Surgical Correction of Scoliosis in Children with Spastic Quadriplegia: Benefits, Adverse Effects, and Patient Selection

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Abstract

Study Rationale Cerebral palsy (CP) is a group of nonprogressive syndromes of posture and motor impairment associated with lesions of the immature brain. Spastic quadriplegia is the most severe form with a high incidence of scoliosis, back pain, respiratory compromise, pelvic obliquity, and poor sitting balance. Surgical stabilization of the spine is an effective technique for correcting deformity and restoring sitting posture. The decision to operate in this group of patients is challenging.

Objectives The aim of this study is to determine the benefits of surgical correction of scoliosis in children with spastic quadriplegia, the adverse effects of this treatment, and what preoperative factors affect patient outcome after surgical correction.

Materials and Methods A systematic review was undertaken to identify studies describing benefits and adverse effects of surgery in spastic quadriplegia. Factors affecting patient outcome following surgical correction of scoliosis were assessed. Studies involving adults and nonspastic quadriplegia were excluded.

Results A total of 10 case series and 1 prospective and 3 retrospective cohort studies met inclusion criteria. There was significant variation in the overall risk of complications (range, 10.9−70.9%), mortality (range, 2.8−19%), respiratory/pulmonary complications (range, 26.9−57.1%), and infection (range, 2.5−56.8%). Factors associated with a worse outcome were a significant degree of thoracic kyphosis, days in the intensive care unit, and poor nutritional status.

Conclusion Caregivers report a high degree of satisfaction with scoliosis surgery for children with spastic quadriplegia. There is limited evidence of preoperative factors that can predict patient outcome after scoliosis. There is a need for well-designed prospective studies of scoliosis surgery in spastic quadriplegia.

Keywords
- cerebral palsy
- scoliosis surgery
- spastic quadriplegia
- outcomes

Study Rationale and Context

Cerebral palsy (CP) refers to a group of nonprogressive, heterogeneous syndromes of posture and motor impairment associated with certain lesions of the immature brain.[1] Spastic quadriplegia is the most severe form of CP characterized by spastic weakness involving the trunk and all extremities. It is the most common type of CP and also has the highest

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rate of scoliosis with a reported incidence of over 60%.[2,3] For children with spastic quadriplegia, the complications of scoliosis can be severe and debilitating. Back pain, poor balance, and compromise of respiratory function are all potential problems with a progressing deformity.[4,5] Furthermore, scoliosis is often associated with obliquity of the pelvis relative to the sitting surface causing uneven weight distribution and resultant pain, decreased sitting tolerance, and increased skin pressure.[6]

Curve progression in children with CP is maximal during puberty and frequently continues to progress even after skeletal maturity. Spinal orthotics are poorly tolerated by children with CP and have not been shown to have a significant impact on scoliosis shape or rate of progression in spastic quadriplegic patients.[7] Surgical stabilization of a progressing curve remains the most effective technique for correcting deformity and restoring sitting posture.[8]

The decision to operate in this group of patients is usually based on multiple factors including the progressive increase in Cobb angle, degree of sitting imbalance, apparent discomfort, and caregiver/patient opinion. This article seeks to review the available literature in this controversial area to facilitate surgical planning.

Objectives

The aim of this study is to determine the following:

- The reported benefits of surgical correction of scoliosis in children with spastic quadriplegia
- The short-term and long-term adverse effects of this treatment
- What preoperative factors, if any, affect patient outcome after surgical correction.

Materials and Methods

Study design: Systematic review.
Search: The databases included PubMed, Cochrane, and National Guideline Clearinghouse Databases, as well as bibliographies of key articles.
Dates searched: The dates were searched up to June 11, 2013.
Inclusion criteria: (1) patients 18 years or younger; (2) surgical correction for scoliosis in spastic quadriplegic patients; and (3) studies with at least 20 patients.
Exclusion criteria: (1) patients older than 18 years; (2) spastic diplegia or hemiplegia, ataxic or athetoid (dyskinetic) cerebral palsy; (3) case reports or case series with fewer than 10 patients; and (4) cadaveric studies, nonhuman in vivo, in vitro, and biomechanical studies.
Outcomes: (1) functional outcomes, including musculoskeletal, respiratory, and gastrointestinal; (2) radiographic outcomes; (3) complications or adverse events; (4) postoperative pain; and (5) patient/parent satisfaction.
Analysis: Descriptive statistics; Complication risks were calculated by dividing the number of patients with a given complication by the total number of patients at risk for that complication. Pooling of data was not done due to concerns regarding heterogeneity of treatments and populations as well as study quality.
Overall strength of evidence: Risk of bias for individual studies was based on using criteria set by *The Journal of Bone and Joint Surgery*[9] modified to delineate criteria associated with methodological quality and risk of bias based on recommendation from the Agency for Healthcare Research and Quality (AHRQ).[10,11] The overall strength evidence across studies was based on precepts outlined by the Grades of Recommendation Assessment, Development and Evaluation Working Group,[12] and recommendations made by the AHRQ.[10,11]

Details about methods can be found in the online supplementary material.

Results

- From 215 citations, 37 articles were evaluated for full-text review. Ten case series (class of evidence [CoE] IV) examining the reported benefits and adverse effects of surgical correction of scoliosis surgery in children with spastic quadriplegia and one prospective cohort study and three retrospective cohort studies (CoE III) reporting predictive factors following scoliosis surgery met the inclusion criteria and form the basis for this report (►**Fig. 1**). No studies were found comparing outcomes from surgery to outcomes from nonsurgical treatment.
- Characteristics of studies investigating the reported benefits and adverse effects of scoliosis surgery are outlined in ►**Table 1** and those examining predictive factors following scoliosis surgery are outlined in ►**Table 2**. Refer to the online supplementary material for critical appraisal, a list of excluded articles, and detailed outcome tables.

