SPINAL STABILISATION IN DUCHENNE MUSCULAR DYSTROPHY

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Of 55 patients with Duchenne muscular dystrophy offered surgical stabilisation of the spine, 32 accepted and 23 refused. We compared both groups pre-operatively and at six-month intervals in respect of survival, forced vital capacity, peak expiratory flow rate and severity of scoliosis. In the nonoperated patients, the forced vital capacity deteriorated by a mean of 8% per annum; in the operated group it remained static for 36 months and diminished slightly thereafter. Spinal stabilisation resulted in an improvement in the peak expiratory flow rate which was maintained for up to five years. In the nonoperated patients the scoliosis progressed from a mean of 37° to a mean of 89° at five years; in the stabilised spines it was improved from a mean of 47° to a mean 34° at five years. There was significantly improved survival in the patients who had undergone spinal stabilisation.

Over 90% of patients with Duchenne muscular dystrophy develop a scoliosis when they become wheelchair-bound at an average age of 9.5 years (Galasko 1977; Gardner-Medwin 1980; Rennie et al 1982). Once in a wheelchair, the scoliosis progresses relentlessly, except in those few patients who develop a hyperextension contracture of the spine (Gibson and Wilkins 1975; Galasko 1987) (Fig. 1).

As the curve progresses, the entire spine collapses to a ‘C-shape’. This is associated with progressive pelvic obliquity which at first makes sitting uncomfortable and eventually results in loss of sitting balance (Figs 2 and 3). Hsu (1983) reported that progression of the curve beyond 40° was associated with diminished sitting tolerance.

Attempts have been made to control the scoliosis by modifications of the wheelchair and the use of spinal orthoses. Although they may slow its progression these measures do not prevent the patient from developing a severe curve (Galasko 1987). Patients do not like orthoses (Robin 1975) as they lack the muscle power to pull away from any painful pressure area. Hsu (1983) reported that orthoses were capable of slowing the progression of curves of less than 25° but had little or no effect on more severe curves. Miller and O’Connor (1985) found that orthoses diminished the already limited lung function in these patients; their forced vital capacity (FVC) fell by an average of 22% (Noble-Jamieson et al 1986). Colbert and Craig (1987) concluded that spinal bracing only slowed progression of the curve and did not ultimately prevent severe scoliosis.

There has been much debate about the role of spinal surgery. Siegel (1978) reported that spinal operations were not well tolerated by these patients, whereas Sakai et al (1977) and Gibson et al (1978) found that spinal fusion, performed early in the disease, gave the patients better sitting balance for the remainder of their lives. Luque (1982) developed a method of segmental spinal stabilisation with sublaminar wiring which seemed particularly appropriate for patients with neuromuscular disease since no postoperative cast or orthosis was required. Rideau et al (1984) found no change in the FVC 24 months after spinal fusion, whereas Swank, Brown and Perry (1982) recorded a reduction after operation. It is not known whether spinal fusion has an effect on life expectancy although Smith et al (1987) thought that it did not.

PATIENTS AND METHODS

In 1990, there were 157 patients with Duchenne muscular dystrophy attending the muscle clinic at the Royal Manchester Children’s Hospital. In every patient postero-anterior erect radiography of the spine and lung-function evaluation were performed routinely every six
months. Spinal surgery was offered if a scoliosis of 20° or more had developed. Patients with Duchenne muscular dystrophy are a considerable operative risk and much time was spent in discussing the potential advantages of surgery and the possible complications. If the patient and his family accepted surgery, the patient was admitted to hospital for repeat lung-function tests, blood-gas analysis and cardiac assessment, which included echocardiography. The final assessment of fitness for operation was made by the anaesthetist.

A total of 58 patients who had developed a scoliosis of 20° or more was offered spinal stabilisation. Three were later found to be unfit for surgery: one had an FVC of less than 20%, one had symptomatic cardiomyopathy, and the third had both a low vital capacity and cardiomyopathy. Twenty-three refused surgery. Of the 32 who underwent spinal stabilisation five were operated on too recently for the results to be included. The mean age of the operated group at the time of surgery was 14 years; of the nonoperated group, 13.6 years.

Because of the benefits of spinal stabilisation on sitting balance, we thought it unethical to randomise the study. We have, therefore, compared the operated group with those who had refused surgery.

**SURGICAL TECHNIQUE**

Spinal stabilisation was carried out using a modified segmental sublaminal wiring technique with facet joint fusion at each level. Sufficient bone was harvested from the spinous processes for grafting and no additional iliac crest or homologous bone was required. In the first nine patients two rods were used which were moulded and fixed into the ilium (Fig. 4). As one of these rods broke where it had been bent to fit into the pelvis (in a case of spinal muscle atrophy, not included in this series), in the next 15 patients the 'L' rods were fixed distally to the sacrum (Fig. 5). A further complication of migration of the rod in two cases (one in this series), necessitated the use of a single 'U'-shaped rod in the remaining patients (Fig. 6). It was trimmed to length and applied to the spine, after suitable moulding, with the closed end fixed to the sacrum.

