



# Cerebral palsy and spinal deformities

## *Beyin felci ve omurilik deformiteleri*

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Cerebral palsy is a common static encephalopathy and can present as multiple musculoskeletal problems. Increased peripheral muscle tone causes joint contractures and decreased functional capacity. The risk for scoliosis increases parallel with the severity of musculoskeletal involvement. Scoliosis adversely affects the functional capacity, daily care, and nutrition in disabled children. Conservative treatments including physical therapy, bracing, and botulinum toxin injections do not prevent the progression of scoliosis in most of the patients and surgical treatment becomes mandatory. With the use of pedicle screws, three-plane fixation is possible, making posterior instrumentation and fusion effective in correction of severe curves and obviating anterior surgery.

**Key words:** Cerebral palsy/complications; child; lordosis/etiology; pelvis/surgery; rhizotomy; scoliosis/etiology/surgery; spinal diseases/surgery; spinal fusion/instrumentation.

Beyin felci, çoklu kas iskelet sistemi sorunları ile ortaya çıkabilen ve sık görülen bir statik ensefalopatidir. Periferik kas tonusunun artması, eklem kontraktürlerine neden olarak hastanın fonksiyonel kapasitesini azaltır. Kas iskelet tutulumunun ciddiyetinin artması ile skolyoz riski de artar. Skolyoz hastaların fonksiyonel kapasitesini, günlük bakımını ve beslenmesini olumsuz etkiler. Fizik tedavi, korse veya botulinum toksin enjeksiyonu gibi konservatif tedavi yöntemleri genellikle skolyozun ilerlemesini engelleyemez ve cerrahi tedavi gerekli hale gelir. Üç planlı tespit imkanı sağlayan pedikül vidalarının kullanımı ile posterior enstrümantasyon ve füzyon, yeterli düzeltmenin sağlanmasında etkili olmuş, anterior cerrahi gereksinimini ortadan kaldırmıştır.

**Anahtar sözcükler:** Beyin felci/komplikasyon; çocuk; lordoz/etyoloji; pelvis/cerrahi; rizotomi; skolyoz/etyoloji/cerrahi; omurga hastalığı; omurga füzyonu/enstrümantasyon.

Cerebral palsy (CP) is an entity defining static encephalopathies that may be due to prenatal, natal, or postnatal causes. Although the primary problem is in the central nervous system, patients need medical treatment due to peripheral symptoms such as increased muscle tone and inadequate muscle control. The treatment focuses on decreasing secondary findings instead of treating the cause of the disease. Thus, physicians treating CP patients are metaphorically trying *to get rid of mosquitoes instead of drying the swamp*. The treatment of spinal deformities and dealing with ambulation of CP patients require totally different ap-

proaches, in that, spinal deformities are static whereas problems with ambulation are dynamic. Additionally, treatment of spinal deformities is technically demanding and surgical treatment is challenging, in contrast to the relatively simpler treatment modalities for problems of ambulation, which are associated with lower morbidity. Although the establishment of treatment indications and selection of treatment modality are rather straightforward for spinal deformities, proper analyses and appropriate surgical treatment, at the correct time, for problems of ambulation require complicated and demanding analyses.

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## Incidence

The incidence of spinal deformities in patients with CP is significantly higher than in the normal population. The incidence and severity increase with the degree of involvement, degree of mental retardation, and worsening functional status. The percentage of patients with spinal deformities is less than 5% for hemiplegics as compared to 60-70% in quadriplegic CP patients. When all patients with CP are evaluated, the incidence of spinal deformities is around 25%.<sup>[1-3]</sup>

## Curve characteristics

The deformities seen in CP patients are different from those of idiopathic deformities in terms of the age of onset of the deformity, the risk for progression, the natural course of the curve, the need for surgical treatment, and the response to nonsurgical treatment. Deformities in CP patients appear at earlier ages, and progression after skeletal maturation generally requires surgical treatment.<sup>[3,4]</sup> Curves that have reached 40 degrees before 15 years of age and thoracolumbar curvatures resulting in pelvic obliquity are accepted to be more likely to progress. Moreover, as the severity of neurological involvement increases, the likelihood that spinal deformities will progress also increases.<sup>[1,3]</sup>

Although in some patients deformity affects primarily the thoracolumbar spine, generally the deformities are rigid, extend to the pelvis, and have greater magnitudes than idiopathic curves. They not only affect the coronal plane, but also affect the sagittal and transverse planes (three-dimensional). Compared to idiopathic deformities, apical rotation and translation are more severe.<sup>[2,4]</sup>

