INTRODUCTION

Patients with cerebral palsy (CP) commonly develop scoliosis. The treatment of scoliosis in these patients can be different from treatment of the idiopathic scoliosis population. Management of the spinal deformity in CP can be challenging and often presents the surgeons and caregivers with many difficult decisions and obstacles. The approach to the care of these children should be multidisciplinary in order to optimize outcomes and decrease the frequent complications.

CEREBRAL PALSY AND SCOLIOSIS

The neuromuscular scoliosis that occurs in CP is typically a C-shaped curve that is often kyphoscoliotic and is associated with pelvic obliquity (Fig. 1). Children with CP have an increased risk of developing scoliosis compared with other patient populations. Muscle weakness, truncal imbalance, and asymmetric tone in paraspinal muscles have long been implicated for the onset of scoliosis in CP, but there is little literature to support this theory.

The authors have nothing to disclose.

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The prevalence of scoliosis in a total population of children with CP is nearly 30%. \(^3\) Curves tend to begin at an earlier age than in idiopathic scoliosis. \(^4\) They are more likely to progress even after the patient reaches skeletal maturity. \(^5\) There is also an increased incidence of increased Gross Motor Function Classification System (GMFCS) level. \(^3\) Children with GMFCS level IV and V CP have a 50% risk of moderate or severe scoliosis by the age of 18 years. \(^3\)

Studies have found correlation between the size of the deformity and the decline in functional activities. Majd and colleagues \(^6\) found an increased curve progression in patients with a decline in function compared with patients who were functionally

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Fig. 1. (A) Anteroposterior/lateral radiographs of 14-year-old boy with scoliosis secondary to CP. The patient has undergone prior baclofen pump placement. (B) The patient has undergone correction of the scoliosis with a T2-pelvis fusion.
stable. They also found that patients developing decubitus ulcers with prolonged sitting were more likely to have larger curves.

SCOLIOSIS TREATMENT IN CEREBRAL PALSY

Nonsurgical Treatment

Nonoperative treatment of scoliosis in patients with CP consists of:

- Observation
- Seating modifications
- Bracing

Observation can be chosen for curves of any size or magnitude. Reasons for continued observation are numerous. The curve size may be of insufficient magnitude to require surgical treatment. In addition, family decisions may be made to avoid operative management or other nonoperative modalities because of various comorbidities or after weighing the risks and benefits.

Molded wheelchair inserts can be used to attempt to improve overall sitting balance. Three-point force configuration support systems have been shown to achieve the best static correction of the scoliosis. However, there is no evidence that this alters the natural history. Modifications of the wheelchair should be individually tailored to the patient.

Bracing goals for CP are often different from the goals in idiopathic scoliosis. There is little evidence that bracing slows curve progression, but there are many studies that show no significant effect of bracing. The goals of bracing are to maintain comfortable, upright sitting, allow functional use of the upper extremities, and allow maximum ability to interact with the environment. Soft braces seem to be tolerated better than rigid orthoses in patients with spastic CP. Soft braces maintain skin integrity and potentially minimize respiratory compromise.

Surgical Treatment

Goals, indications, and benefits of surgery, as well as the risk and potential complications, must be carefully weighed with the patient’s family. There is no absolute indication of surgery (Box 1).

For patients undergoing surgery, a thorough preoperative assessment is required. Assessment needs to include pulmonary function, nutritional status, gastrointestinal evaluation, and neurologic function.

Pulmonary function testing is not easily performed in this patient population, but evaluation with a chest radiograph is often necessary. Patients with poor nutrition (serum albumin <35 g/L and total blood lymphocyte count <1.5 g/L) are known to have increased postoperative infection rate, longer length of intubation, and longer

<table>
<thead>
<tr>
<th>Box 1</th>
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<tr>
<td><strong>Goals of surgery and relative surgical indications for spinal fusion</strong></td>
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<table>
<thead>
<tr>
<th>Goals of Surgery</th>
<th>Relative Surgical Indications for Spinal Fusion</th>
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<tbody>
<tr>
<td>Improve coronal and sagittal balance</td>
<td>Progression of Cobb angle &gt;50°</td>
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<tr>
<td>Improve sitting balance</td>
<td>Deterioration in functional sitting</td>
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<tr>
<td>Halt progression of scoliosis</td>
<td>Age &gt;10 years, with adequate hip range of motion to allow seating, stable nutritional, and medical status</td>
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<tr>
<td>Level pelvis</td>
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<td>Improve positional discomfort</td>
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hospital stays.\textsuperscript{16} Nutrition may need to be optimized with nutritional supplementation or even a G-tube placement. From a gastrointestinal standpoint, assessment for gastroesophageal reflux and aspiration risk is needed because these patients are at risk for aspiration pneumonia.

