Surgical management of spinal deformities in cerebral palsy

A review

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pastic scoliosis is a group of neuromuscular scolioses originating from an upper motor neuron lesion. While CP represents the most frequent form of neuropathic scoliosis from an upper motor neuron lesion, other forms such as spino-cerebellar degeneration (Friedreich disease, Charcot-Marie-Tooth disease), spinal cord tumor, or syringomyelia also have upper motor neuron origins. However, these latter conditions differ in their clinical presentation from the classic patient with CP in that the degree of cerebral involvement in CP may vary from a totally dependent child with developmental delay to an ambulatory child with normal intelligence. Most patients with CP present with muscle spasticity; therefore, this paper will focus on the treatment of patients with the spastic type of scoliosis prevalent in patients with CP.

Epidemiology of CP

Cerebral palsy (CP) spinal deformities encompass a spectrum of deformities that are often initially treated non-operatively, only to result in progression of scoliotic curves and further morbidity. Various surgical interventions have been devised to address the progressive curvature of the spine. This endeavor cannot be taken lightly and at times can be encumbered by prior treatments such as the use of baclofen pumps or dorsal rhizotomies. Care of these patients requires a multidisciplinary approach and comprehensive preoperative and postoperative management, including nutritional status, orthopedic assessment of functional level with specific emphasis on the hips and pelvic obliquity, and wheelchair modifications. The surgical techniques in CP scoliosis have progressively evolved from the classic Luque-Galveston fixation methods, the use of unit rods, and lately the use of pedicle screws, to modern sacropelvic fixation. With the latter method, the spinal deformity in patients with CP can usually be almost completely corrected.

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Abbreviations used in this paper: CP = cerebral palsy; MW = maximum width.
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finding in CP.\textsuperscript{40} The natural history of curve progression has shown that the onset of a flexible curve occurs from 3 to 10 years of age with a rapid progression to a rigid curve. The progression of scoliosis is highest during the adolescent growth spurt, especially when associated with pelvic obliquity, and commonly continues after skeletal maturity for as much as a reported 1.4° per year for curves > 50° (Fig. 1).\textsuperscript{22,27} In considering the timing of the growth spurt, it is important to realize that many children with CP have significantly reduced skeletal maturation, likely from direct and indirect causes including poor nutrition, neurological impairment, or alterations in puberty.\textsuperscript{27} Madigan and Wallace\textsuperscript{39} have found that the increase in the size of the curve was the principal factor for the decline in functional status.

Treatment of the Spinal Deformity

Nonsurgical treatment options of the spinal deformity in CP include a combination of orthotics, physiotherapy, sitting adaptation, and pharmacological substances to decrease the spasticity and the onset of new contractures. Surgical treatment is considered when nonsurgical treatments have failed or are unlikely to provide any benefit to the patient. Observation and nonsurgical management of the curve are warranted when the curve remains < 40° in the lumbar region or remains supple despite moderate worsening, and the patient is able to sit comfortably.\textsuperscript{42} Bracing traditionally has a very limited role in decreasing curve progression in CP.\textsuperscript{44,52} One exception is when ambulatory patients with spastic diplegia develop idio-pathic-type milder curves, in which brace treatment may be more successful. Orthotics and special adaptations such as cushioned seats, wedges, and pads are helpful in achieving sitting balance in an upright and as comfortable a position as possible.\textsuperscript{27} The importance of sitting balance lies in improved respiratory function, ease of feeding, and overall functionality.

In most cases of scoliosis in CP, surgical intervention is considered due to significant curve progression, loss of sitting balance, and comfortable function. Consideration of surgery must take into account that the goals would be to prevent discomfort and achieve an adequately balanced spine to allow the patient to sit and thus improve his or her functional quality of life. For most curves > 40°, progression is likely and surgery is indicated for stabilization. Nevertheless, the complications of surgical treatment of scoliosis in CP remain high, thus stressing the importance of careful preoperative patient selection and preparation.

Preoperative Workup

A thorough preoperative and perioperative medical assessment is mandatory in treating patients with spastic scoliosis due to the high prevalence of medical comorbidities.

Pulmonary Assessment

In patients with severe motor involvement, respiratory compromise is a major cause of morbidity and death. Evaluation of the medical history will focus on the existence of pneumonia in the past and how long the patient had to receive antibiotics to recuperate from these pneumonias. Poor pulmonary reserve as well as ineffective cough increase the risk of morbidity and death with aspiration pneumonias.\textsuperscript{67} Cerebral palsy is associated with increased pulmonary aspiration along with increased risk of upper airway obstruction, including obstructive sleep apnea.\textsuperscript{24,31} Restrictive pulmonary disease secondary to spinal deformity further contributes to poor inspiratory effort. Pulmonary function tests in this setting are usually not helpful as in adolescent scoliosis, mainly due to poor cooperative effort in many children with CP.

