

Does Spinal Fusion Influence Quality of Life in Neuromuscular Scoliosis?

Eyal Mercado, MD, Benjamin Alman, MD, FRCSC, and James G. Wright, MD, MPH, FRCSC

Study Design. Systematic literature review of articles pertaining to quality of life (QOL) in neuromuscular scoliosis patients that underwent spinal fusion.

Objective. To determine if QOL is improved by scoliosis surgery in neuromuscular patients.

Summary of Background Data. The primary focus of most prior studies on neuromuscular scoliosis has been on the technical correction of spinal deformities, and not the child's postoperative performance and function in activities of daily living.

Methods. Computer-based English literature search of Google and PubMed databases.

Results. A total of 198 publications in the English literature between 1980 and 2006 were identified from a PubMed and Google Scholar search of QOL in neuromuscular scoliosis patients that underwent spinal fusion.

Conclusion. Spinal fusion improves QOL in CP (Grade C recommendation). Spinal fusion improves QOL in muscular dystrophy (Grade C recommendation). Spinal fusion does not improve QOL in spina bifida (Grade C recommendation).

Key words: quality of life, spinal fusion, neuromuscular scoliosis, spina bifida, cerebral palsy, Duchenne muscular dystrophy. *Spine* 2007;32:S120–S125

Patients with neuromuscular disorders frequently develop progressive spinal deformities. Clinicians have several therapeutic interventions that may either improve the physical function or slow the deterioration in these children.

However, documentation of the effects on the physical functioning of children in their relevant environments (e.g., at home, at school, and in the playground) has been lacking for many therapies.¹

The usual neuromuscular curve pattern is a long, collapsing curve, with an associated pelvic obliquity, sometimes increased kyphosis, often resulting with the loss of sitting balance. Spinal support braces might help maintain sitting balance but usually fails to prevent progression of deformity. Furthermore, braces are usually poorly tolerated, may limit the lung function in DMD patients,² or cause skin ulceration in spina bifida. Surgical treatment usually involves long instrumentation and frequently fusion

to the pelvis. Amid increasing demand for demonstrated effectiveness, surgical interventions must demonstrate measurable benefit.

Although many studies have reported on the result of surgical treatment of neuromuscular scoliosis, most studies lack a postoperative evaluation of the quality of life (QOL) of the patients or the overall effect on the caregivers. The majority of reports include only data on changes in the scoliosis angle and rates of postsurgical complications, and few have controlled comparisons.^{3–8} Furthermore, surgery has significant rate of perioperative complication rate.^{9,10} Despite a theoretical improvement in the pulmonary and coronary function, there are no reports demonstrating improvement in those functions after spinal fusion in neuromuscular scoliosis.¹¹ Thus, spinal instrumentation and fusion for neuromuscular spine deformity must provide meaningful benefit in their QOL in proportion to the risks involved for patients and their families.

Our purpose was to review the evidence and determine if spinal fusion improves QOL in neuromuscular scoliosis, specifically in cerebral palsy, spina bifida, and muscular dystrophy.

Methods

Clinical publications reporting QOL in neuromuscular scoliosis patients that underwent spinal fusion were identified from a PubMed and Google Scholar search indexed by key words: “quality of life, neuromuscular scoliosis, spinal fusion, scoliosis, spina bifida, meningomyelocele, cerebral palsy, and Duchenne muscular dystrophy,” indexed from 1980 and later.

We draw our conclusions based on the concept of Grades of Recommendation, introduced at the *Journal of Bone and Joint Surgery* (American edition) in September 2005.¹²

Levels of Evidence ratings for multiple studies addressing a clinical care recommendation are to be summarized with use of Grades of Recommendation. Grade A recommendations are based on consistent Level I studies.

Grade B recommendations are based on consistent Level II or III evidence.

Grade C recommendations represent either conflicting evidence or are based on Level IV or V evidence.

A grade of I indicates that there is insufficient evidence to make a treatment recommendations.

Results

A total of 146 publications from a PubMed search and 198 from Google Scholar reported the results of patients that underwent spinal fusion in neuromuscular scoliosis. The reports were from 1980 to 2006 and limited to human studies written in English. Titles and abstracts were exam-

From the Division of Orthopaedic Surgery, Hospital for Sick Children, Toronto, Ontario, Canada.