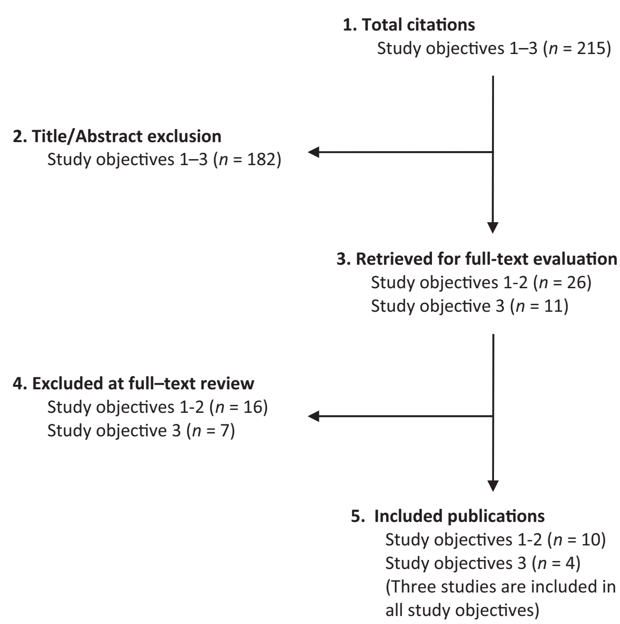


Fig. 1 Flow chart showing results of literature search.

Table 1 Characteristics of included studies for benefits and safety of surgical correction for scoliosis in spastic quadriplegic patients

Investigator (y) Study design CoE	Population[a]	Condition	Intervention	Follow-up (%)
Bohtz et al (2011) Case series CoE: IV	• $N = 50$ • Age (mean): 15.1 y (8.8–33.2) • Male: 46.0%	• Tetra spastic cerebral palsy (GMFCS levels IV and V) • Progressive scoliosis of > 50 degrees	• Spinal fusion with segmental pedicle screw instrumentation in thoracic and lumbar spine • Fusion levels (mean): 13.9 motion segments (range, 7–19)	2 y (% NR)
Keeler et al (2010) Case series CoE: IV	• $N = 52$ • Age (mean): 15.3 y (9.0–19.9) • Male: 59.6%	• Nonambulatory spastic quadriplegic cerebral palsy with scoliosis	• Anterior/posterior spinal fusion (1992–2001) or posterior spinal fusion (1998–2005) with autogenous bone graft (iliac crest and/or rib graft) and additional cancellous allograft, pelvic fixation using Galveston technique or iliac screw, posterior instrumentation: pedicle screw only, pedicle screw/wire, or hook/wire • Fusion levels (mean): NR from T2 or T3 to pelvis	2.9 y (2.0–5.2) (% NR)
Nectoux et al (2010) Case series CoE: IV	• $N = 28$ • Age (median): 16.5 y (12.5–27) • Male: 46.4%	• Nonambulatory spastic quadriplegic cerebral palsy with scoliosis, either single thoracolumbar lumbar curvatures ($n = 27$) or double major scoliosis ($n = 1$) • Progressive scoliosis of > 30 degrees	• One-stage posterior arthrodesis with Moseley rod fixed to the spine by Luque sublaminar wires using the Luque–Galveston fusion technique. • Fusion levels (mean): NR from T2 or T3 to sacrum	• < 1 y (100%) • 3.46 y (2–10) (57%)
Caird et al (2008) Case series CoE: IV	• $N = 40$ • Age (mean): 13.7 y (9–21) • Male: 60.0%	• Spastic quadriplegic cerebral palsy with or without ITB pumps; nonambulatory with limited use of the upper extremities and little or no speech. • Mean preoperative curve: 81 degrees	• Posterior spinal fusion with instrumentation to pelvis using a unit rod or Luque–Galveston construct; patients with large curves also underwent anterior spinal fusion by thoracoscopic thoracotomy, or retroperitoneal • Fusion levels (mean): NR	Follow-up period NR (% NR)
Tsirikos et al (2008) Case series CoE: IV	• $N = 287$ • Age (mean): 13.9 ± 3.3 y • Male: 46.7%	• Spastic or diplegic quadriplegia; no ambulatory function ($n = 249$), stand for assistive transfers ($n = 14$), or community ambulators ($n = 24$); severe mental retardation ($n = 227$), moderate mental retardation ($n = 26$), normal cognitive function ($n = 34$) • Scoliosis, idiopathic or collapsing neuromuscular	• Posterior spine arthrodesis ($n = 242$) or combined anteroposterior fusion ($n = 45$); unit rod instrumentation, Galveston pelvic fixation and sublaminar wires; freeze-dried granulated corticocancellous bone allograft with autogenous bone • Fusion levels (mean): NR from C7 to T3 into the pelvis	• < 2 y (100%) • 8.3 ± 3.0 y (84.0%) • For functional outcome survey: follow-up period NR (66.2%)
Vialle et al (2006) Case series CoE: IV	• $N = 110$ • Age (mean): 16.5 ± 3.5 y • Male: 50.9%	• Nonambulatory spastic quadriplegic cerebral palsy; neuromuscular scoliosis with pelvic obliquity	• Posterior spinal fusion with pelvic fixation; sacral screws and iliac extension connectors; pedicle, laminar, and transverse process hooks; autologous bone graft with or without ceramic substitute; one of the following approaches was used: ◦ Patients in knee-chest position with pelvic obliquity correction by	8.6 y (3–18) (% NR)

Table 1 (*Continued*)

