Management of Spinal Deformity in Cerebral Palsy

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Abstract: Scoliosis is common in cerebral palsy (CP), typified by rapidly progressive curves that impact patient function and quality of life. With age, these curves become rigid, with potential for sitting imbalance, decubitus ulcers, decreased socialisation, increased caregiver demands, and, in some cases, decreased pulmonary function. The use of bracing for scoliosis in CP is to support the collapsing spine rather than to prevent curve progression and should not be expected to alter natural history. Scoliosis correction surgery, however, is indicated for progressive curves greater than $40^\circ$-$50^\circ$, with the primary surgical goals being achieved with a balanced spine over a level pelvis, allowing for a more stable sitting platform and improved quality of life. Hip displacement and scoliosis are often coincident in CP, but the order of surgical management remains controversial. The mainstay of treatment for scoliosis involves posterior instrumentation and fusion from the upper thoracic spine to the pelvis. Though several options are available, the best evidence to date would suggest that segmental pedicle screw fixation achieves better curve correction and an improved risk profile over other implant choices. Often proposed as a benefit of scoliosis surgery in CP, the true impact of curve correction on pulmonary function has not been well studied and is currently unknown. Substantial comorbidities increase the perioperative risk profile—including swallowing difficulties, aspiration risk, recurrent respiratory infections, epilepsy, and malnutrition—necessitating patient counseling and mitigating strategies to optimize surgical outcomes. Optimizing medical and nutritional management pre- and perioperatively is important to tip the balance in favor of benefits over risks.

Key Points:
- Scoliosis is common in cerebral palsy with incidence correlated to disease severity, necessitating clinical and radiographic surveillance based on functional level.
- Scoliosis correction surgery is indicated for progressive curves greater than $40^\circ$-$50^\circ$, with the primary surgical goals being achieved with a balanced spine over a level pelvis.
- Substantial comorbidities increase the perioperative risk profile, necessitating patient counselling, and mitigating strategies to optimize surgical outcomes.
- Identifying and treating the causes of pelvic obliquity—both suprapelvic and infrapelvic in origin—are important to achieve optimal sitting balance.
- The best evidence to date would suggest that scoliosis surgery improves quality of life and is warranted in spite of the risks involved.
Introduction

Scoliosis is a common problem in cerebral palsy (CP) typified by rapid progression that often leads to large curve magnitudes even after skeletal maturity. With age, curves become more rigid, potentially leading to significant functional disabilities, including loss of sitting balance, decubitus ulcers, decreased socialisation abilities, increased caregiver demands, and, in some cases, decreased pulmonary function. Operative intervention is frequently prescribed to improve spinal and pelvic balance allowing for a more stable sitting platform and to improve quality of life. Patients requiring this procedure, however, have a high prevalence of associated comorbidities including swallowing difficulties, inherent risk of aspiration, recurrent respiratory infections, epilepsy, and malnutrition, that serve to increase the perioperative risks associated with surgical management of spinal deformity in CP. For this reason, the decision to proceed with operative intervention should be undertaken after clear communication of the risk/benefit balance so the patient and/or caregiver is aware of what to expect.

Incidence and the Gross Motor Function Classification System

The risk of developing scoliosis in children with CP has been long thought to be related to disease severity but, until recently, few studies have been available that evaluated the related incidence in a population-based study. Specifically, children with severe spasticity and quadriplegic limb pattern have been suggested to have the highest risk of developing scoliosis, while those with decreased neurologic impairment have a lower risk. Given the results of a recent population-based cohort study, the spastic motor type is most commonly seen in CP (86%), with one-third being of quadriplegic topography.

Figure 1. The Gross Motor Function Classification System (GMFCS) for children with cerebral palsy between 6 and 12 years old (expanded and revised). Copyright © Kerr Graham, Bill Reid, Kate Willoughby, and Adrienne Harvey, The Royal Children’s Hospital Melbourne, used with permission.
neurologic impairment, the Gross Motor Function Classification System (GMFCS) was established by Palisano and colleagues.9 (Figure 1). The GMFCS has subsequently been found to be a valid, reliable, stable, and clinically relevant method for the classification and prediction of motor function in CP and the standard by which these patients should be classified.10,11

