Scoliosis in the Child With Cerebral Palsy

Abstract
Scoliosis is common in children with cerebral palsy. The incidence and curve pattern depend on the degree of neurologic involvement. These children carry a higher risk of complications because of the increased presence of associated medical comorbidities. Accordingly, a careful preoperative evaluation is required that should involve assessment of the patient’s pulmonary, nutritional, gastrointestinal, and neurologic systems as well as a thorough evaluation of the spine and musculoskeletal system. Children with progressive curves >40° to 50° are candidates for spinal fusion, especially when the deformity interferes with sitting or is unresponsive to bracing. The goal of surgery is to obtain a stable, balanced, and painless spinal fusion. Although posterior spinal fusion with multisegmental fixation is the most common technique, others, such as anterior release and/or fusion or combined procedures, are now considered. In patients with significant pelvic obliquity or who are at risk of developing pelvic obliquity, instrumentation should extend to the pelvis, particularly in the nonambulatory child.

Incidence
Scoliosis, defined as a Cobb angle >10°, is quite common in children with CP, with an overall incidence of approximately 20%. This incidence varies with the extent and severity of the neurologic involvement. Children with CP who are nonambulatory and have total body involvement have an incidence of scoliosis of 62%6 for bedridden children, the incidence approaches 100%.2

Deformity Pattern and Association With Hip Dislocation
Spinal deformities are varied and common in patients with CP. Scoliosis, kyphosis, lordosis, and pelvic obliquity can occur as isolated deformities or in combination. Unlike idiopathic scoliosis, the classic curve pattern in CP is a long, C-shaped curve (Figure 1, A) that is often kyphoscoliotic but sometimes lordo-
scoliotic (Figure 1, B). The curve may have a left convexity, which is rare in idiopathic scoliosis. In children with lesser degrees of involvement (e.g., children with hemiplegic or ambulatory diplegic CP), the pattern of deformity may resemble that in adolescent idiopathic scoliosis. Curve progression is typically gradual but can be rapid with the onset of puberty, deteriorating neurologic function, shunt malfunction, or prolonged time in a wheelchair.

The association of hip dislocation with scoliosis is not fully understood; however, if a child is sitting or standing with a dislocated hip, the resultant pelvic obliquity will clearly produce a compensatory spinal curve. The corollary has also been described: scoliosis, especially in the lumbar spine, can produce a fixed pelvic obliquity that may result in adduction of the hip on the concave side (and abduction on the other). This “covert windswept deformity” may put the adducted hip at risk for dislocation. Cooke et al demonstrated that the development of scoliosis may put some patients at risk of dislocation. However, whether this is a causal relationship is not clear. In contrast, other authors did not report a correlation between pelvic obliquity and hip subluxation but thought that hip subluxation correlated more strongly with the degree of hip adduction. Although pelvic obliquity can be corrected by spinal surgery, surgery on the hip usually has little effect on pelvic obliquity associated with scoliosis.

Etiology

Muscle weakness and truncal imbalance have been implicated as the primary etiology for the onset of scoliosis in children with CP and other neuromuscular disorders; however, there is little evidence in the literature to support this. The incidence and severity of scoliosis appear to be related to the degree of involvement. Still unknown is whether this is directly related to the primary cerebral injury or to secondary impairments, such as muscle weakness, spasticity, poor balance, or a nonambulatory status.

Several authors have reported the development of spinal deformities such as scoliosis, severe lumbar lordosis, and spinal stenosis after selective dorsal rhizotomy. Turi and Kalen reported 28 spinal deformities in 19 of 43 patients following dorsal rhizotomy associated with wide laminectomies. Deformities included 15 instances of scoliosis, 7 cases of lumbar hyperlordosis, 5 patients with thoracic hyperkyphosis, and one instance of L4-S spondylolisthesis. Conversely, Spiegel et al did not demonstrate significant spinal deformity at a mean radiographic follow-up of 4.2 years after rhizotomy, although 13 of 79 patients had mild curves (mean, 16°). In general, when carefully performed through minilaminectomies, rhizotomies uncommonly result in significant scoliosis.

Natural History

The natural history of scoliosis in patients with CP has been well documented. Larger curves are associated with an increase in hip and pelvic deformity, but the association with a decline in functional activities is less clear. Majd et al found that the curve progressed faster (4.4°/yr) in a group of 10 patients with a decline in function than in a functionally stable group (3.0°/yr). They also found that among patients who developed decubitus ulcers with prolonged sitting, the average curve was much larger. They concluded that there was a correlation between the size of the deformity and the decline in functional activities. Kalen et al evaluated 56 institutionalized adult patients with CP (mean age, 38 years), 14 of whom...
who had curves >45° [mean, 77°] and 42 who had curves <45° [mean, 16°]. Patients in the group with the larger curves had a higher incidence of orthopaedic deformities involving the hip and pelvis but no difference in the number of decubiti, level of function, oxygen saturation, or pulse rate.