Gastrointestinal Assessment

Gastroesophageal disorders in patients with CP are common (15%–75%), likely secondary to neuromuscular incoordination.\textsuperscript{69} Dysphagia and gastroesophageal reflux are common, and these patients are at high risk of bronchoaspiration. A prophylactic gastrostomy and/or anti-reflux procedure such as fundoplication may be chosen prior to spine surgery to prevent aspiration and ensure adequate perioperative nutrition, although these procedures themselves have high complication rates in CP as well as unclear surgical indications.\textsuperscript{67} In some instances the spinal deformity may have consequences on the overall malnutrition of the patient. We have observed 1 case of death before a spine surgery because of a superior mesenteric artery syndrome due to the lordoscoliosis (Fig. 2). It has also been our experience that patients who appeared
to be malnourished have started to put on weight once the spinal deformity was corrected (Fig. 3).

**Malnutrition**

Approximately 46%–90% of children with CP are malnourished. Children with CP are more likely to be underweight and shorter than peers because of malnutrition. Assessment of the malnutrition includes a thorough physical examination, dietary history including assessment of prolonged feeding time and poor oromotor control, as well as laboratory evaluation including total lymphocyte count, albumin, prealbumin, total protein, and transferrin levels. In a retrospective review of patients with CP and spastic quadriplegia who underwent spinal fusion, Jevsevar and Karlin found that patients with serum albumin levels < 3.5 g/dl and total lymphocyte count < 1500 cells/mm³ had an increased rate of infection, longer intubation, and longer hospitalization. The use of total parenteral nutrition in the acute postoperative phase should be considered if no enteral feeding or access is established.

**Infection Management**

Patients with CP are prone to postoperative infection, including wound infections and urinary tract infections. Poor nutritional status, poor skin condition, chronic respiratory problems, and preoperative infections are risk factors for postoperative infection and warrant evaluation in anticipation of surgical correction. It is our routine to check 1 week before the surgery for any urinary tract infections, treat them if present, and administer gentamicin prophylactically during the surgical procedure in all cases, in addition to routine prophylactic antibiotics such as cefazolin.

**Seizure Disorders**

Approximately 30% of patients with CP have a seizure disorder requiring treatment. Most antiepileptic medications used also decrease bone mineral density, which can compound the decreased bone density present in nonambulatory patients. Several studies have associated valproic acid with decreased preoperative platelet count, increased intraoperative blood loss, and increased need for blood transfusions. In a retrospective study of the effects of valproic acid in spine surgery, the relative risk of having > 30 ml/kg blood loss was 23 times greater in patients taking valproic acid than in those not taking the drug. Due to the extensive blood loss expected in deformity spine surgery, preoperative collaboration with a neurologist to adjust valproic acid in anticipation of surgery may be beneficial to reduce blood loss. The presence of severe seizures is perhaps another rationale for achieving a very strong segmental fixation that will, in theory, resist postoperative convulsion due to the rigidity of the construct. It is important to note that the presence of a seizure disorder is a relative contraindication for use of transcranial electric motor evoked potential during surgery. In these cases somatosensory evoked potentials may be reliably used for monitoring, except in cases of severely involved CP in which only 70% have a somatosensory evoked potential that can be monitored at baseline.

**Assessment of Spasticity**

A preoperative assessment of spasticity is necessary in conjunction with evaluation of the spine to achieve desired postoperative posture and sitting balance. Treatment of the spasticity ranges from injection of botulinum toxin in the calves, oral baclofen, insertion of a baclofen pump, or posterior rhizotomy. Selective posterior...
rhizotomy has been shown to decrease spasticity and importantly improve activity of daily living for as long as 5 years after the procedure. The technique of using laminoplasty over laminectomy for rhizotomy does not appear to influence the advent of spinal deformity. Some reports claim increased incidence of scoliosis after dorsal rhizotomy, yet the impact on the natural history of scoliosis in these patients remains unclear. Several large studies have confirmed the efficacy of baclofen pumps for treatment of spasticity in CP. In a recent paper by Shilt et al., the incidence and severity of scoliosis did not appear to be increased in patients who underwent insertion of baclofen pumps. Caird et al. have shown that the presence of a baclofen pump prior to posterior spinal instrumentation is associated with increased reoperations and rehospitalizations as compared with patients without pumps, but no difference has been detected in the surgical correction of the scoliosis between those with and without baclofen pumps. An important note is to make sure that the intrathecal baclofen pump and tubing are working well at the end of the case (Fig. 4) to prevent baclofen withdrawal—symptoms of which include high fever, increased spasticity, tachycardia, seizures, and change of mental status—which could culminate in death.

