Neuromuscular scoliosis — surgical management and outcomes

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Introduction

Neuromuscular scoliosis (NMS) develops in a heterogeneous group of patients with myopathic, upper and lower motor neuron diseases. Spinal surgery is often required to optimise respiratory, sitting and ambulatory function.

Objectives:
Review of management and outcomes of surgically treated neuromuscular scoliosis.

Study design:
Retrospective review of prospectively maintained data, including demographics, intra-operative variables, pre- and post-operative imaging, complications, outcomes and a telephonic follow-up questionnaire.

Results:
Ninety-eight patients (45 male and 53 female) were included in the study. The average operating time was 230 (100–525 ± 60.9) minutes and an average of 15.4 (8–19 ± 2.9) levels were fused. Pedicle screw only constructs corrected the primary curve by 63% initially and 56% correction at last follow-up. Hybrid constructs had an immediate correction of 69% and 47% at last follow-up. Although pedicle screw constructs lost less correction when compared to hybrid constructs, this was not a statistically significant difference. Pelvic obliquity was corrected from 14.02 (0–80 ± 15.54) to 4.06 (0–35 ± 7.69) degrees. The majority of the telephonic responses were positive.

Conclusion:
Corrective spinal surgery in the neuromuscular patient is demanding with a high rate of complications but outcomes are good, with radiographic correction maintained in the long term and high level of patient and parent satisfaction.

Level of evidence:
III

Key words: neuromuscular scoliosis, surgery, management, outcomes

Abstract

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NMS is classified into neuropathic and myopathic groups according to its underlying cause. The neuropathic types are subdivided into upper motor neuron lesions (UMN) and lower motor neuron lesions (LMN). Common UMN causes are cerebral palsy (CP), spinal muscular atrophy and Friederich’s ataxia whereas myelomeningocele and polio are LMN causes. Duchenne muscular dystrophy and Marfan’s syndrome are examples of the myopathic type.

Management of NMS patients requires a multi-disciplinary approach with active involvement of parents, caregivers, physiotherapists, occupational therapists, paediatricians and intensive care physicians. Patients with moderate to severe curves are managed non-operatively if they are too ill to undergo major corrective spinal surgery. Thoracolumbar orthoses are poorly tolerated and restrict the already impaired respiratory function. Custom-moulded wheelchairs are of some benefit to these ill patients.

Surgery is indicated to correct sitting imbalance due to severe deformity as well lift the chest off the abdominal contents as the patient’s trunk collapses. Surgery in this instance may salvage the deteriorating respiratory system and can be life-saving. Discomfort from impingement of the rib cage on the iliac spine causes great morbidity and is relieved by deformity-correcting surgery. Correcting these patients’ coronal balance may improve upper limb function by reducing the need for arm support to prevent slouching/falling over.

Surgical fixation techniques have advanced from previous distracting techniques with Harrington rods and sub-laminar wire fixation of Luque, which had poor rotational control. Modern segmental, pedicle screw-based instrumentation techniques have allowed spinal surgeons to not only halt deformity progression, but dramatically correct it in sagittal, coronal and axial planes.

Materials and methods

Following University of Cape Town Institutional Research Council approval (no.228/2012), a retrospective review of prospectively collected data was undertaken.

The senior author’s (RND) database was interrogated for neuromuscular scoliosis patients undergoing spinal deformity fusion surgery. All underwent their surgery by the senior author between October 2001 and December 2011. All the procedures were performed at Groote Schuur Hospital, Red Cross War Memorial Children’s Hospital, UCT Private Academic Hospital or Constantiaberg Medical Clinic.

Demographics, intra-operative variables, pre- and post-operative imaging, complications and outcomes were analysed. Patients and caregivers/parents were contacted telephonically and asked three questions:
1. Would the patient opt to have the surgery again now that they know what it involved?
2. Would the parent/caregiver opt to have the surgery again if given the same opportunity?
3. Has seating been improved by surgery?

