Spinal fusion and instrumentation for paediatric neuromuscular scoliosis: retrospective review

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ABSTRACT

Purpose. A retrospective study was conducted to review the surgical results among 24 patients with neuromuscular scoliosis, who were treated with spinal instrumentation and fusion at the Department of Orthopaedic Surgery, National University Hospital, Singapore between March 1993 and December 1998.

Methods. We examined complete hospital records of patients who had scoliosis due to aetiologies such as spinal muscular atrophy, cerebral palsy, Duchenne muscular dystrophy, and congenital myopathies. The mean age of patients was 10.6 years (range, 6–14 years) and the mean follow-up duration was 5.5 years.

Results. 18 patients had posterior surgery alone, whereas 4 had an anterior release with posterior instrumentation, and 2 had an anterior fusion with instrumentation. The mean length of stay in the intensive care unit was 2 days and the mean duration of hospital stay was 11 days. The mean correction in scoliosis angle ranged from 75.6° to 25.7°. All patients could at least sit without support postoperatively. The one-second forced expiratory volume and forced vital capacity were, in general, maintained throughout the follow-up. There were 2 major complications and 2 minor ones; these were pseudarthrosis with rod breakage requiring revision, deep infection necessitating hardware removal, superficial infection that responded to antibiotics, and urinary tract infection requiring 3 weeks of antibiotic treatment. There were no deaths or any neurological complications after instrumentation.

Conclusion. Spinal stabilisation and fusion in children with neuromuscular scoliosis is a safe and effective treatment modality. The effect of surgery on long-term pulmonary function, however, remains controversial and needs to be addressed.

Key words: neuromuscular scoliosis; spinal fusion

INTRODUCTION

Patients who have a neuromuscular disorder have a high risk of developing a serious and devastating spinal deformity.1–4 The problems among this group are more complex than those among patients with idiopathic scoliosis. The spinal deformities are difficult to control with a braces and may progress even after
skeletal maturity is achieved.\textsuperscript{5–7} Patients may also have pelvic obliquity, hip dislocation, limited balance and ability to sit, back pain, and, in some instances, a significant decrease in pulmonary function.\textsuperscript{6,8–13} Treatment thus presents a great challenge to the orthopaedic surgeon. In this study, we reviewed our experience with spinal instrumentation and fusion in the treatment of these neuromuscular deformities. We assessed the safety and efficacy of the procedure and compared our results with those in the literature.

\section*{MATERIALS AND METHODS}

Between March 1993 and December 1998, 30 patients with neuromuscular deformity underwent spinal instrumentation with fusion at the paediatric and spine centre of the Department of Orthopaedic Surgery, National University Hospital, Singapore. We examined complete medical and radiographic records of 24 patients (11 boys and 13 girls), whose minimum follow-up duration was 2 years. The mean age at surgery of these 24 patients was 10.6 years (range, 6–14 years).

Seven patients had spinal muscular atrophy (5 thoracic and 2 thoracolumbar, respectively), 6 had spastic cerebral palsy (4 thoracolumbar and 2 long curves, respectively), 5 had Duchenne muscular dystrophy (3 thoracic and 2 thoracolumbar, respectively), and 3 had congenital myopathies (thoracic). Furthermore, there were 2 cases of spina bifida (thoracolumbar) and one of paraspinal neuroblastoma (lumbar).

Preoperative ambulatory status was evaluated according to a modified Rancho Los Amigos Classification system.\textsuperscript{14} 15 patients were able to sit with support and 4 without support; 5 were ambulatory (4 with and one without support, respectively). None of these patients was confined to bed. Some had had a trial of bracing prior to surgery. All patients underwent detailed pulmonary function testing before and after surgery.

Scoliosis, decompensation of the torso, and sagittal alignment were measured on erect X-rays; for sitting patients who required support, X-rays were taken with patients’ hands placed at their side. For those with cerebral palsy who were unable to sit, supine X-rays were taken. X-rays were taken preoperatively, immediately following surgery, and at later follow-up. Traction and bending films were obtained in most cases to look at the flexibility of the spine and for preoperative planning. Spinal curves were measured using the Cobb method; decompensation of the torso was measured using the method of Osebold et al.\textsuperscript{15}