Investigator (y) Study design CoE	Population[a]	Condition	Intervention	Follow-up (%)
			posterior vertebral instrumentation distraction, rotation, and compression ◦ Patients in prone position with asymmetric traction applied between a halo and high pelvic side lower extremity with pelvic correction done before posterior surgery • Fusion levels (mean): NR • Surgery performed before posterior fusion: anterior discectomy and fusion ($n = 50$), hip deformity surgery ($n = 55$)	
Sink et al (2003) Case series CoE: IV	• $N = 41$ • Age (mean): 15 y (9–36) • Male: 68.3%	• Spastic quadriplegic cerebral palsy and progressive spinal deformity (scoliosis and/or kyphosis)	• Posterior spinal fusion using Luque–Galveston instrumentation either alone ($n = 7$) or preceded by anterior release ($n = 34$) • Fusion levels (mean): NR	3.6 y (2–10) (% NR)
Tsirikos et al ("One-stage versus two-stage," 2003) Case series CoE: IV	• $N = 45$ • Age (mean): 14.8 y (9.6–21) • Male: 48.9%	• Spastic quadriplegic cerebral palsy; community or nonambulatory • Progressive neuromuscular scoliosis, associated with severe pelvic obliquity, trunk imbalance, and rigidity of the curve	• Anteroposterior spinal fusion in one stage ($n = 30$, from 1992–2000) or two stages ($n = 15$, from 1988 to 1991); unit rod instrumentation using Galveston technique; freeze-dried bone graft mixed with autogenous graft • Fusion levels (mean): NR from C7, T1, or T2 to the pelvis	3–3.4 y (% NR)
Comstock et al (1998) Case series CoE: IV	• $N = 100$ • Age (mean): 13.8 y (6–24) • Male: 45.6%	• Spastic quadriplegic cerebral palsy; severely retarded ($n = 67$); seizure disorders ($n = 64$); gastric feeding tubes ($n = 20$); wheelchair users ($n = 73$); ambulatory ($n = 6$) • Thoracolumbar, double major, thoracic, lumbar, or double thoracic scoliosis	• Posterior spinal instrumentation and fusion only ($n = 44$), posterior fusion with anterior release and fusion with ($n = 12$) or without instrumentation ($n = 20$), or anterior fusion and instrumentation ($n = 3$) • Fusion levels (mean): NR from T2 to T4 to pelvis	4 y (median) (2–14) (79% for complications, 60% for patient satisfaction)
Jevsevar and Karlin (1993) Case series CoE: IV	• $N = 44$ • Age (mean): 16 ± 4 y (11–23) • Male: NR	• Spastic quadriplegia; support-sitters (wheelchair necessary to maintain sitting position); institutionalized ($n = 44$); preoperative gastrostomy tube ($n = 4$) • Scoliosis, details NR	• Posterior fusion using Harrington or Luque spinal instrumentation ($n = 38$) or anterior spinal fusion (transthoracic, retroperitoneal, or combined approach with autogenous bone) followed by posterior fusion within 3 wk ($n = 6$) • Fusion levels (mean): NR from T4 to L5	• Follow-up period NR (% NR)

Abbreviations: CoE, class of evidence; GMFCS, gross motor function classification system; ITB, intrathecal baclofen; NR, not reported.
[a]Demographics reported for 79 patients with follow-up (Comstock et al, 1998) or for combined treatment groups (Keeler et al, 2010; Caird et al, 2008; Tsirikos et al, 2008; and Vialle et al, 2006).

Table 2 Characteristics of included studies evaluating potential predictive factors affecting outcomes following surgical correction for scoliosis in spastic quadriplegic patients

Investigator (y) Study design CoE	Population	Condition	Intervention	Follow-up (% followed)	Predictive factors evaluated	Outcomes evaluated
Bohtz et al (2011) Retrospective cohort CoE: III	• $N = 50$ • Age (mean): 15.1 y (8.8–33.2 y) • Male: 46.0%	• Tetra spastic cerebral palsy (GMFCS levels IV and V) • Progressive scoliosis of > 50 degrees	• Spinal fusion with segmental pedicle screw instrumentation in thoracic and lumbar spine • Fusion levels (mean): 13.9 motion segments (range, 7–19)	2 y (% NR)	• Demographic factors: none • Surgical factors: fusion levels (extending to sacropelvic region) • Other factors: none	• Complications • HRQOL (CPCHILD)
Vialle et al (2006) Retrospective cohort CoE: III	• $N = 110$ • Age (mean): 16.5 ± 3.5 y • Male: 50.9%	• Nonambulatory spastic quadriplegic cerebral palsy; neuromuscular scoliosis with pelvic obliquity	• Posterior spinal fusion with pelvic fixation; sacral screws and iliac extension connectors; pedicle, laminar, and transverse process hooks; autologous bone graft with or without ceramic substitute; one of the following approaches was used: ◦ Patients in knee-chest position with pelvic obliquity correction by posterior vertebral instrumentation distraction, rotation, and compression ◦ Patients in prone position with asymmetric traction applied between a halo and high pelvic side lower extremity with pelvic correction done before posterior surgery • Fusion levels (mean): NR • Surgery performed before posterior fusion: anterior discectomy and fusion ($n = 50$), hip deformity surgery ($n = 55$)	8.6 y (3–18) (% NR)	• Demographic factors: none • Surgical factors: augmentation with ceramic substitute • Other factors: none	• Pseudarthrosis • Instrument failure
Tsirikos et al ("Life expectancy," 2003) Prospective cohort CoE: III	• $N = 288$ • Age (mean): 13.9 ± 3.33 y • Male: 46.5%	• Spastic quadriplegia; living in family home ($n = 202$) or residential home ($n = 86$);	• Posterior spinal fusion ($n = 242$) or combined anteroposterior fusion; unit rod instrumentation;	• 0.5–1.0 y (% NR)	• Demographic factors: sex, age, level of ambulation, cognitive ability, degree	• Survival time after surgery

Table 2 (*Continued*)

