

Posterior positional plagiocephaly treated by cranial remodelling orthosis

Lausanne's experience with 260 children

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Summary

Principles: Since the recommendation that infants sleep in the supine position, there has been an increase in cases of posterior positional plagiocephaly. Even though this condition is a purely cosmetic problem, if it is severe it may affect the child psychologically. Positioning may help in mild or moderate cases, but more active treatment may be necessary in severe cases.

Methods: A prospective study of 260 children treated by dynamic orthotic cranioplasty for posterior positional plagiocephaly was conducted in Lausanne from 1995 to 2001. Construction of these cranial remodelling helmets is described in detail.

Results: The treatment lasted 3 months on av-

erage, was effective, well tolerated, and had zero morbidity. The ideal period for initiating this therapy is between the ages of 4 and 6 months.

Conclusion: The remodelling helmet is a convincing option which can be recommended in infants with posterior positional plagiocephaly whose skull deformity is not satisfactorily corrected by physiotherapy. It should always be used before surgery is considered for patients with recognised positional plagiocephaly in the first year of life.

Key words: positional plagiocephaly; helmet; orthotic cranioplasty

Introduction

In 1992 the American Academy of Pediatrics (AAP), on the basis of a multinational investigation, established a direct correlation between prone sleeping position and the incidence of sudden infant death syndrome [1]. In this report the AAP recommended that infants be henceforth positioned to sleep either on the back or side. After this publication and the subsequent "Back to Sleep Campaign" [2], an increasing number of children with craniofacial deformities was observed. Following an initial period of diagnostic and surgical roaming in which surgical management had increased to epidemic proportions [3], it became obvious that this abnormal skull shape was not the result of craniosynostosis, which is the premature fusion of cranial sutures, but rather the conse-

quence of unrelieved pressure on the occiput during infant sleep. This cranial deformity was called posterior positional plagiocephaly (PPP). In about two thirds of cases it may correct spontaneously after regular changes in sleeping position or following physiotherapy aimed at correcting neck muscle imbalance [4, 5]. In severe cases which fail to improve despite counterpositioning treatment, the hazards of surgical correction may be avoided by cranial remodelling orthosis, originally described in 1979 [6, 7] and more extensively employed in the nineties [8, 9]. The purpose of this paper is to review our experience in treating children with posterior positional plagiocephaly by dynamic orthotic cranioplasty.

Patients and methods

From March 1995 to November 2001, 260 children presenting with PPP were consecutively treated by dynamic orthotic cranioplasty. They were initially examined by the senior authors at the Paediatric Neurosurgery Clinic of Lausanne University Hospital, Switzerland (CHUV). Only patients with obvious cranial deformation were proposed for a cranial remodelling helmet. For mild cases, parents were reassured and instructed to place the bulging side of their infant's head toward the crib mattress. Physiotherapy was proposed where appropriate. In every child anthropometric measurements were taken, including head circumference, distance from external auditory meatus to external canthus (EAM – EC) and fronto-occipital (FO) diagonals (Fig. 1). In unilateral posterior positional plagiocephaly there is typically an occipito-parietal flattening, ipsilateral to this flattening the ear is anteriorly displaced, and frontal bossing is observable. The ipsilateral external auditory meatus to external canthus distance, as well as the diagonal measured from the occipital flattening to the contralateral forehead, are therefore shorter than their contralateral counterparts (Fig 1, lines A and B). In a vertex view, the head exhibits the shape of a parallelogram.

All cranial remodelling helmets were made in the Ergotherapy Service of the Hôpital Orthopédique de la Suisse Romande, Lausanne, Switzerland. These orthoses are thermoplastic constructs consisting of a semi-rigid styrene outer shell thermobonded to a polyurethane foam inner lining (Polyform, Smith & Nephew, Germantown, USA). The first step was to cover the infant's head with a jersey sock in order to make a plaster of Paris impression (Fig. 2 – left). This impression is then filled with plaster to create a positive cast which is afterward corrected by applying a crescent-shaped silicone module over the occipital flattening. A cranial remodelling orthosis is then created over this corrected cast (Fig 2 – centre). A few days later the helmet is applied to the infant (Fig. 3). Pressure is directed to constrain growth at the ipsilateral frontal and contralateral parieto-occipital bulging, whereas a cavity is created over the adjacent flattened area to promote remodelling. The