Since its introduction, the GMFCS has shown tremendous utility in assessing the risk of developing musculoskeletal deformities. Using Kaplan-Meier survival curves, it has been determined that the diagnosis of scoliosis most often occurs after age 8 for all GMFCS levels.12 Children in GMFCS levels I or II had almost no risk of developing scoliosis, while GMFCS levels IV and V had a 50% risk of developing moderate to severe deformity. Although the authors did not specifically suggest a scoliosis surveillance program based on their results, for these GMFCS levels, it would be reasonable to suggest radiographic screening commence at age 8 years with an erect AP scoliosis X-ray taken annually until age 16 years when the risk of curve development plateaus. Under age 8, visual assessment of the back with the child in a supported sitting position should be performed with radiographs taken only with clinical suspicion. Future studies are required to confirm the utility of this suggested surveillance program.

Pathophysiology and Natural History

From a pathophysiological point of view, scoliosis associated with CP may be secondary to problems with weakness, coordination, and/or hypertonia of truncal musculature coupled with a lack of effective compensatory mechanisms.13 The most common curve type is long and C-shaped, often with associated pelvic obliquity, collapsing kyphosis, and loss of sitting balance (Figure 2).14 The natural history of scoliosis progression is thought to vary according to disease severity. Scoliotic curves associated with lower GMFCS levels tend to behave more idiopathic-like while higher GMFCS levels display a more typical ‘neuromuscular’ pattern and prognosis. Although flexible and easily accommodated by seating adjustments in younger children, the associated truncal deformity becomes more difficult to handle as the child matures and the scoliosis becomes more rigid.

In addition to the problems caused by deformity of the trunk, increased pelvic obliquity can also have a detrimental impact on seating comfort and, therefore, quality of life.15 The pelvic obliquity commonly associated with scoliosis in CP often coincides with a so-

Figure 2. ‘Long C’ shaped thoracolumbar scoliosis with associated pelvic obliquity in a child with spastic quadriplegia (GMFCS level V).
called “wind-swept hip” deformity, whereby the high-side hip is adducted and displaced (i.e., subluxated or dislocated) and the low-side hip is abducted and deeply seated into the acetabulum (Figure 3 a).16 Like the spine, this obliquity becomes fixed as the child advances through adolescence, a situation that can further compromise seating comfort with development of pain and decubitus ulceration. Concomitant impingement of the ipsilateral inferior costal margin against the ‘high-side’ iliac crest can also be a substantial source of pain for these children.17

The effect of scoliosis on respiratory function has not been well studied. Theoretically, development of neuromuscular scoliosis can deform the thoracic cage resulting in a decrease in effective lung volume, causing a restrictive pattern of pulmonary dysfunction. Indeed, children with scoliosis in higher GMFCS levels do experience a high frequency of respiratory infections but it is not known if this frequency is increased over children without scoliosis with the same demographics. One large prospective study from Sweden, however, reports improved respiratory function for children with non-progressive neuromuscular disorders (including CP) following scoliosis surgery.18

Recently, the use of ‘growth-friendly’ implants have been promoted for early-onset scoliosis in CP, with improvements of radiographic parameters evident, albeit with a high risk (30%) of deep wound infection.19 The impact of growing rods on respiratory function, however, is not currently known.

Figure 3. Addressing both infrapelvic and suprapelvic obliquity, a 12-year-old girl with spastic quadriplegia (GMFCS V) and windswept hip deformity (top left). She underwent bilateral releases of adductor longus, gracilis and iliopsoas, varus proximal femoral derotational osteotomies, and a left San Diego acetabuloplasty (top right), to address her ‘infra-pelvic’ obliquity. Following her hip surgery, she had persistent pelvic obliquity which affected her seating along with symptomatic right costo-pelvic impingement (bottom left). This ‘suprapelvic’ obliquity was addressed with a posterior instrumentation and fusion from T3 to the pelvis using segmental pedicle screw fixation and S2-alar sacroiliac screw fixation (bottom right). Following scoliosis correction, the child achieved comfortable wheelchair seating facilitated by a reasonably balanced spine over a level pelvis. Note the improved alignment of the hip joints post spinal surgery. Whether one should address the hips or spine first in these cases remains controversial.
Clinical Assessment

Clinical assessment of children with CP should start with determining their GMFCS level. From the clinical history, the child’s general health status should be elucidated, as it has relevance regarding their overall fitness for surgery and the expected perioperative risk profile. Specifically, the patient’s nutritional status should be ascertained as many of these children have related comorbidities including: malnutrition, gastroesophageal reflux disease (GERD), and swallowing dysfunction. As a result, these children are at significant risk of aspiration pneumonia, which can result in a cumulative decline in pulmonary status and frequent admissions to the intensive care unit (ICU). The initiation of G-Tube feeding and/or Nissan fundoplication may be warranted to improve nutritional status and decrease the risk of aspiration.