In patients with CP, as in those with idiopathic scoliosis, the larger the curve, the more likely it is to progress. Thometz and Simon found that the rate of curve progression in skeletally mature patients with CP was 0.8°/yr when the curve was <50° and 1.4°/yr when the curve was >50°. Saito et al showed that 85% of patients who had a spinal curve >40° by age 15 years progressed to 60°, whereas only 13% of those with a curve <40° by age 15 progressed to 60°.

**Assessment**

The medical evaluation of a child with CP should be extensive and requires a multidisciplinary approach. The clinical history is important; key elements include the child’s perinatal history, growth, and development as well as previous medical treatment. Evidence of neurologic progression, normal early developmental milestones, or a positive family history should raise concerns about the diagnosis.

Assessment should include a general physical examination and a complete musculoskeletal evaluation. Important parameters to be measured are the patient’s nutritional status, respiratory function, sitting/standing posture, gait, and upper extremity functional capacity. Assessment of the patient’s functional status and related changes is critical, including ambulatory ability, ability to swallow, communication skills, and acuity of hearing and vision. The musculoskeletal examination should include evaluation of range of motion, tone, and motor strength. Often, children with tight hamstrings will have decreased lumbar lordosis, especially when sitting. Conversely, contractures of the hip flexors may result in excess lumbar lordosis, particularly evident in standing or supine radiographs. Abduction and adduction contractures can influence the lumbar coronal curve and result in pelvic obliquity. The spinal examination includes evaluation of the degree and flexibility of the spinal deformity, assessment of spinal balance (ie, shoulder height), and evaluation of pelvic obliquity.

Radiographs are required to evaluate scoliosis fully in children with CP, but no consensus has been reached as to the frequency and technique [sitting, standing, or supine]. The technique should be consistent for each patient and determined by his or her functional ability. Standing erect or sitting radiographs should be made when possible. Radiographs can be obtained with the patient in a wheelchair or in a chair with armrests provided there is minimal [noncorrective] assistance. Patient position and bracing, when applicable, are important considerations in comparing radiographic measurements. Mild curves may be positional and can vary in degree from one evaluation to the next. Bending or traction radiographs may help assess stiffness. If the patient is unable to cooperate enough to actively bend, then traction, push-pull radiographs or fulcrum-bending radiographs are very useful for preoperative assessment and planning. Radiographs should be evaluated for spinal rotation, rib deformity, and wedging, which may help define the character of the curve; these elements are rarely found in positional curves.

The radiographic assessment should also include an evaluation for spondylolisthesis, especially in children with spastic diplegia. The incidence of spondylolisthesis in children with spastic diplegia is between 4% and 21%.14

Preoperative magnetic resonance imaging [MRI] is not routinely performed in children with CP who require surgery for scoliosis. The incidence of intraspinal pathology is unknown in children with CP but is thought to be uncommon. Indications for MRI screening include recent major alterations in neurologic function or rapid curve progression.

**Nonsurgical Management**

Numerous modalities—including physical therapy, electrical stimulation, botulinum toxin injections, bracing, and surgery—have been suggested for the treatment of scoliosis in children with CP. Electrical stimulation and physical therapy are ineffective, but a study of 12 patients demonstrated short-term benefit of botulinum toxin injections for neuromuscular scoliosis.15

Molded wheelchair inserts are widely used in an attempt to improve sitting balance. Clinical evaluation, as well as radiographs while the patient is sitting in the chair, can be used to demonstrate improvement in position and curve magnitude. Unclear, however, is whether molded inserts have any role in preventing or slowing curve progression.

Although brace wear is not generally thought to be effective for the treatment of scoliosis in children with CP, many clinicians use bracing to improve sitting balance or to slow the rate of progression in skeletally immature children. Bracing does not seem to negatively affect pulmonary function, and it may decrease the work of breathing by improved positioning.3 Olafsson et al found that curve progression was prevented in only 23 of 83 patients with neuromuscular disorders [38 with CP] treated with bracing. Failure to prevent progression was attributed to discontinuation of brace wear in 41 of the 60 patients in whom the curve progressed. The au-
Surgical Management

Indications for Surgery

The primary surgical goal for the patient with CP is to achieve a solid spinal fusion that will result in a corrected, well-balanced spine with a level pelvis. Several fundamental prerequisites should be established before considering surgery: scoliosis >40° to 50° that is either progressive or interfering with sitting; age >10 years; adequate hip range of motion to allow for proper seating postoperatively; and stable nutritional and medical status.