The rods were fixed from D3 or D4, to the pelvis or sacrum, with wires around both laminae at every level. Both a lumbar lordosis and a dorsal kyphosis were moulded into the rods. Some correction of the scoliosis was obtained when the patient was placed prone on the table and further correction was achieved during the wiring. The operations were carried out with spinal-cord monitoring.

Postoperatively, the patients were kept in the intensive care unit for 24 to 72 hours. They were mobilised, sitting in a wheelchair, by the fifth to sixth day and the time spent sitting was gradually increased thereafter. They were usually discharged from hospital on the fourteenth postoperative day.

The FVC, peak expiratory flow rate and spinal curvature were measured pre-operatively and at six-month intervals and compared with similar measurements in the nonoperated group. The results were calculated by repeated measured analysis using GLIM.
The survival data were analysed using a Log Rank programme with chi-square analysis.

RESULTS

There were no peri-operative deaths and all patients were mobilised within seven days. One patient required a temporary tracheostomy and one had a postoperative bronchoscopy. Another developed a mild chest infection which responded to physiotherapy and antibiotics. One patient developed a 2:1 heart block. There was one case of superficial wound infection which settled with antibiotics. There were no pseudarthroses and no postoperative failures of instrumentation. During tightening one wire broke and another cut through a D4 lamina. There were no neurological sequelae. Two patients experienced temporary pain in a lower limb postoperatively which
disappeared in two to three weeks. The measurements of FVC are shown in Figure 7. The FVC remained static in the spinal stabilisation group during the first 36 months following surgery and then fell slightly. In the patients who had refused surgery it diminished progressively. The difference is significant ($p < 0.0002$). The yearly decrease in the FVC in the nonoperated patients was 8%.

The results for the peak expiratory flow rate are shown in Figure 8. It increased significantly ($p < 0.02$) in the patients who had undergone spinal stabilisation, but remained the same in those who had refused surgery.

The measurements of the spinal curves are shown in Figure 9. Initially, the patients who had undergone surgery had more severe curves (mean 47°), than did the nonoperated patients (mean 37°), but this difference was not significant. The mean postoperative correction of the curve was to 30°, deteriorating to 34° during the first 36 months following surgery. In the group who had refused surgery the curves progressed during the first 36 months to a mean of 93°. Using repeated measure analysis of variance it was found that there was a highly significant increase in scoliosis with time in those who had refused surgery ($p < 0.001$).

The survival data for the two groups of patients (Fig. 10) showed a significantly higher mortality in those who had refused surgery ($p = 0.0029$). Of the 23 patients in the nonoperated group, 17 have died, compared with 12 of the 32 in the surgically stabilised group.

**DISCUSSION**

Our results show that, providing the pre-operative surgical and anaesthetic assessment is carefully done, extensive spinal stabilisation can be carried out safely in patients with Duchenne muscular dystrophy. However, the assessment needs to be stringent: a survey of members of the British Scoliosis Society showed a reported perioperative mortality rate of 6% in such patients (Galasko 1988).

The results confirm that progressive scoliosis is associated with deterioration of lung function, as measured by the FVC and the peak expiratory flow rate (Kurz et al 1983), and that spinal stabilisation can prevent progressive scoliosis for at least five years. The most important result in our study, however, is that spinal stabilisation can improve survival for several years following operation. Our results suggest that it would be unethical to randomise any future studies on spinal stabilisation in this disease.

Sakai et al (1977) suggested that muscular dystrophy patients should have elective pre-operative tracheostomy, but only one of our patients required a temporary tracheostomy postoperatively, and one needed bronchoscopy.

Some patients had very mobile scolioses so that the spine could be readily straightened at surgery, while others, with more severe curves, had a rigid deformity which allowed little correction. In theory, those with rigid severe scoliosis should have undergone anterior release followed by posterior correction and stabilisation, but patients with Duchenne muscular dystrophy are not fit for such an extensive procedure.

The implant should be sufficiently rigid so that no postoperative orthosis or plaster jacket is required. In our series, one rod migrated and required trimming, but there have been no failures of stabilisation. Careful excision of the facet joints and bone grafting probably contributed to this good result. Spinal-cord monitoring was used during all the operations since, although the patients had severe motor weakness and were wheelchair-bound, peripheral sensation was intact and they all had normal bowel and bladder control.

There is some debate as to when stabilisation of the spine should be advised in these patients. Our policy is to propose surgery as soon as the scoliosis measures 20°. Both Rideau, Jankowski and Grellet (1981) and Sussman (1985) suggested that the curve should be no greater than 40° to 50°. It is essential, however, that surgery be offered when the patient still has adequate respiratory and cardiac function to allow him to undergo the procedure safely.

REFERENCES


