Surgical management of neuromuscular scoliosis: a review of 24 cases

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Abstract
Aim: To review the surgical management of neuromuscular scoliosis within the Division of Orthopaedic Surgery, University of Cape Town.

Method: The spinal surgical database was searched for neuromuscular scoliosis cases managed operatively. Their case notes and X-rays were reviewed with respect to procedures employed, complications and radiographic results.

The patients and families were then contacted and asked whether they would undergo the surgery again and whether seating had improved.

Results: Over a three year period, 24 patients underwent surgery by a single surgeon for neuromuscular scoliosis.

The case mix was post-spinal cord injury with paraplegia and subsequent scoliosis in nine, meningomyelocoele in seven, cerebral palsy in three, polio in two, other in three.

The average age at surgery was 13.6 (8.5 - 22.3 ± 3.2) years. There were 13 females and 11 males.

Procedures employed were anterior only in two, posterior only in 18 and combined in four. Fixation to the pelvis was done in 20 cases.

The average pre-operative Cobb angle was 80.0° (30 - 129 ± 27.8), with an average postoperative Cobb of 27.7° (5 - 80 ± 22.5).

Technical complications included two cephalad failures of instrumentation, viz. one screw pullout and one screw-cap loosening. There was one asymptomatic iliac crest screw loosening.

There was one CSF leak due to a medial thoracic pedicle breach in a meningomyelocoele with spontaneous resolution.

One wound dehiscence required debridement and vacuum dressings.

One case developed postoperative neurological deterioration requiring urgent instrumentation removal with full recovery.

Sitting ability improved in all. All but one parent and two patients would have the surgery, knowing what they know now.

Conclusion: Surgical management of neuromuscular scoliosis is rewarding in terms of radiological correction, improved seating, and cessation of brace wear. The meningomyelocoele group continue to provide the major challenge with high incidence of complications.
Neuromuscular disorders are a diverse group of conditions that have a propensity to develop scoliosis. Although muscle weakness and loss of flexibility is often given as the cause, it is more likely due to altered spinal muscle tone and balance control.

As opposed to idiopathic scoliosis, neuromuscular scoliosis is difficult to control with bracing. The curve often extends into the pelvis with obliquity and continues to progress beyond maturity. These patients often have discomfort from large curves, seating difficulties and resultant skin pressure problems. Many are forced to support themselves with their upper limbs during sitting, with further loss of function.

The underlying condition makes their management challenging due to respiratory compromise and insensate skin.

The aim of this study was to review the surgical management of neuromuscular scoliosis in our service.

Material and methods

Over a three-and-a-half year period (October 2001 to April 2005), 24 consecutive patients underwent surgery by a single surgeon for neuromuscular scoliosis. These cases were operated on at Groote Schuur, Red Cross and UCT Private Academic Hospitals. Notes and X-rays were reviewed with respect to procedures employed, complications and radiographic results. The patients and parents were questioned as to whether they would undergo the surgery again and whether seating had improved.

The underlying diagnosis is shown in Table I.

<table>
<thead>
<tr>
<th>Table I: Underlying neuromuscular condition</th>
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<tr>
<td>Cerebral palsy (CP)</td>
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<td>Post spinal cord injury (SCI)</td>
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<tr>
<td>Meningomyelocoele (MM)</td>
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<tr>
<td>Polio</td>
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<tr>
<td>Duchenne’s muscular dystrophy</td>
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<td>Spinal muscular atrophy</td>
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Twenty patients underwent single stage procedures, and four two-stage procedures. The indications for two stages were severe rigid curves or concerns regarding crankshaft phenomena.

The first stage was an anterior release with an average of seven discectomies and an average operative duration of 132 (105 - 150 ± 20.2) minutes with an average of 262 (100 - 600 ± 228.6) ml estimated blood loss.

In addition, 20 underwent a posterior high thoracic to pelvic fusion, two a shorter posterior thoracolumbar fusion and two an anterior instrumented fusion.

The long posterior fusions were done with segmental fixation utilising a combination of pedicle screws and monofilament wires, all titanium. Fixation to the iliac crest was achieved with placement of a long pedicle screw from the posterior crest just superior to the sciatic notch. This tricortical trough provides good purchase. To assist with placement, the lateral crest was exposed subperiosteally to visualise the notch, and if too small to place a digit, a McDonald or Watson Cheyne utilised to palpate the notch and plan the screw insertion pathway. In most cases a 7 mm x 60 mm screw was used, but in the smaller children this was reduced to appropriate sizing.

