in patients with an apex to their curve below L1.

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