Spinal deformity is a common musculoskeletal problem for individuals with cerebral palsy. Severe scoliosis may impair physical function and may be a source of pain. Spine braces and carefully constructed seating arrangements may moderate the behavior of these deformities but do not seem capable of stopping progression, which often continues in adulthood. Spine fusion surgery can produce a stable, durable trunk shape that improves sitting and positioning but the process of surgery is arduous and outcomes can be compromised by numerous serious complications. Despite complications, many families and caregivers express satisfaction with the results of surgery. Careful patient evaluation, studious attention to surgical planning and performance, and a good relationship with patients, family members, and other providers is essential.

The justification for any medical intervention must be based on a clear understanding of the outcome of a condition left untreated (often referred to in the medical community as ‘natural history’). If the unfolding of a condition is unacceptable now, or if the final outcome of the condition will be unacceptable, then it is reasonable to consider intervention (treatment). But treatment is justified only if its outcome will improve the patient’s status and if the processes that surround each intervention are tolerable.

The challenges of this dry conceptual framework come to life when it is applied to individuals with cerebral palsy (CP) and scoliosis. Figure 1 displays the radiograph of a 21-year-old with CP who is able to sit only with support. What intervention, if any, should be recommended for this patient? The answer to that question requires examination of other questions. How often do scoliosis or other spine deformities develop, and how do the curves behave? What are the effects of scoliosis in CP? What is the role of non-operative intervention? What are the goals, processes, and outcomes of operative intervention? This paper provides a summary of the answers to these questions.

**HOW OFTEN DO SCOLIOSIS (OR OTHER SPINE DEFORMITIES) DEVELOP, AND HOW DO THEY BEHAVE?**

If the traditional definition of scoliosis (10° of lateral curvature calculated by Cobb’s method on a radiograph) is used, then scoliosis is common for individuals with CP. Its prevalence far exceeds that of the general population. Idiopathic scoliosis (scoliosis that develops without a clear cause) is found in 1 to 2% of the population. Neuromuscular scoliosis (scoliosis related in some way to altered neurological or muscle function) is found in 15 to 80% of people with CP. The wide range in prevalence is due to variations in the populations being studied – variations that include age, nature, and severity of neurological dysfunction; the extent of impairment of physical function; and the method employed during radiography (supine vs upright image). Balmer and MacEwen reviewed the radiographs of 100 children receiving care in the outpatient clinic at the AI duPont Institute (Delaware) and found scoliosis greater than 10° in 21 children. Studies based on adults residing in institutions, and therefore likely to be more impaired in cognitive and physical function, have reported a much higher prevalence of scoliosis. Madigan and Wallace found scoliosis of at least 10° in 64% of their study group, Thometz and Simon reported 61%, Koop et al. reported 77%, and Saito et al. reported 68%. Troublesome sagittal deformities are less common, occurring in 7 to 10% of individuals with severe impairment.

Nearly all published studies of the prevalence of scoliosis in persons with CP predate the Gross Motor Function Classification System (GMFCS), but the likelihood of scoliosis developing in an individual with CP is clearly
linked to neurological impairment and its effects on physical function. The factors most strongly associated with large curvatures appear to be severe neurological impairment and young age at the time of curve appearance. An individual with severe spasticity that affects the entire body is at the greatest risk of developing scoliosis. Koop et al.\(^4\) found scoliosis greater than 40° at maturity in 30% of individuals with quadriplegia, 10% of those with diplegia, and 2% of those with hemiplegia. The majority of these curvatures started forming before 10 years of age. By comparison, idiopathic scoliosis more than 30° occurs in no more than 0.1 to 0.2% of the population.

Increases in curve size after skeletal maturation are common among individuals with quadriplegia. Madigan and Wallace\(^2\) found the average curve size in institutionalized adults to be 54°. Thometz and Simon\(^3\) demonstrated a relationship between curve size and functional status at maturity when estimating the risk of curve progression. Curves less than 50° increased an average of 0.8° per year, while those of 50° or more increased 1.4° per year. Individuals able to walk in the community demonstrated progression of 0.9° per year, while those who were bedridden demonstrated progression of 2.4° per year. Majd et al.\(^6\) followed 56 adults with scoliosis of 20° or more for an average of 8 years. The average rate of curve increase was 3° per year for individuals with stable function, 4.4° per year for those who had demonstrated a loss of function, and 9.2° per year for those with those with kyphoscoliosis. Saito et al.\(^5\) reported that 85% of individuals with scoliosis greater than 40° at age 15 years progressed to curve magnitudes of 60° or more.

