Spinal deformity in paediatric patients with cerebral palsy

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KEYWORDS
Cerebral palsy; Scoliosis; Spasticity; Pelvic obliquity

Summary
Spinal deformity in children with cerebral palsy is common. The incidence is higher with spasticity, inversely proportionate to the level of ambulation. Scoliosis is commonly associated with significant pelvic obliquity, which decreases sitting tolerance, and causes pain from pelvic impingement on the thorax. There may be cardiopulmonary complications.

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Introduction
Cerebral palsy (CP) is a static encephalopathy affecting the immature brain which leads to permanent motor dysfunction. It is probably the most common neurological condition causing permanent physical disability in childhood. The classification of CP most commonly used by orthopaedic surgeons is based on the anatomic pattern of involvement.

- monoplegia: one limb is primarily affected;
- diplegia: predominantly affecting the lower extremities;
- hemiplegia: one side of the body is predominantly affected;
- triplegia: a hemiplegic pattern exists on one side with diplegia in the lower limbs;
- quadriplegia: all four limbs are affected.

Whole or total body involvement describes a patient with profound quadriplegic involvement, marked cognitive disability, and severe learning difficulties.

CP can be also categorized according to the most prominent motor disorder:

- spastic
- flaccid
- athetoid
- mixed types

Spasticity is a velocity-dependent increase in resistance to motion and represents the most common abnormality in motor tone that occurs in children with CP. Spastic and mixed forms account for more than 85% of cases.

Spinal deformity is a significant problem in children with CP. It most commonly affects those with spasticity, with an incidence directly proportional to the severity of neurological involvement.1–9 The overall incidence of spine deformity in CP is as high as 20–25%.10 It is considerably higher in non-ambulators and in patients with total body involvement.11,12 The curves usually do not respond to brace treatment, especially during the adolescent growth spurt, are progressive, and lead to both coronal and sagittal imbalance, often associated with pelvic obliquity.5,10,11,13–15
Pathophysiology

It is unclear which component of neurological deficit is more responsible for the development of spine decomposition. Spasticity, muscle weakness, incompetent muscle control, and poor balance are all contributory factors.

Functional deficits

The combination of abnormal spinal curvature and neurological impairment can significantly affect function; trunk imbalance and pelvic obliquity may impair walking capacity or sitting tolerance. In ambulators, spinal malalignment and trunk decompensation affect standing balance, limiting walking ability, and cause sitting intolerance in wheelchair-bound patients, converting them to hand-dependent sit- ters. Ambulators may need walking aids or become wheelchair-dependent. In a wheelchair-bound patient, pain may result from impingement of the ribs against the iliac crest on the concavity of the scoliosis. Pressure sores may develop especially if the child has insensitive skin. Non-ambulators have to rely increasingly on their upper limbs for body support, markedly decreasing their functional abilities and thus an increasing need for nursing care.

Additionally, scoliosis in these severely disabled children can cause cardiopulmonary complications, particularly when the curve magnitude is such that the patient can spend little time erect or sitting. The symptomatology in children with severe gastro-oesophageal reflux, swallowing disorders, and a history of aspiration may also get aggravated as the spine deformity progresses.

The purpose of this systematic review is to investigate the characteristics of spine deformity that develops in children with CP, to give guidelines for treatment, and to increase awareness of the complexity of these deformities and the coexistence of a multitude of associated medical problems that arise in this particular patient population.

Coronal deformity

Scoliosis is the most frequently encountered spine deformity in CP and can be associated with a sagittal imbalance ranging from hyperkyphosis to hyperlordosis.

Epidemiology

The incidence of scoliosis in patients with spastic diplegia is 5% compared to 65–74% in patients with spastic quadriplegia. In a review of 272 institutionalized patients with CP, Madigan and Wallace found an overall incidence of radiographically confirmed scoliotic curvatures of 64%.

Types of scoliosis

Lonstein and Akbarnia have defined two distinct scoliotic deformity patterns in patients with CP (Table 1).

Group 1 curves are single thoracic or double thoracic and lumbar curves resembling idiopathic scoliotic curvatures with a level pelvis. These are more common in patients with good ambulatory ability and less severe neurological deficits.

Group 2 curves are long thoracolumbar or lumbar C-shaped curves associated with pelvic obliquity, the elevated side of the pelvis being on the concavity of the curve. These curves correspond to the characteristic pattern of spinal malalignment usually seen in non-ambulatory patients. Group 2 curvatures can be further subdivided into 2A with the curve continuing into the sacrum, and 2B when the sacral vertebrae do not form part of the curve.

Sagittal deformity

Sagittal spine deformity in CP is rare and usually occurs with scoliosis. The prevalence has been quoted at 7% in patients with scoliosis. Patients with hamstring spasticity or fixed hamstring contracture followed by posterior pelvic inclination and decreased lumbar lordosis develop a compensatory thoracic hyperkyphosis. Children who have undergone dorsal rhizotomy through extensive laminectomies to address persistent lower limb muscle spasticity tend to develop a severe thoracolumbar kyphosis, which gives rise to pain. An additional cause of excessive kyphosis is the presence of generalized trunk hypotonia. As the kyphotic deformity progresses, the patient’s sitting ability and head control in a functional position become affected.

