The management of scoliosis in children with cerebral palsy: a review

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Children who suffer with cerebral palsy (CP) have a significant chance of developing scoliosis during their early years and adolescence. The behavior of this scoliosis is closely associated with the severity of the CP disability and unlike idiopathic scoliosis, it continues to progress beyond skeletal maturity. Conservative measures may slow the progression of the curve, however, surgery remains the only definitive management option. Advances in surgical technique over the last 50 years have provided methods to effectively treat the deformity while also reducing complication rates. The increased risk of surgical complications with these complex patients make decisions about treatment challenging, however with careful pre-operative optimization and post-operative care, surgery can offer a significant improvement in quality of life. This review discusses the development of scoliosis in CP patient, evaluates conservative and surgical treatment options and assesses post-operative outcome.

Keywords: Cerebral palsy (CP); scoliosis; child; spine; treatment outcome

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Introduction

Cerebral palsy (CP) is defined as a permanent, non-progressive abnormality of motor function that is a result of injury to the developing brain (1). It can occur pre, peri or post-natally from a variety of causes. The term encompasses a heterogeneous group of conditions characterized by abnormal muscle tone, movement and posture. The incidence of CP is estimated at 2.0 per 1,000 live births in the UK (2). There is a strong link between CP and the development of scoliosis. It is estimated scoliosis occurs in between 21% and 64% of patients with CP (3-5). Spinal deformity is thought to occur before 10 years of age (5-7) and unlike in idiopathic scoliosis, has been shown to progress beyond skeletal maturity (8).

Risk factors for development of scoliosis

The development of scoliosis is strongly linked to the level of global disability caused by the CP. There are classification systems that describe the spectrum of disability that can occur under the umbrella term of CP. The most widely used of these is the Gross Motor Function Classification System (GMFCS), which splits children into 5 categories depending on their functional capacity (Table 1) (9). A large study conducted by Persson-Bunke et al. (5) highlights the statistically significant relationship between GMFCS level and development of scoliosis with 50% of children GMFCS IV–V developing a severe scoliosis. A Cobb angle of greater than 40° at an early age has been found to predict significant progression of a CP scoliosis (6,7). Furthermore an inverse relationship between development of scoliosis and ambulation has been suggested, with the least mobile patients at greatest risk (4,6). Gu et al. (7) suggested age was the most important risk factor and found no relationship between height and weight of children and curve progression.

The rate of progression of the scoliotic curves is variable, which in the adult, largely non ambulatory population, can range between 3.0° and 4.4° per year (10). Rate of progression was found to vary according to size of curve; larger curves (>50°) have been shown to progress almost twice as fast than smaller curves (<50°) (8). Curves were most likely to progress in non-ambulatory, quadriplegic patients (GMFCS IV and V) (8).
Etiology

The etiology of scoliosis in CP has yet to be well defined. Spinal deformity is thought to be associated with muscular imbalance around the spinal axis from either spastic or flaccid muscular weakness (11). In studies focusing on CP, the factors contributing to the development of spinal deformity have been suggested to include spasticity, muscle weakness and poor muscle control (12). In a meta-analysis of the risk factors for the development of scoliosis in CP the authors were unable to draw firm conclusions (13). The authors highlighted the lack of evidence and the poor methodological quality of the research.

Pattern of deformity

Two distinct patterns of scoliotic curves have been described in patients with CP (14). Group-I curves can be considered double curves with a thoracic and lumbar component and occurred most often in ambulatory patients, with minimal pelvic obliquity (14). Group-II curves are single curves in either the thoracic or lumbar spines and were of greater magnitude. They occur more frequently in quadriplegic patients and almost all display significant pelvic obliquity (14) (Figure 1). Pelvic obliquity may be defined as an angulation of the pelvis to the horizontal plane (15). Spinal deformity, hip contractures, leg length discrepancy or a combination of these factors will contribute to the cause of pelvic obliquity (16). Pelvic obliquity can be usefully categorized as supra-pelvic (spine/trunk disorder), pelvic or infra-pelvic (hip joint and lower limb) (17) aiding both diagnosis and management. CP has long been associated with the formation of pelvic obliquity (16,18) with large thoracolumbar curves contributing a supra-pelvic component and asymmetrical lower limb contractures recognized as an infra-pelvic cause. Consequences of untreated pelvic obliquity include the development of pressure points and decubitus ulceration, impaired sitting balance and significant hip joint deformity (17,19), hence making its diagnosis and consideration in treatment planning extremely important.

