

Postural deformity in children with cerebral palsy: Why it occurs and how is it managed

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ABSTRACT. Despite the fact that children with cerebral palsy may not have any deformities at the time of birth, postural deformities, such as scoliosis, pelvic obliquity, and windswept hip deformity, can appear with increasing age. This may lead to respiratory function deterioration and, in more severe cases, affects survival. To date, postural care is believed to help improve the health and quality of life of children with cerebral palsy. This review provides an overview of the cause and clinical management of postural deformity that is seen in children with cerebral palsy.

Key words: Scoliosis, windswept hip deformity, pelvic obliquity, hip dislocation

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Cerebral palsy (CP) does not refer to a specific disease but is an umbrella term that refers to a group of disorders affecting a person's ability to move that occurs in the developing fetal or infant brain^{1,2}. Despite the fact that the musculoskeletal status of children with CP is usually normal at birth, postural deformities can progressively arise with development³⁻⁶.

Postural deformity has a significant impact on not only the quality of life in children with CP⁷, but also on their mortality⁸. Severe scoliosis and deformed chest cage lead to lung compression⁹. Respiratory problems are frequent among patients with CP and are a significant cause of death^{8,10,11}.

These secondary problems, which can lead to functional decline, should be avoided. Various studies have been conducted to investigate the cause of the deformities and how to prevent them; however, there are still unknowns¹²⁻¹⁵. This review provides an overview of the cause and the clinical management of postural deformities that are seen in children with CP.

Postural Deformity in Children with CP

Posture in this review will refer mainly to a sitting or lying position. Deformity refers to an abnormality of the position of the body compared to a normal position. Children with CP tend to show a specific pattern of deformity, a so-called "postural deformity"¹⁶ or "positional deformity"¹⁷. This review uses the term "postural deformity" that includes the following deformities: scoliosis, pelvic obliquity, windswept hip deformity, and hip subluxation/dislocation. A typical image of the postural deformity seen in children with CP is illustrated in Fig. 1. The head is turned to one side, and there is a marked pelvic tilt with spinal scoliosis. Windswept hip deformity is also seen, where one leg falls into abduction and external rotation, and the opposite leg into adduction and internal rotation. Such deformations can cause difficulties in the daily activities of children with CP, such as that in maintaining a sitting position and/or rolling over on a mat^{7,17,18}. Moreover, postural deformity accompanied with deformed chest cage can decrease respiratory function⁸. In CP, recurrent aspiration and impaired airway clearance due to respiratory muscle weakness and/or poor coordination, combined with spinal and thorax deformities, can lead to both acute respiratory tract infections as well as chronic lower airway inflammation, ultimately resulting in bronchiectasis^{8,11}. Once bronchiectasis occurs, this further impairs clearance of sputa, resulting in repeated bronchitis^{8,11}. Respiratory disease is the most significant factor in the causal pathway of CP-related mortality¹⁹. Knee flexion contracture and equinovarus foot are also

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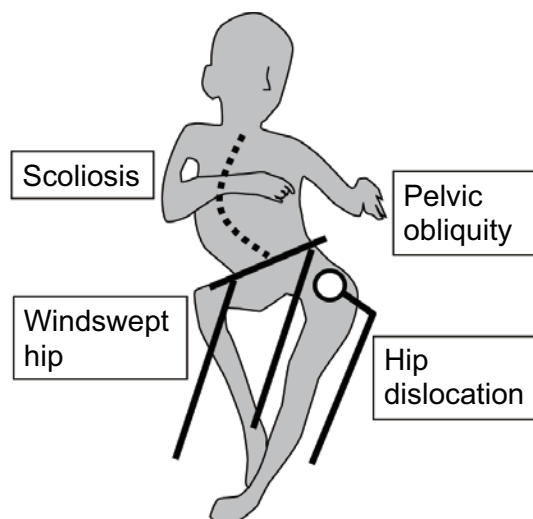


Figure 1. Postural deformity seen in children with CP.

common deformities noted in CP, but these affect more functional aspects such as gait²⁰⁾. Since postural deformity affects respiratory function and indirectly affects mortality, more attention must be paid to this than to other deformities.