Many children with CP will have epilepsy, often requiring one or more anticonvulsants to achieve adequate seizure control. Some of these medications can impair both Vitamin D and calcium metabolism, that can lead to significant osteopenia. Preoperative administration of bisphosphonates may be considered to enhance bone quality in high-risk situations.

Valproic acid has also been reported to have an anticoagulation effect, which may lead to an increased risk of intraoperative blood loss and the need for perioperative transfusion (i.e., blood and clotting factors) during spinal surgery. Despite conflicting results in the available medical literature regarding the bleeding risk associated with anticonvulsant use, it is prudent to screen for coagulation abnormalities in these instances and, if present, consider alternative anticonvulsive medications where appropriate to normalize clotting prior to any surgical intervention.

Although difficult to ascertain in nonverbal patients, it is important to understand if the child has back pain associated with scoliosis. A recent study suggested that back pain in CP may be more prevalent than once thought, likely secondary to paraspinal muscle spasm and/or costopelvic impingement. Pain may be measured by parent proxy via a validated outcome measure such as the FLACC score.

For the musculoskeletal examination, the primary objectives are to: (1) determine the magnitude and flexibility of the spinal deformity, (2) identify the presence of fixed pelvic obliquity, (3) determine the presence of substantial lower limb muscle contractures, and (4) assess the adequacy of seating.

Determining the flexibility of the scoliosis, where present, is an important part of the clinical assessment since a flexible curve can often be initially managed by wheelchair supports and/or thoracolumbar bracing to allow for comfortable, upright sitting. Increasing curve magnitude during adolescence is often the trigger for...
surgical discussion, while intervention is most beneficial before increasing stiffness presents; often necessitating ancillary procedures (e.g. osteotomies, anterior releases) which increase the risk profile in this fragile population.24,25

To assess curve flexibility, the child is placed in a sitting position on the examining table with a parent facing and supporting the patient by gentle axial traction on the spine at the axillae bilaterally. With the back exposed, the clinician observes the spine from the opposite side of the examining table and notes the components of the spinal deformity, including the direction and location of the curve apex, any associated pelvic obliquity, and/or impingement of the lower rib cage on the iliac crest. Following this, the parent is asked to apply axial traction at the axillae while the clinician applies manual pressure at the apex of the deformity and at the ipsilateral pelvis (Figure 4). This technique allows for the application of 3-point pressure to reduce the truncal deformity. If the curve is easily straightened, it is said to be flexible. If not, the curve is said to be rigid, with variable severity.

Pelvic obliquity can be defined as a coronal rotation of the pelvis resulting in iliac crest height asymmetry which may be secondary to a supra-pelvic (i.e., thoracolumbar scoliosis) or infra-pelvic (i.e., wind-swept hip deformity) cause (Figure 3).26 The clinical assessment must include a thorough examination of the hips, specifically assessing restrictions in range of motion (primarily abduction) and for the presence of hip flexion contractures.

The type of wheelchair, its fit, and its associated supports should be assessed. The ability of these supports to maintain erect positioning and comfortable seating should be elucidated. The skin overlying ischial tuberosities, and the sacrum should be examined for signs of decubitus ulceration.

Radiographic Assessment
For the assessment of scoliosis, AP and lateral erect long film spinal x-rays should be performed. As these patients are typically non-ambulatory, these films are often taken in a sitting position. Cobb angle measurements should be performed for the major curve identified, to determine the magnitude of the scoliosis. Concomitant pelvic obliquity should also be measured, with an angle greater than 15° being considered clinically significant.27 Preoperative supine traction radiographs are useful to determine spinal flexibility for patients with severe scoliosis.28

Supine anteroposterior (AP) pelvis x-rays are also required to assess for the presence of hip displacement, both to identify a possible infra-pelvic cause of pelvic obliquity and to initiate early surgical treatment due to the adverse natural history (i.e., painful osteoarthritis) associated with untreated hip displacement (Figure 3).29,30,31