Investigator (y) Study design CoE	Population	Condition	Intervention	Follow-up (% followed)	Predictive factors evaluated	Outcomes evaluated
		nonambulatory ($n = 250$), stand for assisted transfers ($n = 14$), ambulatory ($n = 24$); profound mental retardation ($n = 228$), moderate mental retardation ($n = 26$), cognitive abilities close to normal ($n = 34$) • Severe neuromuscular scoliosis	spinous process autograft with freeze-dried granulated cortico-cancellous allograft bone • Fusion levels: NR		of deformity • Surgical factors: intraoperative blood loss, surgical time, days in hospital, days in intensive care • Other factors: none	
Jevsevar and Karlin (1993)[a] Retrospective cohort CoE: III	• $N = 44$ • Age (mean): 16 ± 4 y (11–23 y) • Male: NR	• Spastic quadriplegia; support sitters (wheelchair necessary to maintain sitting position); institutionalized ($n = 44$); preoperative gastrostomy tube ($n = 4$) • Scoliosis, details NR	• Posterior fusion using Harrington or Luque spinal instrumentation ($n = 38$) or anterior spinal fusion (transthoracic, retroperitoneal, or combined approach with autogenous bone) followed by posterior fusion within 3 wk ($n = 6$) • Fusion levels (mean): NR from T4 to L5	• Follow-up period NR (% NR)	• Demographic factors: preoperative nutritional status (malnourished, $n = 20$ or nonmalnourished, $n = 24$) • Surgical factors: none • Other factors: none	• Blood loss • Duration of postoperative intubation, hospitalization • Complications

Abbreviations: CoE, class of evidence; CPCHILD, caregiver priorities and child health index of life with disabilities; GMFCS, gross motor function classification system; HRQOL, health-related quality of life; NR, not reported.

[a]Preoperative nutritional status classified as nonmalnourished (preoperative serum albumin level ≥ 35 g/L and total blood-lymphocyte count ≥ 1.5 g/L) or malnourished (preoperative serum albumin level < 35 g/L and total blood-lymphocyte count < 1.5 g/L) (Jevsevar and Karlin, 1993).

Table 3 Patient-reported outcomes in included studies of surgical correction for scoliosis in spastic quadriplegic patients

Investigator (y)	Satisfaction with surgery[a]
Bohtz et al (2011)[b]	Satisfied with outcome of procedure (parents or caregivers) Highly satisfied/satisfied: 91.7% Slightly dissatisfied: 5.6% Dissatisfied: 2.7% Would repeat procedure under same conditions (parents or caregivers) Definite yes: 91.7% Unsure: 8.3% Definite no: 0%
Tsirikos et al (2008)	Benefits of surgery offset the risks Satisfied (parents): 95.8% Satisfied (professional caregivers): 84.3%
Comstock et al (1998)	Satisfied with results of surgery Satisfied (parents or caregivers): 85%

Abbreviation: CPCHILD, caregiver priorities and child health index of life with disabilities.
[a]Survey instruments included CPCHILD (Bohtz et al, 2011), a nonvalidated survey assessing patients' functional improvement after surgery (Tsirikos et al, 2008), and a satisfaction with surgery survey (Comstock et al, 1998).
[b]The preoperative and postoperative surveys were both administered at the 2-year follow-up (Bohtz et al, 2011).

Benefits of Scoliosis Surgery (►Table 3, supplementary ►Table 1)

- Three studies reported on satisfaction with surgery. In two studies, 85 to 91.7% of parents or caregivers reported being satisfied with the outcome of the surgery.[13,14] In one study, 91.7% of the parents or caregivers reported that they would repeat the procedure under the same conditions.[13] In another study that asked if the benefits of the surgery offset the risks, 95.8% of parents and 84.3% of the caregivers reported being satisfied.[15]
- One study reported that the Caregiver Priorities and Child Health Index of Life with Disabilities (CPCHILD) scores were significantly better ($p < 0.0014$) at the 2-year follow-up compared with the preoperative scores.[13] (CPCHILD is a questionnaire that measures caregivers' perspective on health-related quality of life of child with cerebral palsy. Scores range from 0 to 100, with higher scores indicating worse quality of life.) However, the preoperative and postoperative surveys were both administered at the 2-year follow-up, which puts these results at increased risk of bias.

Adverse Effects of Scoliosis Surgery

Various types of complications were reported in included studies at follow-ups ranging from less than 1 year to 8.6 years in patients with varying degrees of disability. Patients may have experienced more than one complication (►Table 4, supplementary ►Table 1).

- Overall complication risk at last follow-up ranged from 10.9 to 70.9% in six studies (►Fig. 2).[13,14,16–19]
- Mortality risk for overall study periods ranged from 2.8 to 19% in five studies,[14,15,17–19] with the majority of patients dying from cardiac or respiratory problems. If the oldest study is excluded,[14] the mortality risk for the remaining studies was less than 5% (►Fig. 3).
- Across four studies, risk of respiratory/pulmonary complications ranged from 26.9 to 57.1%, with the most common being pneumonia, pneumothorax, or atelectasis.[16–18,20]
- Risk of infection ranged from 2.5 to 56.8% as reported in six studies.[15–17,19–21] The majority of infections were urinary tract or deep wound infections.
- Four studies reported the risk of a hardware-related complication ranging from 7.5 to 43.8%.[15,17,19,22] Prominent hardware, screw failure, or wire breakage/failure/pullout were most often reported.
- Reoperation risk ranged from 19.5 to 32.5% in three studies.[14,20,22]

Factors Affecting Patient Outcome after Scoliosis Surgery

Two preoperative factors were found to be associated with various poor outcomes following scoliosis surgery (►Table 5, supplementary ►Table 2).