Preoperatively the mean major spinal curve, measured on anterior-posterior erect film of the whole spine, was 75.6° (range, 25.0°–103.0°). In 6 patients, we performed anterior release, using anterior instrumentation in 2 patients; the other 4 had posterior instrumentation and fusion at the same stage, while they were in the prone position. The indication for anterior release was a rigid curve in cerebral palsy, which was defined as a residual curve of 40.0° or more on bending films. 18 patients received only posterior instrumentation (16 had multisegmental fixation with wires and 2 with Harrington fusion). 14 of these underwent a fusion to the sacrum using the Galveston technique. All vertebrae in the curve were fused in neuromuscular scoliosis to correct pelvic obliquity, and avoid the adding-on phenomenon in the future. Facet fusion was performed bilaterally in all patients. Grafts of local bone, rib, and iliac crest were used for the arthrodesis; no allograft was used. The mean blood loss during surgery was 990 ml (range, 120–2000 ml)—a cell saver was used for most patients in the later period of our series. The mean transfusion requirement was 3.5 units, and the mean time taken for surgery was 275 minutes (range, 145–540 minutes).

No external immobilisation was used postoperatively, except in one patient after revision surgery had been performed for a broken rod. The mean postoperative duration of stay in the intensive care unit was 2 days (range, 1–5 days), the mean duration of hospital stay was 11 days (range, 7–16 days), and the mean follow-up was 5.5 years (range, 2–8 years).

Pulmonary function tests including the one-second forced expiratory volume (FEV\textsubscript{1}) and forced vital capacity (FVC) were performed preoperatively and postoperatively, as well as at follow-up. The tests were mainly performed for patients with spinal muscular atrophy and Duchenne muscular dystrophy; patients with cerebral palsy were deemed uncooperative.

\section*{RESULTS}

\subsection*{Degree of scoliosis}

The scoliosis improved from a preoperative mean of 75.6° to a postoperative mean of 25.7°, which corresponded to a 65% correction (Figs. 1 and 2). This is comparable to the extent of correction achieved in most other series.\textsuperscript{16–18}

In 3 patients, there was progression of their curves after surgery. One of these patients, who had Duchenne muscular dystrophy, had a pseudarthrosis and broke his rod. His curve deteriorated from 15° in the
immediate postoperative period to $30^\circ$ after revision surgery. The second patient had a deep infection that required hardware removal; she had a loss of correction from $40^\circ$ immediately after surgery to $100^\circ$ after implant removal, at latest follow-up. The third was a 6-year-old girl with a congenital myopathy, who had a $65^\circ$ thoracic curve, which was corrected to $40^\circ$ with a single Harrington rod fixation on the concave side. As a result of pseudarthrosis, her curve progressed to $85^\circ$ at the 3-year follow-up visit, and it required further revision. None of the other patients had significant ($>5^\circ$) loss of correction during follow-up.

**Functional ability**

According to the modified Rancho Los Amigos Hospital system for classifying functional ability, patients are classified as follows:

**Figure 1** (a) A 7-year-old boy with spinal muscular atrophy and poor sitting posture before surgery; (b) sitting radiograph of the patient, showing correction in Cobb angle immediately after surgery.

**Figure 2** (a) Follow-up radiograph, and (b) postoperative clinical photograph after 8 years, showing maintenance of curve correction and good seating ability.
(1) Class 1 refers to patients who are able to walk about in the community without aids;
(2) Class 2, to patients who are able to walk in the community but require aids;
(3) Class 3, to patients who do not walk but are capable of independent sitting;
(4) Class 4, to patients who are dependent sitters; and
(5) Class 5, to patients who are essentially confined to bed.

In our series, the functional status improved postoperatively in 15 (63%) of the 24 patients. All patients could at least sit with hands free after surgery (Table). 19 patients were able to sit without support and 5 were ambulatory: 4 with aid and one without. The most notable improvement was seen in the dependent sitters. All of them could at least sit independently and use their upper limbs for various activities such as feeding, transfer from place to place,

<table>
<thead>
<tr>
<th>Functional status</th>
<th>Preoperative (n)</th>
<th>Postoperative (n)</th>
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<tbody>
<tr>
<td>Class 1 (independently ambulating)</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Class 2 (ambulatory with support)</td>
<td>4</td>
<td>4</td>
</tr>
<tr>
<td>Class 3 (sitting without support)</td>
<td>4</td>
<td>19</td>
</tr>
<tr>
<td>Class 4 (sitting with support)</td>
<td>15</td>
<td>0</td>
</tr>
<tr>
<td>Class 5 (confined to bed)</td>
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**Figure 3**  (a) Preoperative and (b) postoperative photographs of a 12-year-old girl with spinal muscular atrophy, showing ability to sit without hand support.
and helping in their own care (Fig. 3). The personnel who helped in their management reported that the time needed to care for them was dramatically reduced.

