

Position as a Cause of Deformity in Children with Cerebral Palsy

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Introduction

Deformities in the child with cerebral palsy have been ascribed to muscle imbalance (Sharrard 1961) and increased tone (Pollock 1959) or to the type of cerebral palsy (Bobath and Bobath 1975). As far as we know, the position in which the child is nursed, especially during the first year of life, has not been considered as a cause of deformity.

It is generally accepted that position in the postnatal period can be a cause of deformity in the normal baby. Paine (1961) suggested that plagiocephaly was caused by postnatal head posture, and Hay (1971) found that plagiocephaly was present in 10 per cent of normal babies. Scott (1956) reported that infants commonly had lateral curvatures of the spine which could be seen on x-rays but not on clinical examination, all of which had resolved by the age of two years. Other asymmetries associated with plagiocephaly are unilateral flisting, asymmetrical groin creases, apparent shortening of one lower limb and asymmetry of gait (Robson 1968). We accept the asymmetrical deformities of plagiocephaly, unilateral bat ear, facial and thoracic asymmetry, pelvic obliquity and apparent shortening of one leg—some or all of which may be present in

normal babies — as forming the 'squint' baby syndrome. Because asymmetrical deformities also occur in children with cerebral palsy, we thought it worthwhile to compare the pattern of deformity in a group of 'squint' but otherwise normal babies with a group of cerebral-palsied children with asymmetrical deformities, to see if there is any relationship.

Method and Material

Any baby attending the well-baby clinic at the Simpson Memorial Maternity Pavilion who was noticed to have four or more signs of the 'squint' baby syndrome was examined by one of us and was included in this review. Over the last two years the names of 20 babies were obtained in this way. Recently all these babies have been re-examined at home by a physiotherapist to establish the child's progress and to find out whether any of the signs of the 'squint' baby syndrome were still present. A proforma was filled in at the time the baby first entered the review and again when seen by the physiotherapist. We have also reviewed 20 children with cerebral palsy who are known to have asymmetrical deformities, to find out the incidence of the signs that are associated with the 'squint' baby syndrome. All of

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these children have had radiographs of their hips and spine, usually including films of the spine taken with lateral flexion to each side.

Results

'Squint' Children without Cerebral Palsy

The mothers of the 20 babies in this group had normal pregnancies, except for three who had an antepartum haemorrhage. Three births were by caesarean section and two were assisted with low forceps, otherwise the confinements were normal.

In the neonatal period 11 of the 20 babies preferred to lie with their faces to the right, three with their faces to the left and six did not have a preferred position. Five of the babies slept supine, three slept prone, seven slept lying on their right side, three slept lying on their left side and two were always put down to sleep on alternate sides. One of these last two had a persistent asymmetrical tonic neck reflex and the other had a preferred head-position. In the early months 13 babies were kept in the lying position most of the time; the other seven were put into a semi-reclining position some of the time. The average age at which the babies entered this study was

three months, by which time all but two had plagiocephaly and facial moulding, 11 had thoracic asymmetry and seven had asymmetrical passive hip-movements (Fig. 1). This moulding was more common to one side than the other (Fig. 2).

At review, the average age was 16 months with a range from three months to 4½ years. It was found that 12 of these children had sat and walked at the expected times, five were still too young, and the remaining three had delayed development: two of these were shufflers and walked at 18 months. The third child was neurologically normal but although he had been well fed he had been psychosocially neglected and left to lie in his bed day and night; he showed all the features of the 'squint' baby syndrome. During his stay in hospital he rapidly developed both emotionally and physically; it was during this time, at 10 months, that he started sitting.

Only one of the 20 children had lost all the signs of positional moulding at the time of review, but most children over the age of nine months had lost some of the signs that had been present when they were first seen. The asymmetry of the thorax and the asymmetrical position of the legs had

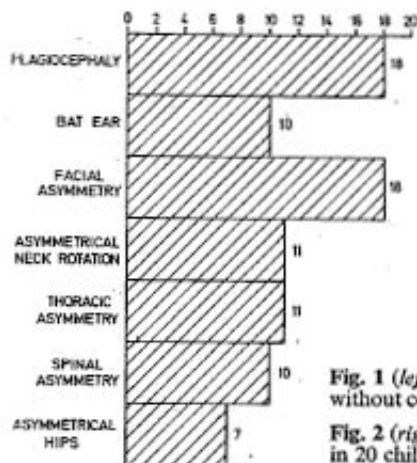


Fig. 1 (left). Frequency of signs of positional moulding in 20 children without cerebral palsy.