Non-surgical management

The aim of non-surgical management of scoliosis in CP is to improve sitting control and reduce or modify curve...
progression without the need for surgical intervention. Historical reports suggest the use of supportive bracing in children with CP was poorly tolerated and ineffective (3). There is a paucity of evidence for the use of modern bracing techniques. However, more recent studies suggest bracing improves sitting balance and trunk support, which provides better control of the head and neck as well as enhanced use of the upper limbs (20,21) as they are not required to support the trunk in the sitting position. Evidence for the use of orthoses to prevent scoliotic curve progression is mixed. Some authors have suggested bracing may slow curve progression (20), especially in younger patients with curves less than 40 degrees (20,22). Other groups have reported less success (21,22) and suggest braces may be beneficial as an interim measure before definitive surgical correction.

In non-ambulatory patients, methods of optimizing seating position have been shown to provide increased support and improve functional outcomes (23). Few studies have focused on the effect of seating systems on correction of spinal deformity. The placement of a 3-point system of lateral support pads was shown to offer a more symmetrical trunk posture and correct curve angles by 35% in non-ambulatory CP patients with scoliosis (24). Botulinum toxin injection has been used as an effective method for reducing spasticity in the limbs of patients with CP (25,26). Nuzzo et al. (27) administered botulinum toxin to a small patient population as a supplement to planned surgical therapy for CP patients with scoliosis. Reportedly, it did not worsen scoliosis and provided some reduction in magnitude of the curve in all patients.

Intrathecal baclofen (ITB) has been used for the treatment of global spasticity in a number of neuromuscular conditions and has been proven to be efficacious in patients with CP (28). The use of ITB pumps, which deliver a continuous infusion, offers improvement in spasticity, ease of care and reduction in pain (29). There is conflicting evidence as to whether ITB pumps can cause progression of a scoliosis. In a number of case series, a significant increase in Cobb angle was observed following ITB pump insertion (30,31). On the contrary, cohort studies using matched patients have shown there to be no difference in progression of scoliotic curves (32,33). The effect of an ITB pump at the time of scoliosis surgery is also controversial. There is an increase in the risk of infection, re-operation and re-hospitalization when compared to matched controls undergoing the same procedure (34). However, in another study by Borowski et al. (35) the insertion of ITB pumps before, during or after posterior spinal fusion had no significant effect on outcome. An economic analysis of the use of ITB has demonstrated it to be a cost effective method of reducing spasticity in CP patients in both the UK (36) and US (37,38) healthcare systems. The use of ITB remains controversial for the reasons outlined above but may be beneficial in patients with severe spasticity in which non-invasive treatments have failed.

Surgical management

Surgery remains the only option for the definitive management of scoliosis in CP. The aims of surgical correction include achieving a balanced spine, prevention of curve progression and improvement in functional quality of life. The timing of surgery should be considered on an individual case basis. Nonetheless surgery should be considered in those patients with large curves (>50°), in those continuing to progress beyond skeletal maturity and in significant curves resulting in functional or physiological disturbance.

Pre and peri-operative considerations

Pre-operative planning is an important consideration before embarking on scoliosis surgery. The complex nature of CP will often cause the child to have concurrent multi-system pathology that requires optimization. A comprehensive preoperative assessment is required including history, physical examination, laboratory and imaging investigations as well as discussion amongst a multi-disciplinary team (MDT). In our center the MDT comprises a scoliosis surgeon, general pediatrician, pediatric respiratory physician and physiotherapist, anesthetist and cardiologist.