In addition, seizure disorders should be controlled, and the medications used to control the condition should be noted. Some medications, such as phenytoin, phenobarbital, and valproic acid, can cause reduced bone mineral densities.\textsuperscript{17} Valproic acid has also been implicated for increased intraoperative blood loss and the need for blood transfusions.\textsuperscript{18}

The technical aspects of surgery for spinal fusion are beyond the scope of this article. There have been major advancements from a surgical standpoint from Luque rods, Luque rods with Galveston technique, Cotrel-Dubousset and Isola instrumentation, and more recently pedicle instrumentation. Pedicle instrumentation has been shown to have improved curve correction rates, lowest loss of correction, and greatest apical vertebral translation.\textsuperscript{19}

The complication rate in the perioperative period for these patients is nearly 30%.\textsuperscript{20} Patients with CP undergoing surgery for spinal deformity are at high risk for complications secondary to medical comorbidities.\textsuperscript{8} Reames and colleagues\textsuperscript{21} and Hod-Feins and colleagues\textsuperscript{22} found surgical complications rates to be much higher among this patient population compared with the rates in idiopathic scoliosis. Perioperative complications including delayed extubation and length of hospital stay are significantly higher in neuromuscular scoliosis compared with idiopathic scoliosis.\textsuperscript{22} This difference is likely directly related to longer surgeries, higher blood loss, and other medical comorbidities such as seizure disorders.\textsuperscript{20,22}

Major complications in the postoperative period have also been described. Death is reported in 0% to 7% of patients.\textsuperscript{23,24} Sponseller and colleagues\textsuperscript{25} reported a deep infection rate of 6% and superficial rate of 4%. Deep infections in patients with CP are often polymicrobial with gram-negative organisms.\textsuperscript{26} Pseudoarthrosis rates have decreased over time from the use of Harrington rods (as high as 40%) to Luque instrumentation (0\%–13\%) to more recent fixation with pedicle instrumentation (1\%) (Box 2).\textsuperscript{26,27}

Outcomes of spinal fusion are difficult to assess in this patient population because there is still no adequate outcome tool to adequately assess improvement in function. Cassidy and colleagues\textsuperscript{28} showed no clinically significant differences in pain, pulmonary medication need, decubitus ulcers, function, or time for daily care in patients who underwent spinal fusion compared with those who had not. In contrast, Larsson and colleagues\textsuperscript{29} evaluated patients 1 year after surgery and found improvement in curve size and sitting balance. Multiple studies have shown high satisfaction in parents and

| Box 2 |
| Complications with spinal fusion in patients with CP |

<table>
<thead>
<tr>
<th>Perioperative Complications</th>
<th>Postoperative Complications</th>
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<tbody>
<tr>
<td>Death</td>
<td>Death</td>
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<tr>
<td>Neurologic complications</td>
<td>Superficial/deep infections</td>
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<tr>
<td>Respiratory distress, atelectasis, pneumonia</td>
<td>Pseudoarthrosis</td>
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<tr>
<td>Delayed extubation</td>
<td>Implant failure</td>
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<tr>
<td>Gastric distension, ileus, obstruction</td>
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<tr>
<td>Seizures</td>
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<tr>
<td>Blood loss</td>
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<td>Hemodynamic instability</td>
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caregivers after surgery. Based on a caregiver questionnaire, Watanabe and colleagues found improvements in sitting balance (93%), cosmesis (94%), and quality of life (71%).

**SCOLIOSIS, SPINAL FUSION, AND INTRATHECAL BACLOFEN PUMPS**

**Indications/Benefits of Intrathecal Baclofen Pumps**

Intrathecal baclofen (ITB) is approved for treatment of spasticity related to several disorders including CP. It inhibits both monosynaptic and polysynaptic reflexes at the spinal cord level thus decreasing excitatory neurotransmitter release from primary afferent terminals to decrease spasticity. Penn and Kroin first reported its use for severe spasticity with immediate reduction of muscle tone to near-normal levels. The efficacy of ITB in children with CP is well documented. Multiple studies have shown improvement in patients with CP. Gooch and colleagues showed improved satisfaction of care providers, ease of care, and decreased pain. Other studies have also found improved ease of care and others have shown improved gait in ambulatory patients with CP. In addition, Gerszten and colleagues showed a decreased need for subsequent orthopedic surgery for lower extremity spasticity. The indications for implantation of ITB pumps are primarily for intractable spasticity (Box 3).