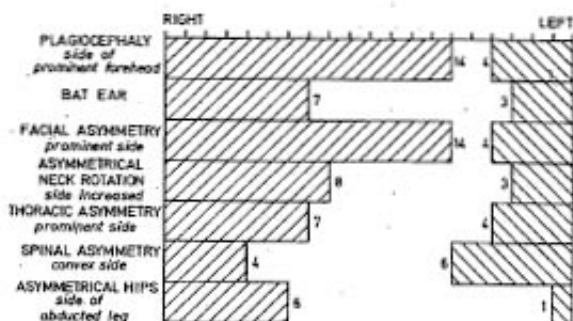


Fig. 2 (right). The side to which signs of positional moulding occurred in 20 children without cerebral palsy.

usually disappeared by one year, but the plagiocephaly and facial flattening was often still present at the age of two years.

Cerebral-palsied Children with Asymmetrical Deformities

The average age of the 20 children with cerebral palsy was seven years, ranging from 14 months to 13½ years. The principal diagnoses were diplegia (10), diplegia with right hemiplegia (1), right hemiplegia (2), bilateral hemiplegia (5), dyskinesia (1) and severe ataxia (1). In all of them motor development was grossly retarded and 10 had not yet gained head control. They were all in the range of severely sub-normal intelligence. One child aged 13 years was functioning at a five-year level but all the others were under a two-year level. It was found that these children had the same signs as are associated with the 'squint' baby syndrome but that the signs were present more frequently and to a greater degree, resulting in a syndrome which we call the 'windswept' child syndrome*. Seven of these children preferred

to lie with their faces turned to the right and eight with their faces turned to the left; the remaining five had no preference. Of the 20 children, all but one had plagiocephaly and all but two had facial asymmetry. Asymmetrical chest moulding was present in 17 children, 15 of whom had a structural scoliosis. 19 of the 20 children had 'windswept' legs with asymmetrical passive hip movements (Fig. 3). The asymmetry occurred in both directions with the preponderance for the same side as in the 'squint' babies (Fig. 4).

Discussion

The natural history of the 'squint' baby syndrome cannot be established from this review, but Hay (1971), in an earlier review of 300 babies, never found plagiocephaly at birth but later it was present in 10 per cent of normal babies. Thistlethwaite (1976) has reviewed 63 low-birth-weight babies at regular intervals during the first year and found that two-thirds of the group showed plagiocephaly. The reason that asymmetry was not usually noted in our babies until three months could be that we were only informed when babies had four signs of the syndrome. Plagiocephaly and facial moulding were the most common signs, possibly because they are the most easily noticed. Both

*The whole syndrome consists of plagiocephaly, facial asymmetry, torticollis with asymmetrical passive neck rotation, unilateral bat ear, lateral asymmetry of the chest often leading to scoliosis, and an asymmetrical position of the hips often leading to dislocation of the hip on the side of the adducted leg.

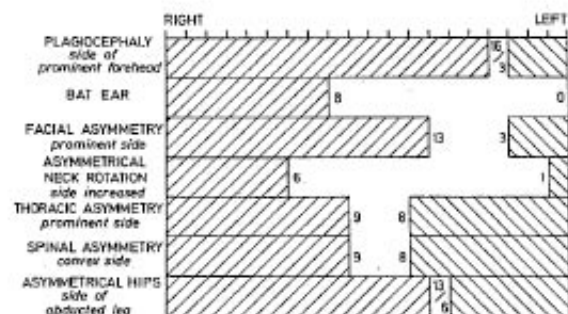
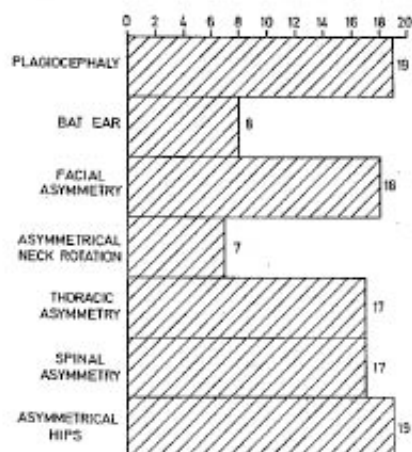


Fig. 3 (left). Frequency of signs of positional moulding in 20 children with cerebral palsy.