The average operative time (skin to skin) for these long constructs was 255 (200 - 345 ± 34.9) minutes, with an average estimated blood loss of 1 955 (700 - 4 200 ± 1 036.7) ml.

A cell-saver was employed in nine cases. This was later fused, which represented 64% of the patient's total blood loss. An average of 504 (200 - 1 065 ± 292.1) ml of packed cells were reinfused, which represented 38% of that collected or 25% of the total blood loss.

Results

Overall the average pre-operative scoliosis as measured by the Cobb angle was 80.0° (30 - 129 ± 27.8). The average postoperative Cobb angle was 27.7° (5 - 80 ± 22.5). This represented an average correction of 68% (18.6 - 93.9 ± 19.4).

Pelvic obliquity was corrected by more than 50% in all but two cases.

At follow-up, all patients were assessed by the clinician and parents to have improved seating.

The parents and patients were asked “Knowing what you know now, would you have had the operation again?” Of the 24 parents consulted, 23 said yes and one no. The negative response was due to the magnitude of the surgery and time in hospital. The rest felt there was tremendous benefit in the seating posture and the cosmesis. Of the patients, 20 said yes, two no and two were unable to comprehend. The two “no’s” were both meningomyeloceles (MM), and one commented that the reason was the length of time away from home. The positive patient responses echoed their parents’.
The complications are summarised in Table II.

There were two postoperative surgical wound infections. One was early in the immediate postoperative period and one late, related to morbidity associated with the condition rather than the scoliosis surgery. The early postoperative sepsis was in an obese MM with a severe posterior scar. This presented initially as a clear fluid leak, thought to be CSF. However the Beta-2 transferrin was negative, and on surgical exploration and wash out, MRSA was cultured. She was successfully managed with vacuum dressings and appropriate antibiotics.

The late sepsis occurred at 6 months post-scoliosis correction in a post-spinal cord injury adolescent following superior rod trimming. He had recently had a MRSA trochanteric sore. The spinal sepsis settled on antibiotics and vacuum dressings, but recurred at one year post original scoliosis surgery. At this point his entire scoliosis construct was removed, as he was fully united.

One patient had radiological iliac screw lucency, despite no clinical symptoms. This case was a severe scoliotic meningomyelocoele. He was an early case in the cohort, where only a posterior procedure was done. There was poor correction and probable failure to fuse the lumbosacral junction, resulting in the loose iliac crest screws. No further surgery is planned as the patient remains asymptomatic.

One hemiplegic cerebral palsy (CP) patient developed left hip pain shortly after spinal correction. She had an associated dysplastic hip. Her sagittal profile had been overcorrected in terms of lumbar lordosis, forcing the hip into relative hyperflexion in the seated position. Radiology confirmed the iliac screws not to be traversing the joint.

There was one neurological complication. A polio case with severe rigid scoliosis and some pre-existing lower limb weakness was distracted to the pre-operative traction X-ray position. Despite a normal “wake up” test, she lost motor and sensory function of the lower limbs postoperatively. She was immediately returned to theatre and the rod removed with full recovery. She was managed with a plaster jacket with a poor scoliosis correction, but remained ambulatory.

There was loss of scoliosis correction in two patients - the one discussed above and another meningomyelocoele that was managed with an anterior only instrumented fusion. The superior screw partially pulled out with a slight loss of correction.

<table>
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<th>Table II: Complications</th>
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<tr>
<td>Wound infection (1 early, 1 late) 2</td>
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<tr>
<td>Lumbar hyperlordosis (hip pain) 1</td>
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<tr>
<td>Neurological deterioration (transient) 1</td>
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<tr>
<td>Loss of scoliosis correction 2</td>
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<tr>
<td>Instrumentation failure 2</td>
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<tr>
<td>Cerebrospinal fluid leak 1</td>
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There were two instrumentation failures, the aforementioned superior screw pullout with subsequent infection and a cephalad screw cap loosening. The latter was the only case where a top-loading locally manufactured system was used due to cost considerations. A side-loading system was utilised in the other patients. There was no loss of correction in this case, but there is a slight prominence.
One case developed a CSF leak. She presented with a fluctuant swelling of the wound postoperatively with a small area of clear fluid oozing through the wound. MRI confirmed a fluid collection, but the patient remained afebrile. This was thought to be iatrogenic from a medial pedicle breach by a thoracic screw. This was managed expectantly and it resolved spontaneously.

