

ETHICS IN PRACTICE

Spinal Fusion for Scoliosis in Patients with Globally Involved Cerebral Palsy An Ethical Assessment

Amanda T. Whitaker, MD, Melinda Sharkey, MD, and Mohammad Diab, MD

Peer Review: This article was reviewed by the Editor-in-Chief and one Deputy Editor, and it underwent blinded review by two or more outside experts. The Deputy Editor reviewed each revision of the article, and it underwent a final review by the Editor-in-Chief prior to publication. Final corrections and clarifications occurred during one or more exchanges between the author(s) and copyeditors.

Clinical ethics is a field of study that promotes the use of rational thought and clinical judgment based on scientific knowledge to reach conclusions regarding complex clinical problems^{1,2}. Common subjects in clinical ethics include informed consent, surrogate decision-making, refusal of care, medical futility, end-of-life care, and quality of life.

Spinal fusion in patients with cerebral palsy who have global involvement, defined as Gross Motor Function Classification System (GMFCS) level V, poses an ethical dilemma³. A child with GMFCS level-V function has severely limited self-mobility, voluntary muscle control, and ability to keep the head and trunk upright. A so-called four-topic model has been developed and validated to assist the clinician in the analysis of ethical dilemmas in patient care (Table I)^{4,6}. This model includes an evaluation of the indications, patient preferences, quality of life, and contextual features that underpin medical decisions. Indications include the problem, history, diagnosis, prognosis, acuity, how the patient may benefit, how harm may be avoided, goals of treatment, likelihood of success, and contingency plan in the event of failure. Patient preferences encompass the stated wishes of the patient or patient-centered decision-making by surrogates. Quality of life emphasizes the goals of restoring, maintaining, or improving the patient's life. Contextual features are concerned with other factors that influence decisions such as religion, culture, the law, confidentiality, research, teaching, economics, and impact on the patient's family and medical team.

Stable balanced posture, improved health and function, increased comfort of the patient, and easing of burden to caregivers have been posited as indications for spinal fusion⁷⁻¹². There are neither randomized controlled trials nor prospective longitudinal studies comparing fusion of the spine with the natural history of the disease. Outcomes measured are mostly observational and based on evaluation by secondary parties rather than within the affected person. It is difficult to determine whether favorable rates of satisfaction are the result of improvement in appearance or whether they represent improvements in comfort and function^{7-9,12}.

Several published studies have used the four-topic model to assist in complex clinical decision-making, such as chronic dialysis and craniectomy for traumatic brain injury^{4,5}. The four-topic model provides an organized framework for evaluation of ethical dilemmas in a logical, simplified manner and serves as the structure for the ethical evaluation of spinal fusion in a child with GMFCS level-V function.

Medical Indications

Indications for the operation include arrest of deformity progression, avoidance of cardiopulmonary dysfunction, improvement of posture, and ease of care.

The development, severity, and progression of scoliosis are associated with the severity of the underlying disease. Two-thirds of institutionalized patients with GMFCS level-V function develop scoliosis, and most (>80%) of them are tetraplegic¹³.

Disclosure: None of the authors received payments or services, either directly or indirectly (i.e., via his or her institution), from a third party in support of any aspect of this work. None of the authors, or their institution(s), have had any financial relationship, in the thirty-six months prior to submission of this work, with any entity in the biomedical arena that could be perceived to influence or have the potential to influence what is written in this work. Also, no author has had any other relationships, or has engaged in any other activities, that could be perceived to influence or have the potential to influence what is written in this work. The complete **Disclosures of Potential Conflicts of Interest** submitted by authors are always provided with the online version of the article.

