Operative treatment for spinal deformities in cerebral palsy

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Abstract The higher the functional impairment, the more likely patients with cerebral palsy (cP) are to develop a scoliotic deformity. This is usually long-sweeping, C-shaped, and progressive in nature, since the causes of the deformity, such as muscular weakness, imbalance, and osteoporosis, persist through adulthood. In contrast to idiopathic scoliosis, not only is the spine deformed, the patient is also sick. This multimorbidity warrants a multidisciplinary approach with close involvement of the caregivers from the beginning. Brace treatment is usually ineffective or intolerable in light of the mostly stiff and severe deformities and the poor nutritional status. The pros and cons of surgical correction need to be weighed up when pelvic obliquity, subsequent loss of sitting balance, pressure sores, and pain due to impingement of the rib cage on the ileum become issues. General risks of, for example, pulmonary or urogenital infections, pulmonary failure, the need for a tracheostoma, permanent home ventilation, and death add to the particular surgery-related hazards, such as excessive bleeding, surgical site infections, pseudarthrosis, implant failure, and dural tears with leakage of cerebrospinal fluid. The overall complication rate averages around 25 %. From an orthopedic perspective, stiffness, marked deformities including sagittal profile disturbances and pelvic obliquity, as well as osteoporosis are the main challenges. In nonambulatory patients, long fusions from T2/T3 with forces distributed over all segments, low-profile anchors in areas of poor soft tissue coverage (sublaminar bands, wires), and strong lumbosacropelvic modern screw fixation in combination with meticulous fusion

techniques (facetectomies, laminar decortication, use of local autologous bone) and hemostasis can be employed to keep the rate of surgical and implant-related complications at an acceptably low level. Excessive posterior release techniques, osteotomies, or even vertebrectomies in cases of very severe short-angled deformity mostly prevent anterior one- or two-stage releases. Despite improved operative techniques and implants with predictable and satisfactory deformity corrections, the comorbidities and quality-of-life related issues demand a thorough preoperative, multidisciplinary decision-making process that takes ethical and economic aspects into consideration.

Keywords Scoliosis · Neuromuscular · Sitting balance · Pelvic obliquity · Cerebral palsy

Introduction

Cerebral palsy (cP) is the leading cause of neuromuscular scoliosis. In contrast to the most frequent type—adolescent idiopathic scoliosis, spinal deformities in nonambulatory cP patients are associated with many obstacles, problems, and complications [1]. Neuromuscular scoliosis is not just a growth disturbance in an otherwise healthy individual, but just one of many other issues in a patient with a chronic illness: spasticity, weakness, imbalance, and inactivity-related osteoporosis (creeping fractures) contribute to curve occurrence and fast progression (collapsing spine). Comorbidities comprise seizures, difficulties with swallowing, recurrent airway and urogenital infections, and poor nutritional status. Most patients have had previous surgery (hips, Baclofen pump, rhizotomies, feeding tubes) [2]. Hence, the risk of further curve progression persists through adulthood. The resulting curve is typically long-

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sweeping, C-shaped, and stiff, with subsequent obliquity of the pelvis and deformity of the thoracic cage. In most cases the sagittal profile is hyperkyphotic [2, 3].

Indications for surgery

The goals of surgery are a level pelvis, brace-free balanced sitting, and the halting of progression through instrumented solid long fusion and pain reduction. Correction should be discussed if positional discomfort and sitting imbalance progress despite seating adjustments and bracing. The loss of coronal and sagittal balance entails a loss of head control. Severely affected cP patients are not able to communicate verbally, but mimics are interpreted by their caregivers. With increasing scoliosis and head tilt, this last resort for communication is also lost. Bracing may delay but does not prevent surgery: in deformities of >50 – 60° , translational forces become increasingly ineffective [4]. Spinal orthosis incorporating axial distraction forces such as Milwaukee braces are not tolerated. The bigger and stiffer the curve, the poorer the nutritional status, and the more pronounced the thoracic deformity and the pelvic obliquity, the more likely it will be that brace treatment is not an option from the beginning. Marked pelvic obliquity leads to painful impingement of the rib cage on the ileum (see Figs. 1 and 2). The skin in the thoracopelvic crease gets moist and superinfected. Delaying surgery results in severe deformities, morbidity, a higher complication rate, and less satisfying outcomes [5].