Fig. 4 (right). The side to which signs of positional moulding occurred in 20 children with cerebral palsy.

were still present at two years of age but were less obvious. Asymmetrical thoracic moulding and asymmetrical legs were less frequent than plagiocephaly and had nearly always disappeared by the time the child was a year old.

Because we have found the same deformities in the 'squint' normal babies and the 'windswept' cerebral-palsied children, we think that they are both part of the same syndrome and only differ in degree. Our findings that the deformities decrease in normal children once they can sit independently but progress in the cerebral-palsied children (half of whom had not gained head control) suggest that once the syndrome has developed the immobility caused by the delayed motor development allows it to progress. Whilst we accept that the intrinsic forces of muscle imbalance and spasticity can cause deformity in children with cerebral palsy, we think that it is the extrinsic force of gravity which causes both the 'squint' deformity in the normal baby and the 'windswept' deformity in the cerebral-palsied child.

We do not know what first causes a baby to turn its face or legs to one side and thus become asymmetrical, but probably there are a number of causes. Dunn (1972) has shown that there is a relationship between plagiocephaly, facial deformity, sterno-mastoid torticollis, postural scoliosis and congenital dislocation of the hip when they are present at birth. He attributes these deformities to extrinsic forces during the prenatal period, usually from the uterus (particularly when there is oligohydramnios) acting on a fetus at a time when it is particularly vulnerable because of its rapid growth and plasticity. This was probably not the cause in either of our groups of babies, as none were noted to have any of these signs at birth. Dunn (1975) also reports a correspondence between the position of the fetus, determined radiographically before delivery,

and the 'position of comfort' (Chapple and Davidson 1941) that the baby prefers after birth.

Robson (1968) mentions as two possible causes of asymmetric motor development the vulnerability of the cerebral cortex beneath the uppermost (non-presenting) parieto-occipital bones during delivery, and obstruction of the vertebral artery when the neck is rotated 35° to the opposite side. Another suggested cause is that the infant's cot is so placed that noises or light always come from one side, causing the face to turn because of primitive acoustic or light reflexes. It is also possible that a mother, for some reason such as getting rid of the baby's wind, always puts her baby down on the same side.

A spot check of 568 normal babies in the lying-in wards of the Simpson Memorial Maternity Pavilion over a period of 10 weeks showed that 50 per cent were lying on the right, 22.4 per cent on the left, and 24.8 per cent lay prone and turned their faces to either side, whilst only 3 per cent lay flat on their backs. These figures are similar to the head-turning preference in our group of normal 'squint' babies. This preference for the right side would also explain the finding of Watson (1971) that in normal people with plagiocephaly the left forehead is flattened twice as often as the right.

Whatever the primary cause of the head turning, the position of the head can influence the position of the rest of the body by initiating the asymmetrical tonic neck reflex which causes unequal muscle tone on the two sides of the body. However, this cannot be the only explanation, as only one of the normal 'squint' babies showed a persistence of the asymmetrical tonic neck reflex and we have seen the syndrome develop in 'floppy' babies, in whom this reflex is not present.

It is our impression that there are two different types of baby who become

'squint'. The first is the 'floppy' baby who will take up any position and for some reason is always laid on the same side so that it becomes 'squint'. The second type is the active baby who prefers to be on one particular side and cries until it is put onto that side.

The 'squint' baby syndrome appears to be unimportant in the normal baby, provided it is recognised as such and not diagnosed as abnormal posture secondary to brain damage, or as congenital dislocation of the hip when the asymmetry occurs in the lower limbs. When the child starts to walk, the syndrome is associated with an asymmetry of gait, as shown by the footprints, but this disappears between the second and third years (Robson 1968). It has yet to be established whether there is a connection between the plagiocephaly which occurs in idiopathic scoliosis and the 'squint' baby syndrome.