Two surgical procedures have been devised for reducing hypertonicity in CP: selective dorsal lumbar rhizotomy (SDR) and intrathecal baclofen pump (ITB) placement. These procedures are performed for spasticity in different populations, and each may alter the natural behavior of the spine. SDR is most commonly performed before adolescence (age range 4–10y) in children with walking ability (typically GMFCS levels II, III, and IV). ITB pumps are most commonly placed near adolescence or in adulthood in individuals with limited or no ability to walk (GMFCS levels IV and V). Studies of the spinal consequences of either procedure are hampered by inconsistent radiographic techniques.

The following four observations about the effects of SDR on the spine seem reasonable: (1) When SDR is performed for individuals with less severe functional impairment, increased lumbar lordosis is common but has few consequences. (2) When SDR is performed in the presence of greater functional impairment, the risk of scoliosis is increased, but severe curves are uncommon. (3) Laminoplasty non-unions are common in the lower lumbar vertebrae. (4) Lumbosacral spondylolisthesis is frequently seen after surgery and is extremely rare before SDR.

Studies of SDR commonly mention the appearance of increased lumbosacral lordosis but do not analyze it in any detail. Scoliosis is described as common but rarely progressive. Spiegel et al.\(^7\) found scoliosis in 17% of their group, with average curve magnitude of 16°. Johnson et al.\(^8\) found scoliosis in 24%, and Golan et al.,\(^9\) in a study that used upright radiographs, found scoliosis in 45% but only 5% had a curve greater than 25°. Spiegel et al.\(^7\) detected non-unions at sites within the laminoplasty used to expose the spinal canal. These became more common in the lowest areas of the lumbar spine: 8% at L3, 35% at L4, and 46% at L5. They reported lumbosacral spondylolisthesis in 12% of their group, all but one grade I in

![Figure 1: This 21-year-old female experiences no pain but her rack graphs show a curve increase of 10° over the last 2 years.](image)
severity. Johnson et al.\textsuperscript{8} found spondylolisthesis in 18%, and Golan et al.\textsuperscript{9} reported this finding in 19%. Nothing is known about the consequences of spondylolisthesis during adulthood.

The following observations about the effects of ITB on the spine seem reasonable: (1) ITB pumps are used to manage the spasticity of individuals with greater functional impairment and tend to be placed near adolescent growth or during adulthood. (2) Scoliosis is often present in individuals undergoing ITB pump placement. (3) It is difficult to separate the effects of ITB from those of accelerated adolescent growth. (4) The effects of ITB are not clearly understood.

In 19 patients, Ginsburg and Lauder\textsuperscript{10} found that curves progressed 1.8° per year before pump placement and an average of 11° per year after pump placement. They expressed concern about the effect of decreased tone and weakness of trunk musculature on the behavior of scoliosis. They recommended spine bracing with ITB. Senaran et al.\textsuperscript{11} matched 26 ITB patients with 25 individuals with CP of similar age, functional impairment, and scoliosis magnitude. Both groups demonstrated curve progression, and there was no difference in the pattern or rate of progression. In a similar study, Shilt et al.\textsuperscript{12} compared 50 individuals with ITB pumps with 50 matched controls. They could not demonstrate a significant difference in scoliosis progression between the two groups. No study has fully examined the timing of ITB placement, baclofen dosages, and details such as the position of the catheter within the dural sac. These studies demonstrate that we do not understand the effect of ITB on the behavior of the spine.

**WHAT ARE THE EFFECTS OF SCOLIOSIS IN CP?**

Severe scoliosis is much more common in individuals with severe functional limitations (GMFCS levels IV and V), the same group prone to lower-extremity contractures and hip dysplasia. Several researchers have found a linkage between the direction of wind-swept deformity and trunk decompensation, with the convexity of the scoliosis to the opposite direction. Despite this observation, a clear association between unilateral hip dislocation and scoliosis has not been established.

Sitting is a fundamental position of function and health for people with CP. The upright position facilitates vision, communication, and mobility. It also facilitates feeding and protects pulmonary function by minimizing gastric reflux and aspiration. Large curvatures are associated with poor sitting. Long, single curves often result in trunk decompensation that requires propping in order to remain upright. The rotational component of scoliosis creates large rib prominences that are prone to excessive skin pressure from seat surfaces and braces. The pelvis may become an end-vertebra for the lowest curvature, resulting in pelvic obliquity. The obliquity can be extreme in very large and long, single curves. Pelvic obliquity creates an unbalanced sitting surface and increases pressure in the tissues over the ischial tuberosity. In the most severe obliquity, pressures will be focused on the greater trochanter. Because sensation is largely preserved in CP, these areas of increased pressure become painful. Because their verbal communication is often compromised and because upper-extremity use is limited, individuals with CP cannot inform others about their pain and cannot move themselves to relieve the pressure. The consequence may be ulceration.