Nonoperative Treatment
Stable support of the scoliotic trunk allows for an upright posture, which facilitates more effective socialization, reduces pain secondary to decubiti, and allows the upper extremities to be free for functional use rather than as support to resist spinal collapse. Adaptive seating systems can be an important nonoperative method to improve patient positioning.32

The decision as to the type of wheelchair supports required depends mainly on the extent of truncal control and the flexibility of the curve. For flexible curves, the use of lateral truncal supports, specialized seating cushions for ischial padding, +/- an anterior chest ("butterfly") strap may be all that is required for a comfortable seating posture to be attained. More rigid curves may not be amenable to this solution, however, and may require a custom-molded seatback to accommodate, rather than correct, the spinal deformity. When this point is reached, these patients are typically
indicated for surgery given the progressive intolerance of wheelchair seating adjuncts.3

Unlike idiopathic scoliosis, the use of bracing for scoliosis in CP is to support the collapsing spine rather than to prevent curve progression and should not be expected to alter natural history. Bracing is used primarily for truncal support, especially in patients with an earlier onset of scoliosis.33 The use of a softer, more flexible, material has been found to be better tolerated with less chance of skin breakdown.34 To facilitate abdominal breathing and to accommodate a G-tube button, the brace should have an abdominal cut-out which allows for these functions.

Operative Treatment

Surgical correction has traditionally been indicated when the major curve exceeded a Cobb angle of 40° -50° and/or there was a significant functional limitations specifically with respect to sitting tolerance.3 Additional surgical goals center on the expectation of improvements in activities of daily living (e.g., dressing, independent ambulation, personal hygiene), absence of pain, ease of care-giving, and social interaction.15

To achieve these goals, the mainstay of treatment for scoliosis in CP involves posterior instrumentation and fusion (PIF) from the upper thoracic spine (typically T2 or T3) to the pelvis.13,14 The pelvis is most often included to reduce its obliquity, where present, making it more horizontal to allow for a better distribution of seating pressure between the ischial tuberosities. Regardless of implant type, the surgical goals remain the same: (1) to achieve a balanced spine over a level pelvis, (2) to arrest curve progression, and (3) to obtain a solid spinal fusion.

The standard posterior approach to the spine for scoliosis correction involves a midline incision from the upper thoracic spine to the sacrum. The paraspinal muscles are then subperiosteally stripped from the posterior vertebral elements, and anchoring implants are placed segmentally at each vertebral level. Rods made from either titanium, cobalt chrome, or stainless steel are contoured to the desired spinal correction and are subsequently reduced and fixed to the implants. Once the desired scoliosis correction has been achieved, the posterior vertebral elements are decorticated and morcellized bone graft is applied to the spine with the goal of achieving a solid bony fusion over all instrumented levels.

Many options for spinal fixation have been previously reported, including the use of sublaminar wires or bands, segmental pedicle screw fixation, and hybrid methods involving more than one implant type.26,35,36 These implants serve as anchor points for rod attachment which facilitate the straightening of the spine (Figure 5).3 The
best evidence to date would suggest that segmental pedicle screw fixation achieves better curve correction and an improved risk profile over sublaminar wire fixation, with decreases in blood loss and pseudoarthrosis rates being most notable.\textsuperscript{37,38}

Pelvic fixation can be reliably achieved by multiple means, with the Galveston technique, iliac screws, and sacral iliac (i.e., S2-alar) screws, being popular choices.\textsuperscript{3} Achieving stable fixation to the sacropelvis that resists loss of correction post-operatively is essential to surgical success.\textsuperscript{39} Though it has been suggested that the use of iliac screws over the smooth tines associated with the Galveston technique may diminish implant pullout and improve pelvic obliquity correction, a comparative study showed no significant differences in pelvic obliquity correction and similar complication rates.\textsuperscript{40} S2-alar iliac screws have been suggested to improve biomechanical stability over iliac screws and may give improved deformity correction with decreased implant failure rates (Figure 5).\textsuperscript{41,42,43}