Hyperlordosis is seen in ambulatory patients, usually diplegics, with flexion contractures of the hips and anterior pelvic tilt. An isolated lumbar lordosis can also be a consequence of multi-level laminectomies performed during a dorsal rhizotomy. In rare cases, a rigid lumbar lordosis may be the result of tethering of the spinal cord. Children with lumbar hyperlordosis often complain of back pain and have an increased risk of developing isthmic spondylolysis and spondylolisthesis.

Clinical Presentation

Scoliosis in CP behaves very differently to the idiopathic type. Scoliotic curves may be structural or postural. Young children present, initially, with a flexible postural curve; a structural component develops with further growth. This distinction is important as surgical management can usually be postponed until late childhood or early adolescence. As children with CP enter their adolescent growth spurt, the rate of progression of the scoliotic curve increases dramatically up to 2–4 degrees/month and a rigid deformity develops rapidly. The rate of curve deterioration and the stiffness of scoliosis also depend on the extent of the patient’s neurological involvement.

Natural history

The progression of the scoliotic curve is not confined to the pubertal growth spurt; it is often influenced by muscle imbalance and gravity, and so significant deformity may appear earlier in life and progress beyond skeletal maturity into adulthood. It is also important to note that puberty may begin much earlier or later in a person with CP. Thometz and Simon reported progression of scoliosis beyond the end of skeletal growth, particularly if the curve...
was greater than 50° (1.4°/year), compared to 0.8°/year for curvatures of less than 50° at skeletal maturity. In other reports, scoliotic curvatures of more than 40° progressed at a rate of 4°/year, or to a mean of 80°.18 Even curves as low as 20° may continue to deteriorate into the adult life at a rate of 0.8°/year.17

Treatment of spinal deformity

The management of spinal deformity in CP is directed at maintaining or improving the functional abilities and quality of life of the child. Thus any treatment must be tailored to the individual patient and consider his/her specific circumstances. Treatment planning must also include a detailed risk–benefit assessment based on the severity of co-existing medical morbidities.

Non-operative treatment

Non-operative management of spine deformity in patients with CP includes seating adaptations and the use of orthotics. Unfortunately, none of these measures has a documented effect on curve progression or the final outcome of the spinal curvature. Thus the aim of all non-operative modalities is not to correct the deformity, but to preserve patients’ functional ability as the curve continues to develop.

Seating supports

These are useful in patients with CP as sitting balance can be improved and an upright posture maintained. They facilitate nursing care and may free the patients’ hands, improving function.9,10,25,26 We believe that appropriate sitting adjustments should be considered the mainstay of managing patients with scoliosis prior to surgical correction. The use of offset chest lateral rests that can be fitted on a wheelchair, which becomes the primary sitting device for those patients over time. Lateral supports need to be revised as the children grow to provide adequate trunk fixation. Shoulder harnesses and straps fitted to the wheelchair can be used for patients with thoracic hyperkyphotic deformities. As the kyphotic curve becomes more rigid, the back of the wheelchair may have to be moulded to accommodate the fixed deformity.

Bracing

A moulded thoracolumbosacral orthosis (TLSO) adapted from the treatment of patients with adolescent idiopathic scoliosis has been commonly used in good initial results.27,28Miller et al.17 followed 43 children with spastic quadriplegic CP and a neuromuscular scoliosis until spinal fusion. Twenty-one of these patients were managed with and 22 without a TLSO. The authors noticed no statistically significant difference between the rates of curve progression or the age at surgical intervention between the two groups demonstrating that there was no impact of the orthosis on the rate of curve progression, or the natural history of the deformity. While a recent study has supported a possible benefit in certain cases,29 the benefits of bracing are disputed in controlling the curve. However, bracing may be useful in the immature child to slow curve progression until the child can safely undergo spine surgery.30

It must not be forgotten that spinal braces are necessarily tight fitting and may cause considerable skin problems and respiratory compromise. Feeding can also be disturbed, particularly in children with pre-existing disorders and gastro-oesophageal reflux. Poor patient compliance is another frequent problem as the braces are uncomfortable and have to be worn for up to 23 h/day.

<table>
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<tr>
<th>Table 1</th>
<th>The classification of scoliotic curves in cerebral palsy.7</th>
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<tr>
<td><strong>Group 1</strong></td>
<td><strong>Group 2</strong></td>
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<tr>
<td>Single thoracic or double thoracic and lumbar curves resembling idiopathic scoliosis</td>
<td>Large C-shaped lumbar or thoracolumbar curves</td>
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<td>Absent or minimal pelvic obliquity</td>
<td>Marked pelvic obliquity</td>
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We believe that a more reasonable consideration would be the use of a soft underarm brace that can be applied and removed easily, and may improve the child’s sitting balance when they are not using their adapted wheelchair. The combination of seating adaptations built on the wheelchair with a non-rigid bi-valved underarm orthosis when the wheelchair is not used is the authors’ preferred method for young patients with curves that still retain their flexibility.