TABLE I The Four-Topic Model*

<p>Medical indications</p> <p>Progressive curve of $>50^\circ$; surgery arrests progression and partially corrects deformity (+)</p> <p>Cardiopulmonary compromise; surgery does not improve cardiac function (-), and may improve subjective pulmonary function (+/-)</p> <p>Pelvic obliquity</p> <p>Surgery improves pelvic obliquity (+)</p> <p>Deformity-causing decubitus ulcers despite chair modifications</p> <p>Surgery does not decrease decubitus ulcers (-)</p> <p>Surgical morbidity (-)</p> <p>Surgery has been associated with decreased life expectancy (+/-)</p> <p>Patient preferences</p> <p>Unable to assess due to inability of the patient to communicate (-)</p>	<p>Quality of life</p> <p>Surgery has a high rate of complications (-)</p> <p>Pain; surgery may reduce pain due to severe spinal deformity (+/-)</p> <p>Surgery may require revision (-)</p> <p>Surgery increases hospitalization (-)</p> <p>Surgery improves posture (+)</p> <p>Surgery may enhance interaction with surroundings; this is immeasurable in the globally involved patient (+/-)</p> <p>Contextual features</p> <p>Altruism, including concern about denial of care (+)</p> <p>Decreased caregiver fatigue (+/-)</p> <p>Cognitive dissonance (-)</p> <p>Influence of industry (-)</p> <p>Cost to health-care system (-)</p> <p>Physician incentive in fee-for-service reimbursement (-)</p>
<p>*The four topics of clinical ethics with features specific to spinal fusion in cerebral palsy are indicated. Each feature is either in favor of spinal fusion in globally involved cerebral palsy (+) or against spinal fusion in globally involved cerebral palsy (-).</p>	

Patients with cerebral palsy can have a delayed skeletal maturity into the fourth decade of life, increasing the period during which the deformity may progress^{7,14}. Scoliosis may progress even after skeletal maturity, with the largest curves progressing most¹⁴⁻¹⁶. Bracing does not influence the natural history^{17,18}. Severity may be lessened and progression may be arrested by spinal fusion^{9,13,19}. The natural history of untreated scoliosis in these patients is obscure. Functional decline is associated with increasing age, but has not been linked causally to curve magnitude¹⁶. In one study, more patients with a curve of $<45^\circ$ demonstrated functional loss than did those having a curve of $>45^\circ$ ²⁰. Spinal deformity, including pelvic obliquity, can be improved with spinal fusion^{7,13,19,21}.

A fundamental challenge to testing cardiopulmonary function as an indication for a surgical intervention is the reliability of patient participation in pulmonary function and other active testing. In a prospective survey of neuromuscular scoliosis (including conditions other than cerebral palsy), and a retrospective study of all levels of cerebral palsy (not limited to GMFCS level V), subjective lung function as assessed by a questionnaire improved after fusion^{9,22}. No difference was found in oxygen saturation or heart rate between patients with scoliosis of $>45^\circ$ and those with mild or no scoliosis^{16,20}. Patients with neuromuscular scoliosis, both those managed with and without spinal fusion, have been reported to be hypoxemic, with oxygen saturations averaging 80%¹⁰. Children who are more affected by cerebral palsy (GMFCS level III) have worse results on pulmonary function tests than those less affected (GMFCS level I), and a positive correlation has been found between activities of daily living and respiratory muscle strength^{23,24}. There are no reported differences in the utilization of pulmonary medication, or in the rate of pneumonia, between patients with cerebral palsy who have had fusion and those who have not²⁵. These findings suggest that

pulmonary function may be substantially affected by the underlying disease and not by operative treatment.

Many studies of spinal fusion in patients with neuromuscular scoliosis have shown subjective improvement in sitting posture, assessed both prospectively by clinical observation using published protocols and retrospectively by radiographs, surveys, and interviews^{7,9,10,13,21,22}. Correction of curve magnitude is consistently achievable²⁶. Patients with scoliosis and GMFCS level-V function require more wheelchair modifications; with fusion, they have improved sitting balance and sitting weight distribution, and they use less support for sitting^{9,20}. While changes in pressure brought about by the pelvic obliquity that accompanies spinal deformity may potentially increase decubiti, studies have not shown a causal connection^{9,20,25}. There are no data, as far as we know, to suggest that spinal fusion improves hip dislocation rates.

Improvements in other functional outcomes are unclear. A prospective functional study in patients with mixed cerebral palsy showed no change in physical function, school absences, or comorbidities⁸. No study, to our knowledge, has described a change in the highest functional level, head control, or feeding ability^{12,20,25}. Another prospective study of patients with mixed cerebral palsy showed that, after surgery, the level of independence deteriorated at six months and walking ability decreased at twenty-four months, demonstrating that spinal fusion may not positively influence functional decline¹³.