Communication barriers make it very difficult to determine whether individuals with severe CP are experiencing pain. If pain is obvious, it can be difficult to locate the source, and many sources are possible. Nonetheless, it appears that pain is very common in adults with CP. Jahnsen et al.\textsuperscript{13} stated that 72% of their study population reported pain. The pain was mild to moderate in 55% of cases and severe in 17%. Pain was present every day for at least 1 year in 28% of individuals. By comparison, only 15% of their normative population reported such significant pain. Andersson and Mattsson\textsuperscript{14} documented pain in 79% of their group, with 22% reporting that the pain was present daily. The neck and back were the most common sites, but pain was possible at every lower-extremity joint, from the hip to foot. Approximately two-thirds of adults with pain report pain in two or more locations.

**WHAT IS THE ROLE OF NON-OPERATIVE INTERVENTION?**

There are two methods to provide external support to the spine in the presence of neuromuscular scoliosis: customized seating arrangements and braces (thoracolumbar-sacral orthoses). This is not an ‘either-or’ choice. Properly devised seating arrangements can support the trunk, distribute sitting-surface pressures, and accommodate contractures around the hip. When combined with mobility bases and communication devices, they can significantly enhance function. Spine braces provide more direct trunk support, which may simplify the seating arrangement. A spine brace may assist sitting outside of a mobility base and may facilitate lifting and transfers by caregivers.

Because there are no agreed-upon criteria among physicians for recommending spine fusion surgery to parents and guardians, and because those physicians use widely divergent criteria for their decisions about surgery, it is not possible to determine whether spine bracing reduces the
need for surgery. During the growth years it is clear that a spine brace will reduce curve size and slow the rate of progression to large curve magnitudes.\textsuperscript{15} It is not clear if bracing is able to stop progression. During adulthood, the role of a spine brace is to improve sitting, a purpose that is valued by caregivers if the brace is properly constructed and does not cause skin pressure problems. Nothing is known about the ability of a spine brace to affect curve progression during adulthood.

**WHAT ARE THE GOALS, PROCESSES, AND OUTCOMES OF OPERATIVE INTERVENTION?**

It is reasonable to consider a spine fusion in the following circumstances: (1) The curvature impairs sitting ability (trunk decompensation, pelvic obliquity). (2) Spine bracing fails to resolve problems with sitting or the brace is intolerable. (3) The curvature is increasing in size despite bracing. (4) Pain, pulmonary dysfunction, or abdominal issues can be clearly attributed to the spine or the brace.

Spine fusion surgery in adolescents or young adults with CP has three major goals: deformity improvement; durable stability of the new spinal shape; and safe and tolerable surgical processes.

Improvement of the spine deformity means achieving a level pelvis with a well-centered or balanced trunk. It does not mean reducing the Cobb angle values to an absolute minimum. The improved spine shape is achieved through a variety of spinal instrumentation techniques and through anterior, posterior, or combined surgical approaches. Supine anteroposterior radiographs performed during the application of longitudinal traction provide a very accurate understanding of spinal column flexibility and assist in surgical decision making. The spine shape achieved through instrumentation is made durable only by the formation of a solid spine fusion. Instrumentation without fusion will deform and break, making the first two goals (improved shape and stability) dependent on each other.

Isolated instrumented anterior procedures are uncommon in CP but may be appropriate for thoracolumbar curve patterns that are similar to those typical of idiopathic scoliosis.\textsuperscript{16,17} Such curves may be found in individuals at GMFCS levels I and II. Anterior procedures with discectomy and fusion (with or without instrumentation) may be combined with posterior procedures. The anterior component of a combined procedure can improve spinal column flexibility and either minimize the residual curvature or assist in achieving the goal of a level pelvis and balanced trunk. Posterior instrumentation consists of two rods attached to the spine through some combination of hooks, sublaminar wires, or pedicle screws.\textsuperscript{18–22} Extension of the instrumentation to the sacrum or pelvis is employed when trunk decompensation or pelvic obliquity is present. The nuances of instrumentation techniques are beyond the scope of this chapter.