Discussion

Scoliosis is common in neuromuscular conditions with an incidence of 25-100% depending on the aetiology.1 The natural history is that of progression, with conditions such as meningomyelocele demonstrating progression rates of 6.2° per year2 as opposed to the 0.5-1° of mature Idiopathic scoliosis. Bracing is generally ineffective, as it requires long-term use as opposed to idiopathic scoliosis where it may be utilised during rapid growth until skeletal maturity. Olafsson3 reported on the use of the pre-fabricated Boston brace in neuromuscular scoliosis. Sixty per cent correction was obtained in the brace, but this represents the curve flexibility rather than a true therapeutic effect. There was only a 23% success rate, i.e. failure to progress 10° at brace cessation. Just under half were non-compliant and 21% progressed despite brace wear. Olafsson concluded that bracing of the patient cannot have a lasting corrective effect although it can be used as sitting support.

The underlying aetiology determines life expectancy, limited with Duchenne's and approaching normal expectancy in traumatic paralysis. Tsirikos4 reviewed 288 cerebral palsy patients who had surgery at the age of 13.1 years, and calculated a mean predicted survival of 11 years postoperatively. Therefore, many of these children will live for significant periods of time, which will result in severe progression. This justifies corrective surgery.
The indications for surgery are diverse. The most pressing factor in our cohort is that of inability to sit without upper limb support. A large number of our cases were post-SCI patients with normal intellectual ability and therefore reasonable social integration prospects. They were frequently hampered by the need to support themselves with their upper limbs, with consequent further functional deficit. Other indications were poor seating due to imbalance and pelvic obliquity, which increases point loading and risk of pressure sores. In the South African environment, there are many patients competing for scarce resources and it is necessary to give priority to those with a reasonable level of intellectual function as well as those with a good social support structure.

The severity of the Cobb angle, rate of progression and impact on present and future respiratory function were also taken into account in favour of offering surgery.

Iliac fixation
All the non-ambulating patients in our study were fused to the pelvis via the iliac blade screw placement. This technique provides excellent control of the pelvis during the corrective procedure. Although peri-screw lucency was seen, it was asymptomatic, and there was no loss of correction as a result. This procedure offers advantages over the more traditional Galveston technique, in that it is technically easier to place the screws, rather than to pre-bend the rod to fit. Also with excessive manipulation of the Galveston rod during the scoliosis correction, a defect in the cancellous bone is developed thereby reducing the fixation. Sink et al. report a 12% posterior migration of the rod from the iliac crest in his cases of Galveston fixation. With the screw thread there is a better screw-bone interface and higher pull out strength. Early compared Galveston with the Colorado II modular system using sacropelvic plates in cadaver spines. He found similar correction but that the Colorado group had less flexion and extension at the lumbosacral level. King reported on the spinopelvic transiliac fixation technique with good results but commented that a well developed iliac apophysis is required, which may not be present in younger patients. We were able to place iliac blade screws in all our patients, and when small, reduced the diameter to 5 mm screws. The apophysis was nibbled to expose the cancellous bone, and the lateral cortex exposed to assist with direction and assessment of violation. The path was made with a flat bladed awl and probed to exclude a cortical breach. Frequently cortical deformation from the screw threads could be palpated laterally but no violation visualised. Marchesi used the Galveston technique with addition of S1 pedicle screws with better reported correction and maintenance thereof.

Extension to the pelvis has been challenged. McCall reports similar correction with constructs to L5, and recommends this as an easier technique with lower intra-operative blood loss. He concludes that consideration for pelvic fixation should be lumbar tilt greater than 15° or an apex of L3 or lower. Whitaker reported on 23 cases with mild pelvic obliquity (6°) and stopped at L5. He found the technique to be safe and effective, again in patients with a spino-pelvic angle of less than 15°.