The decision to perform surgery depends not only on the degree of curve and evidence of progression but also on a careful assessment of the patient’s and family’s goals and evaluation of the risks and perceived benefits. Questions related to the patient’s awareness, ability to interact, tolerance to pain, and ability to express pain should be assessed, although these factors are difficult to quantify. Several attempts have been made to evaluate the benefits of surgery with nonrandomized studies of an intervention group, with or without controls. Surgery should be considered only when the expected benefits outweigh the negative consequences of the natural history and risks of surgery.

Cassidy et al compared 17 patients who had undergone a spinal fusion (mean curve, 35°) to 20 patients who did not undergo fusion (mean curve, 76°). They found no significant difference in the degree of pain, need for pulmonary medication, decubiti, function, or time required for daily care. Askin et al evaluated 20 patients before and after spinal fusion for neuromuscular scoliosis and found no functional gains at 1 year. Conversely, Larsson et al evaluated 94 patients with paralytic scoliosis 1 year after surgery and found improvements in curve magnitude, sitting balance, weight distribution, and skin discoloration.

According to most subjective surveys, the majority of health care workers believe that the patients who underwent fusions were more comfortable and were satisfied with the results of the surgery. Jevsevar and Karlin demonstrated an increased postoperative infection rate, longer length of intubation, and longer hospital stays in patients with CP who had preoperative serum albumin measurement <35 g/L and total blood lymphocyte count <1.5 g/L. Although some studies have failed to show a correlation between preoperative nutritional status and postoperative complications, most surgeons still consider a preoperative nutritional assessment to be important. A simple screening can be done by weighing the patient; if weight is less than the fifth percentile compared with normative weight for a given age, the patient may carry a higher risk of postoperative complications and may require additional evaluation. Nutritional supplementation should be performed parenterally, if possible, for all patients considered to be at risk.

Pulmonary Function

Formal pulmonary functional capacity testing cannot be easily performed in these patients, but respiratory history, clinical examination, and chest radiographs are used to assess pulmonary status. Poor preoperative pulmonary function is associated with prolonged respiratory support after surgery. Thus, awareness of this situation is critical for preoperative planning and discussions with the caregivers.

Nutritional Status

Poor nutritional status has been associated with higher complication rates. Jevsevar and Karlin demonstrated an increased postoperative infection rate, longer length of intubation, and longer hospital stays in patients with CP who had preoperative serum albumin measurement <35 g/L and total blood lymphocyte count <1.5 g/L. Although some studies have failed to show a correlation between preoperative nutritional status and postoperative complications, most surgeons still consider a preoperative nutritional assessment to be important. A simple screening can be done by weighing the patient; if weight is less than the fifth percentile compared with normative weight for a given age, the patient may carry a higher risk of postoperative complications and may require additional evaluation. Nutritional supplementation should be performed parenterally, if possible, for all patients considered to be at risk.

Gastrointestinal Evaluation

Preoperative assessment and treatment of gastroesophageal reflux are important in the prevention of aspiration pneumonia. Many of these children are at risk for aspiration even without surgery because of the...
combination of sedation and supine positioning. If swallowing status has not been determined preoperatively, such a study should be performed to assess the risk of aspiration.

**Neurologic Function**

Patients with CP often have a seizure disorder for which they take antiepileptic medication. The seizure disorder should be well controlled before surgery.

Some antiseizure medications may interfere with coagulation and thus may increase perioperative blood loss. Valproic acid, for example, has been postulated to decrease von Willebrand’s factor. Although studies suggest that valproic acid has little clinical effect on intraoperative blood loss, we feel it is still important to obtain a bleeding time on patients taking valproic acid because routine screening examinations (prothrombin time and [activated] partial thromboplastin time) will not be elevated.

**Technical Considerations**

For internal fixation, multisegmental fixation (usually sublaminar wires) has been used in combination with either Luque rods or a unit rod.26-31 Multisegmental fixation is important because corrective forces can be distributed at all levels in these patients, who are frequently osteoporotic. Other methods of multilevel fixation have also been used with comparable effectiveness, but they may be more costly.32 Newer three-column (pedicle screw) fixation techniques may provide better correction and possibly decrease the need for some anterior procedures. Fixation proximally should extend to the upper thoracic spine (T1 or T2); if this is not done, progressive upper thoracic kyphosis may occur. Fixation level distally depends on the curve pattern but commonly extends to L4 or L5. When pelvic obliquity is >15°, especially if the patient is nonambulatory, fixation should extend to the pelvis.