Neurological

Pre-existing intra-cerebral lesions may cause seizure disorders in children with CP. The child therefore may be taking a combination of anti-seizure medication, which can have side effects and interactions with both anesthetic and analgesic agents that must be considered. Phenytoin, phenobarbital and sodium valproate have been shown to alter calcium absorption, leading to a decrease in bone mineral density (39), which may be significant when selecting placement and type of spinal instrumentation and increase the risk of failure through
implant pull out. Furthermore sodium valproate is known to cause abnormalities in clotting (40). In a study of 114 CP patients undergoing surgery a 26% increase in blood loss was observed in patients taking sodium valproate (41). Considering the major nature of corrective spinal surgery, abnormalities in clotting may risk serious adverse events. There have been reports of coagulopathies including disseminated intravascular coagulation (DIC) developing during major spinal surgery (42) therefore the clotting profile must be closely monitored before, during and after the surgery.

Respiratory

Post-operative complications involving the respiratory system occur frequently in children with CP (43). Abnormalities in pulmonary function secondary to factors such as poor upper airway tone, recurrent aspiration and thoracic cage deformity (44) add to this risk. A comprehensive evaluation of a child's respiratory system including thorough history and examination, laboratory testing and formal pulmonary function tests, are imperative to guide both pre-operative planning and peri-operative management. The use of pre-operative non-invasive ventilation (NIV) training to strengthen respiratory muscles has shown promise in improving outcomes in patients with neuromuscular disease following spinal surgery (45). Current practice demonstrates a movement away from long-term post-operative mechanical ventilation (46) owing to the advances of peri-operative medicine. NIV has been gaining popularity in the management of respiratory disease in pediatric patients with CP (47) and represents a safe and effective option to help mitigate against and manage respiratory complications.

Gastrointestinal

Gastrointestinal disorders in children with CP are common. Gastro-esophageal reflux disease (GORD) is present in up to 70% (48) of CP patients leading to a greatly enhanced risk of bronchopulmonary-aspiration (49), which is often significant on a background of poor respiratory reserve. Optimization of GORD in children with CP using surgical techniques such as fundoplication has been shown to be an effective way of controlling symptoms (50,51). However, the high risk of postoperative complications, morbidity and mortality from anti-reflux procedures must be considered (52-54). Children with CP often experience problems with feeding, leading to an inadequate oral intake and malnutrition (55). A low preoperative serum albumin has been shown to correlate with increased rates of postoperative complications and overall longer hospital stay (56). Thorough nutritional assessment is therefore required with consideration for the use of nutritional supplements or alternative feeding regimens. Intensive nutritional support has been shown to have a significant influence on nutritional status and body composition when administered over a 4 week period (57). Additionally, nasogastric tube feeding may be beneficial in those with swallowing difficulties however should only be used as a short-term measure (58).

Spinal

The past decades have seen major changes in surgery for spinal deformity leading to the development of modern pedicle screw fixation technique; a method that has now been widely adopted. The Harrington rod, initially created for the treatment of spinal deformity secondary to polio (59), represents one of the early methods of spinal fusion in neuromuscular spinal deformity (3). Although this method provided some correction of the spinal curve, complications were both frequent and significant (60,61). This technique was quickly superseded by segmental spinal instrumentation pioneered by Luque et al. (62) which made use of sublaminar wires inserted at each vertebral level to give enhanced immobilization and correction of the curve. The concurrent use of the Galveston method of fixing intramedullary rods into the iliac diaphysis was shown to enhance pelvic fixation and created a hybrid Luque-Galveston instrumentation technique. The initial results reported by Luque demonstrated an average correction of 72% over an 18-month follow up (62). A number of authors have since reported good results with similar instrumentation with a reduction of Cobb angle of between 46–65%, an improvement in pelvic obliquity and reduced rates of pseudarthrosis formation (63-67).

Modification of a single rod into a single U-shaped structure, used alongside Galveston pelvic fixation and sublaminar wires gave rise to the Unit-rod method of fixation. This procedure was first described in a mixed population with neuromuscular scoliosis and achieved an average curve correction of 54.6% (68). Further data has shown curve correction between 62-76% and a reduction in pelvic obliquity of 86-88% (60,64,69) however these studies are limited by relatively small numbers. A large
study by Tsirikos et al. (70) demonstrated a 68% reduction in scoliotic curve and a 71% reduction in pelvic obliquity in a series of 287 patients.