Data was compiled with the aid of a Microsoft Access database and analysed using a Microsoft Excel spread sheet. The Student t-test was employed to calculate p-values with 1-tail significance levels.

Results

Ninety-eight patients were identified of which 45 were male and 53 female. The median age at surgery was 15.3 (6.2-70.4 ± 9.0) years and the average age at surgery was 15.0 (6.2-70.4 ± 9.0) years. Fourteen patients were younger than 10 years at surgery, 78 were between the ages of 11 and 20 years and three between 21 and 30 years of age (Figure 1). The three oldest patients had Parkinson’s disease (99 and 70 years old) and poliomyelitis (56 years old). Their scoliosis was deemed to be due to the underlying disease and not degeneration.

Spinomuscular atrophy and cerebral palsy were the most common underlying diseases (n=18) followed by myelomeningocele (n=15) and paralytic spinal cord injury (n=10) (Figure 2). The average operating time was 230 (100–525 ± 60.9) minutes with an average of 15.4 (8–19 ± 2.9) levels fused per patient. Seventy-two patients had posterior only surgery. Fifteen had anterior only surgery. The anterior only surgery was all for focal thoracolumbar scolioses. They were attributed to neurofibromatosis (n=2), cerebral palsy (n=5) and myelomeningocele (n=7). Fifteen patients had anterior and posterior surgery. This was done on the same day in two patients, and staged in 13 patients.

When an anterior operation was performed for anterior only surgery or for combined anterior/posterior surgery, an average of 6 (2-8 ± 1.2) levels were released. The blood loss for this anterior procedure was 200 (50–850 ± 229.3) ml and duration 130 (60–180 ± 37.9) minutes.

For all the procedures combined, the average total blood loss per operation was 1 812.9 (250–8500 ± 1255) ml, which is 119.5 (25–850 ± 101.0) ml per fused level. There were 18 patients with blood loss greater than 2 500 ml. Patients who were fused to the pelvis had significantly (p=0.006) greater blood loss than those not fused to the pelvis: 2170.1 (600–8500 ± 1389.1) ml vs 1361.8 (250–3600 ± 890.5) ml.

A cell saver was used in 35 cases and 1 800 (500–8021 ± 1396.7) ml blood collected per patient. Of the collected blood volume, 650 (200–3498 ± 588.7) ml or 36% was re-infused. Blood loss when using the cell saver was 2 100 (900–8500 ± 1399.5) ml and without was 1 000 (250–4000 ± 984.9) ml (p=0.00003).

![Figure 1. Age at surgery](image-url)
Posterior spinal fusion was performed on 86 patients. Of these, 38 were with pedicle screw only constructs and 48 with hybrid constructs. A hybrid construct was defined as any construct using transverse process hooks \((n=24)\), sublaminar wires \((n=22)\) or both hooks and wires \((n=2)\) in conjunction with pedicle screws.

Metal density is defined as the ratio of fixation points utilised to those available. In pedicle screw only constructs the metal density was \(0.44 (0-0.75 \pm 0.14)\) whereas in hybrid constructs (hooks and sublaminar wires) the density was \(0.34 (0.06-0.75 \pm 0.13)\).

The most caudal level fused was the pelvis in 48 patients followed by L4 \((n=10)\), L1 \((n=7)\), L5 and L3 \((n=6\) each\) (Figure 3). The pelvis was instrumented in only two walking patients; L4 was the most common inferior level in ambulating patients \((n=8)\) (Figure 4).
Forty-six patients were fused to T2 superiorly, followed by 24 to T4 and nine to T3 (Figure 5).

Six patients died in the post-operative period at an average of 34.2 (20–51 ± 11.92) months post-operatively. All deaths were unrelated to the surgery and due to the underlying neuromuscular condition. Fifty-two patients had a follow-up of greater than two years and 22 are still in the two-year post-operative period. Eighteen were lost to follow-up after their initial post-op visit. This was usually due to patients from distant areas finding it difficult to travel.

