Progression of scoliosis after spinal fusion in Duchenne’s muscular dystrophy

W. J. Gaine, J. Lim, W. Stephenson, C. S. B. Galasko

From The Royal Manchester Children’s Hospital, Manchester, UK

A consecutive series of 85 patients with Duchenne’s muscular dystrophy who underwent spinal fusion over a period of 16 years was followed up with regard to the progression of the scoliosis and pelvic obliquity. Of 74 patients with adequate radiographic follow-up, 55 were instrumented with the Luque single-unit rod system and 19 with the Isola pedicle screw system; seven were instrumented to L3/4, 42 to L5, 15 to S1 and 10 to the pelvis with intrailiac rods.

The mean period of follow-up was 49 months (SD 22) before and 47 months (SD 24) after operation. There was one peri-operative death and three cases of failure of hardware.

The mean improvement in the Cobb angle was 26° and in pelvic obliquity, 9.2°.

Fusion to L3/4 achieved a poorer correction of both curves while intrapelvic rods, achieved and maintained the best correction of pelvic obliquity. Fusion to S1 did not provide any benefit over more proximal fusion excluding the sacrum, with regard to correction and maintenance of both angles. The Isola system appeared to provide and maintain a slightly better correction of the Cobb angle.

The management of the spinal deformity in Duchenne’s muscular dystrophy (DMD) has evolved considerably since the early 1970s. Spinal orthoses were said not to control the curve in 94% of patients and the use of the Harrington rod distraction technique resulted in many complications and failures of hardware. Luque segmental instrumentation with sublaminar wires was introduced in the late 1970s. The L-shaped Luque double rods had a problem of vertical shift and rotation, and a coupled single-unit rod was therefore introduced and widely used in the 1980s to counter these forces. In 1982 a technique using intrailiac rods (Galveston) allowed correction of pelvic obliquity and in the 1990s the Cotrel-Dubousset and Texas Scottish Rite Hospital systems which use laminar hooks, were introduced. The Isola system uses a combination of fixation by pelvic screws with hooks into the lumbar spine (Fig. 1). This has been reported to give good correction of scoliosis and pelvic obliquity when used with Galveston pelvic extension rods.

The trend in the 1980s was to extend the fixation to the pelvis or sacrum. However, the prolonged operating time and increased blood loss persuaded many to limit their surgery to the lumbar spine but fusion only to L3/4 risked retrolisthesis and translation of the L5 vertebra with persistent pain.

The timing of fusion is dictated by the degree of curvature and pulmonary function. Since 95% of curves progress once patients are wheelchair-bound, the trend over the last decade has been to fuse early. Variability in the progression of the curve makes the prediction of spinal deformity difficult. The types of DMD curve have been classified into stable and unstable and into type 1 (progressive ‘C’ lumbar curve), type 2 (double curve with variable progression) and type 3 (which does not progress).

The hyperlordotic lumbar spine tends to be stable, in contrast to a kyphotic spine. The pattern of the curve, the Cobb angle and the vital capacity at the age of ten years are parameters which have been used recently to predict the progression of the curve. A plateau of the vital capacity of less than 1900 ml has been associated with rapid progression of spinal deformity.

Pelvic obliquity is caused in part by the general muscle weakness leading to a general imbalance of the trunk with the pelvis becoming part of the curvature (Fig. 2). Dubousset classified the region of pelvic obliquity as suprapelvic, pelvic and infrapelvic. Gravity accelerates the progression of the spinal
deformity and a flexion contracture of the hip leads to subluxation exacerbating the pelvic tilt.\textsuperscript{17,18} Chan, Galasko and Delaney\textsuperscript{19} showed that 19 of 54 patients with DMD developed subluxation or dislocation of at least one hip. The main aim of correcting the pelvic obliquity is to facilitate the erect sitting position in a wheelchair, without pressure sores.