**Operative treatment**

The only management that addresses effectively spine deformity in patients with CP and has a well-documented positive impact in these children’s quality of life is spine fusion using instrumentation.

**Indications**

When considering surgical treatment, factors to be included are the size and flexibility of the curve in relation to the patient’s age and general medical condition, as well as the desires expressed by the parents or carers.

Surgical correction is indicated when:

- there is documented curve progression of greater than 10°,
- for curves greater than 45–50° in children 10 years of age or older;
- if there is considerable deterioration in the child’s level of function.\(^3\)\(^5\)\(^7\)\(^10\)\(^26\)

Surgery can be postponed for a later stage if the patient is still very young and the curvature remains flexible.

Patients with extremely severe deformities both in magnitude and stiffness, in whom even a combined anterior–posterior spinal arthrodesis would only achieve minimum correction, who also have a host of associated medical problems, including recurrent respiratory infections and poor feeding status, are at a greater risk of developing serious peri-operative complications. In such patients the risk of intra- or post-operative death outweighs the possible benefits from stabilizing the deformity and they should not be considered as good candidates for surgical intervention. On the other hand, spine surgery would be appropriate for patients with a lesser degree of deformity than set out above at skeletal maturity, due to the well-recognized risk of further deterioration into the adult life.

**Aims of surgery**

The objective is to restore trunk alignment, improve respiratory function, alleviate the pain caused by impingement of the ribs against the iliac crest on the concave side of the curve, provide better sitting tolerance in wheelchair-bound patients, prevent the development of pressure sores, and retain standing and walking abilities in ambulatory patients, thus maximizing overall level of function.\(^1\)\(^4\)\(^5\)\(^26\)

A successful outcome would include a balanced spine with his/her shoulders parallel to the pelvis and head and chest centred on top of the pelvis and a normal sagittal balance achieved so that the patient can hold his/her head in an upright position and the body weight is moved forward during sitting.

**Preoperative assessment**

Patients with CP can have multiple medical co-morbidities which need to be addressed prior to any surgical intervention. The surgeon must be a member of a larger multi-disciplinary team, which includes anaesthetists, neurologists, pulmonologists, gastroenterologists, dieticians, physiotherapists, occupational therapists, and intensive nursing care.

**Respiratory**

Pulmonary compromise is common in scoliotic patients with CP. Children with CP may have abnormal hypopharyngeal tone or anatomical abnormalities affecting their upper airway. When these anomalies are combined with the effects of a general anaesthetic (muscle relaxation and increased secretions) there is an increased risk of upper airway obstruction or respiratory failure.\(^3\) These patients will often have a poor coughing mechanism due to weak chest wall muscles. Respiratory failure and pneumonia occur much more frequently in patients undergoing spine fusion for neuromuscular scoliosis when compared to patients with idiopathic scoliosis.\(^3\)\(^1\) This must be considered when an anterior approach to the thoracic or thoracolumbar spine is necessary, as this significantly increases the risk of post-operative pneumonia, particularly if prolonged ventilation is required. Aggressive chest physiotherapy should be routinely used.

Gastro-oesophageal reflux should be suspected if the patient presents with a history of coughing bouts during feeding and recurrent chest infections. Such patients are at a higher risk of aspiration pneumonitis particularly if they have a poorly coordinated swallowing mechanism. The aspiration pneumonia can be silent and lead to progressive lung damage and fibrosis without early clinical symptoms.\(^2\) Hence preoperative chest radiographs should be routinely obtained to identify evidence of pulmonary fibrosis. While lung function tests would be helpful in detecting respiratory compromise, these are often impossible to perform due to poor patient cooperation.

Thoracic spine deformity may critically diminish pulmonary function; a thoracic lordosis will reduce the anteroposterior diameter of the chest, affecting the functional residual capacity and scoliosis may decrease the volume of one lung, impairing gas exchange and expectoration of secretions. Both predispose to recurrent chest infections.\(^9\)

These problems have obvious implications for the planning of major spine surgery in already compromised children. Close cooperation with paediatric pulmonologists is essential. Postoperative care in the Intensive Care Unit (ICU) is standard but prolonged ventilation post-surgery is rarely required. Preoperative tracheotomy is rarely indicated but may aid the care of children with complex upper airway problems. It may be necessary to aid weaning off the ventilator and aggressive pulmonary toilet can prevent chest infection.

**Developmental/nutritional**

Global developmental delay is common in patients with CP. Growth charts often show the child well below the 25th or
Feeding is often a significant problem and must be fully evaluated prior to surgical intervention. Attention must be paid to the previous feeding history, the consistency of foods ingested, and the child’s tolerance of oral feeds. As already mentioned, swallowing may be a problem because of uncoordinated muscle contraction or abnormal pharyngeal muscle tone. If the clinical history is suggestive of aspiration it may be necessary to perform swallowing studies.