- Degree of preoperative thoracic kyphosis. Increased degree of thoracic kyphosis resulted in a slight, but statistically significant, increased risk of death (calculated relative risk, 1.02; $p = 0.023$).[23]
- Preoperative nutritional status. Patients who were malnourished (preoperative serum albumin level < 35 g/L and total blood-lymphocyte count < 1.5 g/L) experienced a significantly higher rate of infection (95%, $p = 0.0001$), a longer period of postoperative endotracheal intubation (mean, 15.7 ± 10.7 hours; $p = 0.002$), and a longer duration of hospitalization (mean, 17 ± 6.7 days; $p = 0.002$) than those who were not malnourished (25%; mean, 7.2 ± 5.6 hours; mean, 13 ± 3.5 days; respectively).[21]

Clinical Guidelines

No clinical guidelines were found.

Table 4 Safety outcomes in included studies of surgical correction for scoliosis in spastic quadriplegic patients

Postoperative outcomes	Follow-up (y)	% (*n*/*N*)
Risk of any complication		
Bohtz et al (2011)	2	16 (8/50)
Keeler et al (2010)	2.9–3.3	46.2 (24/52)
Nectoux et al (2010)	< 1	57.1 (16/28)
	3.46	56.3 (9/16)
Vialle et al (2006)	8.6	10.9 (12/110)
Tsirikos et al ("One-stage versus two-stage," 2003)[a]	3–3.4	Major complications: 37.8 (17/45) Minor complications: 53.3 (24/45) Technical complications: 20.0 (9/45)
Comstock et al (1998)	Early postoperative (time period NR)	13 (13/100)
	4	70.9 (56/79)
Risk of mortality		
Nectoux et al (2010)	< 1	3.6 (1/28)
Tsirikos et al (2008)	For entire study period of 8.3 ± 3.0	2.8 (8/287)
	Intraoperative	1.0 (3/287)
	< 2	1.7 (5/287)
	8.3 ± 3.0	0 (0/241)
Vialle et al (2006)	8.6	4.5 (5/110)
Tsirikos et al ("One-stage versus two-stage," 2003)	3–3.4	4.4 (2/45)
Comstock et al (1998)	For entire study period	19 (19/100)
	Immediate postoperative	1.0 (1/100)
	Early postoperative (time period NR)	3 (3/100)
	4	11.4 (9/79)
	Follow-up period NR	% NR (6 deaths)
Respiratory/pulmonary complications[b]		
Keeler et al (2010)	2.9–3.3	26.9 (14/52)
Nectoux et al (2010)	< 1	57.1 (16/28)
Caird et al (2008)	Follow-up period NR	42.5 (17/40)
Tsirikos et al ("One-stage versus two-stage," 2003)	3–3.4	31.1 (14/45)
Cardiovascular[c]		
Keeler et al (2010)	2.9–3.3	15.4 (8/52)
Infections[d]		
Keeler et al (2010)	2.9–3.3	21.2 (11/52)
Nectoux et al (2010)	< 1 y	3.6 (1/28)
Caird et al (2008)	Follow-up period NR	22.5 (9/40)
Tsirikos et al (2008)	< 2	4.2 (12/287)
	8.3 ± 3.0	2.5 (6/241)
Vialle et al (2006)	8.6	4.5 (5/110)
Jevsevar and Karlin (1993)	Follow-up period NR	56.8 (25/44)
Neurologic[e]		
Keeler et al (2010)	2.9–3.3	5.8 (3/52)
Hardware related[f]		
Nectoux et al (2010)	3.46	43.8 (7/16)

(*Continued*)

Table 4 (*Continued*)

Postoperative outcomes	Follow-up (y)	% (*n*/*N*)
Tsirikos et al (2008)	8.3 ± 3.0	7.5 (18/241)
Vialle et al (2006)	8.6	9.1 (10/110)
Sink et al (2003)	3.6	39.0 (16/41)
Reoperation[g]		
Caird et al (2008)	Follow-up period NR	32.5 (13/40)
Sink et al (2003)	3.6	19.5 (8/41)
Comstock et al (1998)	Follow-up period NR	21 (*n*/*N* NR)

Abbreviations: CSF, cerebrospinal fluid; NR, not reported; NSAIDs, nonsteroidal anti-inflammatory drugs.
Note: overall complication mortality risk reported for entire study period or for all follow-up periods if available.
[a]Major complications include coagulopathy, infection, drug reaction (NSAIDs), pancreatitis, pneumonia, prolonged gastric tube or ventilator, respiratory failure, pleural effusion, and superior mesenteric artery syndrome; minor complications include atelectasis, infection (bowel, central line, superficial wound, urinary tract), diabetes, donor bone graft reaction, drug reaction (Dilantin), gastritis, hematuria, hemothorax, heterotopic ossification, ileus, constipation, persistent fever, pneumothorax, skin breakdown, wound hematoma; technical complications include sublaminar wires cutout, severe skin breakdown, painful protruding spinal instrumentation, persistent sacroiliac inflammation, perforation of the ileum (Tsirikos et al "One-stage versus two-stage," 2003).
[b]Respiratory/pulmonary complications include pneumonia, pneumothorax, segmentary atelectasis, and segmental pneumopathies.
[c]Cardiovascular complications include coagulopathy with or without hypotension, hypotension.
[d]Infections include superficial or deep wound, urinary tract, intravenous central line access, fevers of unknown origin, segmental pneumopathies.
[e]Neurologic complications include intraoperative spinal cord monitoring event, postoperative seizures.
[f]Hardware-related complications include sublaminar wire failure, protrusion of instrumentation, sacral or iliac screw failure, dual-rods connector, wire/hook, pullout, and rod breakage.
[g]Reasons for reoperation include wound infection, persistent CSF leak, baclofen pump, posterior instrumentation, or NR.