Complication rates have been reported to be as high as 33.1% within the first three months after the operation and have ranged from 17.9% to 81%^{7,10,19,27,28}. The most common complications are infection and pseudarthrosis. The rate of infection may be underestimated because of short mean follow-up times, which will not detect delayed presentations¹⁹. Other complications include loss of correction, implant failure, fracture, junctional deformity, and death^{11,19,28}. Revision rates

after spinal fusion have been reported to be as high as 21%⁷. The complication rate increases with greater cognitive impairment, poor hygiene, poor skin, and poor nutrition^{11,19}.

The mean duration of survival after spinal fusion in an uncontrolled, observational study was found to be eleven years and two months²⁹. Mortality after spinal fusion has been reported to range from 0.3% within thirty days to 10% within two years; this is increased from the baseline mortality rate for patients with cerebral palsy^{7,28,30}. The number of days in the intensive care unit after the operation and preoperative hyperkyphosis correlated with mortality²⁹. There was no correlation between preoperative comorbidities and the length of postoperative stay in the intensive care unit²⁹.

Variability in definition and reporting as well as the rapidity of technological advances, such as neural monitoring, instrumentation, imaging, and antifibrinolytics for prophylaxis against venous thromboembolism, make the interpretation of results, and the establishment of indications, difficult^{10,15}. Another factor is the low level of evidence of the literature. There is no study with Level-I evidence, to our knowledge. Most studies are retrospective and rely on surveys in order to measure satisfaction with outcomes. The subjectivity of study participants also manifests in study investigators. One retrospective study with an 85% satisfaction rate excluded patients who died or suffered spinal cord injury⁷. This rate may decline with the inclusion of all patients.

In summary, the medical indications for which a consensus may be reached in the literature for spinal fusion in patients with GMFCS level-V function are improved posture and a reduction in the need for sitting supports. The medical indications against spinal fusion are high complication rates and increased mortality. The evidence is equivocal for other medical indications, including a change in the natural history of functional decline, pain, decubitus ulcer, and cardiopulmonary function.

Patient Preferences

Patients with GMFCS level-V function are incapable of participating actively or directly in decision-making to a meaningful extent. Clinical ethicists recommend decisions be centered around the best interests of the patient^{1,2}. Acting in the best interest of the patient is justified by the ethical guideline of beneficence. Interests of and benefits to secondary parties, such as parents and other caregivers, must be subordinated to those of the patient and must not be the central focus or driving influence². However, in this clinical setting, the preferences of the patient are unknown, and assessment of outcome shifts to secondary parties.

Quality of Life

Quality of life is determined by an individual's perception of what physical, mental, and social sacrifices he or she is willing to make for an improvement in quality of life. A grade-C recommendation (poor-quality evidence) has been given after an evaluation of the literature to determine if spinal fusion for scoliosis influences the quality of life in patients with GMFCS

level-V function and other neuromuscular conditions³¹. At the present time, we know of no higher-quality randomized controlled studies of quality of life from which to make a clear recommendation for or against surgical intervention for scoliosis in these patients.

Quality of life may be affected by pain, most notably that caused by the impingement of the ribs on the iliac crest on the concavity, and a concentration of body weight toward the convexity, in severe curves. In retrospective studies of caregivers, most express a subjective decrease in pain in patients after spinal fusion^{7,12,27}. This effect has not been observed when studied prospectively^{9,22}. Objective measures of pain, such as quantity of analgesics administered and missed days of school, have not shown a treatment effect from spinal fusion^{8,9,20,22}.

Retrospective studies of patients with mixed levels of neuromuscular scoliosis have demonstrated subjective improvement in physical appearance and self-esteem only in the setting of normal intelligence^{7,32}. Parents and other caregivers agree that appearance is improved after spinal fusion¹². These observations may be influenced by detection bias as it would be difficult for individuals involved in making the decision to perform surgery to limit this substantial source of bias.