**Box 3**

**Indications for ITB pump and benefits of ITB pump therapy**

<table>
<thead>
<tr>
<th>Indications for ITB Pump Therapy</th>
<th>Benefits of ITB Pump Therapy</th>
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<tbody>
<tr>
<td>Intractable spasticity</td>
<td>Improved spasticity</td>
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<tr>
<td>Uncontrollable spasticity with drug therapy</td>
<td>Improved range of motion</td>
</tr>
<tr>
<td>Intolerable side effects to oral baclofen</td>
<td>Improved ease of care</td>
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<tr>
<td></td>
<td>Improved pain</td>
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<tr>
<td></td>
<td>Increased caregiver satisfaction</td>
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<tr>
<td></td>
<td>Improved hygiene, transfers</td>
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<td></td>
<td>Improved gait</td>
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**Technique for Insertion of Intrathecal Baclofen Pumps**

A trial test dose is frequently given before proceeding with placement of an ITB pump. This test can be performed on an inpatient or outpatient basis. Typical response is within 1 to 2 hours and lasts up to 8 hours. Based on this, it may be recommended to proceed with ITB pump implantation.

There are 2 components to the ITB pump infusion system: the pump, which commonly is implanted on the abdominal wall, and the catheter, which travels from the pump to the cerebrospinal fluid (CSF). Depending on the reservoir size and the dosing, the pump can hold enough drug for 4 to 6 months of therapy. It is refilled with a subcutaneous injection into the pump. Because many of these patients undergo spinal fusions, options for ITB pump placement include before, during, or after scoliosis surgery. Technical descriptions of placement are present in the literature.

ITB pump placement before spinal fusion is technically less demanding than after spinal fusion. Patients are positioned in a lateral decubitus position. A midline incision is placed at the mid to lower lumbar level at the level of the iliac crest. This placement can easily be incorporated into any future incision needed for a spinal fusion. A separate fascial incision is made approximately 1 cm lateral from midline. The catheter is placed below the fascia to provide an additional layer of closure. A 14-gauge Tuohy needle is directed midline and obliquely in a cephalad direction so as to be placed...
into the dural sac between L2/3 or L3/4. Fluoroscopy is used to confirm intrathecal placement and to guide advancement to the appropriate level. The final catheter placement is guided by diagnosis, the extremities affected, and the desired effect. Most pumps are placed on the right side to avoid current or future gastrostomy tubes. An incision is made in this location and a subcutaneous or subfascial pocket is developed in order to place the pump. In thin patients with CP, the pump is typically placed subfascially to ensure an additional layer of closure. The subfascial placement of the catheter and pump provide an additional barrier to CSF leak, which can complicate the procedure. The catheter is then tunneled from the spine to the pump and is tested to ensure backflow of CSF.

When placing the ITB pump after a spinal fusion, an incision is made through the previous midline scar and a subperiosteal exposure of the fusion mass is performed. Fluoroscopy is used to locate the implants and the level of spine exposed. The technique of using a burr to open a hole in the fusion mass at L2/3 or L3/4 has been described. If a large hole is made, this increases the risk of CSF leak. This risk can be minimized by leaving a thin layer of bone and pushing the Tuohy needle through it to gain access to the spinal canal. At our institution (Rady Children’s Hospital San Diego) we have modified this technique depending on the type of spinal instrumentation used. In cases in which sublaminar wires have been used, the wires at L3 or L4 are removed. This hole in the fusion mass left by the wire is typically large enough for the catheter needle to enter the spinal canal. In patients instrumented with segmental pedicle screws, a 2-mm K-wire is used to create an oblique hole through the lower lumbar fusion mass. Under fluoroscopic guidance, the K-wire is oscillated through the fusion mass. Tactile feedback typically shows when the spinal canal is entered. In many cases, the K-wire has not punctured the thecal sac. Once the catheter is placed and secured similarly to that described earlier, bone wax and/or fibrin sealant is placed around the fusion mass hole to minimize the risk of CSF leak.

Patients with prior ITB pump implantation who undergo a posterior spinal fusion (PSF) can be approached in several ways. It is possible to work around the catheter and keep it intact, although this can be frustrating to the surgeon. The catheter can be removed and the dura can be sealed uneventfully with a new catheter placed after spinal fusion. It is also possible to cut the current catheter and then reanastomose with some systems.

**Complications of Intrathecal Baclofen Pump Placement**

Although the safety and efficacy of ITB pumps has been evaluated, there are still significant complications associated with their placement before, during, or after spinal fusion in patients with CP. Complications related to ITB pump placement are well reported in the pediatric literature.[39-44]

Wound complications and infections are also a common reason for rehospitalization and reoperation in patients with CP. Fjelstad and colleagues[43] found the rate of infection after ITB pump placement to be higher in children than in adults (10% vs 0%), but more of the pediatric patients had a diagnosis of CP compared with the adult population in this study. Overall acute infection rates ranged from 4% to 10%. One study found a significant increase in infection rate with subcutaneous pump placement compared with subfascial placement (20.1% vs 3.6%; P<.001) and thus recommended subfascial placement. When infection occurs, there is a high rate of reoperation and removal of the catheter and pump.[40,44]