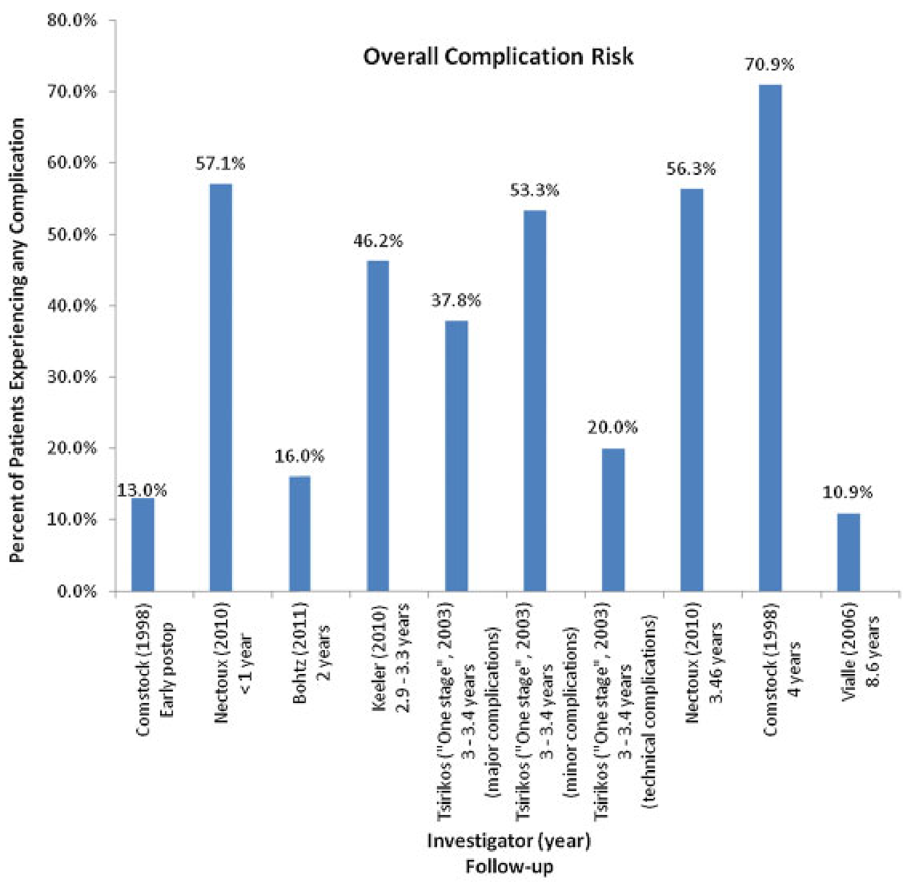


Fig. 2 Overall complication risk following scoliosis surgery in spastic quadriplegic patients.

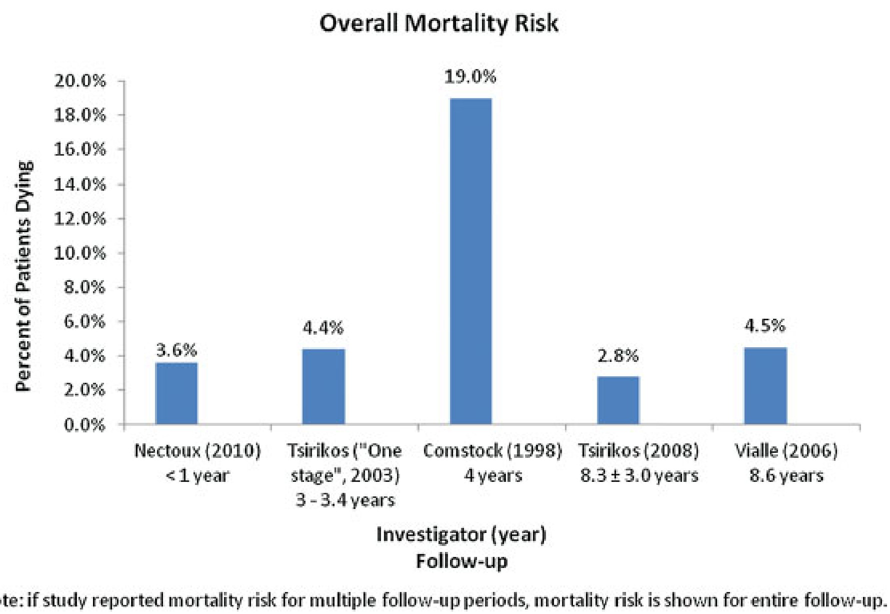


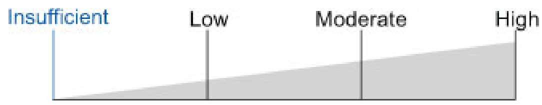
Fig. 3 Overall mortality risk following scoliosis surgery in spastic quadriplegic patients.

Table 5 Summary of demographic and surgical factors evaluated as predictive factors for outcome following scoliosis surgery in spastic quadriplegic patients

	Multivariate analysis to control for confounders	**No multivariate analysis**		
	Tsirikos et al ("Life expectancy, 2003)	**Bohtz et al (2011)**	**Vialle et al (2006)**	**Jevsevar and Karlin (1993)**
Outcome evaluated	**Risk of mortality after surgery**	**Risk of complications/ changes in HRQOL**	**Pseudarthrosis/ instrument failure**	**Increased blood loss Longer duration of postoperative intubation/ hospitalization Risk of complications**
Demographic factors				
Age, sex, cognitive ability	NS			
Level of ambulation	NS			
Degree of preoperative thoracic kyphosis	↑			
Degree of preoperative scoliosis	NS			
Preoperative nutritional status[a]				NS (blood loss) ↑ (duration of intubation and hospitalization) ↑ (infection)
Surgical factors				
Fusion of sacropelvis		NS (complications and HRQOL)		
Augmentation with ceramic substitute			NS (pseudarthrosis and instrument failure)	
Intraoperative blood loss, surgical time	NS			
Days in hospital	NS			
Days in intensive care	↑			

Abbreviations: HRQOL, health-related quality of life; NS, not significant; ↑, increased risk of outcome.
Note: Empty cell indicates that factor was not evaluated.
[a]Preoperative nutritional status classified as nonmalnourished (preoperative serum albumin level ≥ 35 g/L and total blood-lymphocyte count ≥ 1.5 g/L) or malnourished (preoperative serum albumin level < 35 g/L and total blood-lymphocyte count < 1.5 g/L) (Jevsevar and Karlin, 1993).