With the advent of more powerful posterior spine constructs, specifically pedicle screws, there seems to be much less need for anterior surgical spinal releases.\textsuperscript{44} The disadvantages of anterior surgery are well-known, including increased surgical time and blood loss, need for a chest tube, and subsequent pulmonary difficulties due to the takedown of the diaphragm and the lung.\textsuperscript{45} Still, anterior releases are occasionally needed for large stiff curves. Flexibility can be assessed clinically as previously described or with the \textit{Miller flexibility test}.\textsuperscript{26} When anterior surgery is needed, staged procedures rather than single-day surgery may improve outcomes.\textsuperscript{34} Traction or distraction techniques can be used as an adjunctive deformity correction in lieu of anterior approaches.\textsuperscript{46} A combination of femoral-cranial traction, skin-traction, and Gardner-Wells tongs, or intraoperative rib-pelvis or spine-pelvis distraction have been described to avoid anterior releases.\textsuperscript{47} Indeed when compared to PIF alone, the use of intraoperative skull-femoral traction has been shown to significantly improve major curve correction and pelvic obliquity in CP.\textsuperscript{48} For any of these techniques, it is important to remember that the overall surgical goal, however, is to correct spinal balance in this very high-risk population and not to attempt complete Cobb angle correction.

Postoperatively, a short stay (1 to 2 days) in the intensive care unit (ICU) – primarily for fluid management – followed by 4 to 5 more days on the inpatient ward is typically required following these procedures. When medically stable, immediate mobilization out of bed up to the chair is ordered with no need for post-operative bracing or casting.

Patients with CP undergoing spinal fusion also often have a need for spasticity management with many also candidates for intrathecal baclofen (ITB) therapy. A large multicenter series reported that ITB does not significantly add to complication rates.\textsuperscript{49} The options when encountering a previously placed intrathecal catheter would be to either revise the catheter at the time of the surgery or to simply work around it. ITB can also be implanted during the same setting as the spinal fusion, with little change in postoperative course or complication rates.\textsuperscript{50} Keeping the patient lying flat postoperatively for 24 to 48 hours may help decrease the incidence of cerebrospinal fluid leak and/or symptomatic spinal headache after concomitant ITB pump insertion.

\textbf{Outcomes}

Despite high patient/caregiver satisfaction, scoliosis surgery in children with CP is fraught with high complication rates, likely related to the increased prevalence of comorbidities inherent to this patient population.\textsuperscript{3,51} Despite these risks, children with CP seem to be highly tolerant of spinal surgery, with a relatively long-predicted life expectancy postoperatively.\textsuperscript{52} Over the past few years, high-level evidence has been published which supports the role of spinal fusion in the improvement of QOL.\textsuperscript{3}

Recently, the development of the Caregiver Priorities & Child Health Index of Life with Disabilities (CP CHILD) questionnaire has provided a validated disease-
specific outcome measure to apply specifically to patients with CP.53 In a prospective longitudinal cohort multicenter study investigating children with severe CP who underwent scoliosis surgery, the authors found that by 12 months postoperatively, significant improvements in positioning/transfers, health, and overall QOL were achieved.54

These results were corroborated by a recent retrospective case-control study where children with severe CP (GMFCS IV and V) and scoliosis greater than 40° demonstrated significant postoperative improvements in overall CP CHILD scores, personal care/activities of daily living, positioning/transferring/mobility, comfort/emotions, and communication/social interactions, while the observational group deteriorated.22 In the surgical group, the complications included wound infections (22%), pneumonia (17%), reoperations due to post-surgical collections (12%), pneumothorax (6%), and recurrent hip dislocation (6%).

A large international, multicenter, prospective spine study group reported a large study of 212 patients with CP (GMFCS IV/V), where 74% of caretakers rated spine fusion as “the most beneficial intervention in their child's life” at 2-year follow-up.55 CPCHILD scores significantly improved at 2 years postop in 6 of 8 domains. This study group also performed an extensive evaluation for the risk of complications in patients at GMFCS V functional level and subsequently proposed a subclassification system of GMFCS V which allowed for risk stratification based on the following medical comorbidities: seizure disorder, gastrostomy tube, tracheostomy, and nonverbal status. This study showed significantly lower CPCHILD scores, and higher complication rates (specifically wound infection) with patients that had 3 comorbidities. Finally, that study group did a formal “risk-benefit” analysis that compared the improved quality of life with the risks of the complications and concluded that the surgery was worth the risks.

Conclusion
In conclusion, given the best evidence available, scoliosis correction improves quality of life in CP albeit with a high rate of complications. Prioritizing spinopelvic balance over aggressive Cobb angle correction and achieving a solid fusion are the primary goals of surgery. Optimizing medical management and instituting measures that serve to mitigate the risks of surgery are important to tip the balance in favor of benefits over risks.

References