In many children with cerebral palsy the 'windswept' syndrome can be seen to develop during the first few months of life. It is not dependent upon the type of cerebral palsy; it can occur in both the hypotonic and hypertonic types, with and without obligatory primitive postural reflexes. It is seen in both congenital and acquired cerebral palsy and in the latter we have seen the deformity develop rapidly over a period of six weeks. The 'windswept' deformity may override the 'postural' deformity imposed by abnormal muscle tone so that in the lower limbs, whilst the deformities are symmetrical at the knees and feet, they are a mirror image of each other at the hips. When the child is only seen for the first time at the end of the first year of life, the 'squint' position and asymmetrical movements may be well established. They are then thought to be part of the pattern of cerebral palsy and have been described as a 'stage one diplegia' (Bobath and Bobath 1975).

Once the asymmetry has developed it is

self-perpetuating as long as the child is in the horizontal position. Even if the child is made to lie on alternate sides, the effect of gravity on the moulding is not reversed because on one side—usually the preferred side—the child lies half-way between the mid-lateral and the supine positions, whilst on the other side it is half-way between the mid-lateral and the prone positions (Figs. 5 and 6). The increasing asymmetry eventually results in the gross deformities so often found in the child with severe cerebral palsy. The severely affected child has gross plagiocephaly, facial asymmetry, torticollis, a unilateral bat ear (Figs. 7 and 8), lateral asymmetry of the chest (Fig. 9) and 'windswept' legs (Fig. 10). The torticollis causes asymmetrical passive neck rotations, with increased rotation on the side to which the head is turned. The lateral asymmetry of the chest at first causes a rigid segment in the thoracic spine, (as shown by the lateral flexion radiographs in Fig. 11) but later, commonly during adolescence, it can appear clinically as a rapidly-increasing and severe scoliosis (Fig. 12). The abducted leg of the 'wind-



Figs. 5 and 6. *Left*: a 'squint' child lying in his 'position of comfort'. *Right*: the same child lying on his other side, showing that moulding effect is not reversed because he now lies half-way between lateral and prone positions.



Figs. 7 and 8. A 'windswept' child, showing (*left*) facial asymmetry and unilateral bat ear and (*right*) plagiocephaly.



Fig. 9 (*above*). Skyline view, showing thoracic asymmetry.

Fig. 10 (*right*). 'Windswept' legs.