The primary curve was corrected from 77.2 to 32.5 degrees immediately post-operative – a correction of 66% or fraction of 0.66 (0.1–1 ± 0.24). This correction was maintained for the follow-up period (Figure 6) and statistically significant throughout. At last follow-up (27.97 (1–107 ± 23.83) months) the primary curve magnitude was 37.59 (0–110 ± 25.73) degrees. The secondary curves were initially corrected from 44.2 to 25.5 degrees – a correction of 0.62 (~1–1 ± 0.42). This correction was however not maintained in the follow-up period. At 25.52 (1–72 ± 17.78) months the secondary curves measured 29.05 (4–68 ± 15.78) degrees. Only the immediate post-operative secondary curve correction was statistically significant (Figure 7).

Pedicle screw only constructs corrected the primary curve by 63% which was maintained at 56% at latest follow-up. Hybrid constructs (sublaminar wires and/or hooks) had an immediate correction of 69% and 47% at last follow-up. Hybrid constructs with only sublaminar wires corrected by 69% initially, declining to 52% at follow up (Figure 8). Although pedicle screw only constructs lost less correction (11.1%) when compared to hybrid constructs (31.1%), this was not a statistically significant difference.

Pelvic obliquity was corrected from 14.02 (0–80 ± 15.54) to 4.06 (0–35 ± 7.69) degrees. This is a correction of 71% (0–56 ± 0.51) (Figure 9). A correction of 72% was achieved when the pelvis was incorporated in the fusion mass (n=48). When the pelvis was not incorporated in the fusion mass, the pelvic obliquity correction was 67% (n=38).

Eighty-six families were telephonically contactable. Some patients/parents simply would not commit to answering positively or negatively – they were grouped under ‘unsure’ (n=66). The majority of the responses were positive (160 vs 9) and both parent/caregiver and child would opt to have the surgery again if given the opportunity (Table I). Patients who were ambulating (n=20) were excluded from the sitting follow-up question. A patient with post-operative neurological deterioration was the only patient who thought sitting balance was not improved post-operatively.

### Complications

#### Neurological

There were four patients with neurological complications. A 13-year-old boy with hydrocephalus underwent a T2–L2 posterior spinal fusion for cerebral palsy-induced scoliosis. Although he was wheelchair bound pre-operatively, he had some leg control and sensation. During the immediate post-operative period he developed cardio-respiratory instability which was confirmed to be due to ascending spinal oedema. He required inotropic support and mechanical ventilation. At day 15 post surgery he appeared as a C4 complete spinal cord injury, but slowly regained his upper limb neurological function. Unfortunately he remained a T8 complete paraplegic and never regained his lower limb motor function or sensation.

A 15-year-old girl with polio and a severe kyphoscoliosis underwent a T2–L4 posterior spinal fusion. Despite a normal intra-operative wake up test, she developed complete paraplegia a few hours after the procedure. She was taken back to theatre immediately, the instrumentation removed and the spine allowed to return to its pre-operative position. She regained motor function by the morning and returned to normal over the next few months.

A 7-year-old girl with a severe dystrophic kyphoscoliosis from neurofibromatosis with pre-operative myelopathy underwent a T2–T10 posterior spinal fusion and circumferential vertebral column resection to decompress the cord. Her myelopathy worsened post-operatively but slowly recovered in a protracted eight-month hospital stay.

Four patients had unintentional durotomies. These were typically the meningomyelocele patients with thin skin adherent to the expanded thecal sac. There were no neurological consequences.
Massive blood loss

One patient had a massive blood loss of 8,021 ml. This was partly explained by persistent hypothermia. Via the cell saver 3,498 ml was re-infused, which prevented any further consequences.

Respiratory

Pulmonary oedema developed in a patient on post-operative day 10. This was due to hypoalbuminaemia and negative intrathoracic pressure from the expanded thoracic cage. She was ventilated via tracheostomy for three days and recovered uneventfully.