Orthopedic Assessment

Patients with CP should be evaluated for hip stiffness or contracture, hamstring tightness, pelvofemoral muscle contractures, pelvic obliquity, and neck range of motion or contracture. Previous surgery around the hip and range of motion of each hip should be documented, taking care not to inadvertently include the lumbar spine flexion in the assessment of the hip flexion range of motion. Hip stiffness may represent a formal contraindication...
to spine surgery because patients may not be able to sit or lie down after extensive corrective spinal instrumentation. Full pelvic radiographs are necessary before a surgical plan for the spine is determined to evaluate the presence and degree of hip dysplasia, subluxation, or dislocation.

Hamstring tightness should be evaluated to make sure the patient will not have a tendency to be imbalanced and to lunge, or in the worst cases will not be able to sit after corrective surgery of the spine. Hamstring tightness may require a release before or at the time of the surgery at either the hip or the knee level to improve overall outcome of standing or sitting balance after spine surgery. Hamstring tightness should be evaluated to make sure the patient will not have a tendency to be imbalanced and to lunge, or in the worst cases will not be able to sit after corrective surgery of the spine. Hamstring tightness may require a release before or at the time of the surgery at either the hip or the knee level to improve overall outcome of standing or sitting balance after spine surgery.

The pelvic obliquity should be assessed to determine if the cause is from the hips or the spine, is an intrapelvic cause, or is mixed.18 An often-used method by the senior author to determine the cause of pelvic obliquity is to have the patient lie prone with the legs hanging free off the end of the examining table. This maneuver relaxes the hip adductor and abductor muscles, removing infrapelvic causes of pelvic obliquity. This position lends itself to demonstrate that the remaining pelvic obliquity may be from the spinal deformity itself. If a doubt persists, the examiner can extend the hips from the flexed position by adducting the 2 legs together and extending the hips by bringing the legs parallel to the trunk. If there is no pelvic obliquity with the hip at 90° and the pelvic obliquity recurs as the hip is extended, the pelvic obliquity likely is attributable to a pelvofemoral muscle contracture. In the same position, the examiner can determine if the pelvic obliquity originating from the spine is reducible or not. This is extremely important because the ability to reduce the pelvic obliquity by external maneuvers suggests that the patient may not need an anterior release or spinal osteotomies. The caregiver may be asked to hold the trunk still while the surgeon stabilizes the pelvis and moves the legs parallel with the trunk. The surgeon then takes the hip through adduction and abduction. If obliquity persists despite adduction and abduction of the hips in this position, a fixed spinal-pelvic obliquity exists. The same maneuvers can be performed in the radiography suite to measure the amount of correction for the pelvic obliquity.

Once the hips and spine have been assessed separately with appropriate clinical examination and radiography, one can then decide whether to address the hip first, spine first, or both. In various instances it is advisable to address the hip first. An extension contracture or a major hip flexion or adduction contracture should be released first. This may involve a simple soft tissue procedure (adductor tenotomy and/or abductor release) or a more complex surgery including muscle release, open reduction, femoral varus derotation osteotomy, femoral shortening, periacetabular osteotomy, and/or hip resection in severe cases. The spine surgery may follow these procedures after several months to allow for healing and recovery from the initial surgery.

In cases in which hip surgery will bring little or no benefit to the patient in terms of sitting and nursing care, the spine surgery should facilitate sitting and nursing care regardless of whether or not it addresses the hips. For instance, in the case of a mixed pelvic obliquity in which there is an abduction contracture and a suprapelvic obliquity in a CP sitter, the spine surgery will allow the patient to sit straight with the hips going to the side, and the hip surgery will give the patient little benefit.

Certain scenarios may be addressed simultaneously.
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A hamstring contracture may make knee extension very difficult after fusion to the pelvis. In this case it is possible to start the surgery with the patient supine or prone and release the tight hamstrings. This step will add another biomechanical advantage because the lumbosacral instrumentation will be less involved during knee extension in the postoperative period.

The neck position should be assessed both separately and in conjunction with the overall deformity. If the patient has a thoracolumbar kyphoscoliosis and the neck is in a position of fixed extension that compensates for the thoracolumbar kyphosis, the risk is to have the head tilted backward posteriorly after spinal instrumentation and correction of the kyphosis. On the other hand, if the head is in a kyphotic position, there is risk of junctional kyphosis proximal to the instrumented levels.