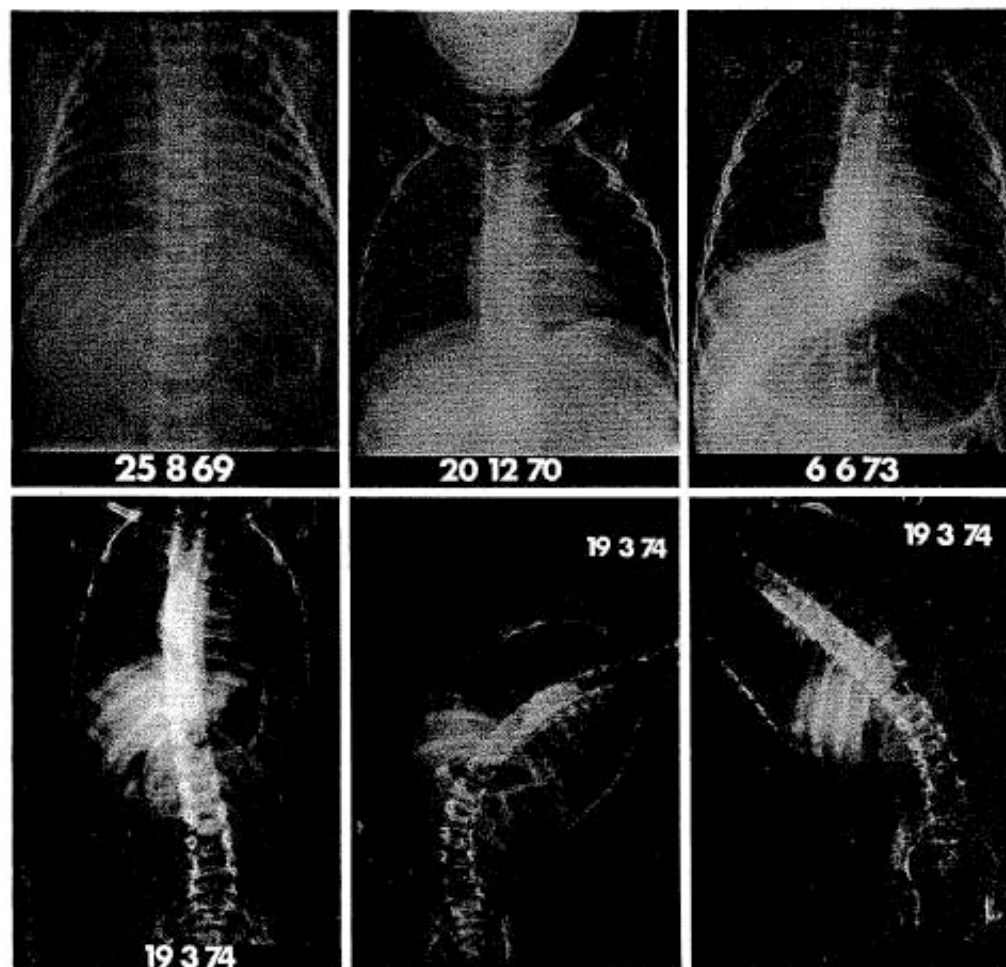


Fig. 11. *Upper, left to right*: the spine of a baby at birth, at 16 months when there were signs of cerebral palsy and positional deformities, and at 3½ years. *Lower, left*: the spine of the same baby at 4½ years, showing an established scoliosis; (*centre*) whilst flexed to the left and (*right*) whilst flexed to the right, showing a rigid segment in the thoracic region.

'swept' legs shows increased abduction and external rotation, and reduced adduction and internal rotation in comparison with the other leg. X-rays show that the acetabulum of the adducted hip fails to develop normally, with increasing subluxation and eventual dislocation of the hip (Fig. 13). By adolescence the combination of severe scoliosis and unilateral dislocation of the hip, with the legs 'windswept' to one side, makes it difficult to place these patients in a comfortable sitting position (Fig. 14)

and they often complain of back pain and pain from the dislocated hip. Eventually they become almost impossible problems of nursing.

Because of the gross deformities that can occur in the 'windswept' child, it is important that this cause of deformity should be prevented. It is possible that this may be one of the major benefits of early diagnosis of cerebral palsy. We have not been able to determine whether we can prevent the syndrome, but there appears

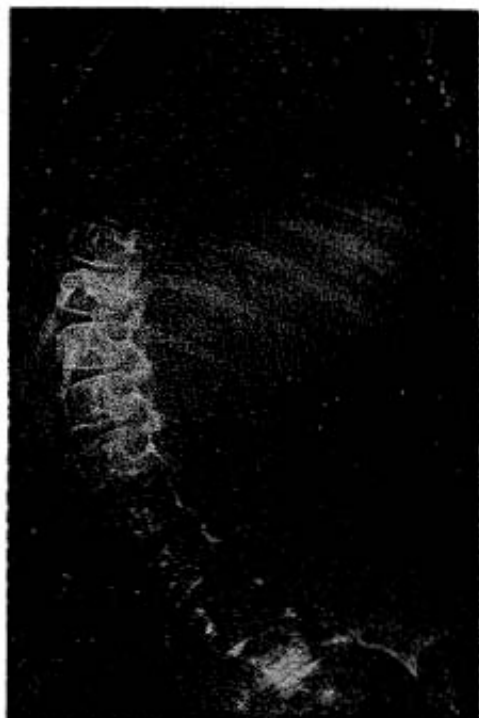


Fig. 14. A 'windswept' cerebral-palsied child with severe deformities.

Fig. 12. (Left) Severe scoliosis in an adolescent 'windswept' child with cerebral palsy.



Fig. 13. Hips of a child with cerebral palsy. *Upper left*: at 7 months, already showing slight asymmetry; *upper right*: at 2½ years, showing unilateral subluxation; and *lower left*: at 4 years, showing unilateral dislocation.

to be a lot of advantages in nursing all babies in the prone position whenever possible. The normal baby cries less, is less vulnerable to 'mass startle', and Moro responses and asymmetric tonic neck reflexes are less obligatory. A baby changes its head position when lying prone, but cannot do so when supine. The prone position allows the legs of the young baby to be placed in symmetrical abduction: in the older baby with delayed development the legs should be kept abducted by double nappies or a simple abduction splint, but once asymmetry has

commenced these measures are not sufficient.