Lumbar curves are the most common curve type in quadriplegic CP and usually they are on the left side. The rate of progression of the curves is not associated with the location of the curve. Thoracolumbar curves seem to be greater in magnitude on initial presentation with the fastest progression, followed by lumbar curves and thoracic curves.<sup>[3]</sup>

## Treatment problems

Progressive CP scoliosis not only causes cosmetic problems, but also decreases utilization of the upper extremities, may result in ischial pressure sores, may adversely affect the circulatory system, intensifies nutritional problems, and compromises hygiene

and transportation.<sup>[2]</sup> Inadequate nutrition adversely affects the immune system resulting in an increased incidence of infections.<sup>[2,5]</sup>

The presence of other multiple system disorders in addition to spinal problems makes treatment more difficult. Contractures of the musculoskeletal system, especially dislocations of the hip joint, can be closely related to spinal deformity. During the preoperative evaluation of spinal deformities, other parts of the musculoskeletal system should also be taken into consideration.<sup>[1,2,4]</sup> Due to the underlying neurological condition resulting in inadequate weight bearing and decreased mobilization, vertebrae are osteoporotic and the pelvis is smaller than in normal patients, and in addition, anticonvulsive medication exacerbates osteoporosis. In total, this condition is associated with an increased likelihood of surgical complications.

As most of these patients have seizures, in addition to spasticity and mental retardation, utilization of external supports after surgical treatment is impossible. As a result, there is a definitive need for rigid fixation at the time of surgery.

Although in the past, pelvic or intrapelvic deformities were held responsible for the development of spinal deformities, it is currently accepted that the etiological cause of spinal or hip deformities is spasticity. The relationship of spinal deformity (pelvic obliquity) and hip dislocation has been studied extensively; a recent study demonstrated that dislocated hips had no additional effect on the progression of scoliosis in CP kids.<sup>[6]</sup> There are examples of normal hips in patients with severe pelvic obliquity, in addition to unilateral hip dislocations with normal spinal alignment. Without any question, the hip joint should be carefully monitored and dislocation should be prevented.<sup>[7]</sup> However, the idea of preventing the development of spinal deformity by preserving the normal hip joint is not realistic.

Selective dorsal rhizotomy (SDR) has recently become a commonly used treatment method in the management of spasticity.<sup>[8,9]</sup> With this method, spasticity is decreased and, as a consequence, functional capacity is increased. However, this technique has been shown to increase mechanical weakness in the posterior spine, resulting in an increased tendency for hyperlordosis and spondylolisthesis. In addition to mechanical instability, the consequent weakness of the posterior musculature and the hip flexor muscle af-

ter surgery has also been shown to increase the risk for spinal deformity. In the first series published, the incidence of spinal deformity after SDR was reported to be up to 36%, a higher percentage than reported in CP patients who were treated nonsurgically.<sup>[10]</sup> Coronal plane deformities are mild, whereas sagittal plane deformities can be more severe. In recent years, the trend has been to limit the extent of laminectomies, aiming to decrease the likelihood of spinal deformities. When SDR is performed based on appropriate indications, it dramatically increases the ambulatory capacity of CP patients. Balancing the risk for development of a relatively easily treated spinal deformity against the difficulty in managing complicated ambulation problems, SDR remains a worthy procedure.<sup>[8,9]</sup>

Another treatment method for spasticity is an intrathecal baclofen pump (ITB). With this method, a subcutaneous pump is implanted to deliver small dosages of baclofen directly into the intrathecal space. It has been shown that spasticity can be decreased markedly and motor development in young children improves as a result of this treatment.<sup>[11]</sup> There is a discussion in the literature about the effect of decreased spasticity of paravertebral muscles on progression of scoliosis. A recent study demonstrated that ITB had no significant effect on curve progression, pelvic obliquity, or the incidence of scoliosis when compared with a matched control group of patients with spastic CP without ITB.<sup>[12]</sup> There are other case series reporting similar results.<sup>[13,14]</sup>

## Nonoperative management

Braces that are generally used in the treatment of spinal deformity are largely ineffective in CP patients.<sup>[15-17]</sup> In addition to having no effect on the course of deformity, patients are not compliant with brace use. Moreover, the occurrence of seizures while patients are in the brace might result in serious problems, in the same way the limitation of chest wall movements associated with brace wearing might exacerbate existing pulmonary problems and may result in pressure sores.