Other segmental systems including the Cotrel-Dubousset instrumentation have a limited evidence base for use in CP patients. A number of small studies have reported good outcomes, Piazzolla et al. (71) showed an improvement in Cobb angle of 57.2% with a reduction in pelvic obliquity of 58.9% which is comparable to Luque and unit rod systems. Furthermore, in a study of 60 patients followed up over 7 years, the authors demonstrated the Cotrel-Dubousset technique to offer effective, sustained scoliosis correction (72).

Advances in methods of spinal instrumentation produced the pedicle screw technique, offering enhanced correction of three-dimensional deformity (73) (Figure 2). Despite being widely accepted as a safe and reliable method of correcting adolescent idiopathic scoliosis, there have been few studies on the use of pedicile screws in neuromuscular scoliosis. Modi et al. (74) reported a satisfactory decrease in the magnitude of coronal and sagittal curves with a reduced rate of major complications over 3 years follow up. In a retrospective analysis of 45 patients on which pedicle screw instrumentation was performed, a favorable correction of scoliotic curve and pelvic obliquity was reported with a high carer/parent satisfaction rate (75). Although this data is promising, more work is required to assess the long-term efficacy of pedicle screw constructs.

Although the majority of patients with CP develop spinal deformity from the age of 10 onwards, there is a population of patients in which significant deformity occurs at an early age. Complete spinal fusion using the above techniques can cause problems when performed on skeletally immature patients, impairing the growth of the spine and thorax with effects on respiratory development and function. More recently technology has been developed to allow for the use of ‘growing rods’ to provide spinal support and correct deformity whilst allowing for growth. A study by McElroy et al. (76) represents the only published use of growing rods in patients with CP. Reports included a sustained improvement in Cobb angle and pelvic obliquity, however a significant complication rate, particularly from infection.

Anterior release and fusion procedures have been traditionally used for large, stiff curves and may, therefore, be considered for use in patients with CP. A number of indications for use of the anterior approach have been suggested including an inability to correct pelvic obliquity on forward flexion (77) and stiff thoracolumbar curves that exceed 70° on radiographic imaging (78,79). Furthermore, the disruption of the growth plate and subsequent inter body fusion achieved using the anterior approach, in skeletally immature patients, has been shown to prevent further growth of the anterior column and subsequent ‘crankshaft phenomenon’ (80). Access to the anterior spinal column can require opening of body cavities (thorax and abdomen) exposing patients to a greater risk of complications. Interestingly, in a small patient series, Auerbach et al. (78) demonstrated there to be no significant difference in the rate of postoperative complication between posterior only and anterior and posterior surgical techniques. Contemporary instrumentation techniques using pedicle screws have challenged the requirement for an anterior approach. The use of posterior-only pedicle screw constructs has been shown to offer excellent curve correction with a minimal complication rate (74). In fact, the increasing popularity of this instrumentation technique is diminishing the need for anterior release procedures. Moreover, alternative, posterior only techniques such as

Figure 2 Postoperative radiograph following pedicle screw instrumentation.
vertebral osteotomy (81) and vertebral column resection (82) have been reported in an attempt to manage large, stiff scoliotic curves.

There are currently no studies directly comparing the current methods of spinal instrumentation in the CP population. Second generation techniques as well as pedicle screw instrumentation have all been shown to provide effective correction of deformity in terms of Cobb angle correction and reduction in pelvic obliquity. Current practice in this center involves the use of all pedicle screw constructs with anterior release procedures reserved for large, stiff thoracolumbar curves aiming to prevent the need for pelvic fixation.