**Respiratory status**

Analysis of preoperative and postoperative pulmonary function showed that the number of episodes of respiratory infections in these children reduced after surgery. There was no significant improvement in the FEV₁ and FVC values postoperatively (FVC% of predicted value preoperatively and postoperatively were 57.3% and 62.7%, and FEV₁% were 58.0% and 60.1%, respectively). Although there was a little increase in these values, they remained almost the same. The findings thus indicate that there was no actual improvement in the respiratory status postoperatively, but the inevitable deterioration of respiratory function with age in patients with spinal muscular atrophy and Duchenne muscular dystrophy seemed to be retarded as a result of the surgical procedure.

**Complications**

There were no deaths in the immediate postoperative period. No neurological complications secondary to the instrumentation were encountered. There were only 4 complications, yielding a complication rate of 17%.

**Infection**

Three of the patients developed infection: 2 had wound infections (one major and one minor, respectively) and one had a minor complication of urinary tract infection. Of the 2 patients with wound infections, one had a superficial stitch abscess, which responded to local treatment and antibiotics. The other had a deep wound infection; this was an 8-year-old boy with spina bifida and a 90° kyphoscoliosis. He had undergone anterior and posterior surgery, but polymicrobial deep infection developed, which was treated with wound debridement, removal of the implants, and prolonged (6-week) antibiotic therapy. Correction was lost after implant removal, namely, from the immediate postoperative angle of 40° to 100° at the last follow-up. The patient with urinary tract infection was found to be infected with *Staphylococcus aureus*, which responded to 3 weeks of therapy with amoxicillin; the patient did well subsequently.

**Hardware complications**

In a patient with Duchenne muscular dystrophy, the rod broke at 30 months postoperatively. He had a pseudarthrosis at the dorsolumbar junction where the

![Figure 4](image-url) **Figure 4** Radiographs of an 11-year-old boy with Duchenne muscular dystrophy (a) immediately after surgery, (b) with breakage of rods at 30 months, and (c) at 5-year follow-up, showing that pedicular instrumentation was only one level above the pseudarthrosis because of previous rod placement; however, this had been mechanically adequate at 5 years' follow-up.
rod eventually broke; his curve progressed from 15° immediately after surgery to 30° at the time of rod breakage (Fig. 4a and b). This patient, with this major complication, required revision surgery in which pedicular screws were fixed, along with revision of the broken rod and bone grafting of the area. The patient responded well after the surgery and showed a good fusion mass with no further loss of correction at the latest follow-up of 5 years after the revision surgery (Fig. 4c).

Not a single death was encountered in the postoperative period in any of the patients. There were no significant pulmonary complications postoperatively, and we did not encounter any neurological complications secondary to the procedure.

DISCUSSION

The surgical management of paediatric neuromuscular scoliosis has often, in the past, been looked at with trepidation because of potentially lethal complications. But now there is little doubt that surgical stabilisation and fusion definitely benefit these patients. Surgical procedures, however, are fraught with difficulties and are not free of complications, the major one being infection. With the introduction of segmental spinal instrumentation and improved postoperative care facilities, surgery is now considerably safer, and results are better and far more consistent.

After the pioneering work of Luque, in 1975, segmental instrumentation systems have been universally accepted as providing better results than the Harrington system. Various studies have reported the correction of scoliosis in patients with idiopathic and neuromuscular scoliosis, with angle improvements of 52% to 72%. Allen and Ferguson designed the Galveston technique to improve fixation to the pelvis and reported good results, with a scoliosis angle correction of 64% for staged scoliosis surgery and intermediate traction. We achieved an angle correction of 65%, which is comparable to the Allen and Ferguson study. However, it should be noted that long collapsing curves in neuromuscular scoliosis are often severe yet flexible.

There have been various reports for single-staged scoliosis surgery, with the focus shifting to single-day anterior and posterior surgery as opposed to 2 separate procedures. Some advantages cited for the one-step procedure are less blood loss, less anaesthesia and operating time, and fewer nutritional problems. Accordingly, all procedures in our series (anterior release with posterior fusion and instrumentation) were performed on the same day. The medical complications we encountered were comparable to those in the literature, the recovery time was shorter, and there would be no significant increase in the morbidity and mortality as compared with the 2-staged corrections, although the procedure is mentally and physically demanding for the surgeon.