The diagnosis of DMD is made on muscle biopsy. Surgery is usually advised when patients become wheelchair-bound and their curves reach 20\(^\circ\) to 30\(^\circ\). In comparison with other centres, in our institution a relatively low percentage of vital capacity (20\% to 25\%) is accepted for surgery provided that the patient does not have a significant cardiomyopathy. Fusion is usually performed using a combination of autologous bone graft from the spinous processes, and freeze-dried allograft. Initially, patients remained intubated in intensive care, but with improved anaesthetic techniques, shorter surgery times and earlier surgical intervention, more recently operated patients have been sent directly back to the ward. Our aim was to review the progress in the scoliosis and pelvic obliquity in 85 patients with DMD who had undergone spinal fusion over a period of 16 years.

**Patients and Methods**

From 1982 to 1998 the records and radiographs of the 85 patients were studied and measurements taken of the Cobb angle and pelvic obliquity, from the date of initial referral to the latest follow-up. Pelvic obliquity was measured from a sitting anteroposterior film, measuring the angle between a line joining the upper level of the iliac crests, and a line horizontal to the film. Different observers took a mean of three sets of measurements. A minimum of three pre-operative and three post-operative readings was required over a minimum period of two years before and two years after operation (four years in total). The number of months before and after the fusion was plotted against the Cobb and pelvic obliquity angles. The kyphotic index was not routinely measured because the lateral radiographs were inconsistent. The rate of progression of the scoliotic deformity was calculated from a series of pre-operative film readings. The correction obtained at the time of instrumentation and the subsequent rate of progression of the curve, if any, were recorded. The level of fusion, the type of instrumentation, the direction and apex of the curve and any post-operative implant failures and complications, were noted.

**Statistical analysis.** A simple statistical model was used to analyse the progression of both the Cobb angle and the angle of pelvic obliquity. The model consisted of a pre-operative trend, the immediate pre-operative angle, the immediate post-operative angle and the post-operative trend. In statistical terms, two aspects of the data required to be taken into account, namely that the dataset was longitudinal, i.e. there were repeated measurements on each individual patient, and that it was unbalanced, i.e. not all
Table I. Details of the patients, the mean spinal deformity and lung function measurements

<table>
<thead>
<tr>
<th></th>
<th>Pre-operative</th>
<th>Post-operative</th>
<th>Last follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean (SD) follow-up in months</td>
<td>23</td>
<td>20 (3 to 65)</td>
<td>10.8 (6 to 35)</td>
</tr>
<tr>
<td>Mean pelvic obliquity (°; range)</td>
<td>26 (0 to 100)</td>
<td>27.3 (6 to 55)</td>
<td>35 (6 to 85)</td>
</tr>
<tr>
<td>Mean Cobb angle (°; range)</td>
<td>6.7 (7 to 106)</td>
<td>1.56* (2 to 2.74)</td>
<td></td>
</tr>
<tr>
<td>Mean vital capacity (litres; range)</td>
<td>228 (96 to 410)</td>
<td>269 (60 to 440)</td>
<td></td>
</tr>
</tbody>
</table>

* best reading achieved at a mean of 10 months after surgery
† best reading achieved at a mean of 14 months after surgery
‡ peak expiratory flow rate

individuals had the same number of measurements, nor were the measurements taken at the same times relative to the date of surgery. To account for these aspects a hierarchical linear regression model (HLM) was fitted to the data using restricted maximum likelihood and the HLM5 program. Separate regressions were performed for the effect of the extent of fusion and type of rod because there were insufficient data to analyse their combined effect. A Box Cox transformation was performed to assess the power transformation of both Cobb and pelvic obliquity models, which best fitted the simple model of a single pre-operative trend, immediate change in angle at the time of operation and single post-operative trend. The statistical hypothesis test was performed using the robust variance estimator of Huber and White. The hypothesis tests were therefore not dependent on the validity of the assumptions of the HLM model. To provide some protection for type-I errors, overall F-tests were performed to test the effect of the extent of the fusion type or type of rod, before performing the individual hypothesis tests.