Contextual Features

Contextual features in surgery encompass social, economic, and legal considerations. In patients without cerebral palsy, contextual factors are considered less important because they are peripheral to the patient. In a patient with GMFCS level-V function, contextual features rise to prominence as proxies, displacing the patient from the center of decision-making. Specific contextual features relevant to spinal fusion in patients with GMFCS level-V function include hospital resource utilization and throughput, technology, caregiver well-being, cognitive dissonance and placebo effect, physician incentive, and the education and training of surgeons.

Economic pressures are increasing worldwide for all aspects of health care. Spine surgery represents arguably the greatest such increase in orthopaedics. A hospital resource utilization review at our institution (unpublished data) demonstrated that spinal fusion for neuromuscular scoliosis consumes more than three times the resources for the same reimbursement compared with idiopathic scoliosis²⁵. Such cost, with increasing scrutiny of complications, measures of quality, and linkage to contracting and reimbursement, impact access to care as institutions shun risky procedures in patients who require resource-intensive care^{33,34}. This unwritten and undeclared distribution of resources, passive and obscure, merits ethical scrutiny.

The parents of children with GMFCS level-V function are at risk for serious health and social consequences. The rates of divorce and maternal depression in families with a child who has cerebral palsy exceed those of the general population and are associated with the degree of neuromuscular involvement^{35,36}. No study of spinal fusion, to our knowledge, includes these important metrics.

While one retrospective study showed subjective improvement in ease of care after spinal fusion, most prospective studies have shown no change in ease of care such as time consumed or physical effort^{9,12,25}. One study described worsening of the caregiver burden at six months after spinal fusion because of the increased spasticity of the patient, suggesting that the overall burden of global disease outweighs the effect of surgical intervention¹³. There had been no reported change in the health of the caregiver⁸. While reducing caregiver fatigue may bestow benefit to a child because of the child's complete dependence, caregiver perception of benefits has not been supported by objective measures.

After spinal fusion in patients with GMFCS level-V function, the literature has described a high level of parent and caregiver satisfaction, ranging from 81% to 95%^{7,8,12,25}. This satisfaction is related to the decision and is above and beyond the patient or the disease: extended caregivers are less likely to recommend the operation than parents, who demonstrate bias by their decision on behalf of their child. The gravity of the decision, related to both the high risk of the procedure and the sole assumption of responsibility by the parent, is a contextual factor unique to this patient population.

The pediatric orthopaedic surgeon plays another critical role in the contextual framework. As long as the prevailing system of reimbursement is based on fee for service, which rewards intensity and volume of care, there will be a conflict of interest that drives spinal fusion and disincentivizes nonoperative care. Operative treatment of patients with cerebral palsy has the support of strong tradition. Disease manifestation is potentially extreme and consistent. Complications, in particular neural injury, in a patient with GMFCS level-V function who is understood a priori to have high risk may be perceived less negatively. These factors may conspire to inform a favorable view of outcomes on the part of the surgeon.

Discussion

The evolution of the four-topic model from medical indications, patient preferences, and quality of life to contextual features represents a progression from the quantitative and patient-centered to the qualitative and environmental. While clinical ethics compel a greater weighting of the former, evaluation of spinal fusion in patients with GMFCS level-V function is more influenced by the latter.

Assessment of indications and outcomes in patients with GMFCS level-V function is more observational than participatory. Observational data pass the test of objectivity, and when obtained from validated quality-of-life instruments, they are informative and relevant. Subjective data, obtained from participants other than the patient, are the least robust ethically and empirically. Evaluation of outcomes and treatment recommendations driven by theory and what appears logical and self-evident are ethically moot.

Medical indications are assembled for a clear picture of the facts and probabilities. The diagnosis, course of disease, goals of treatment, and risks and benefits of the proposed intervention must be carefully considered². At this time, no robust evidence exists for medical improvement of the patient

after attainment of a fused spine. Outcomes are inconsistent and often conflicting, obscuring decision-making. However, the potential for clear harm exists. In other studies using the four-topic model, medical indications are underpinned by powered statistics and control groups, unlike studies on cerebral palsy with global involvement and spinal fusion^{4,5}.