The treatment of spinal deformity and nonsurgical management of spasticity with botulinum toxin (Botox) has been evaluated in a limited number of cases with short follow-ups, but with encouraging results. Nuzzo et al.<sup>[18]</sup> reported that botulinum toxin injection to the concave side in patients who required a delay in

surgery increased the effectiveness of brace treatment and halted curve progression. However, the temporary effectiveness of botulinum toxin and the fact that it cannot be used in later stages, due to mechanical collapse of spinal deformity, should be kept in mind.

In CP patients, sitting supports and wheel-chair adaptations can be used as a means to control spinal deformity.<sup>[19]</sup> Although an increase in functional capacity may be achieved and some functions may be preserved, to a limited extent, the control of spinal deformity progression is impossible.

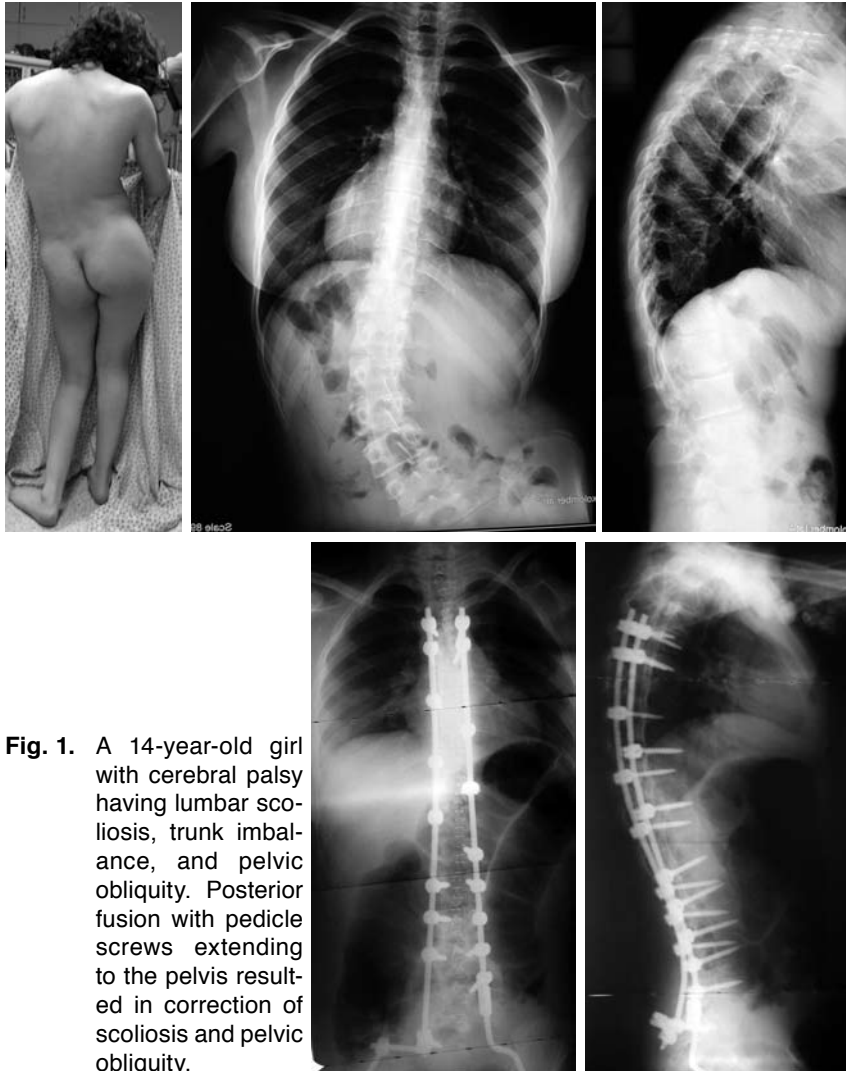
## Surgery

### Indications

The principal indications for surgical treatment are the progression of spinal curvature and the development of functional losses related to spinal deformity. Patients with spastic CP have the highest incidence of scoliosis.<sup>[3]</sup> Almost 70% of these patients have scoliosis that tends to progress even after skeletal maturity. Saito et al.<sup>[3]</sup> found that patients with curves greater than 40° before 15 years of age ended up with greater curves. They recommended that fusion should be considered for curves greater than 40° before age 15 years. Surgical treatment may result in balanced sitting and allow independent use of the upper limbs. Other beneficial effects include effective respiration and pulmonary clearance, and the facilitation of good nutrition. The restoration of spinal alignment may, in some conditions like hyperlordosis, also help the management of gastroesophageal reflux or superior mesenteric artery syndrome. Finally, spinal surgery makes nursing care much easier.<sup>[20,21]</sup>