Poor nutritional state predisposes the patient to delayed wound healing and a poor immunological response to infection. As malnutrition is a common finding in these children due to the combination of a poor diet, often deficient in essential vitamins, and a high metabolic demand due to recurrent chest infections or other co-morbidities, their nutritional status must be assessed prior to surgery with a detailed history, a thorough clinical examination, supplemented by haematological and biochemical tests. Readily available gross markers of nutrition include serum protein, albumin, and transferrin levels as well as peripheral lymphocyte count. Jevsevar and Karlín reported a significantly lower infection rate after spine arthrodesis, a shorter period of endotracheal intubation and less hospitalization time in patients with a preoperative serum albumin of 35 g/L or above and a total blood lymphocyte count of 1.5 cells x 10^9/L or above. Thus, surgical intervention should be postponed in the malnourished patient and preoperative supplemental nutrition may be necessary to optimize a candidate for surgery, preferably administered through the gastrointestinal route using nasogastric or nasojejunal feeding tubes. Night-time feeding by nasogastric tube may also be beneficial. A feeding gastrostomy may be required if tube feedings are not tolerated. In the presence of persistent gastro-oesophageal reflux, a Nissen fundoplication may alleviate the problem. However, if the reflux is not very severe to necessitate immediate treatment it might be better to postpone any surgical repair until after the spine is corrected, because the effect of the spine fusion on the gastro-oesophageal reflux can be totally unpredictable.

Gastritis and peptic ulceration are quite common in this group of patients. Gastrointestinal dysmotility is also frequent among children with CP, which again predisposes to severe gastro-oesophageal reflux and constipation. All these factors combined with a long general anaesthetic using neuromuscular blockade will contribute to severe gastro-oesophageal reflux and constipation. All patients with identified coagulopathies, those on anti-epileptic medication such as sodium valproate, have been shown to decrease von Willebrand’s factor causing a prolonged bleeding time. Phenobarbital has a similar effect and increases the risk of severe peri-operative bleeding. Liaison with a neurologist is important as these medications may be discontinued and an appropriate alternative commenced prior to surgical intervention.

When children are scheduled for surgery there must be at least one blood volume of blood typed and cross-matched. The intra-operative use of a cell saver system may limit the need for transfusion. Controlled hypotensive anaesthesia and meticulous haemostasis during tissue dissection are required to reduce blood loss. Early administration of fresh frozen plasma should be considered in the presence of a gradually increasing intra-operative blood loss, especially in patients with identified coagulopathies, those on anticonvulsant medication and patients with severe neurological involvement.

During surgery, transfusion of a significant volume of packed cells and clotting factors may be required. There are, however, many problems associated with large-volume blood transfusions; these include anaphylaxis and lesser febrile reactions, hyperkalaemia, hypocalcaemia, and hypothermia. A consumptive coagulopathy (disseminated intravascular coagulation) may be the result of large volume transfusions and must be avoided with the appropriate administration of clotting products.

**Haematological**

Intra-operative blood loss is a well-described problem in children with CP. Tsirikos et al. have previously reported a mean intra-operative blood loss of 2.9 L, corresponding to 1.2 blood volumes in a review of 288 consecutive patients undergoing spine fusion. Patients with CP may also have inherent or acquired disorders of coagulation. This needs to be identified preoperatively as blood loss during spine arthrodesis will be considerable. Anti-seizure agents, such as sodium valproate, have been shown to decrease von Willebrand’s factor causing a prolonged bleeding time. Phenobarbital has a similar effect and increases the risk of severe peri-operative bleeding. Liaison with a neurologist is important as these medications may be discontinued and an appropriate alternative commenced prior to surgical intervention.

**Neurological/metabolic/urinary**

Mental retardation, learning difficulties and behavioural problems are common in children with CP. These may complicate the postoperative care in patients undergoing spine surgery, as good compliance is generally required for chest physiotherapy, mobilization in and out of bed, toileting, and feeding. Considerable effort will be required to nurse these patients effectively and safely. Patients with ventriculo-peritoneal shunts should be assessed before and after spine surgery to confirm adequate function of the shunt.

Seizures are another frequent clinical feature of CP. Liaison with a paediatric neurologist is essential to optimize the patient for surgery and prevent uncontrolled seizure activity in the peri-operative period. The side effects of the chronic administration of anti-epileptic medication include the development of osteomalacia. Bone quality is often inherently poor, especially in quadriplegic patients because of disuse osteopenia. Poor bone quality increases the risk of intra-operative blood loss and instrumentation failure during spine fusion. Preoperative management of osteoporosis with the administration of intravenous bisphosphonates might be
a consideration, especially in non-ambulatory quadriplegics with marked osteoporosis, to maximize their bone quality before spine surgery.

Intrathecal baclofen therapy has been introduced to control severe spasticity unresponsive to oral anti-seizure agents. A recent study by Shah et al. has shown that the administration of botulinum toxin through an intrathecal baclofen pump does not increase the risk of developing progressive scoliosis in this group of patients.

Patients with CP have a greater prevalence of recurrent urinary tract infections due to urinary incontinence. The presence of an active urine infection at the time of the spinal fusion may increase the risk of a deep wound infection, with attendant risk to the surgery. Doubly incontinent patients also have a significant risk of developing deep wound infections caused by direct contamination.