A fundamental problem that pervades the literature on this topic is heterogeneity and the lack of strict inclusion criteria. For example, one of the most widely quoted studies showing improved outcomes after the operation combined disparate diagnoses, including multiple neuromuscular conditions⁹. Even within the cerebral palsy cohort, ambulatory patients, who function at a higher level than patients with GMFCS level-V function, were included. This exposes the dangers of extrapolation and a generalized, non-nuanced approach to neuromuscular scoliosis.

Technological advances in spinal fusion complicate the assessment of outcomes of spinal fusion. The greater potency of implants and effectiveness of perioperative care may improve and encourage spinal fusion. The lack of standardization in surgical technique and the surgeon experiential difference between those who publish their results and those who do not adds to confusion in the interpretation of results.

An essential factor to consider in the treatment of children with severe cerebral palsy is cognitive dissonance. Cognitive dissonance describes the tension created from attempting to reconcile two contradictory beliefs or realities, a tension that may be relieved by changing one of the two to be consistent. Randomized controlled trials have been performed in orthopaedics exposing the strong role of a placebo effect in surgery, as exemplified by knee arthroscopy in osteoarthritis and vertebroplasty for osteoporotic spine fractures³⁷⁻³⁹. Cognitive dissonance is one component of the placebo effect⁴⁰. Several factors predispose parents and caregivers to this mechanism. They consent to an intervention that exposes their child to high risk, and such exposure is justified by a good outcome. Postdecision analysis encourages a perception of benefit. Caregivers have high expectations before the operation⁸. The cost to the family and time off work may increase the desire for a successful outcome⁴⁰. A poor or fair objective outcome is dissonant with the decision, preparation, expectations, and increased work, whereas good subjective outcomes will be harmonious. This results in a perceived positive outcome by the decision maker. The physician also is vulnerable. Length of training, complexity of procedures, and conflict of interest represent a substantial investment in a good outcome. Acceptance of cognitive dissonance is disempowering, devaluing a medical intervention, i.e., surgery, which is presumed effective.

While we continue to lack high levels of evidence for surgical management of spine deformity in patients with GMFCS level-V function, other forces will drive care. Despite the ethical imperative to avoid them, we rely on observational data obtained from secondary parties in decision-making, because this is expedient. Since the patient with GMFCS level-V function is dependent on others for care, increasing others' quality of life to improve the patient's quality of life is compelling.

Our discipline is dynamic and responsive to the challenges of caring for these patients. Surgical techniques have advanced to reduce morbidity, including antifibrinolytics for prophylaxis against venous thromboembolism, prophylactic antibiotic powder in the surgical site, and more stable fixation. These represent steps in an iterative process designed to improve care. The development of validated health-related quality-of-life instruments such as CPCHILD (Caregiver Priorities and Child Health Index of Life with Disabilities) is a response to the ethical imperative for patient-centered decision-making, care, and assessment⁴¹.

It is debated whether Level-I evidence is necessary or indicated or even feasible for patients with this condition. Challenges include poor accrual, shared decision-making with patients who undermine randomization by treatment preference, unfeasibility of blinding surgery, and variability introduced by the surgeon's skill and experience⁴². Balancing this view are randomized controlled trials of selective dorsal rhizotomy and botulinum toxin in patients with GMFCS level-V function, which have provided clear and standard guidelines for the use and assessment of such treatment modalities, as well as robust evidence for their efficacy^{43,44}.

Our review is a call for prospective and controlled evaluation of spinal fusion for the treatment of scoliosis in patients with GMFCS level-V function. We need to measure objective data for objective goals that will determine patient-centered outcomes. We need to eliminate or limit bias wherever possible. We need consistency in inclusion and in analysis. We need to acknowledge that the patient in this setting does not participate in decision-making. We need to control contextual features. We need to be accepting that we may not succeed, because of the multiple factors involved and the complexity of disease. If so, we need to acknowledge the burden-of-proof razor: lack of

evidence for a procedure requires no evidence to dismiss it. It is incumbent on those who would introduce, apply, or advance a medical treatment to prove benefit.