**Surgical Planning**

**Proximal Fixation**

It is common for patients with CP to have kyphotic deformity of the thoracic spine along with poor head control and spastic trunk muscles complicating sagittal balance. Fixation should extend proximally at least to T-3 to prevent upper thoracic junctional kyphosis or loss of proximal fixation due to the poor muscular control in these patients. However, despite fixation proximal to T-3, for patients with thoracic T5–12 kyphosis > 50°, it is strongly recommended to use alternative instrumentation to Luque-Galveston techniques due to the 36%–62% reported proximal fixation failure or junctional kyphosis when using this type of instrumentation. It should be noted, however, that there have been no reports that favor a distinct type of technique or instrumentation for the patient with hyperkyphotic CP.

**Fusion to Pelvis**

Most of the spastic deformities in this patient population would benefit from long segmental instrumentation to counteract the often sweeping and progressive deformity. A pertinent question then becomes whether to extend the fusion to the pelvis. The decision to fuse to the pelvis depends on the patient’s ambulatory status and the degree of pelvic obliquity present. Pelvic fixation to the sacrum, however, is technically challenging in the CP population due to osteopenia that is commonly found in nonambulatory patients. The complications of pelvic fixation include failure of instrumentation, loss of fixation, and pseudarthrosis, although modern techniques are available to address these outcomes.

For nonambulatory patients, fusion to the pelvis is indicated if there is significant pelvic obliquity, as there will be no functional loss related to ambulation and an improvement in sitting balance is expected. The advantage of fusing to the pelvis instead of L-5 is an improved ability to correct and maintain correction of pelvic obliquity. It has been our experience that stopping short of the sacrum in nonambulatory patients with CP and severe pelvic obliquity may lead to recurrence of pelvic obliquity (Fig. 5). However, stopping the fusion at L-4 or L-5 is indicated in very selected cases for nonambulatory patients with mild pelvic obliquity (< 15°) and little spasticity, as clinically stable spontaneous correction of mild pelvic obliquity has been shown with the use of lower lumbar pedicle screw instrumentation without pelvic fusion in patients with neuromuscular scoliosis.

If the patient is ambulatory and does not have pelvic obliquity, there is no indication to extend instrumentation to the pelvis. A mobile lumbosacral junction is believed to be essential to absorb angular and rotational movements of the trunk during gait and transfer activities. In most cases the patient requires the mobility of the lumbosacral junction to power gait. In this case, the fusion should follow the rules of idiopathic scoliosis with distal fixation proximal to the sacrum, with the exception that selective thoracic fusion as in Lenke Type 1C or 3C curves do not apply in neuromuscular patients due to the broad and progressive nature of the curve (Fig. 6).

If the patient is ambulatory and there is pelvic obliquity, extension of the fusion to the pelvis depends on the curve characteristics and the patient’s gait patterns. When

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**Fig. 5.** Radiographs and photographs obtained in an adolescent girl with a previous fusion down to L-5 performed 3 years prior. Postoperatively the pelvic obliquity was well corrected. A: Because the prior instrumentation did not include the sacrum, the pelvic obliquity recurred (posteroanterior radiograph). B: Correction with anterior release from L-2 to S-1 was performed. Multiple Smith-Petersen osteotomies posteriorly and the use of iliosacral screws according to Dubousset allowed complete correction of the pelvic obliquity (posteroanterior and lateral radiographs).
the pelvic obliquity is due to the thoracolumbar curve but the lumbosacral curve is not rotated and still supple, fusion may end caudally at the nonrotated vertebra, which is often L-4 or L-5, and occasionally L-3 (Fig. 7). The pelvic obliquity tends to disappear once the thoracolumbar curve is corrected in this case, as the lumbosacral curve is not structural and the “pelvic vertebra” is in line with the low-lumbar spine. When the pelvic obliquity is secondary to the lumbosacral curve, it is essential to observe how the patient ambulates. If the patient powers gait through the lumbosacral junction with significant lumbar flexion and extension moments to walk, fusion to the pelvis is not advisable to preserve mobility. Radiographic criteria can also assist in decision making; McCall and Hayes41 showed equivalent Cobb angle and pelvic obliquity correction with a unit rod fixated distally to L-5 via pedicle screws, compared with a unit rod fixed to the pelvis in patients with <15° of L-5 tilt (a line across the top of L-5 referenced to the intercrestal line). Furthermore, several authors79-80 have demonstrated the efficacy of stopping short of the pelvis

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**Fig. 6.** Radiographs of progressive scoliosis in a 13-year-old girl with CP who has remained ambulatory. A: Analysis of the patient’s images revealed no pelvic obliquity and the caudal lumbar spine shows no spinal rotation (posteroanterior and lateral views, left). The bending posteroanterior views (right) revealed excellent lumbar flexibility. There was thus no indication to extend the fusion to the sacrum. B: In this case the fusion was extended to L-4 (and could have possibly stopped at L-3). The patient’s walking capacity remained unchanged after the surgery (posteroanterior and lateral views).