Change in Postural Deformity with Age

Generally, no structural abnormality is apparent in the trunk or extremities at the time of birth^{3,6,21)}. However, problems arise in the course of postnatal development, and postural deformities gradually become more prominent^{22,23)} (Fig. 2). In severe cases, the deformities continuously progress beyond the growth period²⁴⁻²⁶⁾.

Among the postural deformity types, hip dislocation is likely to be detected early in childhood. Hip dislocation in CP often appears at 2-3 years of age²⁷⁾. Scoliosis appears at 5-6 years of age in children with severe CP^{28,29)}, and at around 8 years of age in children with mild CP³⁰⁾. Windswept hip deformity becomes prominent by the age of 10 years, and the risk of deformity continues thereafter²²⁾.

Relationship between Postural Deformity and Gross Motor Function

The clinical features of children with CP vary. Classifications of CP have focused on types of tone abnormalities such as spasticity, athetosis, and dystonia; however, the classifications do not provide prognostic information³¹⁾. Gross motor function classification system for CP (GMFCS) is a useful tool to predict limits of motor function for a given level of CP severity^{32,33)}. It divides children with CP into five groups based on the overall functional capability (Table 1): no functional impairment (level 1); functional limitation when walking in crowds, may need assistive device (level 2); walks with an assistive device (level

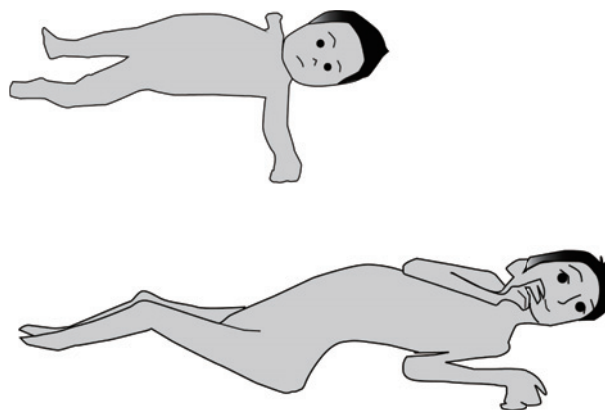


Figure 2. Example of a change in lying posture with increasing age in a child with CP. At 1 year of age, the head is rotated to the left side, but the trunk is straight and both hips and knees are extended in a neutral position (above). At 10 years of age, an asymmetrical postural deformity is prominent; the head is rotated to the left, the spine is curved towards the right side (scoliosis), and both hips and knees are flexed and rotated to the right side of the body (windswept hip deformity) to the right (bottom).

3); relying more on wheeled mobility (level 4); and cannot sit independently, bedridden (level 5)^{33,34)}. Gross motor development curves, which correspond to each of the 5 GMFCS levels of severity, are widely recognized. Once a child is assigned to a GMFCS level, the corresponding motor function change curve shows the average pattern of change in the motor score. The majority of children with CP reach a peak motor function score at 5-7 years of age^{35,36)}. Patterns of gross motor development by level of severity have been explained by a stable limit model (SLM) or a peak and decline model (PDM)^{37,38)} (Fig. 3). Children at GMFCS level 1 and 2 have no functional decline, thus the SLM was embraced. In contrast, at level 3, 4, and 5, they show significant decline after a peak at 7 years of age; thus, the PDM was embraced³⁷⁾. As mentioned above, trunk deformity becomes noticeable around 7 years of age, but it is thought that deformation may also be involved in such functional decline.