Fig. 1 A 16-year-old boy with pain, a pressure sore, and marked deformity of the left hemithorax due to pelvic obliquity and subsequent rib-to-pelvic impingement

Diagnostic workup

A thorough history should include questions regarding previous treatment, pain, recurrent urinary and pulmonary infections, seizures, current medication, and sleep disturbance. Questions regarding choking and the ability to cough and swallow may reveal a tendency to aspirate. A multidisciplinary comprehensive approach is of the utmost importance. Caregivers and therapists (physio-, ergo-, and speech therapists) ought to be involved in the decision-making process since they experience the daily struggles and hazards encountered by the patient much more than the orthopedic surgeon does. Special emphasis should be placed on the assessment of functional level by an experienced physiotherapist. The probability of developing a progressive spine deformity is directly linked to motor ability. Gross motor function testing (GMFM) allows for risk assessment and observation over time. A thorough neurological exam serves as a preoperative baseline. In addition, it determines the amount of active, targeted function of the upper extremity. Both of these inform the decision of whether to use intraoperative spinal cord monitoring or not. Nutritional evaluation and bleeding studies are important, particularly for those on valproic acid medication, known for its interaction with platelet function [2]. Pulmonary function testing is not possible in those noncompliant patients. Polysomnography and echocardiography may therefore be helpful. Surgical site infection is the most frequent postoperative complication. Potential sources such as feeding tubes, urinary catheters and recurrent pulmonary infections should be taken into account. Skin sores or moist skin creases need to be recognized and treated. Many patients are in puberty and display acne vulgaris. *Propionibacterium* acne is found in low-grade late infections after spinal fusion [6, 7]. We routinely recommend eradication by topical or systemic pre-treatment. The core medical team, including the orthopedic surgeon, intensive care specialist, pulmonologist, and anesthesiologist, should be frank about the risk of death, prolonged intubation, tracheostoma, and home ventilation.

Clinically, the patient is assessed in their wheelchair, sitting on the bedside, and in a supine position. In the wheelchair, the overall balance of the trunk, head control, the flexibility of the cervical spine, and the pressure distribution on the sitting surface is checked. Severe pelvic obliquity leads to pressure concentration, pain, and even pressure sores. Longitudinal gentle traction under the shoulders provides an approximate idea of the curve correctability in both the coronal and sagittal planes. Long-standing severe curves lead to asymmetric shortening of the neck muscles, which may become more apparent when the trunk is corrected. Radiography includes long cassette

Fig. 2 An 11-year-old girl with cerebral palsy, loss of sitting balance, and painful rib–pelvic impingement **a**. Progressive pelvic obliquity (40°) and right convex thoracolumbar stiff curve (120°) with a compensatory left convex thoracic curve **b**, **c**. Two-stage (1 week

interval) anterior lumbar release and posterior multi-segmental hybrid construct consisting of polyaxial lumbar pedicle screws, iliosacral screws, thoracic sublaminar (Luque) wires, and a proximal claw (pedicle and transverse process hook)

standard anteroposterior and lateral radiographs, if possible in a sitting position to display the spinal collapse. Supine traction view (manual gentle traction on the legs and countertraction below the shoulders or with a head halter and pelvic straps) yields information about spine flexibility and correctability of the pelvic obliquity (angle between a horizontal line and the line to the top of the iliac crests) and the need for osteotomies or an anterior release. We do not recommend routine spine or skull magnetic resonance tomography. A CT scan with 3D reconstruction is helpful in case of revision surgery, to ease the planning of osteotomies, or to assess the sacropelvic anatomy in severe curves. In the supine position, contractures of the hips and knees as well as compensatory lumbar hyperlordosis become apparent. Windshield wiper deformities, adductor contractures, and dislocated hips are at least partially secondary to the spine and pelvic asymmetry. Pelvic balance needs to be restored before open hip reduction and multi-level releases to provide a basis that is as anatomic as possible.

Technical considerations for instrumented fusion

Curves of $>60^\circ$ respond very well to axial traction. Severe curves ($>90^\circ$) may warrant preoperative halo traction [3]. Prone positioning and intraoperative skull–femoral traction (Gardner–Wells tongue, supracondylar Schanz screws) with asymmetric pull on the legs provide a 30–60 % curve