Several authors have reported the technical results of spinal fusion for treatment of scoliosis in children with neuromuscular disorders.31,36-31 Curve correction varied from 45% to 76%; pelvic obliquity improvement was between 49% and 88%. Curve progression after fusion is typically small (<10°) and may be related to the magnitude of the residual curve and remaining growth of the anterior spine. Typically, no postoperative bracing is needed, but an orthosis may be used when fixation is tenuous, bone quality is poor, or patients are unable to comply with restrictions. Typical postoperative activity restrictions may include limitations on lifting, bending, manual wheelchair propulsion, and/or self-transfers. These limitations depend on bone quality and whether fixation is extended to the pelvis.

Anterior release and fusion are often performed for larger, more rigid curves (ie, those that do not bend out to <60°),16 as well as in children who have not reached skeletal maturity. Anterior spinal procedures have a higher complication rate in children with neuromuscular disorders than in those with idiopathic scoliosis.33 Performing both anterior and posterior procedures on the same day (single-stage spinal fusion) resulted in a lower complication rate, decreased blood loss, better nutritional status, and shorter length of hospital stay.34 Patient morbidity and surgical time of the anterior portion of the procedure may decrease with use of thoracoscopic techniques. In some centers, anterior releases are being performed less frequently; the indications for anterior release may be changing, especially as more mechanically effective posterior constructs are being implemented.

Some surgeons believe that children who are skeletally immature should have an anterior spinal fusion to prevent progression that can occur even after successful posterior spinal fusion. This continued progression is thought to be the result of continued anterior spinal growth and has been termed the crankshaft effect. Comstock et al21 found a 30% rate of late progression, and >75% of their patients who progressed after surgery were skeletally immature. Conversely, other investigators found minimal progression in skeletally immature children who had a posterior spinal fusion alone with a unit rod.32 Factors other than skeletal maturity may play a role in curve progression after fusion, such as type of fixation, degree of correction, and level of fusion.

The indication for fusion to the pelvis is still under debate. Whitaker et al35 retrospectively reviewed 23 patients with neuromuscular scoliosis after posterior spinal fusion (with pedicle screws) ending in the lumbar spine. They found that when pelvic obliquity was <15° (mean, 6°) before surgery there was no deterioration or pseudarthrosis at an average follow-up of 61 months. In a study of patients with severe scoliosis (mean, 70°) and severe pelvic obliquity (mean, 27°), fusion to the pelvis corrected the scoliosis and pelvic obliquity to 24° and 5°, respectively; there was a 9% pseudarthrosis rate.32

The Galveston method of pelvic fixation31 has been used with good success (Figure 2). The posterior column of the pelvis usually has good bone quality. The stable fixation that it offers also facilitates better correction of pelvic obliquity and provides a fixed point over which to balance the head and spine. The Galveston unit rod is a precontoured double rod that combines a U-shaped bend proximally with two distal bends to accommodate insertion into the iliac wing of the pelvis. The instrumentation combines the strength of the Luque and Galveston techniques with the convenience of prebent rods. The design also inhibits migration of the rod, which was a problem with Luque rods.

Several additional methods to supplement fixation to the pelvis have been shown to provide satisfac-
Scoliosis correction, including bilateral sacral screws, iliosacral screws [Figure 3], spinopelvic transiliac fixation,36 and an S-contoured rod that wraps over the sacral ala (the Dunn-McCarthy technique). The advantages of S-contoured rods are that they do not cross the sacroiliac joint, take less time to insert, and result in less loosening.37 Arlet et al38 described the “MW” configuration, combining iliosacral screws with iliac screws for a very strong fixation posterior spinal fusion. This fixation gives an “M” appearance on the pelvic radiographs and a “W” appearance in the axial plane.

Halo Traction
Preoperative halo-gravity traction for patients with neuromuscular scoliosis is an option when the curve is severe and rigid. The efficacy of this practice was questioned by Flierl and Carstens39 after they evaluated 32 patients with neuromuscular scoliosis. Patients with diagnoses of myelomeningocele and poliomyelitis had 12.8% and 16.9% correction, respectively; however, patients with CP had minimal improvement from traction. Other investigators have also concluded that traction between staged anterior-posterior surgeries was not useful for curve correction in patients with CP.