The gross motor function of children with CP is closely related to the occurrence of postural deformity, and more severe deformity is observed in individuals with higher GMFCS levels^{22,27,28,30,39)}. Persson-Bunke *et al.*³⁰⁾ found that the proportion of children with scoliosis increased with GMFCS level and that those with GMFCS level 4 and 5 had a 50% risk of having moderate or severe scoliosis by the age of 18 years. Hagglund *et al.*²⁸⁾ reported that 75% of children had severe scoliosis with a Cobb angle of 40 degrees or greater at the age of 20 years. No child developed hip dislocations with migration percentage exceeding 40% in GMFCS level 1, while 10% of children with level 2,

Table 1. Summary of the Gross Motor Classification System (GMFCS) for children with cerebral palsy developed by Palisano et al. ³²⁾

GMFCS level	Description
Level I	Walks without limitation; performs running and jumping; but speed, balance, and coordination are reduced.
Level II	Walks without assistive devices; limitations in walking outdoors and in the community.
Level III	Walks with handheld assistive devices; limitations in walking outdoors and in the community.
Level IV	Children can sit but are usually supported; more reliance on wheeled mobility in the community.
Level V	Lack of independence even in basic antigravity postural control; self-mobility is severely limited, even with the use of assistive technology.

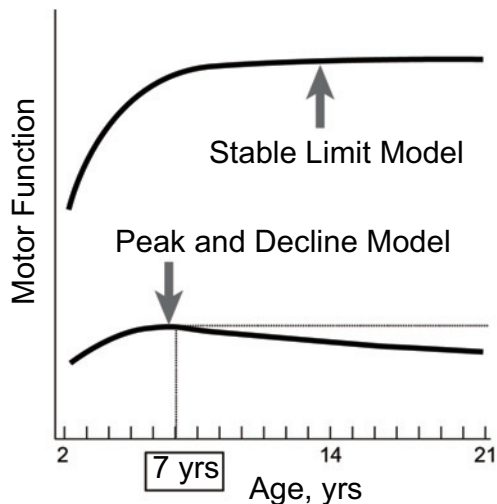


Figure 3. Gross motor development curves explained by the stable limit model and the peak and decline model. The stable limit model was embraced for children with mild CP; in contrast, the peak and decline model was embraced for children with severe CP. 7 years was the average age that peak of motor development was reached. (adapted with permission from Hanna SE, Rosenbaum PL, et al.: Stability and decline in gross motor function among children and youth with cerebral palsy aged 2 to 21 years. *Dev Med Child Neurol.* 2009; 51: 295-302)

18% with level 3, 45% with level 4, and 64% with level 5 developed severe hip dislocation²⁷⁾. The windswept hip deformity was also absent in level 1 and 2, but was detected in level 3, and was seen in 52% of children in level 5²²⁾.

Persson-Bunke *et al.*³⁰⁾ also analyzed the relationship between postural deformity and types of muscle abnormalities, but there were no significant differences regarding the CP subtype such as the spastic, dyskinetic, ataxic, and mixed types.

Risk Factors for Emergence and Progression of Postural Deformity

Abnormalities in muscle tone and/or muscle imbalance have been established as the primary cause of postural de-

formity in children with CP. Typically, hip dislocation is explained by the muscular imbalance in which strong hip flexors and adductors overpower the hip extensors and abductors^{20,40)}. However, asymmetrical muscle activity does not clearly explain the patterns of deformity. For example, if the right side of the spinal muscle contracts more to the left side, the spine bends to the right. However, no relationship between the direction of dominant muscle tone and the direction of scoliosis has been established⁴¹⁾. Moreover, Porter *et al.*²¹⁾ found that a particular asymmetrical recumbent posture in the first year of life was associated with the direction of the subsequent pattern of postural deformity. Therefore, postural deformity may start as the simple tendency of an individual to spend a lot of time in a particular asymmetrical position.

Environmental factors, such as remaining in asymmetrical recumbent postures for prolonged periods due to poor ability of a child to move spontaneously, should be considered^{42,43)}. Children with severe CP tend to remain in the same body configuration for extended periods when sleeping^{44,45)}. Several systems have been developed to monitor posture in the daily life of children with CP, by determining the angular orientation with respect to gravity of the body part to which an accelerometer is attached⁴⁵⁻⁴⁷⁾. With this method, body posture can be monitored at all times, in the midst of both daily and nocturnal activities in the life of a child with CP (Fig. 4). As a risk factor for emergence and progression of postural deformity, an abnormal muscle tone, in addition to prolonged asymmetric posture, should be considered.