Mechanical

A 59-year-old female with Parkinson’s disease developed a positive sagittal balance despite extensive anterior release (T10–L4) and L5/S1 transfarominal interbody fusion at the initial procedure. She required revision surgery with a pedicle subtraction osteotomy of L3 seven months after index surgery and is now doing very well.

Five patients had asymptomatic rod breakages or pedicle screw loosening. These were left alone. One patient had early pull-out of his superior screws. He was managed with a thoracolumbar sacral orthoses.

Infection

There were two early post-op wound infections. These were managed with repeated washouts, vacuum dressings and secondary closure under the cover of appropriate antibiotics for six weeks.

A 6-year-old arthrogrypotic patient presented 10 months post-operatively with late onset sepsis and skin breakdown over the prominent screw heads. This resolved with removal of instrumentation, debridement and six weeks of antibiotics.

Discussion

Scoliosis develops in the majority of neuromuscular patients. The mainstay of management of the scoliosis is surgical arthrodesis of the spine in a functional position. Bracing causes further deterioration of the already compromised respiratory system in the established severely scoliotic patient and is only recommended for a select few NMS patients. Corrective spinal surgery in patients with a severe physical handicap results in deterioration of function in the initial six months, but is then followed by return to pre-operative function in the next six months. Surgery has been shown to improve quality of life in patients with scoliosis related to CP and muscular dystrophy.

Timing of surgery is a contentious issue with two opposing factors to be taken into account: skeletal growth (thoracic height) and pulmonary development on the one hand and deformity progression with pulmonary restriction and compromise on the other. In normal children, the pulmonary function growth spurt lags behind the height growth spurt on average 0.6 to 0.9 years. This is independent of age and gender. Children reach 66.7% of adult T1–T12 height at age 5 and 81.5% at pre-adolescence. The first eight years of life have been termed the golden period for thoracic growth and pulmonary development. Considering all these factors, the exact age at surgery is often a subjective decision influenced by many factors including age, gender, underlying neuromuscular condition and neurological function or loss thereof.

Pre-operative pulmonary function testing is a reliable indicator of post-operative pulmonary complications and prolonged (>3 days) post-operative mechanical ventilation. However, pulmonary function testing in young children, even with normal mental capabilities, is often not reliable. Neither is poor pre-operative pulmonary function (FVC < 30%) or nocturnal non-invasive ventilation a contra-indication to surgery in the NMS patient. Because of this, we do not routinely perform pulmonary function tests as a pre-operative assessment on our NMS patients but judge surgical suitability by subjective pre-operative respiratory symptoms, respiratory effort and exertion tolerance.
The transphysiologic anterior approach allows for release of rigid, severe, short segment curves, but carries greater postoperative respiratory morbidity. A combined anterior and posterior approach is required when the anterior approach alone will not allow sufficient surgical access. Many NMS patients preclude chest violation due to underlying respiratory compromise, and the posterior approach is the mainstay. It is however, not without complications. Modi et al reported on haemo/pneumothorax, pleural effusion, pulmonary oedema, complete spinal cord injury, deep wound infection and death in a series of 50 patients. Unusual complications included coccygodinia.

Blood loss in NMS surgery is the focus of many recent articles. Most agree that NMS is an independent risk factor for increased blood loss during spinal surgery. Tranexamic acid has been shown to reduce the blood loss in posterior scoliosis surgery by up to 45% but high dosage is required to be effective (100 mg/kg before incision and 10 mg/kg/hour during surgery). The cell saver is a vital instrument in the armamentarium of the spinal surgeon to decrease or negate allogenic blood transfusion. Consumables are however expensive and this cost needs to be weighed up against the cost of allogenic blood and the associated risk of infection transmission and adverse reactions. In this series of NMS patients, blood loss when using the cell saver was 2 100 ml and 1 000 ml without (p = 0.00003). This difference indicates that the pre-operative blood loss risk assessment was accurate and the cell saver used appropriately. Of course absolute blood loss is not easily interpreted as many of these children have small blood volumes. This may lead to a situation where a proportionally large blood loss is still not large enough to allow cell saver collection as the minimum collection for spinning down at our institution is 800 ml.