Complications

Postoperative complications from spinal surgery in patients with CP are common. The reported overall complication rate in the literature is variable, ranging between 17-68% (67,83-86). High risk of complication is associated with non-ambulatory status and greater angle of scoliosis curve (85,87). These patients often suffer from the greatest physical disability and a number of preoperative medical comorbidities, which may account for this increased risk. Complications have been reported to affect many body systems including respiratory, gastrointestinal and neurological. In a recent meta-analysis of complications following surgery for neuromuscular scoliosis, pulmonary complications were found to be most common (22.7%), followed by implant complications (12.5%) and infections (10.9%) (88). The authors suggested age at the time of surgery (<13 years) was associated with higher rates of neurological (15.1% compared to 3%) and pseudarthrosis (11.6% compared to 1.7%) (88). No explanation of this association was offered, however younger, smaller patients may cause an increased difficulty in placement of instrumentation therefore increasing risk of neural injury and failure of fusion.

Strategies to reduce complication rates in CP patients undergoing spinal fusion are sparsely reported. Contemporary evidence has suggested the use of vancomycin powder may reduce the rate of wound infection in spinal surgery. In a large retrospective review the overall infection rate was reported to be less than 1% (89) when using vancomycin powder. Interestingly, the only randomized control trial has shown there to be no significant difference in the rate of infection when using combination intravenous and intra-wound antibiotics compared to intravenous alone (90). Recent best practice guidelines advocate the use of intra-wound vancomycin in high-risk patients (91) such as those with CP.

Outcome

Measurement of outcome following corrective surgery in CP can be difficult. Assessing the opinion of children who have varying degrees of learning difficulty makes the use of traditional methods of measuring postoperative outcomes problematic. Surveys of patients and parents in the postoperative period suggest a high level of satisfaction following surgery, with a large majority of parents willing to recommend the procedure to others (92). Up to 99% of parents reported being satisfied with the outcome of the procedure with 85–94% willing to consider surgical intervention for their children again (83,93). The retrospective nature of these studies does open them to the influence of bias, with few reporting a pre-operative assessment as a comparator. Furthermore, targeting the opinions of parents who will often have made the ultimate decision about their child’s treatment may not provide an objective measure of outcome. Interestingly, in a comparative study between opinions of parents and caregivers (education professionals, therapists) of children with CP, Tsirkos et al. (92) demonstrated both groups noted significant improvement in both appearance and function following surgery.

On the other hand there are prospective studies that have shown that whilst parents remain satisfied with the postoperative outcome, surgery provides no improvement in function, school attendance or co-morbidities (94,95). Askin et al. (95) prospectively measured functional outcome in patients following scoliosis surgery and found there to be a decline in function over the first 6 months and no overall improvement 12 months postoperatively. Small patient numbers, a short follow up period and heterogeneous patient group may have influenced this data set.

Quality of life is perhaps the most important outcome measure in any postoperative CP patient. In a systematic review, the evidence suggests an improvement in postoperative quality of life in CP patients who underwent scoliosis surgery (96). Nevertheless, the authors commented there are conflicting reports and the literature is currently lacking well-controlled, prospective studies (96). It is therefore imperative that careful consideration of the risks and benefits of surgery takes place on an individual patient basis, with involvement of the patient, family and wider
members of the multi-disciplinary team.

**Conclusions**

Neuromuscular scoliosis is a common manifestation in children with CP. Without timely therapeutic intervention, scoliotic curves will continue to progress and cause impairment in function and increased risk of poor health. Management options are available that include the use of external bracing through to modern surgical techniques using segmental spinal fusion and pedicle screws. Current spinal instrumentation techniques offer a significant decrease in the magnitude of scoliotic curves and pelvic obliquity, which is sustained throughout long term follow up. As CP is a multi-system disease, careful consideration must be given to the preoperative optimization and the postoperative management of the child. A multi-disciplinary approach involving pediatric specialists will allow for this. Nonetheless, these patients remain at high risk of postoperative complications. Outcome following surgery is difficult to assess, however, parents and caregivers report satisfaction with the positional and functional improvements gained. The risks and benefits of all options must be extensively discussed with patients, their families and their caregivers before a decision is made. Surgically, posterior spinal fusion, which in the modern era is based on the pedicle screw construct, should be offered to children with large, progressive curves, which limit function and risk further morbidity. Thorough preoperative assessment should precede surgery to mitigate potential complications with pharmacological changes, respiratory support and anti-infective agents used where appropriate.

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**Footnote**

*Conflicts of Interest:* The authors have no conflicts of interest to declare.

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