CURRENT CONCEPT REVIEW

Management of spinal deformity in cerebral palsy: conservative treatment

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Abstract

Introduction The incidence of scoliosis in Cerebral Palsy (CP) is directly related to the Gross Motor Function Classification System (GMFCS) level. The natural history of untreated scoliosis in patients with CP is one of progression and factors implicated in deterioration include type of involvement (quadriplegia), poor functional status (nonambulatory, GMFCS levels IV and V), and curve location (thoracolumbar). The generally accepted incidence in the overall CP population is 20–25 %.

Materials and methods We recently published our short term results for 31 children treated with a short lumbar brace. In cases of a "positive hands up test" we recommend a short lumbar brace, and in patients with scoliosis with a Cobb angle $>20^{\circ}$ a double shelled brace.

Results In our study, there was a correction of 37 % for the lumbar Cobb angle and 39 % for the thoracic Cobb angle at a mean follow-up of 28 months.

Conclusion The incidence of scoliosis in the overall CP population is 20–25 % and is directly related to the GMFCS level. Therefore, we recommend early treatment and prescribe a short lumbar brace in patients with dynamic instability of the trunk, and in scoliosis with a Cobb angle $>20^{\circ}$ a double shelled brace.

Keywords Spinal deformity · Scoliosis · Cerebral palsy · Conservative treatment · Short lumbar brace · Double shelled brace

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Introduction

The incidence of scoliosis in cerebral palsy (CP) varies greatly, from 6 % to almost 100 % [1–3]. The generally accepted incidence in the overall CP population is 20–25 % [3]. It is highest in patients with spastic CP (about 70 %) and lowest in those patients with athetoid type (6–50 %) [4]. In the same study [4], patients with subluxated or dislocated hips (an indicator of disease severity) were also found to have an incidence of scoliosis of approximately 75 %. The incidence of scoliosis is also directly related to the Gross Motor Function Classification System (GMFCS) [5] level.

The cause of scoliosis in CP is not entirely clear, but thought to be a combination of muscle weakness, truncal imbalance, and asymmetric tone in paraspinous and intercostal muscles [3]. The natural history of untreated scoliosis in patients with CP is one of progression, and factors implicated in deterioration include type of involvement (quadriplegia), poor functional status (nonambulatory, GMFCS levels IV and V), and curve location (thoracolumbar) [1-3, 6]. According to our experience, scoliosis mainly results from inadequate gravity control which typically leads to lateral flexion of the thorax over the relative mobile spine, thus a thoracolumbar curve. Saito et al. [1] describe the risk factors for progression of scoliosis in spastic CP as follows: having a spinal curve of 40° before age 15 years, having total body involvement, being bedridden, and having a thoracolumbar curve. Patients with these risk factors might benefit from early surgical intervention to prevent severe scoliosis. It is, however, known that bone growth is reduced under pressure and increased under tension. Thus, lateral bending due to inadequate trunk control can already be regarded as a risk factor for a scoliosis if given sufficient time.

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Fig. 1 Short lumbar brace



Fig. 2 Double shelled brace

Nonoperative treatment options consists of observation and bracing, but also include seating modifications and medical management. There are two methods to provide external support to the spine in the presence of neuromuscular scoliosis: customized seating arrangements and braces [2]. Most of these patients with neuromuscular



Fig. 3 Sufficiant correction of a neuromuscular scoliosis in a 12 years old girl with spastic quadriplegia. **a** the Cobb angles were 35° (lumbar) and 25° (thoracic). **b** the angles were 12° (lumbar) and 4° (thoracic)

problems suffer from poor balance control and dynamic instability of the trunk. At the early stage, the patient presents only with dynamic instabiliy of the trunk and this can be easily observed by asking for "hands up" while sitting. The goals of any intervention are to maintain comfortable sitting and to allow functional use of the upper extremities. There is some evidence that the use of braces may slow down curve progression in quadriplegic CP [7], but it is unclear whether spinal bracing reduces the need for surgery [2]. During growth, however, spinal bracing reduces curve size and slows down progression to severe curves [7]. Nevertheless, it is unclear whether bracing is able to stop progression [1-3,7-11]. Thus, spinal bracing in children with CP is a well-described and established method of conservative management [7, 9, 10, 12–17]. Although botulinum toxin A is increasingly used to treat limb spasticity in patients with CP, there is scant evidence for its use in treating scoliosis [3].