An interesting finding of this series was that patients with multiple level sublaminar wires (and pedicle screws at the bottom and/or top of the construct) maintained their correction better than constructs supplemented with hooks. A median of 2.17 (1–6 ± 1.09) hooks were used in 24 patients. These were at the superior end of the construct (transverse process hooks) when pedicle screws could not be inserted confidently. There was, however, not a statistical difference between the loss of correction of pedicle screw only constructs and hybrid constructs using sublaminar wires (Figure 8). The cost of pedicle screws is of concern, as is the risk of neurological injury when passing sublaminar wires.

Pre- and postoperative primary curve Cobb angles in this cohort are comparable to similar series in international literature and good correction was achieved throughout the follow-up period (Figure 6). Secondary or compensatory curves were not always included in the fusion mass and subsequently tended to deteriorate (Figure 7). Pelvic obliquity is a cause of great morbidity, and correction thereof often results in improved sitting function, pressure distribution and personal hygiene. It also frees the upper limbs from supporting the trunk while sitting, to allow for more regular activities like feeding and playing. A similar pelvic obliquity correction was achieved with and without incorporation of the pelvis into the fusion mass (71% compared to 67%).

The use of spinal cord monitoring has become the standard practice of care in spinal deformity correction. It has, however, a limited role in NMS patients. Somatosensory-evoked potential (SSEP) monitoring with multiple recording sites has been shown to be effective in early detection of intra-operative spinal cord injury in NMS surgery but it requires a trained neurophysiologist to be present during surgery. We currently use surgeon-controlled, transcendural, motor-evoked potential neuromonitoring for all idiopathic corrections, but have found it ineffective in many NMS cases.

Understandably, the patients themselves were less enthusiastic about the surgical procedure than the parents or caregivers. Most of the parents, who responded negatively to the questionnaire, cited the surgery simply being too much for the child and parent to warrant the outcome. Some thought the rigid spine and child made transferring and caring more difficult.

Conclusion

Although corrective spinal surgery in the neuromuscular patient is demanding and has a high complication rate, few complications have long-term sequelae. Primary curve deformity correction is obtained in the long term. Sitting and walking balance is improved by correcting the pelvic inclination and there is a high satisfaction rate from parents and patients. Corrective spinal surgery in the neuromuscular patient can halt and correct deformity and prolong life by maintaining or even improving pulmonary function.

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No benefits of any form have been derived by the author from any commercial party related directly or indirectly to the subject of this article.

References

CRITERIA FOR AUTHORSHIP AND CO-AUTHORSHIP OF ARTICLES

The following are internationally acknowledged criteria for authors/co-authors.

With the increase in faculty and in research projects, there is a potential for increased confusion and conflict regarding appropriate authorship credit on manuscripts and presentations. The following are some relatively standardised criteria that can be helpful. These may be overstrict when considering clinical studies in which surgeons often do the “hands on work” that create the study but may not perform major analysis and writing functions. However, all authors should read and contribute editing comments prior to submission.

Relman criteria for authorship

In particular, to qualify as an author a person should fulfill at least three of the following five requirements:

1. Conception of idea and design of experiment
2. Actual execution of experiment; hands on lab work
3. Analysis and interpretation of data
4. Actual writing of manuscript
5. Be able to present to a learned gathering a lecture on the work; interpret it, defend it and take responsibility for it.

These are just guidelines. On the other hand it is probably far worse to leave someone off the list who feels they may have contributed than to include someone who did a bit less.

We should all be as inclusive as possible, offer our interested colleagues the opportunity to provide input, analysis and editing of our works to support each other and improve our papers.