**Fig. 7.** Radiographs showing mild pelvic obliquity in a male patient with CP who was ambulatory. A: The patient showed a significant sagittal imbalance with thoracolumbar kyphosis (posteroanterior and lateral views). Because he was able to walk we wanted to preserve the lumbosacral junction, but the kyphosis made an extensive fusion down to L-4 or L-5 mandatory. B: Because of the patient’s low physical demands and the fear of recurrence of pelvic obliquity, the fusion was extended down to L-5. Two years later the patient’s gait improved. The gait was made easier because the patient was rebalanced in the coronal and sagittal planes (posteroanterior and lateral views).
when using pedicle screw instrumentation in neuromuscular patients with limited pelvic obliquity (< 15°). On the contrary, if the patient does not power gait via the lumbar-sacral junction, fusion to the pelvis should be considered as it may improve correction of the pelvic obliquity and the overall stable balance of the patient.

Anterior Release

Another crucial question to consider is whether the patient would benefit from an anterior release. Traditionally, anterior release is considered in cases of severe kyphosis, large curves with a fixed spinal pelvic obliquity, and large stiff curves that are not correctable with traction or bending. Although the decision is made mostly on clinical judgment of curve stiffness, several authors have proposed discrete recommendations, including Vialle et al. who recommended anterior release if pelvic obliquity does not correct on frontal bending radiographs. Auerbach et al. and Swank et al. recommended anterior release for stiff thoracolumbar curves exceeding 70° or residual curve exceeding 50° on radiographs of side-bending. Another consideration is to perform an anterior release of the spine in a skeletally immature patient by disrupting the growth plate to prevent continued growth of the anterior column (the crankshaft phenomenon).

In extensive anterior spinal surgery, significant post-operative pulmonary complications are relatively common, including pneumonia, pneumothorax, atelectasis, and respiratory failure. Anterior surgery carries the risk of opening the chest cavity, which may often compound baseline compromised lung function. One-stage anterior and posterior spinal surgery also increases operative time and, hence, the amount of intraoperative bleeding. For anterior and posterior surgery in CP, the literature is inconclusive as to whether complication rates are improved when comparing 1-stage versus 2-stage surgeries. Tsirkos et al. performed a retrospective review exclusively of patients with CP, comparing the complication rates of 1-stage versus 2-stage surgeries, and found no significant difference in overall complications. However, the authors suggested that there was increased risk of surgical technical complications in the 1-stage group, and recommended 2-stage surgery for this patient population. Due to complications of prolonged surgery, careful patient selection for anterior release should be made. The need for anterior release should be reserved for highly stiff curves with significant pelvic obliquity, as Auerbach et al. concluded that posterior instrumentation with unit rods without anterior release in patients with curves < 72.1° and an average correction of 39.7% on radiographs showing bending demonstrated an excellent 74% pelvic obliquity correction without increased complications, compared with an anterior release and posterior instrumentation group of larger curve magnitude and stiffness.

The indications for anterior release are decreasing with the use of newer instrumentation and techniques. The use of posterior-only pedicle screw instrumentation in CP may decrease the need for anterior release, as Modi et al. concluded. A similar correction, maintenance of correction over a minimum 2-year follow-up, and similar complication rate were observed with all posterior pedicle screw instrumentations when compared with traditional unit rod instrumentation with or without anterior release in patients with a mean curve of 76.8° and pelvic obliquity of 9.2°. In an attempt to avoid the complications of anterior spine surgery, several surgeons have reported on techniques including posterior vertebral column resection and posterior multilevel vertebral osteotomies for use in rigid curves with > 50% curve correction and minimal complications. Nevertheless, the data are limited on these techniques and their application for patients with CP, and more information is needed to compare posterior-only osteotomies and posterior instrumentation to traditional anterior release and posterior instrumentation.