Table 6 Evidence summary

Baseline quality: High = majority of articles level I/II; low = majority of articles level III/IV *Upgrade:* Large magnitude of effect (1 or 2 classes); dose–response gradient (1 class) *Downgrade:* Inconsistency of results (1 or 2 classes); indirectness of evidence (1 or 2 classes); imprecision of effect estimates (1 or 2 classes)		
Outcome	**Strength of evidence**	**Conclusions/comments**
What are the reported benefits of surgical correction of scoliosis in children with spastic quadriplegia?		
Patient satisfaction with surgery		Three case series reported satisfaction with surgery. Parent or caregiver satisfaction with surgery ranged from 85 to 91.7% in two studies. In one study, 91.7% of the parents or caregivers reported that they would repeat the procedure under the same conditions and another study reported that 95.8% of parents reported that the benefits of the surgery offset the risks. CPCHILD scores were significantly better ($p < 0.0014$) at a 2-y follow-up compared with the preoperative scores, but the preoperative and postoperative surveys were both administered at the 2-y follow-up.
What are the short- and long-term adverse effects of surgical correction of scoliosis in children with spastic quadriplegia?		
Adverse events		Overall, the evidence of adverse effects of scoliosis surgery is insufficient. There was wide variation in overall complication risk (10.9–70.9%) and infection risk (2.5–56.8%) in six studies, respiratory/pulmonary complications (26.9–57.1%) and hardware-related complications (7.5–43.8%) in four studies, and reoperation risk (19.5–32.5%) in three studies. The risk of mortality ranged from 2.8 to 19% in five studies.
Are there any factors affecting patient outcome after surgical correction of scoliosis in children with spastic quadriplegia?		
Demographic and surgical factors affecting patient outcome		Overall, the evidence that factors predict patient outcome after scoliosis surgery is insufficient. Four studies examined predictive factors for different outcomes. Only one study performed a multivariate analysis to control for confounders: this study found that increased degree of thoracic kyphosis and number of days in the ICU increased the risk of dying. Another study found that patients who were malnourished experienced a higher risk of infection and longer duration of intubation and hospitalization. And two studies found no significant predictive factors for pseudarthrosis, instrument failure, complications, or HRQOL.

Abbreviations: CoE, class of evidence; CPDHILD, caregiver priorities and child health index of life with disabilities; HRQOL, health-related quality of life; ICU, intensive care unit.
Notes: All AHRQ "required" and "additional" domains[a] are assessed. Only those that influence the baseline grade are listed in table.
Baseline strength: Risk of bias (including control of confounding) is accounted for in the individual article evaluations. High = majority of articles level I/II; low = majority of articles level III/IV.
Downgrade: Inconsistency[b] of results (1 or 2); indirectness of evidence (1 or 2); imprecision of effect estimates (1 or 2); subgroup analyses not stated a priori and no test for interaction (2).
Upgrade: Large magnitude of effect (1 or 2); dose–response gradient (1).
[a]Required domains: risk of bias, consistency, directness, precision. Plausible confounding that would decrease observed effect is accounted for in our baseline risk of bias assessment through individual article evaluation. Additional domains: dose–response, strength of association, publication bias.
[b]Single study = "consistency unknown."

Evidence Summary

Overall, the limited evidence available suggests that parents (or caregivers) of patients with spastic quadriplegia are satisfied with the outcomes of surgery. However, studies in this area are limited and of poor quality so that the strength of the evidence is low. Studies that have examined the adverse effects of scoliosis surgery are highly variable with widely differing reported incidences and study designs thus preventing any definitive conclusions. Similarly, there is limited evidence of preoperative factors that can predict patient outcome after scoliosis surgery. In all areas, there is an undoubted need for further well-designed prospective studies (►**Table 6**).

Fig. 4 Anteroposterior radiograph: preoperative.

Fig. 5 Lateral radiograph: preoperative.

Illustrative Case

A 16-year-old female patient with spastic quadriplegia and wheelchair dependency presented with significant spinal pain. She had been reviewed 4 years previously at which stage she had been comfortable in her chair. Over time, she had developed more truncal shift and worsening spinal deformity. Despite multiple wheelchair adaptations, her curve had progressed and she was able to verbally complain of increasing pain and difficulty sitting (►**Figs. 4** and **5**). Following preoperative cardiac and respiratory assessments, informed consent was obtained and the patient underwent a posterior instrumented scoliosis correction (►**Figs. 6** and **7**). No posterior osteotomies were performed. As there was no major pelvic obliquity, the instrumentation was performed to L5 rather than the pelvis. Surgery was routine, with an operative time of 176 minutes, a blood loss of 16% of estimated blood volume (11.5 mL/kg) and a salvage blood return of 8% of estimated blood volume. Single dose antibiotics at induction and three postoperative doses were administered. The patient was extubated immediately postoperatively and observed overnight on pediatric intensive care. She made an uneventful recovery and was discharged at day 6 postoperatively. One month later, she was reviewed and a small superficial wound collection was noted. She was returned to theater and a superficial wound collection was debrided. Wound swabs subsequently cultured *staphylococcus aureus* and diphtheroids and she received targeted oral antibiotics for 6 weeks as an outpatient with optimal wound healing. At 2 years of follow-up, there are no implant-related problems, no spinal pain, and no signs or symptoms of infection. She has gained weight and has no respiratory complications. The child and caregivers are extremely pleased with the final result.