Spine fusion procedures for individuals with CP can be arduous for the surgical team and for the patient and the following questions must be answered: (1) What constitutes appropriate preoperative evaluation and preparation? (2) What approach should be used? Anterior, posterior, or combined? (3) If a combined approach, should it be done in a single event or as separate procedures? (4) What type of instrumentation should be used? (5) What type of bone graft is appropriate? Should bone graft alternatives be used? (6) What prophylactic antibiotics should be used and for how long? (7) How should ITB catheters be managed? (8) What blood products should be used? (9) Should somatosensory-evoked potentials and transcranial motor-evoked potentials be attempted? (10) Should endotracheal intubation and mechanical ventilation be done postoperatively? (11) When should computer-guided surgical navigation be used? (12) How should postoperative nutrition be accomplished?

**AND THE LIST GOES ON!**

While the goals of surgery are clear, there are many possible complications. The report of a single institution, given in meticulous detail, reflects the experience of many workers. Tsirikos et al.\textsuperscript{21} described the outcomes of spine fusion for 287 consecutive individuals with CP undergoing surgery performed by two highly experienced surgeons at a single institution over a 12-year period.\textsuperscript{21} Of these patients, 242 underwent a posterior-only procedure and 45 underwent a combined anterior and posterior procedure. The unit rod was used for instrumentation in all procedures. In the posterior-only group, the mean preoperative scoliosis was $74^\circ$(SD 21). Mean surgical time was 3.9(SD 0.8) hours, blood loss was 2.8(SD 2.4) L, intensive care unit (ICU) stay was 4.9(SD 5.3) days, and total hospitalization was 18.6(SD 13.0) days. In the combined anterior and posterior group, the mean preoperative scoliosis was 86$^\circ$(SD 40). Mean surgical time was 7.1(SD 1.6) hours, blood loss was 3.4(SD 1.7) L, ICU stay was 6.7(SD 5.1) days, and total hospitalization was 24.5(SD 24.3) days. In the 287 patients there were three deaths (1%), 18 deep wound infections (6%), and four dural tears. Instrumentation problems were seen: rod penetrations of the ilium in 17 (6%) patients; sublaminar wire cutout in 14 (5%); and painful protrusions in 14 (5%). Overt pseudarthrosis and neurological change were rare (one of each) and less frequent than reported in other studies.

Complications appear to occur in 40 to 80% of patients who undergo spine fusion, with death in approximately...
The major complications are severe bleeding, pulmonary compromise, deep wound infection, spinal cord dysfunction (including paraplegia), and pseudarthrosis. Total blood loss (during and after surgery) equal to or greater than calculated body blood volume is common. Postoperative respiratory compromise occurs in as many as 25% of all patients, with prolonged mechanical ventilation (>3d) required in 5%. Such problems are most common when preoperative pulmonary function tests show a forced vital capacity less than 40% of predicted. Deep wound infections have been reported in 1 to 10% of cases. While staphylococcal species are a common cause, at least half of deep wound infections are polymicrobial and include organisms such as Enterobacter, Enterococcus, and Escherichia coli. Deep infections often require multiple surgeries, wound vacuums, and prolonged antibiotics. Dramatic changes in lower-extremity neurological function can occur as a result of surgery. The exact rates may not be known, and what is reported may represent only the most obvious paraplegias. Intraoperative ‘wake-up’ tests are impractical in CP and questions have been raised about the efficacy of neurodiagnostic monitoring of somatosensory-evoked potentials and motor-evoked potentials. Useful data can be obtained in 70 to 90% of cases, and most authors recommend intraoperative monitoring while recognizing its limitations (transcranial motor evoked potentials are not recommended in individuals with a seizure disorder). While it may seem that loss of lower-extremity neurological function is of little importance to someone who cannot walk, loss of existing bowel and bladder control and loss of protective skin sensation do matter.

Most studies report average preoperative scoliosis of more than 70º, and many cases exceed 100º. Many of the curves are rigid. These facts play a role in the failures of spinal instrumentation and the resultant risk of fusion pseudarthrosis (up to 20% of cases). Since spinal instrumentation acts as an internal prop while the body forms the solid fusion that makes the new spine shape permanent, large residual curves (>50º) challenge the strength and stability of the metal devices. Rod bending and fracture occur. Hyperkyphosis and ‘pull-out’ occur at the upper end of the rods, and rod-bone, rod-screw, and rod-rod failures occur distally. Prominent fixation devices can result in skin breakdown.

The long list of potential complications (major and minor) suggests that preoperative planning and preparation are very important. The issues that must be addressed are nutrition, pulmonary function, gastroesophageal reflux, bowel motility, seizures, coagulopathies, and the effects of current medications. The adage that many postoperative complications and problems started before surgery should be taken seriously.
has become more difficult than it would have been if performed years earlier. These points out the value of stable and long-term relationships between physicians and their patients and families. Such relationships foster communication, including passage of information and time for listening. That work may result in more timely decisions for a difficult problem.

REFERENCES