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Address correspondence and reprint requests to James G. Wright, MD, Division of Orthopaedic Surgery, Hospital for Sick Children, 555 University Avenue, Toronto, Ontario, M5A 1X8 Canada; E-mail: james.wright@sickkids.ca

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Defining and Measuring QOL

QOL measures are increasingly used to supplement objective clinical or biologic measures of disease to assess the effectiveness of interventions, including surgery. Health-related QOL measures reflect a growing appreciation of the importance of how patients feel and how satisfied they are with treatment in addition to the traditional focus on disease outcomes. The challenge in measuring QOL lies in its uniqueness to the disease and to the individuals. Patients with different neuromuscular disorders have different overall function, needs, and expectations; therefore, assessment of their QOL should be done on a disease-specific basis rather than as a group. Many of the existing measures of QOL impose standardized models of QOL and preselected domains. Another issue particularly relevant to children is that surgery is often performed to address long-term issues in QOL. No study has demonstrated long-term effect on QOL in neuromuscular patients.

There are 2 main approaches to the measurement of health status: general measures, attempting to address all aspects of health, and disease-specific measures, addressing only those aspects of health relevant to the disease or condition.

Measuring QOL in patients with disabilities is difficult and should take into consideration issues like:

1. Hopes, ambitions, and expectations.
2. Individuals' perceptions in the context of their culture and value systems where they live and in relation to their goals, expectations, standards, and concerns.
3. Individuals' issues they regard as important in their lives.¹³⁻¹⁵

The disease-specific outcome scales that have been developed for patients with neuromuscular disease include parameters such as sitting balance, feeding, hygiene care, ambulation, pain, and appearance.¹⁶⁻¹⁸ Few of these scores, however, focus on those aspects of physical disability related to spinal deformity, or the measures are specifically designed for young children in which scoliosis is not relevant (like Pediatric Evaluation of Disability Inventory).

An example of a generic measure is the Activities Scale for Kids, which was developed specifically to measure physical disability in 5- to 15-year-old children with musculoskeletal disorders. It does so by directly asking children what they have been doing at home, at school, and in the playground, and with what degree difficulty. Although this instrument was restricted to child self-report, and therefore excluded children with significant cognitive impairments, previous research has shown high concordance between child-report and parent-report (ICC 5 = 0.94).^{1,19}

Two specific measurements have been developed to assess QOL in scoliosis: The Scoliosis Research Society and the American Academy of Orthopedic Surgeons Mo-

deems evaluate spinal deformity. The second developed by Climent *et al*²⁰ measures QOL in adolescents with spine deformities (both structural and postural curves). These instruments evaluate idiopathic scoliosis in an active ambulatory population and were not intended for patients who sit full time in wheelchair with very weak upper and lower extremities. Thus, there is no QOL measure appropriate for all neuromuscular patients. Furthermore, because of the heterogeneity of patients with neuromuscular scoliosis, it is important to group them according to their specific disease.

Goldberg²¹ commented on the lack of a comprehensive, well-accepted outcome study for patients with cerebral palsy and stated that, until one is designed, "we can conclude only that cerebral palsy surgery makes patients different but not always better."

Only one measure has been developed specifically for patients with neuromuscular scoliosis; Wai *et al*²² developed a valid and reliable instrument, the Spina Bifida Spine Questionnaire, to evaluate those aspects of physical disability related to scoliosis and important to children with spina bifida and their families and found it reliable and valid disease-specific self-administered questionnaire assessing children with spina bifida and scoliosis.

Duchene Muscular Dystrophy (DMD)

Motor strength and pulmonary function are key issues QOL for boys with muscular dystrophy. Spinal fusion for scoliosis should be early enough in the course of the deformity, when pulmonary and cardiac function are sufficient, so the patient can be anesthetized and operated relatively safely and in order to reduce the likelihood of major complications. Prevention of pulmonary function deterioration is one of the important goals of scoliosis surgery in those patients. However, pulmonary function deteriorates despite the spinal fusion.^{2,23,24} It is unclear whether patients who undergo fusion deteriorate more slowly than those who do not. Because of lack of controlled studies, it is uncertain to whether surgical stabilization of the deformity improves pulmonary function.