Surgical Options

Classically, the surgical treatment of neuromuscular scoliosis has been based on the use of Luque rods or a unit rod along with extension to the pelvis using the Galveston technique. If a Luque rod is used for the Galveston technique, it is bent such that approximately 6–8 cm of length is inserted along the thickest portion of the iliac wing, a path from the posterior superior iliac spine toward the anteroinferior iliac spine. Sublaminar wires are passed and secured to the rods, initially to the convex rod, then to the concave rod to provide gradual correction of the spinal deformity. A reduction of the concavity is performed by having an assistant manually push the trunk at the convexity, while the surgeon carefully pulls the affixed sublaminar wire at the apex and tightens it to a stabilized rod. This type of instrumentation has proven efficacy, is inexpensive, and is relatively simple and safe in experienced hands. The amount of correction provided by the Luque rods or the unit rod has in most series averaged 50%–75% of the Cobb angle, for an average initial Cobb angle of 60°–75°. Several studies compared the efficacy of using unit rod instrumentation for correction of neuromuscular scoliosis, and generally advocated the use of unit rods versus Luque rods due to improved curve and pelvic obliquity correction for flexible scoliosis. Sponseller et al. found that although unit rods compared with custom bent rods provided superior correction of pelvic obliquity, their use had higher complication rates of infection and length of intensive care unit stays. The unit rod instrumentation is a single rod with a proximal U-bend that is precontoured to include standard coronal and sagittal profiles. Reduction using the unit rod is obtained after bilateral pelvic fixation of the distal ends is achieved, and the affixed rod is used like a rudder to correct the coronal profile of the curve. Overall, the advantage of the Luque-Galveston techniques is that they are inexpensive compared with third-generation segmental instrumentation with pedicle screws and have a long history of proven stable outcomes. However, the disadvantages of the Luque-Galveston techniques should be acknowledged. The technique of taking down the ligament flavum at each level to pass multiple sublaminar wires for segmental fixation onto the rods may cause small epidural bleeds that can accumulate into significant blood loss. Furthermore, decortication is limited by the sublaminar wires and fusion may not ensue. With the Galveston tech-
nique of pelvic fixation, regardless of the type of rod used, potential long-term problems include loosening of distal fixation presenting as a windshield wiper sign at the level of the iliac wing, and risk of pseudarthroses from distal fixation failure. In cases of severe pelvic obliquity, robust fixation to the sacropelvis via sacral and iliac screws is needed to perform a cantilever maneuver to align the pelvis perpendicular to the trunk. The Luque-Galveston technique is not the ideal instrumentation for hyperkyphotic spinal deformities, secondary to its association with an increased risk of proximal and distal loss of fixation due to biomechanical cantilever forces in the kyphotic spine. Due to the need for robust pelvic fixation in cases of severe pelvic obliquity, along with the use of third-generation instrumentation, several alternative pelvic fixation methods to the Galveston technique have been developed. Iliosacral screws were first introduced by Dubousset’s group and were shown to offer stable fixation for as long as 5 years postoperatively. In this technique, 7-mm screws are inserted from the iliac wing into the S-1 pedicle at a medially oriented angle of 45°–60°. Medial orientation of S-1 screws is associated with increased pullout strength, making this a strong option for pelvic fixation, although the neurological complications can be as high as 10%. The use of iliac screws for pelvic fixation has been analyzed by several authors and has shown that iliac screws provide comparable correction of spinal curve and pelvic obliquity to Luque-Galveston techniques. The surgical principles of iliac screw fixation are similar to the Galveston technique, although in most cases S-1 pedicle screw fixation supplements iliac screws. Although a direct biomechanical comparison of current iliac screws and the Galveston technique has not been performed, Schwend et al. have confirmed biomechanical superiority in flexion failure loads of long iliac screws inserted in a way that spans the greater part of the length of the ilium when compared with the Galveston technique. Phillips et al. have also proposed that 2-iliac-screw fixation in each ilium decreases the complication of both proximal instrumentation failure and distal rod disengagement compared with single-screw fixations bilaterally. Arlet et al. devised another pelvic fixation method, called MW instrumentation, to provide a stable construct for pelvic obliquity in severe neuromuscular scoliosis (Fig. 8). The technique is a combination of iliosacral screws along with iliac screws inserted in a Galveston fashion. Using the MW technique, the authors have been able to manage extremely stiff pelvic obliquity without any anterior release. Using the MW fixation of pelvic obliquity correction, we achieved a correction rate similar to that of Galveston techniques (80% curve correction) for very large curves (90° average Cobb angle; Fig. 9). This was confirmed by Carroll et al. who found that patients undergoing a posterior spine fusion with MW fixation had a 30% better correction of the pelvic obliquity than the traditional Galveston construct.