There is a concern that by fusing to the sacrum/pelvis in ambulatory patients that there will be a deterioration of gait function, possibly leaving them non-ambulatory. Tsirikos reviewed 24 CP patients at 2.8 years postoperatively. Despite fusions to the sacrum with pelvic fixation, he found no alteration in their ambulatory status except one. This patient had heterotopic calcification of both hips. He had pre- and postoperative gait analysis in 12 of the patients, which showed no change in gait.

Anterior versus posterior surgery
The choice of anterior or posterior surgery is often determined by the underlying condition and the rigidity of the curve as assessed clinically and radiographically with traction and bending views. In our series, anterior releases were avoided if possible due to respiratory concerns in the post-SCI patients with poor intercostal muscle function as well as the myopathy cases. Where there were severe rigid curves such as in neglected CP patients, releases were considered. In MM patients it is useful to achieve an anterior interbody fusion due to the absent posterior elements. The concerns regarding crankshaft phenomena are probably overlayed now that strong segmental fixation is available. Smucker showed no significant post-surgical radiographic change at 2 years in 50 CP patients with open tri-radiate cartilage at the time of surgery. A subgroup of 29 had 4.6 years follow-up without deterioration. He concluded that posterior spinal fusion was adequate to prevent crankshaft. This was echoed by Westerlund with 28 immature patients, with nine having open tri-radiate cartilage and five less than 10 years old.

Chang reported on the use of cantilever correction forces as opposed to the traditional distraction manoeuvres in 41 patients with a pre-operative Cobb angle of 98° and a correction of 67%. He concluded that cantilever correction is an effective management of large and rigid curves and could avoid anterior release. This is the manoeuvre used in our cohort.

The rods are first attached to a good distal lumbar and pelvic fix, and then reduced into the cephalad screws. This leverage provides enormous power of correction. Once the fixation points are applied to the rod, further correction is done with some distraction between fixation points in the concavity.
Viviani’s compared one-stage or two-stage procedures, and reported a shorter total ICU and ward stay, lower blood loss and shorter total theatre time for one-stage procedures. This article needs to be interpreted with caution, as it reflects a longitudinal case series where the surgeon utilised one-stage procedures only after a learning curve on the two-stage procedures. In other words the learning curve was at the expense of the two-stage cohort, which skews the results. However, Ferguson et al. reported similar findings in their 29 patients. They had 36 (124%) major and minor medical complications in the staged patients, whereas 14 (88%) major or minor complication were present in same-day surgery patients.

In contrast, Tsririkos et al. reported that there was no statistically significant difference between same-day and two-stage patients when evaluating scoliosis correction, hospitalisation time, and intensive care unit stay. However same-day surgery was associated with increased intra-operative blood loss, prolonged operative time, and a considerably higher incidence of medical and technical complications, including two peri-operative deaths. He concluded that two-stage anteroposterior spinal fusion provides safer and more consistent results with several advantages over the single-stage procedure in the management of patients with cerebral palsy and neuromuscular scoliosis.

There is an argument for anterior surgery alone, especially in the meningomyelocele group where there is poor soft tissue cover and altered bony anatomy. Basobas reviewed 21 patients retrospectively, the majority being MM. He reported good correction and maintenance thereof with anterior fusion with few complications. Sarwahi reported on 111 cases of anterior only surgery with an overall complication rate of 44% (21.2% major).

Blood loss
Our average blood loss was 1 955 ml. This is large and frequently represented more than the patient’s total blood volume, but is in keeping with other reports. We did not utilise hypotensive techniques and aimed for a mean blood pressure of between 60 and 70. Edler compared neuro-muscular and non-neuromuscular scoliosis surgical blood loss in 163 cases. He found that neuromuscular patients had a seven times greater risk of a greater than 50% total blood volume loss than the other group. Kannan found that there was an average of 78% total blood volume loss in neuromuscular patients as opposed to 20% loss in idiopathic scoliosis. He also noted a higher INR and reduced factor 7 suggesting activation of the extrinsic coagulation pathway in the neuromuscular group. Murray performed a retrospective review of patients undergoing posterior spinal fusion for scoliosis assessing the success of autologous pre-donation in preventing allogeneic blood use. He found that more than 90% of autologous donors successfully avoided receiving allogeneic blood.