correction, which reduces the correction forces at the implant–bone interface [8]. Most cP patients display flexion contractures of their hip and knee joints. Traction is not recommended in severe cases since it extends the pelvis and provokes lumbar hyperlordosis. Positioning a severely contracted patient on a standard table will pose problems. Therefore, a frame-type spine table (e.g., Jackson table) where the knee can be positioned below the table is mandatory to achieve the desired sagittal balancing. The utmost care must be taken to avoid any pressure sores. Apart from intraoperative skull–femoral traction, forces between the implant and the osteoporotic bone are reduced by implementing an extensive posterior release, including supra- and interspinous ligaments, the flavum, and the facets. In very severe and stiff curves, osteotomies (Ponte, Smith–Peterson) and—in selected cases (sharp angular kyphoscoliosis) vertebrectomies (vertebral column resections)—are valuable alternatives for preventing prior open release (periapical discectomies) [2, 3, 9, 10]. The long lever arm of the rigidly instrumented spine (upper T-spine to pelvis) places high demands on the fusion technique, particularly at the lumbosacral junction. Meticulous facetectomies and autologous bone harvested from laminae and spinous processes (burr or chisel) in combination with stable multi-level fixation prevent pseudarthrosis and revision surgery. The classic Galveston–Luque technique—introduced in the 1970s—consists of multilevel sublaminar steel wires in combination with a triple bent smooth steel rod that are anatomically placed in the same area as iliac screws are

placed nowadays. Though the corrective potential in the coronal plane is comparable to that of modern screw constructs, the stability is less, as witnessed by windshield wiper osteolysis beneath the iliac rods. Less stability is associated with a higher pain level, delayed mobilization, and a higher risk of pseudarthrosis and implant failure compared to stiff all-pedicle-screw constructs. However, it is still the cheapest and most low profile construct available. The bigger and stiffer the curve, and the softer the bone, the more advantageous multipoint fixation with polyaxial screws becomes [11]. Implant profile height is an important issue at the level of the thoracic spine and at the ileum, since most of the cP patients are underweight. Hybrid strategies unify the advantages of sublaminar wires or acrylic bands for the thoracic spine and polysegmental screw fixation of the ileum, sacrum, and lumbar spine [12]: the former provides easy continuous rod reduction and a low profile, and the latter strong fixation for optimal reduction of pelvic obliquity, which is crucial to regaining trunk balance and for controlling the long lever arm of a fused spine. In order to achieve optimal screw purchase, surgeons must be aware of any pelvic asymmetries (thickness and orientation of the ileum), mainly in patients with windswept hips [13]. Traditional screw paths enter at the posterior iliac spine and pass 1–2 cm above the sciatic notch, where thickness and density is optimal. Double iliac screws provide more stability but may be difficult to place [14]. The screw diameter is usually 6–7 mm and the length 50–80 mm. In any case, careful burial of the screw head beneath the level of the crest prevents skin breakdown. However, the ileum is sometimes too thin and distorted to allow for straightforward screw placement. In such cases, the sacroiliac technique along a trajectory from the posterior sacral surface towards the iliac wing or the outside-in technique from the outer table of the ileum into the sacrum provide alternatives [9, 15]. Mean corrections of coronal plane deformity and pelvic obliquity are 50–70 % in most series [2, 3, 10, 15].

Complications

Among all types of scoliosis, neuromuscular deformities entail the highest morbidity and mortality rates [1]. Though second-generation instrumentation and improved perioperative management have lowered the risk, the endeavor of spine surgery in cP patients is still burdened with average complication rates of 8–33 %, including death (around 1 %) and deep infections, increased hospital stay, and revision surgery [1, 3, 9, 16]. The risk of surgical site infection is 4–10 %, which is about threefold higher than for idiopathic scoliosis [1, 17]: risk factors are long operation time, significant bleeding (reported averages are around

700–2,000 ml, but can reach up to 5,500 ml, particularly for vertebral column resections or 260 % of total blood volume) with the need for transfusion, use of allograft bone, a ventriculoperitoneal shunt, severe cognitive impairment and neurological involvement, as well as malnutrition (serum albumin <3.5 mg/dl, lymphocyte count <1,500/ml), poor soft tissue coverage, and concomitant urinary, skin, or pulmonary infection [2, 10]. In the case of wound dehiscence, a dural tear (cerebrospinal fluid = CSF leak) has to be considered. Perioperative antibiotic coverage with repeat doses every 6 h and continuation until the suction drains are removed, repeat irrigation during surgery, careful soft tissue handling, meticulous surgical hemostasis supported by antihemorrhagic agents and changes of gown, masks, and gloves after 3–4 h are among the most important preventive measures [2]. Careful correction of hyperkyphosis and instrumentation up to T2/T3 with a local hook clamp (pedicle hook and transverse process hook) for smooth transition between fused and unfused areas support the prevention of junctional kyphosis [2]. The pseudarthrosis rate was around 10 % in the first-generation dual or unit Luque–Galveston technique. This represented the most frequent late complication and was usually related to implant problems [2]. With modern pedicle screw fixation and stable sacropelvic instrumentation it has become rare and is usually associated with deep infection.