In conclusion, evaluation of spinal fusion for scoliosis in a patient with GMFCS level-V function by the four-topic model of clinical ethics is challenging, demonstrating the current complexity of surgical decision-making and the lack of robust evidence. Medical indications are conflicting, with variable benefit but a risk of substantial complications. Patient preferences are impossible to determine. Quality of life is measured through secondary parties. Contextual features, least important in an ethical framework, disproportionately drive care. Spinal fusion in children with cerebral palsy who have GMFCS level-V function poses an ethical dilemma because of insufficient and conflicting evidence of benefit, substantial risks, inability of the patient to participate in decision-making or the evaluation of outcome, and disproportionate emphasis on caregiver perceptions. ■

Amanda T. Whitaker, MD
 Mohammad Diab, MD
 Department of Orthopaedic Surgery,
 University of California San Francisco,
 MU-325W, 500 Parnassus Avenue,
 San Francisco, CA 94143-0728.
 E-mail address for M. Diab: Mohammad.Diab@ucsf.edu

Melinda Sharkey, MD
 Department of Orthopaedics and Rehabilitation,
 Yale School of Medicine, Yale Physicians Building,
 800 Howard Avenue,
 New Haven, CT 06510

References

- Jonsen AR, Siegler M, Winslade WJ. Clinical ethics: a practical approach to ethical decisions in clinical medicine. 4th ed. New York: McGraw-Hill; 1998. p 206.
- Lo B. Resolving ethical dilemmas: a guide for clinicians. 5th ed. Philadelphia: Lippincott Williams & Wilkins; 2013. p 372.
- Palisano R, Rosenbaum P, Walter S, Russell D, Wood E, Galuppi B. Development and reliability of a system to classify gross motor function in children with cerebral palsy. *Dev Med Child Neurol*. 1997 Apr;39(4):214-23.
- Honeybul S, Gillett G, Ho K, Lind C. Ethical considerations for performing decompressive craniectomy as a life-saving intervention for severe traumatic brain injury. *J Med Ethics*. 2012 Nov;38(11):657-61. Epub 2012 Jul 17.
- Schumann JH, Alfandre D. Clinical ethical decision making: the four topics approach. *Semin Med Pract*. 2008;11:36-42.
- Sokol DK. The "four quadrants" approach to clinical ethics case analysis; an application and review. *J Med Ethics*. 2008 Jul;34(7):513-6.
- Comstock CP, Leach J, Wenger DR. Scoliosis in total-body-involvement cerebral palsy. Analysis of surgical treatment and patient and caregiver satisfaction. *Spine (Phila Pa 1976)*. 1998 Jun 15;23(12):1412-24; discussion 1424-5.
- Jones KB, Sponseller PD, Shindle MK, McCarthy ML. Longitudinal parental perceptions of spinal fusion for neuromuscular spine deformity in patients with totally involved cerebral palsy. *J Pediatr Orthop*. 2003 Mar-Apr;23(2):143-9.
- Larsson EL, Aaro SI, Normelli HC, Oberg BE. Long-term follow-up of functioning after spinal surgery in patients with neuromuscular scoliosis. *Spine (Phila Pa 1976)*. 2005 Oct 1;30(19):2145-52.
- Lonstein JE, Akbaria A. Operative treatment of spinal deformities in patients with cerebral palsy or mental retardation. An analysis of one hundred and seven cases. *J Bone Joint Surg Am*. 1983 Jan;65(1):43-55.
- Sponseller PD, LaPorte DM, Hungerford MW, Eck K, Bridwell KH, Lenke LG. Deep wound infections after neuromuscular scoliosis surgery: a multicenter study of risk factors and treatment outcomes. *Spine (Phila Pa 1976)*. 2000 Oct 1; 25(19):2461-6.
- Tsirikos AI, Chang WN, Dabney KW, Miller F. Comparison of parents' and caregivers' satisfaction after spinal fusion in children with cerebral palsy. *J Pediatr Orthop*. 2004 Jan-Feb;24(1):54-8.
- Askin GN, Hallett R, Hare N, Webb JK. The outcome of scoliosis surgery in the severely physically handicapped child. An objective and subjective assessment. *Spine (Phila Pa 1976)*. 1997 Jan 1;22(1):44-50.
- Thometz JG, Simon SR. Progression of scoliosis after skeletal maturity in institutionalized adults who have cerebral palsy. *J Bone Joint Surg Am*. 1988 Oct; 70(9):1290-6.
- Madigan RR, Wallace SL. Scoliosis in the institutionalized cerebral palsy population. *Spine (Phila Pa 1976)*. 1981 Nov-Dec;6(6):583-90.
- Majd ME, Muldowny DS, Holt RT. Natural history of scoliosis in the institutionalized adult cerebral palsy population. *Spine (Phila Pa 1976)*. 1997 Jul 1;22 (13):1461-6.
- Olafsson Y, Saraste H, Al-Dabbagh Z. Brace treatment in neuromuscular spine deformity. *J Pediatr Orthop*. 1999 May-Jun;19(3):376-9.
- Terjesen T, Lange JE, Steen H. Treatment of scoliosis with spinal bracing in quadriplegic cerebral palsy. *Dev Med Child Neurol*. 2000 Jul;42(7):448-54.
- Benson ER, Thomson JD, Smith BG, Banta JV. Results and morbidity in a consecutive series of patients undergoing spinal fusion for neuromuscular scoliosis. *Spine (Phila Pa 1976)*. 1998 Nov 1;23(21):2308-17; discussion 2318.
- Kalen V, Conklin MM, Sherman FC. Untreated scoliosis in severe cerebral palsy. *J Pediatr Orthop*. 1992 May-Jun;12(3):337-40.