Historical

The introduction of Harrington instrumentation was a tremendous evolution in the operative management of these complex deformities, but the incidence of pseudarthrosis and consequent curve recurrence remained high. Harrington instrumentation was used in the surgical management of neuromuscular scoliosis until the late 1970s but the distraction techniques applied through the Harrington system were difficult and often impractical in the soft bone of patients with quadriplegic CP.

In 1977, Luque developed the concept of segmental spinal fixation with the application of translational corrective forces through the use of multiple-level sublaminar wires and two single rods. This achieved a wide distribution of forces over each vertebra, with an increased initial spinal stabilization and a low risk of instrumentation failure.

This gave greater correction and decreased risk of implant failure. However, later reports indicated a rate of pseudarthrosis associated with the Luque system of up to 10%, instrumentation-related complications up to 21%, and curve progression postoperatively in up to 30% of the cases.

The Galveston technique of intramedullary placement of the rod in the iliac bed was developed by Allen and Ferguson and accomplished a secure pelvic fixation. However, it was soon recognized that the two unconnected soft rods introduced by Luque were moving independently, which was the reason why they failed to provide a stable fixation. The use of postoperative immobilization did not prove to resolve the problem. The Unit rod, a further development of the Luque-Galveston technique, introduced rigidly connected rods, which allowed improved correction of both spinal curvature and pelvic obliquity, together with restoration of coronal and sagittal trunk balance.

Other techniques

More recent instrumentation systems using pedicle screws have been introduced in the management of neuromuscular scoliosis. These are based on the same principle of segmental fixation with pedicle screws or hooks and the alternative of iliac bolts for lumbo-pelvic or sacro-iliac plates for sacro-pelvic fixation.

A common problem in the application of third generation instrumentation that use pedicle hooks or screws in patients with CP and marked associated osteopenia is poor vertebral fixation, which can limit the extent of corrective manoeuvres and may significantly increase the risk of pseudarthrosis. In our experience, in this group of children with considerably poor bone mineral quality, the laminae provide the strongest point of fixation compared to the pedicle or the vertebral body and can withstand segmental translational forces applied through sublaminar wires necessary to achieve correction of the deformity and balancing of the spine in both coronal and sagittal planes. On the other hand, if repeat surgery is required, e.g. to address a non-union, it is technically much easier and safer to revise an instrumentation system that uses pedicle screws with or without hooks as opposed to sublaminar wires.

As these modern techniques become common and widely utilized, it is essential to establish benchmarks for degree of deformity correction and complication rate related to the technical aspects of the individual procedure. As third generation spinal instrumentation is being widely utilized, there is also a significant increase in implant cost; this has to be balanced against possible benefits obtained by the use of these latest techniques.

We believe that, even in the advent of these more modern instrumentations, the Unit rod still remains the primary system for the treatment of patients with CP because it is simple to use, it is considerably cheaper than most other systems and can achieve good deformity correction with a low loss of correction, as well as a low prevalence of associated complications and a low re-operation rate (Fig. 1).

In the largest reported series of 288 patients with CP and spine deformity, spinal arthrodesis using the Unit rod instrumentation achieved correction of the scoliosis by 68% and of the pelvic obliquity by 73% with restoration of a normal sagittal alignment in both the thoracic and lumbar spine. A very satisfactory correction of spinal curvatures was maintained at a mean follow up of 3.2 years. There were no detected pseudarthroses and the overall number of reoperations for technical reasons was very low. It is also important to note that the Unit rod instrumentation construct used in this series costs our hospital the equivalent of less than two pedicle screws.

Surgical technique using the unit rod instrumentation

General

Thorough preoperative assessment (see earlier in this article) of the patients is performed, including evaluation of their respiratory capacity, cardiac function, immune system, coagulation mechanisms, nutritional status, feeding disorders, seizures, urinary system and their overall level of functional impairment. All patients receive prophylactic antibiotic treatment with the administration of a first-generation cephalosporin immediately before and for 24 h after surgery. Before anaesthesia induction, arterial and central venous lines are placed, and the central venous line is maintained until the second stage in patients who will have a two-stage procedure. A nasogastric tube is used to
decompress the stomach and a Foley catheter to monitor urinary output. Cell-saver is used intra-operatively, and the patients receive homologous blood transfusions at the discretion of the anaesthesiologist. Spinal cord monitoring with the use of somatosensory or motor-evoked potentials is applied during surgery in the ambulatory patients only.

We perform the anterior procedure in the lateral decubitus position, using a thoracic or thoracolumbar retroperitoneal approach, through the bed of a removed rib, depending on the apex of the deformity and the levels of anterior release that are required. The anterior approach allows for an extensive release of the anterior longitudinal ligament, complete annulectomy and discectomy, with the intention to provide angular and rotational mobility of the spinal segments, while at the same time enhancing anterior fusion in the excised disc spaces with the application of morsellized rib graft. No anterior instrumentation is used.