Bridwell *et al*,²⁵ in their work on patients with progressive flaccid neuromuscular scoliosis, found that most patients/families believed that the spinal surgery improved their function, cosmesis, sitting balance, and QOL. However, the most negative outcomes concerned function, where 18 patients of the 48 studied were worse, 6 of 48 were neutral, and only 24 of 48 were better relative to preoperative function. The authors' explanation for these negative scores was that it reflects the progressively deteriorating nature of these conditions but does not take into account what the patient's function would have been (presumably even worse) had surgery not been performed. The most favorable evaluations (virtually all positive scores) concerned cosmesis, QOL (patient and parent), and overall satisfaction through an average follow-up period of 7.8 years after surgery.

Granata *et al*² similarly assessed outcome and QOL in their group of 30 DMD patients at mean follow-up of 4

years, by an open-ended questionnaire; 92% of patients were satisfied with the results and 100% of parents thought their son's QOL had improved. However, only 54% believed their son's care became easier (27% thought it became worse), which emphasize the difficulties in measuring the results with nonvalidated questionnaire. They also reported that patients with curve $<40^\circ$ were doing better than those with curve $>40^\circ$.

Ramirez *et al*²⁶ reviewed retrospectively the charts of 30 DMD patients and performed a telephone survey subjectively assessing the patient's outcome after surgery, in particular, the patient's current condition, complaints (patient or caretaker) related to surgery, postoperative respiratory problems, sitting tolerance, back pain, upper extremity use, effect on QOL, and whether they would recommend the surgery to others. It is not mentioned whether the questionnaire was validated. Fifteen of the 21 families contacted thought that spinal surgery resulted in an overall improvement in the patient's QOL. Specifically, families noted improved sitting ability, better posture, simplified nursing care, less back discomfort, and improved self-esteem. These comments included half of the patients with severe curves who had major complications. Six families thought that no improvement resulted from spinal fusion; of the 6 families, 4 experienced a major complication and 1 other had died in his second postoperative year. There were several other concerns or complaints that families expressed in the survey. Three patients were thought by family members to have increased difficulty with eating because of their sitting position. Because they sat more upright, they had more difficulty getting their food to their mouths.

In summary, the need to operate relatively early in the course of the deformity frequently when the curves are relatively mild, as well as the change in the course of the disease caused by steroids, makes measuring the impact of scoliosis surgery on the QOL in those patients more complex.

To further confound issues of QOL in DMD, the use of steroids to attenuate the progressive muscle weakness also alters the rate of development of spinal deformity, and it is not clear whether it influences the patient's QOL.

Since there is a reported decline in the rate of surgery in boys treated by long-term steroids, perhaps QOL related to the spine may be worse in these patients.

These issues illustrate how change in medical management for neuromuscular disorder produces profound differences in QOL issues related to the spine.

Based on self-report, the surgery may improve sitting ability, posture, simplify nursing care, and improve self-esteem. The effect on the respiratory and cardiac function is unknown.

Given the large emotional family investment in surgery, and the uncontrolled nature of the surgical series, the evidence in support of improving QOL of these patients is weak (Grade C recommendation¹²).

Cerebral Palsy (CP)

Spinal deformity constitutes a significant problem in pediatric patients with CP, more commonly affecting children with spasticity, and demonstrating an incidence directly proportionate to the severity of neurologic involvement.

The wide spectrum of cognitive and motor function in CP patients makes it hard to draw conclusions and, therefore, at a minimum requires distinction between ambulators and nonambulators and between cognitively intact and impaired patients. Spinal malalignment and trunk decompensation may affect standing balance in ambulators, possibly limiting their walking ability. Spinal imbalance may contribute to sitting intolerance in full time wheelchair users, converting them to hands-dependent sitters.