- 21.** Karampalis C, Tsirikos AI. The surgical treatment of lordoscoliosis and hyperlordosis in patients with quadriplegic cerebral palsy. *Bone Joint J.* 2014 Jun; 96-B(6):800-6.
- 22.** Watanabe K, Lenke LG, Daubs MD, Watanabe K, Bridwell KH, Stobbs G, Hensley M. Is spine deformity surgery in patients with spastic cerebral palsy truly beneficial?: a patient/parent evaluation. *Spine (Phila Pa 1976).* 2009 Sep 15;34(20):2222-32.
- 23.** Kwon YH, Lee HY. Differences of respiratory function according to level of the Gross Motor Function Classification System in children with cerebral palsy. *J Phys Ther Sci.* 2014 Mar;26(3):389-91. Epub 2014 Mar 25.
- 24.** Wang HY, Chen CC, Hsiao SF. Relationships between respiratory muscle strength and daily living function in children with cerebral palsy. *Res Dev Disabil.* 2012 Jul-Aug;33(4):1176-82. Epub 2012 Mar 22.
- 25.** Cassidy C, Craig CL, Perry A, Karlin LI, Goldberg MJ. A reassessment of spinal stabilization in severe cerebral palsy. *J Pediatr Orthop.* 1994 Nov-Dec; 14(6):731-9.
- 26.** Tsirikos AI, Lipton G, Chang WN, Dabney KW, Miller F. Surgical correction of scoliosis in pediatric patients with cerebral palsy using the unit rod instrumentation. *Spine (Phila Pa 1976).* 2008 May 1;33(10):1133-40.
- 27.** Mohamad F, Parent S, Pawelek J, Marks M, Baström T, Faro F, Newton P. Perioperative complications after surgical correction in neuromuscular scoliosis. *J Pediatr Orthop.* 2007 Jun;27(4):392-7.
- 28.** Reames DL, Smith JS, Fu KM, Polly DW Jr, Ames CP, Berven SH, Perra JH, Glassman SD, McCarthy RE, Knapp RD Jr, Heary R, Shaffrey CI; Scoliosis Research Society Morbidity and Mortality Committee. Complications in the surgical treatment of 19,360 cases of pediatric scoliosis: a review of the Scoliosis Research Society morbidity and mortality database. *Spine (Phila Pa 1976).* 2011 Aug 15;36(18):1484-91.
- 29.** Tsirikos AI, Chang WN, Dabney KW, Miller F, Glutting J. Life expectancy in pediatric patients with cerebral palsy and neuromuscular scoliosis who underwent spinal fusion. *Dev Med Child Neurol.* 2003 Oct;45(10):677-82.
- 30.** Brooks JC, Strauss DJ, Shavelle RM, Tran LM, Rosenbloom L, Wu YW. Recent trends in cerebral palsy survival. Part I: period and cohort effects. *Dev Med Child Neurol.* 2014 Nov;56(11):1059-64. Epub 2014 Jun 26.
- 31.** Wright JG, Einhorn TA, Heckman JD. Grades of recommendation. *J Bone Joint Surg Am.* 2005 Sep;87(9):1909-10.
- 32.** Mercado E, Alman B, Wright JG. Does spinal fusion influence quality of life in neuromuscular scoliosis? *Spine (Phila Pa 1976).* 2007 Sep 1;32(19)(Suppl):S120-5.