Neuromonitoring
Somatosensory spinal-evoked potentials (SSEPs) and the Stagnara wake-up test have become the gold standard for intraoperative monitoring of scoliosis surgery. In the presence of a neuromuscular disease, SSEPs are often abnormal or may be absent. Noordeen et al40 felt that SSEPs were useful; however, a higher incidence of false-negative results was detected than in patients with idiopathic scoliosis. Langeloo et al41 found that transcranial electrical motor-evoked potential monitoring worked well in patients with neuromuscular disease.

Although there is no consensus regarding the need for and degree of monitoring in children with CP, we use SSEP monitoring. If the SSEP values fall beyond the predetermined levels (50% drop in amplitude), then the surgery is stopped and the anesthesia is optimized. If the amplitude does not improve, alterations are made in the instrumentation or the curve correction is reduced. A wake-up test is performed if necessary. We feel that SSEP evaluation is important even in nonambulatory patients to monitor upper extremity intraoperative positioning. If SSEP values fall, then the upper extremity should be repositioned.

**Postoperative Complications**
Complications in patients with CP undergoing corrective surgery for scoliosis are common and can at times be devastating. Complications include implant or bone fixation failure; pulmonary, gastrointestinal, and neurologic complications; wound healing problems; and death. Complication rates have been reported to range from 44% to 80%, with a perioperative death rate of 0% to 7%.27 Recently reported complication rates have been much lower.17 In a study of 111 patients, Sarwahi et al33 found that 21.6% had major and 22.5% had minor complications; pulmonary insufficiency was the most frequent complication.
Implant Failure

Implant failure does not always necessitate revision surgery. In a study of 74 patients undergoing spinal fusion and Luque rod instrumentation, Broom et al.\(^2^7\) described six cases of broken rods, four of which were asymptomatic and required no additional treatment. They felt that the larger 1/4-in (6.4-mm) rods were less likely to fail than the 3/16-in (4.8-mm) rods.

Another complication is penetration of the pelvic limb of the unit rod into the pelvis. Depending on the location of the rod and clinical symptoms, revision surgery may be necessary. Of 10 patients with CP who underwent revision surgery (after posterior spinal fusion with a unit rod), 3 had perforation of the inner wall of the pelvis. All three were revised, two at the time of surgery and one 13 days later. Five of the patients underwent revision because of progression below the curve (failure to fuse to the pelvis); five underwent revision for pseudarthrosis with implant failure and curve progression (three rod failures and two cross-link failures).\(^4^2\)

Pulmonary Complications

Postoperative respiratory problems are common in children with CP because of pain and the inability to participate in pulmonary care. In one study, postsurgical pneumonia occurred in 1 of 15 children with neuromuscular scoliosis and restrictive lung disease;\(^4^3\) 2 patients required tracheostomy. Staging anterior-posterior procedures, however, does not seem to decrease the incidence of respiratory complications.\(^3^4\)

Gastrointestinal Complications

Significant gastrointestinal problems can occur in patients with CP. Severe gastric distension, dysmotility with resulting obstruction, gastroesophageal reflux, and pancreatitis have been described. Two of seven patients in one study died be-
cause of bowel obstruction. Treatment initially consisted of left lateral positioning and enteral feeding beyond the obstruction.

Neurologic Complications

Neurologic complications are uncommon after spinal fusion in children with neuromuscular disorders, but changes in SSEPs are often encountered. Modifying the procedure may be required. The passage of sublaminar wires, when carefully performed, does not carry a high incidence of neurologic injury.

Wound Complications

Wound complications (including superficial and deep infections) after surgical correction of scoliosis in children with CP occur more often than in those with idiopathic scoliosis. Szüke et al described an 8.7% rate of postoperative wound infection; 7 of 15 were deep infections, but only 1 late infection required removal of the instrumentation. Six of seven deep infections were treated with debridement with healing by secondary intention; one was closed over drains and required re-debridement because of a recurrent abscess.

Summary

Children with CP frequently develop spinal deformity and are at higher risk of complications because they commonly have significant comorbidities. The approach to the care of these children should be multidisciplinary to ensure a successful outcome. The goals of the surgery are to establish a stable, balanced, painless spinal fusion. Outcomes have improved with heightened awareness of complications, improved medical care, and improved surgical instrumentation for deformity correction. The type and methods of internal fixation should be individualized for each patient based on the severity and stiffness of the curve, degree of neurologic involvement, skeletal maturity, and ambulatory status.

References

Citation numbers printed in bold indicate references published within the past 5 years.