The incidences of catheter-related and pump-related complications are variable in the literature. Complications include pump malfunction and catheter dislodgement/breakage/malfunction. Rippe and colleagues[41] reported a total of 264 catheter
complications in 785 patients. Borowoski and colleagues\(^42\) reported 16% to 27% device complications in their patients, most which were catheter malfunctions. Armstrong and colleagues\(^36\) reported 10 complications in 19 patients over 568 months. Similar to infections, the complications related to catheters and pumps are a common reason for rehospitalization and reoperation.\(^45\)

An additional complication reported with ITB pump placement before scoliosis surgery is progression of scoliosis. Significant controversy exists as to whether insertion of ITB pumps causes progression of scoliosis in patients with CP,\(^46–52\) and some clinicians think that the progression is related to patients who receive an ITB pump being more severely involved and thus their natural history is to have progressive scoliosis.\(^8\)

Two series of patients reported accelerated scoliosis progression after ITB pump placement.\(^47,48\) Segal and colleagues\(^47\) initially presented their series of 5 patients with a mean progression of 44° over 11 months leading to spinal fusion. Burn and colleagues\(^50\) found an annual progression of Cobb angle of 18° and an increase in progression per year in patients less than 15 years old at the time of ITB pump placement. Ginsburg and Lauder\(^46\) found an increase in scoliosis progression from 1.8°/y before implantation to 10.9°/y after implantation.

However, these studies supporting progression of scoliosis after ITB pump placement were small series. In contrast, 2 studies that compared matched cohorts of patients with CP with and without ITB pumps found no difference in the rate of scoliosis progression.\(^51,52\) In the study by Shilt and colleagues,\(^52\) patients were matched by diagnosis of CP, age, sex, topographic involvement, and initial Cobb angle. No difference was found between the mean change in Cobb angle in the patients with ITB pumps (6.6° per year) compared with the controls (5.0° per year).

In addition, there is also concern and debate about whether prior placement of an ITB pump can further complicate CP scoliosis surgery and increase the risk for wound complications. There are 2 studies that have compared the outcomes of PSF in CP with and without ITB pumps.\(^38,45\) Caird and colleagues\(^45\) compared 20 patients with spastic quadriplegic CP with ITB pump who underwent PSF with 20 matched patients without an ITB pump. They found increased reoperation and rehospitalization, as mentioned previously, and a higher infection rate in the ITB pump group (20% vs 0%; \(P = .063\)). However, the patients in the ITB pump group had more preoperative comorbidities.

Borowski and colleagues\(^38\) compared 4 groups of patients with CP: PSF before ITB pump placement (\(n=26\)), PSF and ITB pump placement concurrently (\(n=11\)), PSF after ITB pump placement (\(n=25\)), and ITB pump placement only (\(n=103\)). In all four

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### Box 4

#### Perioperative and postoperative complications of ITB pumps

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<thead>
<tr>
<th>Perioperative Complications of ITB Pumps</th>
<th>Postoperative Complications of ITB Pumps</th>
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</thead>
<tbody>
<tr>
<td>Bleeding</td>
<td>CSF leak</td>
</tr>
<tr>
<td>CSF leak</td>
<td>Infection: pump, CSF</td>
</tr>
<tr>
<td>Initial alteration in bladder control</td>
<td>Wound dehiscence</td>
</tr>
<tr>
<td>Apnea, respiratory depression</td>
<td>Catheter-related complications: catheter</td>
</tr>
<tr>
<td>Baclofen overdose: lethargy, seizures, respiratory depression</td>
<td>breakage/dislodgement/malfunction</td>
</tr>
<tr>
<td>Apnea, respiratory depression</td>
<td>Pump failure</td>
</tr>
<tr>
<td>Baclofen overdose: lethargy, seizures, respiratory depression</td>
<td>Seroma at pump site</td>
</tr>
<tr>
<td>Risk of scoliosis progression</td>
<td>Need for reoperation for one of the</td>
</tr>
<tr>
<td>complications listed earlier</td>
<td>complications listed earlier</td>
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groups, they found an infection rate of 8% to 9% with no differences between groups. There was also no difference in device or catheter complications between groups. They concluded that ITB pumps are able to be implanted and managed without any increase in complication rate before, during, or after spinal fusion.

SUMMARY

The incidence of scoliosis in patients with CP is high, particularly in those with more involvement. Many of these patients undergo spinal fusion for the scoliosis. However, complication rates are extremely high. The risks and benefits must be thoroughly discussed with the patient’s family and caregiver before proceeding with surgery. Because of severe spasticity, many of these patients undergo ITB pump placement before, during, or after PSF. The complication rates are high with ITB pump placement, but many patients have significant benefit. Despite high complications, it seems equally safe to place ITB pumps before, during, or after spinal fusion.

REFERENCES