Narayanan *et al*²⁷ developed a disease-specific measure useful for evaluating the effectiveness of various interventions for the severe CP patients population (GMFCS IV-V). The questionnaire is based on recommendations from caregivers, healthcare professionals experienced in the management of children with severe CP, and a review of other questionnaires. The 36-item questionnaire, called the Caregiver Priorities and Child Health Index of Life with Disabilities, has 6 domains: 1) Personal Care, 2) Positioning, Transfer, and Mobility, 3) Communication and Social Interaction, 4) Comfort, Emotions, and Behavior, 5) Health, and 6) Overall Quality of Life. The questionnaire is a reliable and valid disease-specific measure of the caregivers' perspective on activity limitations, health status, well-being, and ease of care for children with severe CP. Its responsiveness (sensitivity to change) to therapeutic interventions has not been examined yet.

Previous studies²⁸⁻³⁰ suggested that fusion extending to the pelvis should be avoided for ambulatory patients in whom the scoliotic deformity is associated with pelvic obliquity. In CP patients with scoliosis, the aim of the surgery is to restore trunk alignment, improve respiratory function, alleviate the pain caused by impingement of the ribs against the iliac crest on the concave side of the curve, provide better sitting tolerance in wheelchair-bound patients, and retain standing and walking abilities in ambulatory patients, thereby maximizing their overall level of function.³¹

Tsirikos *et al*³² found no alteration in the ambulatory capacity in 23 of 24 spastic CP patients that underwent spinal fusion T1-T2 to sacrum with pelvic fixation. One patient in whom bilateral hip heterotopic ossification developed during the postoperative period after the spinal surgery developed bilateral hip ankylosis and unfortunately progressively lost the ability to ambulate. Twelve of their 24 patients underwent video gait analysis before and after surgery, which showed no change in their ambulatory function and a gait pattern similar to the preoperative pattern by 6 months after spine surgery.

Other studies^{11,33-35} have found that the benefits of spinal fusion for globally affected patients with spasticity are uncertain, emphasizing that the proposed benefits should be weighed against the increased incidence of po-

tential surgical complications, including perioperative death.¹¹

Tsirikos *et al*³⁶ delineated parents and professional caretaker satisfaction after spinal fusion in children with spasticity by sending a questionnaire to the patients' caregivers. Most of the parents who responded to the questionnaire suggested that the surgery was beneficial and increased their children's level of function. The parents also appreciated the dramatic correction of the patients' spine deformity and the consequent improvement of their physical appearance. There were also many negative responses in the section that referred to the deleterious effects of the spinal deformity on different functional skills of children with spasticity. A limitation of this study was that, although the answers received during the survey clearly demonstrated that most of parents and caretakers consider the surgical outcome of the spinal fusion beneficial for the patients, it does not evaluate actual function after spinal fusion but only the perceived improvement in the physical abilities of children with CP who underwent this procedure as reported by the parents and caregivers.

Jones *et al*¹⁰ prospectively examined both functional and caregiver satisfaction of 25 total body involvement CP patients, using the pediatric psychosocial and physical function assessment designed by the Pediatric Orthopedic Society of North America in conjunction with the Musculoskeletal Outcomes Data Evaluation and Management System of the American Academy of Orthopedic Surgeons.³⁷⁻³⁹ There were no significant changes between preoperative and postoperative assessments of physical function, school absence, comorbidities, and parental health. Although patients had a high rate of complications, pain, happiness, frequency of feeling sick and tired, and parental satisfaction improved significantly by 1 year after surgery. They suggested to aiming for achievable expectations such as improvement in the patient's looks, sleeping comfort, pain relief, and self-attitude.

In summary, spinal fusion in CP patients with scoliosis, based on self report, corrects the patients' spine deformity and consequently improves their physical appearance and self-esteem. Ambulation does not appear to decline after spinal fusion. Level of function was not improved. The proposed benefits should be weighed against the increased incidence of potential surgical complications. In view of the lack of well-controlled studies (Grade C recommendation¹²), the improvement in QOL is uncertain.

Spina Bifida

The prevalence of scoliosis in children with spina bifida has been estimated to be as high as 50%. Progressive scoliosis often results in loss of truncal stability, especially in curvatures $>40^\circ$, and when associated with pelvic obliquity $\geq 25^\circ$.