### Advantages and disadvantages

It is important to keep in mind that surgical treatment is complicated, demanding, and expensive. Although remarkable results can be obtained following surgery, we should not forget that functional recovery is very limited due to the nature of the disease. There is no controversy as to the role of surgery in hemiplegic and diplegic patients with a spinal deformity and no mental retardation; however, the indications for surgery in patients with total body involvement are more controversial.<sup>[1,22]</sup> The cost-benefit ratio has been extensively discussed worldwide in terms of medical, economic, and ethical aspects. Major spinal surgeries conducted in major hospitals by experienced surgeons may not add years to the lives



**Fig. 1.** A 14-year-old girl with cerebral palsy having lumbar scoliosis, trunk imbalance, and pelvic obliquity. Posterior fusion with pedicle screws extending to the pelvis resulted in correction of scoliosis and pelvic obliquity.

of CP patients, but add life to their years. Therapists, caregivers, and families report meaningful changes in the quality of patients' lives, and that they become very happy after corrective spinal surgeries.<sup>[11,23-25]</sup> Although most CP patients have serious handicaps, both mentally and physically, it is not and must not be the responsibility of doctors to withhold the benefits of modern medicine from these patients who may benefit from more adequate care.

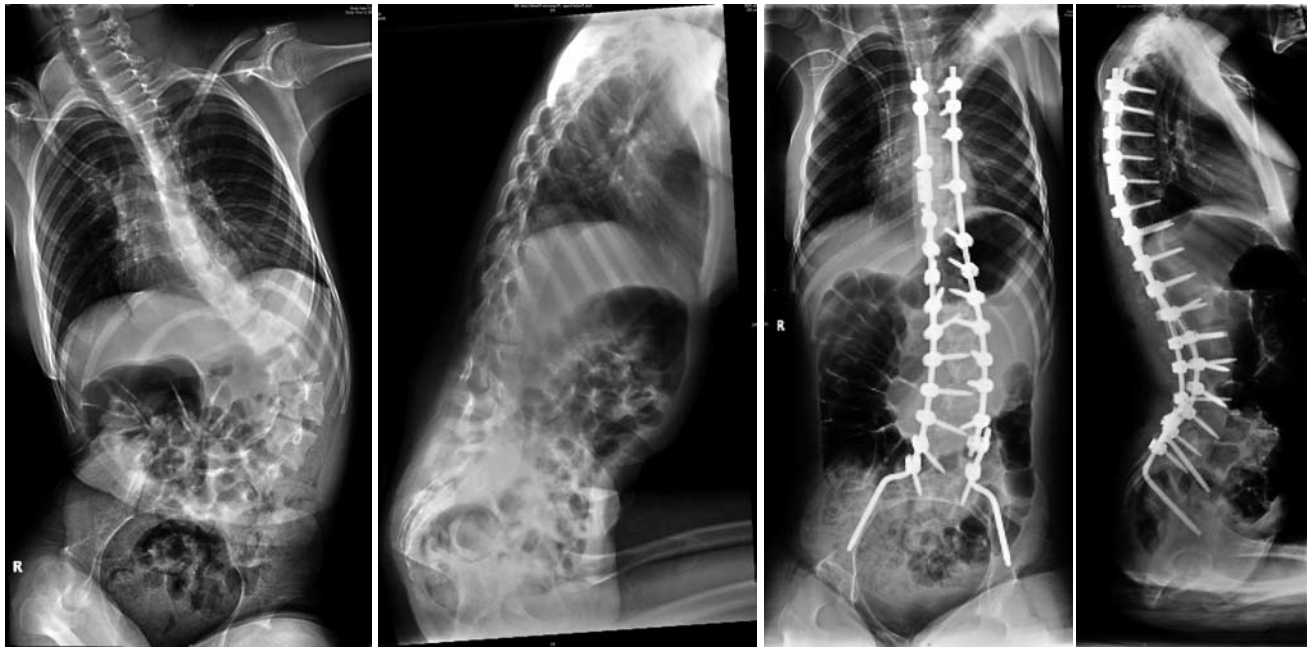
In patients requiring surgery, early intervention, when spinal deformity has not progressed and become rigid, increases surgical success and decreases postsurgical complications.