The economic cost of third-generation spinal instrumentation is significant in the treatment of patients with neuromuscular scoliosis, compared with the less expensive Luque wires and Galveston instrumentation. However, the impact of adding an anterior release in stiff curves,
an imperfect correction, increased bleeding, and possible revisions for pseudarthroses may justify the choice of lumbar pedicle screws along with sacropelvic fixation, including iliac screw fixation or the MW construct. Further biomechanical and long-term prospective studies are needed that directly compare newer sacropelvic fixation systems including iliac screws or MW constructs to traditional Galveston techniques to draw conclusions for justification of newer techniques, which often translates into increased cost.

**Intraoperative Considerations**

Decreasing bleeding during surgery has become a major concern in these large operations. Patients with neuromuscular diseases have an almost 7-times higher risk of losing > 50% of their estimated total blood volume during scoliosis surgery. The use of vasodilators, desmopressin, hemodilution, cell saver, or thrombin activator medication should always be considered in these high-risk patients.

Positioning is the key in avoiding increased venous pressure. Not only must the patient’s abdomen be free of compression, but the surgeon must also anticipate any changes of the positioning that will occur during the surgical correction. The use of the different posts must therefore be judiciously positioned and often in an asymmetrical fashion to optimize the venous return. Traction can be used in the form of halo traction or even halofemoral traction to help reduce the pelvic obliquity. The presence of a hip flexion contracture may require positioning with the hips at 90°. Vialle et al. evaluated 110 patients and compared patients with their hip flexed using halo traction versus patients in the prone position using halo and lower-extremity traction to reduce pelvic obliquity preoperatively. These authors concluded that asymmetrical prone traction improved final scoliosis correction as well as pelvic obliquity correction. Intraoperative reduction by asymmetrical traction decreased the total duration of the anesthetic procedure.

The approach must be meticulous and use cautery rather than sharp dissection with the Cobb elevator. The use of sublaminar wires may increase epidural bleeding, and the surgeon should be cognizant of the increased bleeding risk with this technique. Another way to decrease the amount of bleeding is to perform a segmental exposure of the spine to implement compressive hemostasis. One may expose the lumbosacral spine first, insert the pedicle screws where appropriate, and apply temporary sharp towel clips to close the muscle fascia over sponges. Then, the thoracolumbar spine is exposed and instrumented and the spine is temporarily closed as in the lumbosacral spine. Lastly, the upper thoracic spine is approached and instrumented. Once the entire spine is instrumented, the towels clips are removed and rods are ready to be inserted in a less hemorrhagic surgical field.

A solid fusion can be achieved by different means in this group of patients. It is usually not possible to rely on the local bone to achieve a fusion. The iliac crest usually cannot be harvested because of the intrapelvic rods or screws and significant osteopenia in neuromuscular spine patients. It is therefore advised to use either a femoral head allograft that will be morselized or the medial cortex of the tibia as advocated by Zeller and colleagues. Zeller et al. reported no pseudarthrosis in his group of patients with neuromuscular scoliosis using a tibial graft and there was little morbidity from the tibial bone graft harvest, as these patients are nonambulatory. The use of an allograft has been widespread in neuromuscular scoliosis and appears to be effective. However, a recent report by Sponseller et al. identified an allograft as a significant risk factor for infection in neuromuscular scoliosis. The advent of bone morphogenetic protein, not yet FDA-approved for children, may play a role in the future for these extensive fusions to the pelvis. To date, concern for potential long-term side effects in children and prohibitive costs have limited its use.

When the patient has a baclofen pump, one should be careful about the positioning of the patient on the operative table. The shunt may have to be cut and the use of a shunt connector may be required at the end of the surgery. In most instances we prefer to slide the rods under

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**Fig. 9.** Radiographs obtained in a 14-year-old girl with CP and severe and stiff pelvic obliquity. **A:** The spinal curve was measured as 105° (posteroanterior view, left), and the pelvic obliquity as 45° (lateral view, right). **B:** On the side-bending posteroanterior radiographs obtained by the senior author, the curve corrects to only 70°; such a curve is a classic indication of anterior release. **C:** Thanks to a radical posterior release and an MW construct, a pure posterior correction manages to bring the pelvis back to a neutral position. After 2 years the correction was maintained (posteroanterior and lateral views).
the shunt so they do not interfere with the shunt itself. Naturally this is not possible if one uses a unit rod. In the case of a posterior laminectomy performed for selective rhizotomy, the only instrumentation option that remains is the use of pedicle screws, because hooks or sublaminar wires cannot be inserted.35

Postoperative Complications

The postoperative complication rate for scoliosis surgery in neuromuscular patients has been high, ranging from 44% to 80%, with a perioperative death rate of 0% to 7%.8,35 Postoperatively these patients are at risk for pulmonary complications including bronchoaspiration, atelectasis, and respiratory failure. The patients are also at risk for gastrointestinal complications including gastrointestinal bleeding, reflux, and bowel obstruction.