- 33.** Skaggs DL, Lehmann CL, Rice C, Killelea BK, Bauer RM, Kay RM, Vitale MG. Access to orthopaedic care for children with Medicaid versus private insurance: results of a national survey. *J Pediatr Orthop.* 2006 May-Jun;26(3):400-4.
- 34.** Cho SK, Egorova NN. The association between insurance status and complications, length of stay and costs for pediatric idiopathic scoliosis. *Spine (Phila Pa 1976).* 2014 Dec 9. Epub 2014 Dec 9.
- 35.** Joesch JM, Smith KR. Children's health and their mothers' risk of divorce or separation. *Soc Biol.* 1997 Fall-Winter;44(3-4):159-69.
- 36.** Sawyer MG, Bittman M, LA Greca AM, Crettenden AD, Borojevic N, Raghavendra P, Russo R. Time demands of caring for children with cerebral palsy: what are the implications for maternal mental health? *Dev Med Child Neurol.* 2011 Apr;53(4):338-43. Epub 2011 Jan 14.
- 37.** Buchbinder R, Osborne RH, Ebeling PR, Wark JD, Mitchell P, Wriedt C, Graves S, Staples MP, Murphy B. A randomized trial of vertebroplasty for painful osteoporotic vertebral fractures. *N Engl J Med.* 2009 Aug 6;361(6):557-68.
- 38.** Moseley JB, O'Malley K, Petersen NJ, Menke TJ, Brody BA, Kuykendall DH, Hollingsworth JC, Ashton CM, Wray NP. A controlled trial of arthroscopic surgery for osteoarthritis of the knee. *N Engl J Med.* 2002 Jul 11;347(2):81-8.
- 39.** Tsutsui S, Pawelek J, Baström T, Lenke L, Lowe T, Betz R, Clements D, Newton PO. Dissecting the effects of spinal fusion and deformity magnitude on quality of life in patients with adolescent idiopathic scoliosis. *Spine (Phila Pa 1976).* 2009 Aug 15;34(18):E653-8.
- 40.** Homer JJ, Sheard CE, Jones NS. Cognitive dissonance, the placebo effect and the evaluation of surgical results. *Clin Otolaryngol Allied Sci.* 2000 Jun;25(3):195-9.
- 41.** Narayanan UG, Fehlings D, Weir S, Knights S, Kiran S, Campbell K. Initial development and validation of the Caregiver Priorities and Child Health Index of Life with Disabilities (CPCHILD). *Dev Med Child Neurol.* 2006 Oct;48(10):804-12.
- 42.** Touijer KA. The promise and challenges of randomized controlled trials for surgical interventions. *Eur Urol.* 2013 Apr;63(4):615-6; discussion 616-7. Epub 2012 Aug 19.
- 43.** Copeland L, Edwards P, Thorley M, Donaghey S, Gascoigne-Pees L, Kentish M, Cert G, Lindsley J, McLennan K, Sakzewski L, Boyd RN. Botulinum toxin A for non-ambulatory children with cerebral palsy: a double blind randomized controlled trial. *J Pediatr.* 2014 Jul;165(1):140-146.e4. Epub 2014 Mar 12.
- 44.** Farmer JP, Sabbagh AJ. Selective dorsal rhizotomies in the treatment of spasticity related to cerebral palsy. *Childs Nerv Syst.* 2007 Sep;23(9):991-1002. Epub 2007 Jul 21.