How We Care the Postural Deformity

Spasticity reducing: Reducing spasticity seems to be the most direct treatment for improving or preventing deformities but has no effect in the long term⁴⁸⁻⁵⁴⁾. For example, intramuscular injections of botulinum toxin A can be effective in reducing muscle tone over a long period, but does not prevent development of contractures in spastic muscles⁵²⁾. The results of an 11-year follow-up study indicated that hip dislocation in children with CP is related to the severity of motor function classification, rather than the

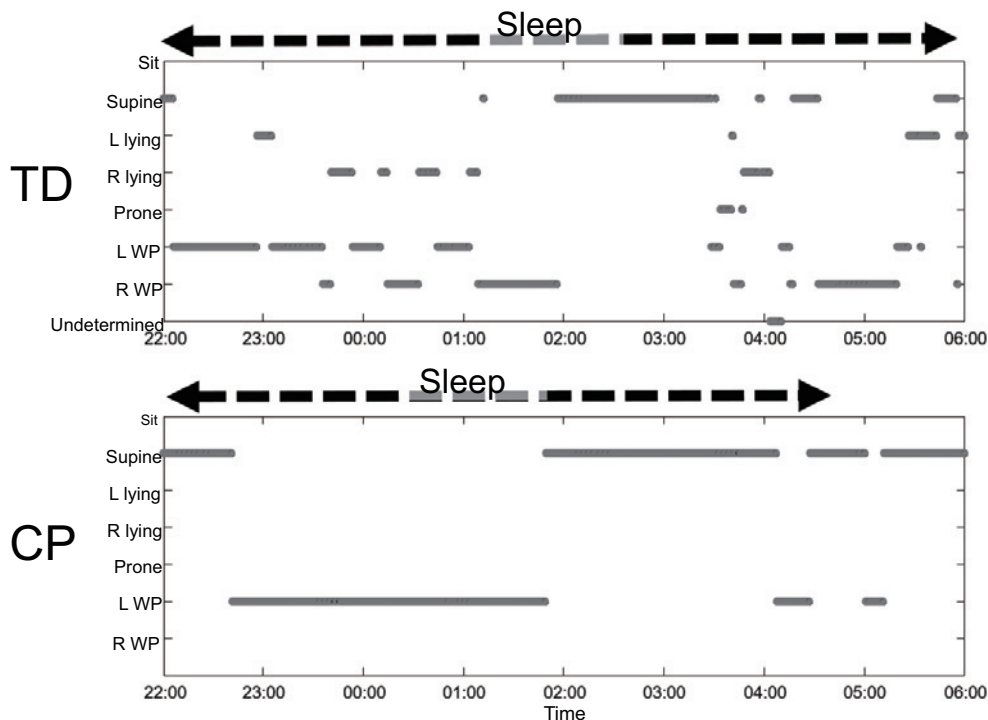


Figure 4. Example of body posture during night-time sleep in a child with typical development (TD: 7 years) and a child with severe cerebral palsy (CP: 8 years) (unpublished data). Note that the prolonged period spent in the WP position was observed in CP. WP: windswept hip position.

definition of whether the muscle tone is spastic, dystonic, or mixed⁵⁵). Moreover, several studies demonstrated a significant increase in the rate of scoliotic curve progression after spasticity-reducing treatment^{48,49,54}). One possible reason for this is that the loss of muscle tone leads to difficulty in maintaining or changing posture, which causes development and/or progression of deformity⁵⁰.

Surgery: Postural deformity mostly occurs in children with severe CP (GMFCS level 4 and 5), and the degree of the deformity is also more severe in these children. Risk of complications is high^{13,56}), and thus, the indications for surgery should be carefully discussed⁵⁷). One major type of surgery for deformity is spinal fusion for scoliosis. The expected effects of the surgery include the arrest of the deformity progression, avoidance of cardiopulmonary dysfunction, improvement of posture, and ease of care⁵⁶). However, the effects actually obtained are mainly related to functional aspects, such as improved posture and a reduction in the need for sitting supports, and the evidence of the effect is equivocal for arrest of deformity and improvement of cardiopulmonary function^{12,56,58}).