Discussion

This systematic review is limited by the following:

- All studies were CoE III or IV and the majority of studies had small sample sizes.
- The majority of studies considered for inclusion reported outcomes without specifying subtypes of CP. Spastic quadriplegia is the most severe form of CP and is associated with the highest incidence of scoliosis. It is likely that outcomes for scoliosis surgery in spastic quadriplegia differ significantly from those for other subtypes. To best inform

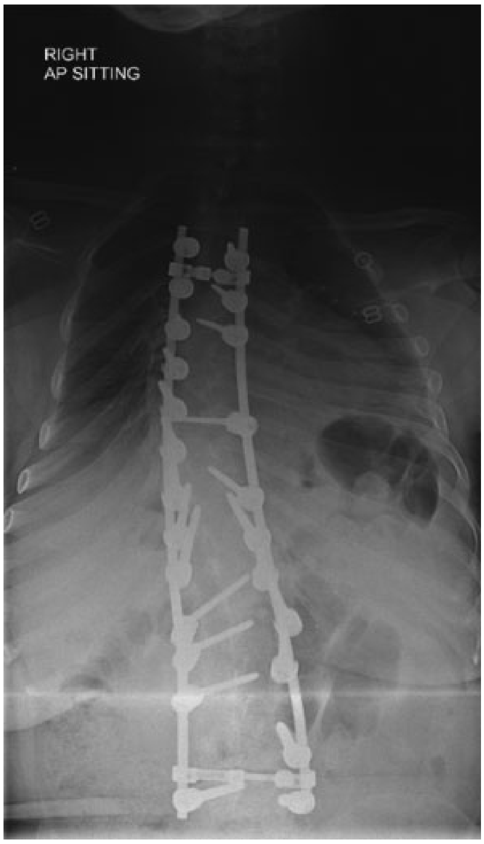


Fig. 6 Anteroposterior radiograph: postoperative.

surgical practice, it is important that CP subtypes are accurately delineated in future studies.

- Surgical outcomes were poorly delineated with limited or no use of validated outcome instruments.
- Those studies that reported adverse events had highly variable design and identified a wide range of risks for various complications.
- Only four studies were identified that examined predictive factors for outcome after scoliosis surgery, with only one of them conducting a multivariate analysis that controlled for potential confounders. Further studies that comprehensively characterize patients preoperatively control appropriately for confounding factors are required to establish optimal treatment based on individual patient factors.

Given that the overall strength of evidence was insufficient for all studies, firm conclusions regarding the benefits of scoliosis surgery and various risk factors are not possible. It is therefore difficult to make any definitive recommendations regarding the preoperative assessment and appropriate selection of patients for surgery. Generally, preoperative evaluation includes respiratory, cardiac, and anesthetic assessments with the aim to determine the patient's fitness to undergo surgery. Nutritional disorders are addressed by the gastroenterology and dietetic teams. The assessments are subsequently reviewed and discussed in a multidisciplinary team setting, before the final treatment decision is made.

Demonstrating robust, clinically meaningful outcomes for this group of patients remains a significant challenge. Future studies should employ both objective and subjective measures of satisfaction and functional status to fully appreciate the impact of scoliosis surgery.

Fig. 7 Lateral radiograph: postoperative.

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Disclosures
None

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Editorial Perspective

The EBSJ reviewers congratulate the authors on selection of this interesting topic and a thorough assessment of the numerically large (>200 peer-reviewed studies) but qualitatively and relatively poor level of evidence. It is quite apparent that the current level of evidence basis is insufficient to clearly guide us in selecting the optimal treatment for this emotionally very challenging care dilemma affecting this frequently very compromised patient population. A priori, this article leaves no doubt that the very major surgery necessary for the management of the neuromuscular scoliosis of spastic cerebral palsy (CP) patients is fraught with substantial morbidity and mortality. The other main insight is that for a majority of these patients, the patient-reported outcomes can only be determined by third-party observers—care providers, parents, social workers, etc.—raising the ethical question of who should be in charge of the decision-making process for very involved spastic CP children and how to actually rate outcomes. Are patient-reported outcomes determined by third-party observers to be given the same level of validity as those reported by patients? How are such surrogate outcomes measures different from surgeon-derived patient outcomes? What are the factors that lead third parties to score patients as successes or as failures? This subject area deserves a lot more attention.

In light of the large number of studies that were reviewed, one reviewer suggested use of a Jadad scale or an Oxford scoring system to quantify the quality of the available literature more.[1] EBSJ felt that by adhering to the present-day Evidence base Pyramid the authors sufficiently tiered the available studies to help readers find a meaningful weighting of studies.

Another clear finding worth highlighting is the potential need for improvement in the areas of preoperative patient preparation and risk optimization. Specifically, the assessment of the preoperative nutritional status is traditionally and commonly overlooked in spine surgery, but based on the findings of Legg et al, there appears to be clear reason to routinely include nutritional parameters in preoperative decision-making. To look for ways to preoperatively improve this variable beyond a yet-to-be established threshold before going for major reconstruction surgery would appear to be a promising next step in minimizing perioperative infection risks for patients.

Another important practical finding of this systematic review was the impact of kyphosis on complications. Based on the authors' findings, it appears worthwhile to focus more on kyphosis as a dependent variable in decision-making for surgery and case severity determination. EBSJ believes that Dr. Legg and the co-authors have contributed very meaningful insights to the spine community and wishes to thank them for their efforts.

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