Patients who are in a poor medical state and have associated co-morbidities are selected to undergo a two-stage procedure under different anaesthetic sessions. In this group, after the anterior stage is completed, the patients are taken to the ICU, where hyperalimentation and aggressive pulmonary care are initiated until the posterior procedure 7–8 days later. The chest drain is removed when the drainage is less than 150 mL/day, usually on the third or fourth day after surgery.

Patients whose general medical condition will tolerate the second stage under the same anaesthetic are immediately rolled into the prone position after the anterior procedure is completed, and posterior fusion with the Unit rod instrumentation is performed. The Unit rod instrumentation is used to give segmental posterior fixation, with the spinal arthrodensis always extending from C7, T1, or T2 to the pelvis using the Galveston technique (Figs. 2–4). Decortication of the transverse processes and lateral laminae with facetectomy is performed and the posterior instrumentation is reinforced with abundant cadaveric bone graft mixed with autogenous graft harvested from the spinous processes. Iliac crest bone cannot be obtained in most of these patients due to various degrees of osteoporosis. Drains are not routinely placed, but meticulous closure of the lumbosacral fascia is essential to obliterate dead space and facilitate wound healing.

No postoperative immobilization or external trunk support is used. The patients are mobilized early to an upright position and are engaged to a physical therapy program. Their wheelchair is modified to adapt with the corrected seating posture.

Surgical considerations

Levels of fusion

The extent of the spinal fusion in patients with CP depends on the type of the scoliotic curve. For group 1 curves, the same fusion-level criteria are used as for idiopathic scoliosis. In idiopathic scoliosis it is better to fuse the curvature short using instrumentation, in order to achieve satisfactory curve correction and spinal stabilization, but also preserve as many mobile spinal segments as possible.

Long C-shaped thoracolumbar or lumbar curves that cause severe pelvic obliquity and usually occur in non-ambulators, require extension of the spinal fusion from the upper thoracic spine to the sacrum with pelvic fixation with the...
aim to restore truncal balance.\textsuperscript{1,4,10,26} The arthrodesis should not stop proximally in the mid-thoracic area, but should extend to T1 or T2 to prevent the development of an add-on kyphotic deformity proximal to the most cephalad instrumented level.\textsuperscript{10,26,49}

Whenever significant pelvic obliquity follows the scoliotic curvature, the instrumented fusion should include the pelvis to provide satisfactory correction of the spinal decompensation and reduce the risk of recurrent deformity.\textsuperscript{3–5,26} Recurrent distal scoliotic deformity requiring revision surgery and extension of the arthrodesis to include the lumbosacral joint has been previously reported in children with CP who underwent short initial fusions.\textsuperscript{47}

It has been traditionally believed that fusion to the pelvis should be avoided in ambulatory patients in whom the scoliotic deformity is only occasionally associated with pelvic obliquity.\textsuperscript{1,7,26,50–52} There are no published reports documenting that fusion to the pelvis will decrease the ambulatory capacity of children with CP. This widespread orthopaedic myth probably arose from the early attempts to instrument to the pelvis using straight Harrington rods, which removed all lumbar lordosis and created “lumbar flatback”. Additionally, many of the instrumentation systems initially used in the surgical management of neuromuscular scoliosis were not rigid enough to achieve a solid spine fusion without external trunk support and required long periods of bed rest and body casting.

A recent study of 24 patients has demonstrated that in ambulatory paediatric patients with spastic CP and neuromuscular scoliosis, spinal fusion with the Unit rod segmental instrumentation provided stable distal fixation to the pelvis, achieved satisfactory correction of the deformity, and restored trunk balance in both the coronal and sagittal plane, without compromising the patient’s ability to ambulate.\textsuperscript{53} We believe that with immediate postoperative mobilization and by preserving normal sagittal plane alignment, ambulatory status is not affected by spine fusion extending to the pelvis.

Pelvic obliquity

Pelvic obliquity is a fundamental difference between an idiopathic and a neuromuscular scoliosis and has to be addressed during the surgical management of the scoliotic curve. It can arise from spinal–femoral, pelvic–femoral or spinal–pelvic muscle contracture.\textsuperscript{26} It may develop as the result of extension of the scoliotic deformity into the pelvis\textsuperscript{54} or due to asymmetric contractures of the hip abductors that creates hip imbalance, as part of the hip “wind-swept” deformity. A combination of both factors can occasionally co-exist. Previous investigators have postulated that adductor contracture and hip subluxation may lead to pelvic obliquity, which is followed by the development of scoliosis.\textsuperscript{9}

Anterior surgery

Anterior release is intended to maximize the flexibility of the curvature and improve the correction using the posterior instrumentation. Thus it is indicated in the presence of severe curve stiffness. The flexibility of the curve can be assessed by obtaining longitudinal traction radiographs with the patient in the supine position or suspension anteroposterior views of the spine. Side-bending radiographs are not

\textbf{Figure 2} Preoperative anteroposterior (a) and lateral (b) radiographs of the spine of a 12-year-old patient with total body involvement CP showing a severe collapsing thoracolumbar C-shaped scoliosis measuring 130° with associated marked pelvic obliquity (black line). The patient had very poor sitting tolerance due to significant spinal pain as well as pain from impingement of the ribs against the left iliac crest on the concavity of the scoliosis.
reliably reproduced in this population of patients due to poor compliance. Radiographs with the patient in trunk hyperextension can be obtained to evaluate the flexibility of kyphotic deformities. Estimation of the rigidity of the curve can be also achieved using the physical examination side-bending test introduced by Freeman Miller with the patient bent against the examiner’s knee.