Infections are common occurrences postoperatively, including those of the respiratory tract, urinary tract, and the surgical wound. Lipton et al.36 reported a 24% rate of pulmonary infection, an 18% rate of urinary tract infection, and a 5% rate of wound infection in a series of 107 patients with CP who underwent spinal fusion. Similar rates were reported in other studies.5,6,8 In a recent multicenter review, Sponseller et al.8 identified significant wound infection rates of 6% deep and 4% superficial. The use of unit rods was associated with significantly higher rates of infection than custom bent rods, although a clear reason for this finding was unknown. The implications of deep infection may expand to also affect final coronal curve correction (53% correction for deep infection, 67% for no infection) possibly due to pseudarthrosis or compromised fixation secondary to infection. Borkhau et al.7 have proposed the use of an allograft impregnated with gentamicin to decrease the rate of deep infection from 15% to < 5%.

Appropriate nursing and prophylactic measures should be taken to prevent such complications. At our institution special mattresses are used to prevent pressure ulcers, and physical therapy is initiated the next day to promote early mobilization to prevent respiratory complications. The patient is encouraged to sit up to avoid aspiration and there is a low threshold for placing nasogastric tubes until adequate bowel function has returned. Depending on their nutritional status, patients are often provided with supplemental nutrition preoperatively through placement of a gastrostomy tube, which can be used effectively postoperatively. Foley catheters that are placed preoperatively should be left in place for 3–5 days and a barrier placed between the perineum and the incisional wound to avoid contamination. Aggressive preoperative treatment of urinary tract infections should be undertaken, and perioperative antibiotics should be administered and redosed appropriately throughout the surgical procedure. Gastrointestinal prophylaxis should be instituted early with proton pump inhibitors or H-2 blockers.

Other Postoperative Considerations

Modification of the home and sleeping arrangements should be started prior to discharge. The wheelchair will need adjustment after the surgical correction with a change in the Roho cushion, change in head straps, and arm supports and cushioning. Attention to padding is important to avoid complications such as coccygodynia.47 Reassessment of the hips should be performed to determine the need for tenotomies to address contractures that may become more prominent after surgery.

Postoperative pain control is paramount for patient and family satisfaction, as undertaking the surgical endeavor is demanding and often cannot be effectively communicated by the patient. The use of ketorolac can be implemented in the context of normal kidney function along with judicious use of narcotics that are dosed based on weight and prior patient exposure. As these patients often require a multidisciplinary approach to their care, a low threshold for a pain management consult would be wise.

Surgical Outcome

There have been serious questions regarding the validity of performing extensive operations on patients with CP who were totally dependent, and for whom benefit of the surgery could only be appreciated by their caregiver. In a survey of 37 institutionalized patients, Cassidy et al.14 found no clinically significant differences in pain or pulmonary medication utilization, therapy, decubitus ulceration, function, or time for daily care between surgical and nonsurgical groups. Nevertheless, the majority of health care workers believed that the patients who underwent fusion were more comfortable. More recent assessment by Jones et al.31 found that patient pain, happiness, frequency of feeling sick and tired, and parental satisfaction improved significantly by 1 year postoperatively. However, there were once again no significant changes between preoperative and postoperative assessments of physical function, school absence, and comorbidities. It therefore appears that postoperative improvements are mostly perceived by the caregiver for whom surgery represents a significant improvement. For the patient, we are still lacking an outcome tool that would demonstrate definitive improvement. However, it is the senior author’s opinion that patients with CP and severe scoliosis who have preserved or mildly deficient IQ benefit greatly from surgery because they are able to express a perceived benefit after the surgery. There is, therefore, no rational reason to deny these surgeries for more profoundly involved types of CP under the assumption that we cannot perceive the benefit of the surgery.

Conclusions

The surgical treatment of patients with CP has greatly benefited over the past 2 decades from improved patient selection, surgical technique, and stronger instrumentation. The Luque-Galveston instrumentation and technique that used to be the mainstay of treatment of pelvic obliquity is now being progressively replaced by third-generation instrumentation, although further comparative studies are needed, including a cost-effectiveness analysis. In the decision-making process regarding an operation, the non-ambulatory patient with CP can usually safely undergo fusion to the pelvis, and the ambulatory patients should
always be approached with caution in regard to sacropelvic fusion due to the potential of gait impairment if they use the lumbosacral junction to power gait. The results of surgery include improved patient balance and caretaker satisfaction, particularly when the overall coronal and sagittal balance of the spine is restored.

Disclosure

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