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SUMMARY

The asymmetrical deformities in 20 children with various types of cerebral palsy are compared with 20 children without cerebral palsy who have the so-called 'squint' baby syndrome (asymmetrical deformities of plagiocephaly, unilateral bat ear, facial and thoracic asymmetry, pelvic obliquity and apparent shortening of one leg). It is suggested that the 'squint' baby syndrome and the 'windswept' child syndrome in children with cerebral palsy are stages of the same syndrome and that in both the deformities are caused by the effect of gravity on an immobile growing child, rather than spasticity or muscle imbalance. Asymmetrical deformity should therefore be amenable to physiotherapeutic intervention, rather than trying to modify maturation of the damaged brain. As the 'windswept' cerebral-palsied child can develop some of the most severe deformities seen in cerebral palsy, it is important that asymmetrical deformities should be prevented.

RÉSUMÉ

La position, cause de déformations chez les enfants I.M.C.

Les déformations asymétriques chez 20 enfants présentant des types variés d'infirmité motrice cérébrale ont été comparées à celles de 20 enfants sans infirmité motrice cérébrale mais présentant le syndrome appelé 'bébé dévié' (aplatissement asymétrique du front, oreille en chauve-souris d'un seul côté, asymétrie faciale et thoracique, bassin oblique et raccourcissement d'une jambe). Il est suggéré que le syndrome du 'bébé dévié' et la déformation 'en coup de vent' chez les enfants IMC sont dûs au même mécanisme et qu'il y a un effet de la pesanteur sur un enfant immobile en croissance à l'origine de la déformation, plutôt qu'une spasticité ou un déséquilibre des muscles. La déformation asymétrique releverait d'une intervention physio-thérapique plutôt que d'une tentative de modification de la maturation du cerveau lésé. Comme un enfant IMC 'en coup de vent' peut présenter les déformations les plus graves observées au cours de l'infirmité motrice cérébrale, il est important de prévenir ces malformations.

ZUSAMMENFASSUNG

Pathologische Haltung als Ursache für Deformierungen bei Kindern mit Cerebralparese

Die asymmetrischen Deformierungen bei 20 Kindern mit verschiedenen Manifestationen einer Cerebralparese wurden mit den Deformierungen bei 20 Kindern ohne Cerebralparese

verglichen, die ein sogenanntes 'Squint' Baby Syndrom hatten (einseitige Schädeldeformierungen, einseitiges Fledermausohr, Gesichts- und Thoraxasymmetrien, Beckenschiefstand und deutliche Verkürzung eines Beines). Vermutlich beruhen das 'Squint' Baby Syndrom und die 'windswept' Mißbildung bei Kindern mit Cerebralparese auf demselben Mechanismus und man nimmt an, daß der mehr Effekt der Schwerkraft, die auf das bewegungsarme wachsende Kind wirkt diese Deformierungen verursacht als die Spastik oder die Imbalanz der Muskulatur. Die physiotherapeutische Behandlung ist daher sinnvoller als die Reifung des geschädigten Gehirns beeinflussen zu wollen. Da das cerebralparetische Kind mit einer 'windswept' Mißbildung die schwersten Deformierungen entwickeln kann, die bei der Cerebralparese beobachtet wurden, ist es wichtig, daß asymmetrische Deformierungen verhindert werden.

RESUMEN

Posición como causa de deformidad en niños con parálisis cerebral

Se comparan las deformidades asimétricas de 20 niños con varios tipos de parálisis cerebral, con las de 20 niños sin parálisis cerebral, y que tienen el llamado síndrome de 'estrabismo' (deformidades asimétricas con plagiocefalia, oreja en murciélago unilateral, asimetría facial y torácica, oblicuidad pélvica y acortamiento aparente de una pierna). Se sugiere que el síndrome del 'niño estrábico' y la deformidad en 'golpe de viento' en niños con parálisis cerebral se deben al mismo mecanismo, a saber que es el efecto de la gravedad sobre un niño en crecimiento e inmóvil lo que causa estas deformaciones más que la espasticidad o el desequilibrio muscular. La deformidad asimétrica debería por lo tanto ser orientada hacia una intervención fisioterapéutica, más bien que tratar de modificar la maduración del cerebro dañado.

Puesto que el niño paralítico cerebral con síndrome 'del golpe de viento' puede desarrollar alguna de las más severas deformidades vistas en la parálisis cerebral, es importante la prevención de las deformidades asimétricas.

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