Another measurement used was the estimation of the projected time at which the post-operative curves would deteriorate to the pre-operative measurements. The null hypothesis tested was that there was no difference between the Luque or Isola implants in terms of correction achieved and maintained (p < 0.5) and that the level of fusion, including pelvic fixation, did not affect the correction and maintenance of the Cobb or pelvic obliquity angles.

Results
Of the 85 patients operated on the radiographs were missing or had been destroyed in five. In another six the measurements of angles were inadequate because of poor quality or insufficient numbers of radiographs. Details of 74 patients are given in Table I. Their mean age at the time of fusion was 13.8 years (12.5 to 18.5). Table II gives details of the fusion sites and type of rod used.

Double curves were present in six patients; these did not progress significantly irrespective of the type or level of instrumentation. Right-sided curves were present in 58% of the patients. Curve apices at L2 or below accounted for 48%, apices at T12 or L1 for 32%, and at T11 or above for 20% of patients.

Major complications included one peri-operative death because of cardiorespiratory insufficiency, three failures of the implant, three wound infections and two cases of symptomatic pseudarthrosis. The metal implant was removed from one patient because of excessive prominence. Fusion at the L5/S1 level was difficult to assess on radiographs but with the deterioration of pelvic obliquity after instrumentation at this level, it is likely that failure of fusion is common. The Isola system achieved a slightly better initial correction of the Cobb angle of 55% (median reduction, from 53° to 25°) as opposed to 42% (median reduction, from 46° to 27°) for the Luque system (not significant; p = 0.103). For both types of rod, the immediate post-operative reductions, however, were statistically different from zero (p < 0.0005) and implant surgery significantly reduced the trend in the progression of the Cobb angle with respect to time (p < 0.0005). The Isola system appeared to maintain the correction more effectively over time (p = 0.096). However, some deterioration occurred in the first two years after surgery with both systems before the curves stabilised.

With regard to an optimal level of fusion, patients in whom fusion was to no lower than L4 did less well in terms of correction of the curve and maintenance of the correction. Although pelvic fixation gave the greatest immediate correction of the Cobb angle, more proximal fusion to L5 had a similar long-term effect and maintenance. Fusion to levels L5, S1 or the ilium gave comparable results with an initial curve of around 50° being maintained at 35° at four to five years (Fig. 3).

Fixation of an intrailiac pelvic rod achieved the best immediate correction of the pelvic obliquity. However, fusion to the sacrum or L5 gave similar long-term correction.

With regard to the level of fusion, analyses suggested that fusion to the pelvis gave a significant (p = 0.028) immediate

<table>
<thead>
<tr>
<th>Fusion site (distal limit)</th>
<th>Rod type</th>
<th>Luque</th>
<th>Isola</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>L4</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>L5</td>
<td>6</td>
<td>1</td>
<td></td>
<td>7</td>
</tr>
<tr>
<td>Sacrum</td>
<td>26</td>
<td>16</td>
<td>42</td>
<td></td>
</tr>
<tr>
<td>Pelvis</td>
<td>13</td>
<td>2</td>
<td>15</td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>56</td>
<td>20</td>
<td>75</td>
<td></td>
</tr>
</tbody>
</table>

THE JOURNAL OF BONE AND JOINT SURGERY
post-operative change in the Cobb angle compared with that to both L3/L4 ($p = 0.019$) and to the sacrum ($p = 0.016$).

Extrapolation of the regression lines to predict when the Cobb and pelvic obliquity angles returned to their immediate pre-operative values provided another tool for statistical analysis (Table III). The results showed again that fusion to the pelvis appeared to maintain pelvic obliquity better than more proximal fusion.