### Techniques

The instrumentation that will be used in surgery should facilitate three-dimensional correction of the defor-

mity, be strong enough to eliminate the need for external support after surgery, and should be low-profile in design, as the paravertebral muscles are atrophic.<sup>[21]</sup> Instrumentation should always be supplemented with fusion, and the instrumentation and fusion should be extended from the upper thoracic vertebrae to the pelvis.<sup>[2,21,26]</sup> In the past, there have been serious concerns about the effect on ambulatory capacity of instruments extending to the pelvis. It is now known that, in patients with successful restoration of the sagittal contours, ambulatory capacity is unchanged (Fig. 1).<sup>[21,26-28]</sup> To be more precise, failures are related to insufficient lordosis restoration, long-term immobilization, and cast application, but not to fusion.<sup>[2,27]</sup>

Pedicle screws provide three-column fixation so that greater corrective forces can be applied, which helps achieve greater correction of rotation, and have



**Fig. 2.** In severe deformities, posterior fusion with pedicle screws resulted in good curve correction and anterior surgery became unnecessary.

been shown to be effective to obtain posterior fusion. In CP patients, because of osteoporosis, larger diameters should be used, preferably, all levels should be instrumented, and the use of allografts is generally necessary to obtain fusion.<sup>[20,29]</sup>

In many cases, anterior surgery can be added for a better correction and effective fusion.<sup>[21,30,33]</sup> In the spinal literature, anterior surgery is advocated for the prevention of the development of the crankshaft phenomenon in young patients undergoing posterior fusion. It is still debatable whether the crankshaft phenomenon is really a problem in patients with CP. Although Smucker and Miller<sup>[7]</sup> reported that crankshaft was not a problem in children with CP, the possibility cannot be definitely ruled out since in their series, the number of patients operated on before the age of 10 years and followed-up till maturity was too small.

It should not be forgotten that the decrease in lung capacity of these patients, due to spinal deformity, might increase the risk associated with anterior surgery. Anterior surgery is generally planned for patients with advanced deformities who are considered at high risk for surgery, as these patients have other comorbidities;<sup>[30,31]</sup> therefore, the addition of anterior surgery increases the risks in already complicated cases.

With the use of pedicle screws, which are capable of producing three-dimensional correction, and of

controlling the growth of the anterior column, it may be possible to obtain an effective correction with a posterior approach in young children without encountering the crankshaft phenomenon, therefore making anterior surgery unnecessary. The main goal with instrumentation is to make the pelvis parallel to the ground and to center the head over the pelvis.<sup>[20]</sup> The correction of sagittal plane deformities and maintenance of the achieved correction is more difficult than corrections in the coronal plane. It is not easy to restore lordosis with only posterior surgery in hyperlordotic patients, or to maintain upper thoracic alignment in hyperkyphotic patients with only sublaminar wires or hooks; particularly in sagittal plane corrections, the use of pedicle screws has been shown to decrease these problems (Fig. 2).<sup>[21,27]</sup>

### Complications

During the pre- and postoperative periods, nutritional support is essential; also, special attention should be paid to maximizing lung capacity.<sup>[5]</sup>

Appropriate anesthesia is important and provision should be made for supporting mechanical ventilation, should there be a need. As these patients are prone to coagulopathies, expected blood loss is higher,<sup>[32]</sup> this tendency being more prominent in patients using valproate. Additionally, dilution phenomenon may occur due to acute rehydration in patients with contracted

intravascular volume. Since bleeding generally originates from the venous system, hypotensive anesthesia is usually not effective.

The risk for neurological deficits related to surgery is not excessively high. The utilization of neuromonitorization in order to decrease this risk can result in high false negatives or positives, and from a practical point of view, is useless.

The propensity for surgically-related infections after spinal surgery is higher in CP patients.<sup>[5,33,34]</sup> This risk increases particularly in cognitively impaired, epileptic patients, those in whom allografts were utilized, and patients with low albumin and lymphocyte levels.

## Conclusion

The severity of spinal deformities increases with the severity of the body involvement in cerebral palsy. Unlike idiopathic scoliosis, progression of the curves does not stop with skeletal maturity. Severe curves decrease functional capacity, cause sitting imbalance and pressure sores, and adversely affect pulmonary and cardiac functions. Conservative treatment usually does not stop curve progression in long term and most of the patients require spinal fusion. With the use of pedicle screws, three-dimensional correction is possible and anterior fusion is unnecessary, but extending the fusion to the sacrum is needed in patients with severe pelvic obliquity.

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