Our strategy

There is evidence of asymmetrical bone growth under asymmetrical load as applied by habitual trunk side lean due to lack of balance control, and there is a high risk in developing a scoliosis in all patients with neuromuscular disorders. Furthermore, trunk instability impedes head control and hand function. For these reasons, we recommend starting treatment at the very early stage when the patient proves to be incapable of keeping an upright posture while sitting (postive hands up test). In these situations, we start with a short lumbar brace (Fig. 1) for functional indications, and in patients with scoliosis with a Cobb angle >20° double-shelled brace (Fig. 2) worn when unpright.

Early treatment with short lumbar brace

We recently published our short-term results for 31 children (14 boys and 17 girls with a mean age of 12.6 years, range 3-24 years), with dynamic trunk instability due to neuromuscular diseases treated with a short lumbar brace [18]. Of these, 21 children suffered from spastic quadriplegia and the remaining 10 patients from different neuromuscular disorders. The mean lumbar Cobb angle at study onset was 25.3° (range 10–48°) and the thoracic Cobb angle 9.9° (range 2–15°). At a mean follow-up time of 28.3 months (range 12-38 months), the lumbar Cobb angle was 16.0° (range: 0-52°), significantly corrected (p < 0.001), and the mean thoracic Cobb angle was 7.1° (range: 0-35°). This was a correction of 36.8 % for the lumbar Cobb angle and of 39.4 % for the thoracic Cobb angle. In only 3 cases was a deterioration of the Cobb angles found during the study period. Figure 3 outlines a sufficient correction of a neuromuscular scoliosis in a 12-year-old girl with spastic quadriplegia (GMFCS level V). Without the brace (Fig. 3a), the Cobb angles were 35° (lumbar) and 25° (thoracic). With the brace (Fig. 3b), the angles were 12° (lumbar) and 4° (thoracic). In our study [18], an average 2.0 lumbar short braces per patient were used during the study period of 28.3 months. In our opinion, this treatment option is appropriate for mildly affected cases, but we recommend close meshed observation both clinically and radiologically.

All patients with scoliosis of a Cobb angle over 20° get double-shelled braces at our department, but they are not always well accepted by the patients, parents, and caregivers. We strongly recommend checking the correct fitting of the brace: a tight fit at the pelvis, an adequate spacer between pelvis and ribs, and a wider fit at the thorax for breathing. There should be a large open window at the belly to enable eating while wearing the brace. Further, the brace should be easy to handle. An important part is taking the mould. We emphasise taking it in overcorrection (bending to the convex side) of the patient in order to get immediate and optimal correction. This method is superior to working with corrective pads at least in our hands. The mould in overcorrection provides large contact areas in a corrective postion without local pressure as there is from pads. Nevertheless, one should be aware that the soft tissue structures remain compressible and thus the brace always has a tendency to be too wide, as at the pelvis dorsally over the glutei and the waist, and too tight at the ribs. Tight fitting, however, additionally provides a larger contact area and better control of the included segments. Especial attention should be given to the pelvis as a wide fitting does not allow proper control of the lumbar spine. The braces should always be worn under gravity and upright posture (in sitting and standing). The final correction in the brace always needs to be checked by radiographs (at least an anterior-posterior view under load without and with the brace) in order to check any correction, which should be at least 30 %. Only long-term use of this strategy will show whether it will be possible to reduce the incidence of at least severe scoliotic deformities.

Conclusions

We suggest using spinal braces for functional reasons in order to improve head and hand function, and to prevent spinal deformities which are prone to develop in frequent side leaning positions. In dynamic instability of the trunk we recommend early treatment with a short lumbar brace for functional reasons. In cases of scoliosis (Cobb angle >20°), braces need to be applied early and consistently in order to prevent deterioration. Short bivalve braces with optimal correction seem to fit best with all these requirements.

Conflict of interest None.

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