Passive stretching: Passive stretching is performed with the aim of preventing a decrease in the range of joint motion due to a decrease of muscle extensibility. Although passive stretching may lead to a temporary improvement in passive range, there is lack of evidence supporting that it can prevent a decreased range of motion i.e. contracture^{59,60}).

An interesting study conducted by Tardieu et al⁶¹), investigated how many times a muscle should be stretched for preventing contractures in children with CP. They recorded the range of ankle joint motion continuously for a 24-hour period using their own apparatus. After mean duration of 7 months, they compared the total stretch time between children with CP who showed progressive contracture and those who did not. All children without progressive contracture showed a minimum of 6 hours' stretch time during a 24-hour period. In contrast, all children with progressive contracture showed a shorter stretch time of approximately 0 to 3 hours. This indicates that even if a therapist can maintain a child's muscle in the stretched position manually, he or she should maintain it more than 6 hours a day. Wearing a night-time splint may be reasonable to keep the soleus muscle stretched. However, it was shown that a child failed to keep the soleus muscle stretched despite wearing a night-time splint. The child also developed contractures, indicating that a splint is useless if not properly worn.

This implies that the duration of stretching, whether it is done passively or actively, may be the key to prevent contracture. However, during passive stretching, the child is literally in a passive state, exerts very less effort, and tends to get bored. They would experience a much longer period of stretching from self-motivated activities, such as going to school, horseback riding, and swimming programs, that in turn lead to long-lasting effects of stretching⁶²).

Bracing: Spinal bracing does not influence the natural course of scoliosis progression^{63,64}. Rather, it is effective in improving stability in sitting, which results in better overall function^{63,64}. One reason for bracing not being able to prevent scoliosis progression might be that children with CP do not use the orthosis long enough, especially during the night⁶⁴. Children with severe CP tend to spend more time in a lying position, which might limit the bracing time. Spinal orthoses may be more suitable for use by children with mild CP who focus on increasing sitting stability, which promotes upper limb use instead of preventing deformity.

Postural care: An expert multidisciplinary group developed a consensus statement on postural management of children with CP based on evidence from clinical experience and scientific literature⁶⁵. In the consensus statement, a 24-hour posture management program is recommended for children with severe CP (GMFCS level 4 and 5) that focuses on the role in preventing deformities^{65,66}. One example of the postural management is the use of night-time postural equipment^{67,68}. This consisted of supportive mattresses and padded supports, which promote a symmetric anatomically-correct lying position^{67,69}. Some studies show that postural care may be effective in preventing hip dislocation^{14,70,71}; however, evidence regarding the efficacy of postural care is still lacking¹⁵.

Cerebral Palsy Follow-Up Program: A surveillance program for CP called the Cerebral Palsy Follow-Up Program (CPUP) was established in 1994 in southern Sweden⁷². This hip surveillance method includes a standardized follow-up of gross and fine motor function, clinical findings, and treatment⁷². The gross and fine motor function of a child is examined by a local physical therapist and occupational therapist twice a year until the age of 6 years, and once a year thereafter⁷². The survey involves a standardized radiographic screening of the hip even in children with no symptoms⁷². The radiographic examination is conducted once a year for children in GMFCS levels 3 to 5, and at 2 and 6 years of age in those with level 2⁷². Those with level 1 are not examined⁷². This follow-up program aims for early detection of contractures and deformities that will then allow for early, preferably non-surgical, treatment²².

After the CPUP program was started, the incidence of hip dislocation and windswept hip deformities in children with CP was reduced^{12,72}. An early detection of deformities would be necessary for postural management for those children.

Conclusion

Postural deformity exhibiting complex aspects, such as scoliosis, windswept hip deformity, pelvic obliquity, and hip dislocation, is frequently seen in children with severe cerebral palsy, who can hardly move by themselves. This deformity affects the prognosis of such children. The author

believes that the key to preventing postural deformity is to detect it at an early stage and to apply postural care for the at-risk children.

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