Surgery can improve or impair sitting balance, ambulation, and activities of daily living.⁴⁰

Surgery is generally successful at correcting the scoliosis with average improvements of approximately 50% from preoperative levels. The morbidity of surgery, however, is significant, with complication rates in excess of 40% to 50%.^{28,41,42}

The functional benefit of surgery is uncertain. Previous studies evaluating the functional outcomes after surgery were inconclusive,^{7,43-45} in part, due to the lack of reliable and valid questionnaires. Moreover, these studies used outcome measures that were clinician derived and therefore may not have reflected the activities that are important to the patients and their families.

Wai *et al*²² developed a valid and reliable instrument to evaluate those aspects of physical disability related to scoliosis and important to children with spina bifida and their families and developed the Spina Bifida Spine Questionnaire.

Wai *et al*⁴⁶ found that coronal imbalance was the only aspect of spinal deformity found to affect one aspect of physical function (*i.e.*, sitting). On average, sitting balance decreased 1 point (on the 10-point scale) for every 17-mm shift in coronal imbalance, or for a 50° increase in the primary scoliosis Cobb angle. After adjusting for neurologic level, this study showed no significant association between any aspect of spinal deformity with self-perception and overall physical function as measured by the Activities Scale for Kids. The absence of association shown in this study suggests that spinal deformity may not have a substantial effect on overall physical function or self-perception in children with spina bifida. Furthermore, those children who have had spinal fusion controlling for all other factors had lower overall function.

Schoenmakers *et al*⁴⁰ reported that it takes about 18 months before patients and their parents experience the beneficial effects that spinal fusion can have on functional abilities. After surgery, walking abilities often become difficult, especially for exercise walkers, although other factors such as ongoing neuraxial disease or natural history cannot be ruled out.

The importance of these studies is that perhaps simple interventions, such as chair modifications, should be investigated as possible means to shift the trunk to improve coronal balance and sitting function. In patients in whom surgical therapy is chosen, specific attention should be directed toward achieving coronal balance, including correction of all curves and leveling pelvic obliquity.

In summary, simple interventions, such as chair modifications, should be investigated as possible means to shift the trunk to improve coronal balance and sitting function before surgical intervention. Furthermore, no clear association was found between spinal deformity and overall physical function or self-perception.

The role of routine surgery for patients with spina bifida requires a reevaluation. At this time, surgery may be appropriate for those with sitting balance difficulties, not addressed through simple interventions such as chair modifications.

■ Conclusion

QOL measurement in children with disabilities is a complex and dynamic issue. One should not assume that children with neuromuscular scoliosis necessarily share the same expectations, needs, or goals in life and therefore cannot measure a QOL with the same instruments. To date, general satisfaction, as assessed by open-ended self-report questions, is generally high. For full-time wheelchair users, the sitting balance appears to be improved, while the coronal balance is the most important parameter affecting sitting. For dependent patients, surgery is based on self-report, simplified nursing care. Ambulation ability does not appear to deteriorate in CP patients. Given the wide array of sources for pain, in neuromuscular patients, and the cognitive difficulties of many of the patients, the role of surgery in relieving pain is uncertain.

Another issue in interpreting these studies is that, in the course of time, technology may improve and the disease state may change: for example, motorized mobility devices, which enable relative freedom of mobility, and increased independency for severe CP patients and their caregivers, or the substantial change in DMD patients with the introduction of steroids, prolonging their ambulation and delaying the need of ventilators. We do not know what the future will bring, perhaps an ability to cure the damaged foci in the brain of a CP child or the ability to use gene therapy to repair the genetic defect in boys with DMD; therefore, the goals of treatment and the aims for improving QOL in those patients would be different.

Based on grades of recommendation, the current literature shows that there is poor quality evidence (Grade C) that spinal fusion improves QOL in CP, and in muscular dystrophy and does not improve QOL in spina bifida (Grade C recommendation).

■ Key Points

- QOL measurement in children with disabilities is a complex and dynamic issue.
- Most studies regarding QOL in neuromuscular patients after spinal fusion are of low level of evidence (IV or lower).
- To date, general satisfaction, as assessed by an open-ended self-report questionnaire, is generally high.
- Spinal fusion improves QOL in CP, and in muscular dystrophy and does not improve QOL in spina bifida (Grade C recommendation).

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