Combined anterior release/posterior fusion is indicated when curve correction to 40°–50° cannot be obtained, when there is a fixed pelvic obliquity of more than 10° or when the thorax cannot be balanced over the pelvis. Recent studies have proved the advantages of combined anterior–posterior procedures to obtain better deformity correction in more severe and rigid curves, through a circumferential spinal arthrodesis, minimizing the risk of pseudarthrosis and curve progression. Since anterior surgery is intended to increase flexibility of the curve and not to provide a fusion, the use of anterior instrumentation is not recommended, because this will limit the amount of correction than can be achieved through the posterior instrumentation.

Crankshaft phenomenon can occur in a very young patient who undergoes a posterior only spinal fusion for scoliosis. Although the back of the spine is fused, the front of the spine continues to grow causing the twisting characteristic of the fusion mass. The use of the original Luque instrumentation did not prevent the development of crankshaft. However, Smucker and Miller have followed 29 children with open triradiate cartilages who underwent spine fusion with the Unit rod system until the end of their growth and found no progression of the deformity.

**One or two-stage procedures**

There is much debate as to whether the anterior and posterior spinal arthrodesis should be staged. While there have been comparative studies of combined versus staged anterior–posterior spinal surgery in patients with neuromuscular scoliosis which mostly favour the single-stage procedures, they are all of mixed populations with different underlying neurological pathologies. Tsirikos et al. performed the only study on an isolated group of patients with CP. They showed that even though one-stage anterior–posterior spine operations in patients with spastic quadriplegia provided comparable results with the two-stage procedures in correcting spinal malalignment, they were
associated with a considerable mortality rate including three immediate postoperative deaths, increased operative morbidity and higher risk of technical complications. We believe that staged spinal procedures on different days is preferable in individuals with very large curves and concomitant medical illness.

**Sagittal deformities**

Lipton et al. have recently reported their experience in correcting sagittal plane spinal deformities without concomitant scoliosis in 24 patients. The authors identified deterioration in sitting balance and back pain as the principal indications for surgery. Children with sagittal curves of 70° or greater were more likely to develop severe symptoms; they reported two patients with hyperlordosis presenting with superior mesenteric artery syndrome and one with loss of bowel and bladder function. All of these problems resolved after surgical correction of the deformity.

Hyperkyphotic deformities associated with hamstring tightness are considerably improved after hamstring lengthening at an early stage. In the presence of an established kyphotic deformity causing functional problems posterior spine arthrodesis should be considered. This should extend from the cervico-thoracic junction to the pelvis to achieve maximum correction of the kyphotic curvature and prevent risk of proximal recurrent deformity and to control the posterior pelvic tilt. Rigid kyphotic curves need to be treated with anterior release followed by posterior spinal fusion.

Excessive kyphosis may affect venous return to the heart and attention must be paid intraoperatively to maintain haemodynamic stability while correcting the deformity. In an earlier previous study, Tsirikos et al. have shown that the presence of severe preoperative thoracic hyperkyphosis was the only deformity factor affecting survival rates and demonstrated statistically significant predictability for a poor life expectancy after spine fusion in children with CP.

**Lumbar hyperlordosis** secondary to hip flexion contractures can be treated initially by muscle lengthening procedures. Fixed lordotic curves require anterior closing wedge osteotomies prior to instrumented fusion. If there is an increased anterior pelvic tilt, the chance of the Unit rod penetrating the inner iliac table is significantly increased, either during insertion of the pelvic leg of the rod or whilst bending the rod in situ to correct the spinal curvature. This can be aggravated by the poor bone quality encountered in these patients. The use of pedicle screws in the lumbar spine may be necessary to allow for a better correction of the increased lordosis or to provide bony fixation in patients who underwent previous laminectomies, but placing pedicle screws if there have been previous dorsal rhizotomies is very demanding, as the posterior vertebral anatomy is distorted due to the multiple laminectomies.

In the presence of increased lordosis insertion of the Unit rod into the pelvis can be particularly difficult. To deal with this problem, the rod is cut on one side at the thoracolumbar region and the two pelvic legs inserted to the iliac bones separately. Then when pelvic fixation is achieved bilaterally, the two parts of the Unit rod can be then brought together and secured with rod connectors. When correction of a sagittal deformity is performed, the length of the Unit rod must be adjusted by 1–2 cm to allow for intra-operative lengthening (in hyperlordosis) or shortening (in hyperkyphosis) of the spinal column.