Conclusions

Progressive structural scoliosis with pelvic obliquity and loss of sitting balance in a nonambulatory cP patient warrants long posterior fusion from the upper T-spine to the sacrum and pelvis. Modern sacropelvic fixation and the cantilever double-rod reduction technique is the most effective and safe way of correcting pelvic obliquity [18]. One- or two-stage anterior surgery should be avoided to minimize additional morbidity. Multipoint polyaxial pedicle screw fixation to distribute forces and skull–femoral traction, posterior release techniques, osteotomies, and vertebrectomies to diminish corrective forces yield satisfying corrections and outcomes. Severe deformities and late surgery provoke more complications and less satisfaction [5].

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References

1. Reames DL, Smith JS, Fu KM et al (2011) Complications in the surgical treatment of 19,360 cases of paediatric scoliosis: a review of the Scoliosis Research Society Morbidity and Mortality Database. *Spine (Phila Pa 1976)* 36:1484–1491

2. Lonstein JE, Koop SE, Novachek TF, et al (2012) Results and complications following spinal fusion for neuromuscular scoliosis in cerebral palsy and static encephalopathy using Luque–Galveston instrumentation: experience in 93 patients. *Spine (Phila Pa 1976)* 37(7):583–591
3. Piazzolla A, Solarino G, De Giorgi S et al (2011) Cotrel–Dubousset instrumentation in neuromuscular scoliosis. *Eur Spine J* 20(Suppl 1):S75–S84
4. White AA 3rd, Panjabi MM (1976) The clinical biomechanics of scoliosis. *Clin Orthop Relat Res* 118:100–112
5. Watanabe K, Lenke LG, Daubs MD et al (2009) Is spine deformity surgery in patients with spastic cerebral palsy truly beneficial? A patient/parent evaluation. *Spine (Phila Pa 1976)* 34:2222–2232
6. Bemer P, Corvec S, Tariel S et al (2008) Significance of *Propionibacterium acnes*-positive samples in spinal instrumentation. *Spine (Phila Pa 1976)* 33:E971–E976
7. Hahn F, Zbinden R, Min K (2005) Late implant infections caused by *Propionibacterium acnes* in scoliosis surgery. *Eur Spine J* 14:783–788
8. Jhaveri SN, Zeller R, Miller S et al (2009) The effect of intra-operative skeletal (skull femoral) traction on apical vertebral rotation. *Eur Spine J* 18:352–356
9. Sponseller PD, Zimmerman RM, Ko PS et al (2010) Low profile pelvic fixation with the sacral alar iliac technique in the paediatric population improves results at two-year minimum follow-up. *Spine (Phila Pa 1976)* 35:1887–1892
10. Sponseller PD, Jain A, Lenke LG, et al (2012) Vertebral column resection in children with neuromuscular spine deformity. *Spine (Phila Pa 1976)* 37(11):E655–E661
11. Wang X, Aubin CE, Crandall D et al (2011) Biomechanical comparison of force levels in spinal instrumentation using monoaxial versus multi degree of freedom postloading pedicle screws. *Spine (Phila Pa 1976)* 36:E95–E104
12. La Rosa G, Giglio G, Oggiano L (2011) Surgical treatment of neurological scoliosis using hybrid construct (lumbar transpedicular screws plus thoracic sub-laminar acrylic loops). *Eur Spine J* 20(Suppl 1):S90–S94
13. Ko PS, Jameson PG 2nd, Chang TL et al (2011) Transverse-plane pelvic asymmetry in patients with cerebral palsy and scoliosis. *J Pediatr Orthop* 31:277–283
14. Phillips JH, Gutheil JP, Knapp DR Jr (2007) Iliac screw fixation in neuromuscular scoliosis. *Spine (Phila Pa 1976)* 32:1566–1570
15. Chechik O, Fishkin M, Wientroub S et al (2011) A new pelvic rod system for the surgical correction and fixation of pelvic obliquity in paediatric neuromuscular scoliosis. *J Child Orthop* 5:41–48
16. Modi HN, Hong JY, Mehta SS et al (2009) Surgical correction and fusion using posterior-only pedicle screw construct for neuropathic scoliosis in patients with cerebral palsy: a three-year follow-up study. *Spine (Phila Pa 1976)* 34:1167–1175
17. Master DL, Connie PK, Jochen SH et al (2011) Wound infections after surgery for neuromuscular scoliosis: risk factors and treatment outcomes. *Spine (Phila Pa 1976)* 36:E179–E185
18. Jones-Quaidoo SM, Yang S, Arlet V (2010) Surgical management of spinal deformities in cerebral palsy. A review. *J Neurosurg Spine* 13:672–685