**Table III.** Predicted time in months for return to pre-operative curve angles

<table>
<thead>
<tr>
<th>Fusion site (distal limit)</th>
<th>Cobb angle</th>
<th>Pelvic obliquity</th>
</tr>
</thead>
<tbody>
<tr>
<td>L4</td>
<td>50</td>
<td>57</td>
</tr>
<tr>
<td>L5</td>
<td>116</td>
<td>72</td>
</tr>
<tr>
<td>Sacrum</td>
<td>123</td>
<td>53</td>
</tr>
<tr>
<td>Pelvis</td>
<td>109</td>
<td>108</td>
</tr>
</tbody>
</table>

While some mild curves in DMD are destined not to progress, as illustrated by the type-3 hyperextended curve of Oda et al. and the double curves of Smith, Koreska and Moseley, other curves such as the Oda type 1 are associated with unremitting progression of the scoliosis and pelvic obliquity. In our series there were six hyperextended double curves which did not progress beyond a Cobb angle of 25° and pelvic obliquity of 10° before surgery. These patients may not have progressed without surgery. Identifying the unremitting progression or collapsing spine at an early stage is difficult.

The disadvantages of instrumenting the pelvis include a longer operating time, greater blood loss, a more difficult operation technically, a risk of nonunion and pelvic pain. For these reasons Mubarak, Morin and Leach suggested instrumenting only to the level of L5, unless the pelvic obliquity was greater than 10° or the scoliosis curve greater than 40°. Bony fusion to the pelvis is difficult to achieve because of the severe osteoporosis in patients with DMD. Most authors in North America advocate pelvic fixation. Alman and Kim suggested that lumbar curves tended to be associated with greater progression of pelvic obliquity and advocated pelvic fixation in curves with an apex at or distal to L1. Frost et al. recommended early instrumentation of the sacrum in order to prevent pelvic obliquity. Comparing fusion to the pelvis using Luque/Galveston implants (31 cases) with fixation by pedicle screws to L5 (19 cases) Sangupta et al. found that satisfactory control of pelvic obliquity and the Cobb angle was achieved with fixation at L5 provided that the curve was relatively small and patients were operated on early. We found instrumenting to the sacrum to be no more effective at controlling the pelvic obliquity than instrumenting to L5, but pelvic fixation achieved the greatest immediate correction and best maintenance of correction.
A combined anterior and posterior fusion in one or two stages is the most effective way of correcting pelvic tilt in other neuromuscular cases, but such a combined approach poses a high risk of blood loss and hypotension in DMD. From a review of the literature on DMD post-operative progression of the curve after surgery has been recognised. Difficulty in maintaining an initial correction of pelvic obliquity with instrumentation to L5, S1 and even in some cases with pelvic rod has been noted. Radiographic progression of the curve is common in our experience but it is a slow progression and does not cause functional problems. While obvious causes of such progression are fracture or pullout of hardware and failed bony fusion, less easily recognised causes may include asymptomatic pseudarthrosis at one or more levels, plastic deformation of the rods with remodelling of the fused spine over time and controversially, contractures of the iliotibial band and dislocation of the hip.

Rates of complication vary from 32% to 61%. Major complications including cardiopulmonary effects, infection and problems with hardware occurred in 27% in a series of 30 patients reported by Ramirez et al. The documented rate of major complications in our series was 18%, including one peri-operative death and three cases of failure of hardware one of which was associated with obvious progression of the curve. Functional differences in outcome between fusion levels should be a focus in future studies.

Although surgery was performed at a more advanced stage of curve than would be advocated today, our review suggests that the pedicle screw Isola system appears to be marginally better at correcting and maintaining curves than the unit rod system, but is four times more expensive. Fusion to L3/L4 gives inadequate correction. It should be recognised that some pelvic obliquity is acceptable, possibly up to 30°, provided the upper trunk is balanced and the seating is adapted appropriately (Fig. 4). The HML regression analysis technique is useful in assessing the radiographic progression of spinal deformity before and after surgery.

With the use of respiratory support systems and steroid supplements the life expectancy of patients with DMD is likely to increase into the late twenties and early thirties and long-term maintenance of correction of spinal and pelvic deformity will become more crucial. 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