In the presence of lumbar hyperlordosis the surgical management of scoliosis becomes significantly more difficult.
and the posterior instrumentation is very technically challenging. In a recent study, excessive lumbar lordosis was associated with a high incidence of technical problems and an increased morbidity; patients with lordosis of more than 60° developed complications involving pelvic fixation of the Unit rod in 15.1% of the cases compared to 3.6% if the preoperative lordosis was less than 60°. Additionally, there was increased blood loss and operative time was prolonged.

Surgery for hyperlordosis is complicated and is the component of the deformity which is technically most difficult to address using the Unit rod construct.

Postoperative management

The patients are managed post-operatively in ICU and may require on assistive ventilation at least for the first night. This is normally necessary for patients with conditions such as tracheomalacia, pharyngeal dysfunction or after a complicated surgical procedure. Attention must be paid to fluid balance, pain management, and pulmonary care and close monitoring of the blood pressure, haemoglobin and electrolyte levels, blood coagulation times, and body temperature are very important. Nutritional support is essential and feedings should be started soon after surgery by central venous hyperalimentation until the patient is ready for gastrointestinal feeding. At that stage, the patients will need to be placed on nasogastric, nasojejunal or gastrostomy feeding depending on individual circumstances.

Aggressive physiotherapy is necessary to encourage pulmonary toilet, deep breathing, and expectoration.

The patient is mobilized in a newly adapted wheelchair as soon as possible. A reclining wheelchair can be used initially to provide better sitting comfort during the immediate postoperative period. The patient should be ready for discharge when the surgical wound is completely healed, is afebrile, sitting comfortably in a wheelchair, and feeding adequately. This is usually 7–10 days post-operatively. Full recovery should be anticipated by 6 months from surgery.

Postoperative complications

The best predictor for the development of postoperative complications is the degree of neurological disability. The study by Lipton et al. showed that the presence of other disorders such as poor nutrition, gastrostomy feedings, antiseizure medication, tracheal abnormalities or tracheostomy did not constitute additional risk factors. However, other investigators have suggested that inadequate nutritional status increases the risk of urinary infections, prolonged patient intubation and hospital stay.

Life expectancy/outcome after spine surgery

While obviously parents should be counselled about prognosis, there is limited information on the predicted survivorship of patients with CP. Affected individuals were previously assumed to have lower survival rates than the general population, but recent studies have shown substantially better survival rates even for the total-body involved patients, demonstrating clearly the intensive current medical support provided by the modern health systems. Thus CP should be considered as a disease with which one lives in contrast to the old belief that it should be regarded as a condition from which one dies. Nonetheless, predicting life expectancy for children and adolescents with CP is difficult.

One study has demonstrated a relatively long mean predicted survivorship of 11.2 years for children and adolescents with severe spastic CP and neuromuscular scoliosis who underwent spine surgery. The most accurate predictor for survival rates in this group was the number of days spent postoperatively in the ICU. This at least partially reflects general medical status of each patient, particularly related to impaired respiratory function.

The positive impact on the child of correcting the scoliotic deformity and maintaining a good coronal and sagittal spinal alignment is shown by a very high satisfaction rate among parents and professional caregivers, who appreciate the impact of spinal realignment surgery on the child’s level of function, sitting ability, physical appearance, comfort, and the ease of nursing care. In more recent studies, the vast majority of the patients (95.8%) and caregivers (84.3%) who were interviewed considered that the benefits from correcting scoliotic deformity undoubtedly offset the possible risks from surgery, and would have no hesitation in recommending this procedure for children with CP who develop significant scoliosis interfering with their level of function and their overall quality of life.

Summary

Spine surgery in paediatric patients with CP who have severe neurological compromise and complex medical problems is technically difficult and there is an increased risk of life-threatening complications. However, there is a well-documented positive impact on these children by correcting their deformity and maintaining a good coronal and sagittal spinal balance. Spinal arthrodesis is the only surgical procedure that has such a high satisfaction rate among parents and caregivers, especially for quadriplegic patients. With improved medical management and a multidisciplinary approach, life expectancy for this group is higher than previously reported and operative procedures to correct the spine have a definitive effect in improving the patients’ quality of life.

Key points

- Spinal deformity is a frequent orthopaedic problem in children and adolescents with CP
- The risk factors for developing scoliosis in patients with CP are spastic quadriplegia, absence of ambulatory function, early onset, and thoracolumbar C-shaped curve patterns.
- The only treatment that addresses effectively spine deformity in these patients and has a well-documented positive impact in their quality of life is spinal arthrodesis using instrumentation.
- Patients with CP may have a host of medical comorbidities, which will need to be addressed prior to...
scoliosis surgery and will require a well-coordinated multidisciplinary approach.

- Segmental spinal fixation with the use of the Unit rod instrumentation is simple to use, is considerably cheaper than newer third generation systems, and can provide optimum correction of both scoliosis and the associated pelvic obliquity in this group of patients with a low rate of complications and re-operations for technical problems.
- Life expectancy in children with CP is higher than previously reported and operative procedures to correct the spine have a definitive impact in improving their quality of life and are associated with a very high satisfaction rate among parents and professional caregivers.

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