However, these were more frequently idiopathic scoliosis patients. Patients with neurological causes of scoliosis more commonly used allogeneic or directed donation, even if they pre-donated.

Cell saver techniques remain controversial due to cost. Despite not preventing allogeneic transfusion in our cohort, an average of 25% of the volume lost was rein infused as packed cells, which reduced the degree of allogeneic transfusion. This may have advantages in avoiding immunosuppression as caused by large allogeneic transfusions.

Complications
There is a high incidence of complications in this group of patients with reports from 44-124% depending on the sensitivity of reporting, with pulmonary complications around 30% and infection up to 14%. These are often related to the underlying condition with poor pulmonary function, insensate skin, reduced intellect and the insult of significant surgery. Yuan assessed pre-operative lung function tests and found that a FEV1 of less than 40% and neuromuscular scoliosis were predictors of requiring ventilation for longer than three days. Padman also found that patients who developed postoperative pulmonary complications had pre-operative vital capacities less than 50% of that predicted. In those who did not develop pre-operative complications the average VC was 64.6% of that predicted.

Yazici reported on 47 patients with a re-operation rate of 11% (i.e. five complications). This was for infection in two cases, one pseudoarthrosis, and two cephalad instrumentation prominence. Sponseller in a multi-centre 10-year retrospective review, reported on 210 MM and CP patients. They had 25 infections, and found cognitive function and the use of allograft to be risk factors. Eleven cases were successfully managed by debridement and closure, two were allowed to granulate by secondary intention, and seven required instrumentation removal for control of sepsis. Szoke reported an 8.7% infection rate in 172 CP patients, with half deep and half superficial. The majority occurred in the distal part of the wound, and were diagnosed within two months of surgery. All the superficial wound infections were treated successfully by local wound care and intravenous antibiotics. The removal of instrumentation was necessary in the one late deep wound infection that occurred two years after the spinal fusion. The remaining six deep infections were treated by irrigation and debridement with the wound left open, allowing it to heal by secondary intention. All their wound infections occurred in severely neurologically involved spastic quadriplegics who were non-ambulatory and severely mentally retarded and had seizure disorders.
Instrumentation problems have been reduced with the use of modular systems. Windscreen-wiper effects (lateral translation of the spine due to rotation around the fixation points) of up to 38% have been negated by rigid segmental fixation with screws as opposed to sub-laminar wiring. The incidence of rod breakage is reported at 5%. This was not experienced in our series and the improvement is attributed to the more rigid fixation and lower rate of pseudo-arthrosis which was previously reported at 10%. Sink found that the cases of both cephalad and caudal failure were associated with kyphosis and that this could be avoided with anterior release and fixation. Our only case of cephalad failure was kyphotic, but probably due to a misplaced pedicle screw.

Outcome

Frischut evaluated the morphologic and functional outcome of 41 patients with progressive neuromuscular scoliosis. Despite a mediocre radiographic correction of 50%, functional improvement could be demonstrated in 20 patients. Seventeen patients remained unchanged, whereas four patients showed deterioration. Cosmetic results were rated as excellent by 25 patients, good by seven, and poor by one. For eight patients the appearance was unimportant. Pain relief was experienced in all cases (n = 4) of the pre-operative low-back or abdominal pain.

Whitaker confirms that 87% of his cohort was very satisfied, with only 4% dissatisfied. This is similar to our experience with a high level of satisfaction in terms of improved seating and cosmesis.

Askin found that it took six months for patients to return to their pre-operative functional level and improvement was not seen after 12 months.

Conclusion

Although surgical correction of neuromuscular scoliosis is an expensive procedure with a significant complication rate, it is rewarding in terms of improved seating, cosmesis and patient/parent satisfaction. Meningomyelocoele remains the most challenging group, and anterior techniques should be considered to avoid the associated problems via the posterior route.

References

8. Frischut evaluated the morphologic and functional outcome of 41 patients with progressive neuromuscular scoliosis. Despite a mediocre radiographic correction of 50%, functional improvement could be demonstrated in 20 patients. Seventeen patients remained unchanged, whereas four patients showed deterioration. Cosmetic results were rated as excellent by 25 patients, good by seven, and poor by one. For eight patients the appearance was unimportant. Pain relief was experienced in all cases (n = 4) of the pre-operative low-back or abdominal pain.


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