Pulmonary function after surgery has been a controversial issue. Most studies were only applicable to patients with Duchenne muscular dystrophy. Kennedy et al. retrospectively studied patients with Duchenne muscular dystrophy who had surgical stabilisation, and compared them with patients who did not have surgery. The researchers concluded that there was no significant difference in the rate of deterioration of FVC% among scoliotic patients with Duchenne dystrophy, whether or not they had undergone the fusion procedure. There are, nevertheless, a few reports that surgery actually slows down the deterioration of pulmonary function. Kurz et al. in their retrospective and longitudinal study of patients with Duchenne muscular dystrophy, demonstrated a slower rate of decline of FVC% in operated patients, and hence recommended early spinal instrumentation and fusion for these patients. Galasko et al. analysed 2 groups of scoliotic patients with Duchenne muscular dystrophy, and found that the FVC deteriorated by 8% in the non-operated group while in the operated group, it remained static for 36 months and thereafter diminished slightly; surgical stabilisation also resulted in an improvement in the FEV1, and there was significantly improved survival in patients who had undergone spinal stabilisation. We were able to analyse the postoperative pulmonary function in 12 patients with Duchenne muscular dystrophy and spinal muscular atrophy. During a mean follow-up period of 3 years, there was no deterioration of their pulmonary function. But the sample number was too small for the finding to be statistically significant.

The beneficial effects of the surgical stabilisation are less controversial. It is universally agreed that surgery definitely improves the functional status of these patients. Although the overall level of function depends on the natural course of the underlying condition, surgical stabilisation of the spine provides numerous benefits. These benefits include freeing of the hands from the task of trunk support for other beneficial activities; improvement of pelvic obliquity
(we could not provide an objective assessment, because pelvic obliquity could not be clearly documented in our series of radiographs), hip subluxation in cases of cerebral palsy, and thus sitting balance; easier transfer of the child from bed, thereby reducing the burden on the care provider; elimination of pain arising from the spine, and may even retardation of the progressive deterioration of pulmonary function. All these benefits help improve the self-esteem of the disabled child.

In our series about two-thirds of patients demonstrated improvement in the functional status, mainly among those who needed to use their hands for support while sitting (class 3 of the modified Rancho Los Amigos Hospital classification). This is an excellent result for these children. Our overall complication rate of 17% compares favourably with that of most studies. Literature in the late 1980s and early 1990s often quoted an overall complication rate of 44% to 62%.16 Benson et al.16 in 1998 reported in their case series of 20 complications (17 minor and 3 major) in 14 (of 50) their series with neuromuscular scoliosis. The 3 major complications were deep infections in patients with meningomyelocele; there were also 2 rod breakages, but the researchers mentioned that none of the complications affected the outcomes. In our series, we had one deep infection that necessitated hardware removal, which resulted in curve progression and an unsatisfactory outcome.

In a recent review of deep wound infection in operated patients, Sponseller et al.21 reported deep wound infections in 25 (12%) of 210 patients studied. They analysed possible risk factors and concluded that the only 2 statistically significant risk factors were the degree of cognitive impairment and the use of allograft. Moreover, they also found that Gram-positive infections were more amenable to debridement and closure as compared with Gram-negative and polymicrobial infections, which often require hardware removal, as was observed in a patient in our series.

The rate of pseudarthrosis has significantly decreased following the use of anterior as well as posterior fusion techniques. We find it difficult to make a diagnosis of a pseudarthrosis based on plain X-rays alone. We also believe that there is no role for observation in asymptomatic patients with hardware failure; this was illustrated in the patient whose curve progressed after the rod breakage.

The short stay in the intensive care unit and the absence of any significant pulmonary complications in the postoperative period are indicative of the high level of perioperative care, which is provided by a modern setup with an experienced team.

**CONCLUSION**

The data from this study support our belief that spinal stabilisation and fusion in children with neuromuscular scoliosis is a safe and effective treatment modality. In experienced hands, the risk of complications, both intra-operative and postoperative, is low. Surgery provides significant functional benefit to these children, but the issue of the effect of surgery on long-term pulmonary function remains controversial and needs to be addressed.

**REFERENCES**