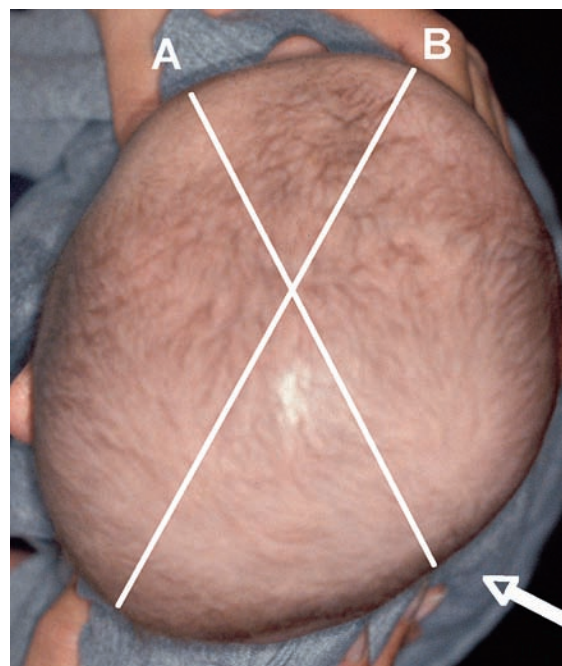


Figure 1

Right posterior positional plagiocephaly. Vertex view of the head. Right occipital flattening (arrow). Short (A) and long (B) fronto-occipital diagonals.

helmet was worn progressively during the first week of treatment up to 21 hours a day. Every 2–3 weeks, the children were followed up in the Ergotherapy Service for adjustment of the orthoses where necessary. In addition, the patients were reevaluated at the end of the treatment by the senior authors. At every examination anthropometric measurements were repeated. Data are presented as the mean \pm SD. Statistical comparisons were performed by the Student paired *t* test analysis. Significant differences were reported when the *p* value was <0.05 .

Figure 2

Plaster of Paris impression from the infant's head covered with a jersey sock.



Results

From March 1995 to November 2001, 260 infants (63 girls and 197 boys) were consecutively treated with a cranial orthosis for PPP in the Hôpital Orthopédique de la Suisse Romande, Lausanne, Switzerland. Most of the patients had been unsuccessfully treated by osteopathy and/or physiotherapy before referral to the CHUV Paediatric Neurosurgery Clinic. Mean age at the beginning of treatment was 6.1 ± 2.0 months (range: 3–18 months). As demonstrated in table 1, there was a

progressive increase in the number of cases treated during the period of study.

There were 117 children with right-sided posterior plagiocephaly (45%), 100 with left-sided (38%), and only 43 with a bilateral form, without lateralisation of the occipital flattening (17%). On examination there were 36 infants (14%) showing a contraction of the sternocleidomastoid muscle, contralateral to the side of posterior plagiocephaly. Most of the patients, however, even in the absence

Table 1

Epidemiology of patients treated with cranial remodelling orthosis in Lausanne from 1995 to 2001.

	1995	1996	1997	1998	1999	2000	2001
N (girls/boys)	15 (3/12)	20 (0/20)	14 (0/14)	38 (8/30)	44 (13/31)	50 (17/33)	79 (22/57)
Mean age at treatment onset (months)	5.8	7.1	5.1	6.2	6.0	6.1	6.0
Mean duration of treatment (weeks)	21	18	17	15	12	12	9
Mean number of helmets used per child	1.4	1.5	1.6	1.8	1.6	1.7	1.2

of clear-cut torticollis, had a tendency to turn the head preferentially on one side, as previously described [10].

Average duration of treatment was 13 ± 8 weeks (range: 4–31 weeks). There was a tendency toward shorter treatment over the years. When

therapy was initiated before 6 months of age its mean duration was 12 ± 6 weeks, and 13 ± 10 weeks when patients were aged over 6 months at treatment onset. This difference was not statistically significant.

The results were subjectively considered good by the parents and the investigators (Fig. 4). Objectively, this favourable impression was confirmed by anthropometric measurements showing symmetrisation of the ipsilateral and contralateral EAM – EC distances as well as FO diagonals. Therefore, the ratios of these left and right distances and diagonals rapidly tend toward 1.0. Indeed these ratios improved to statistically significant levels after as little as 1 month of treatment (figures 1 and 2).

In this series good results were obtained with a single helmet in 147 patients, whereas 98 and 15 infants required 2 and 3 cranial orthoses respectively. The cranial orthosis did not restrict cranial growth, since serial head circumference measurements showed that every patient remained on his usual percentile curve.

Figure 3

Side view of an infant wearing a cranial remodelling helmet.

**Figure 4**

A. Pretreatment vertex view of the head of a 4-month-old infant with right posterior positional plagiocephaly. **B.** Results after 2 months of treatment by cranial orthosis.



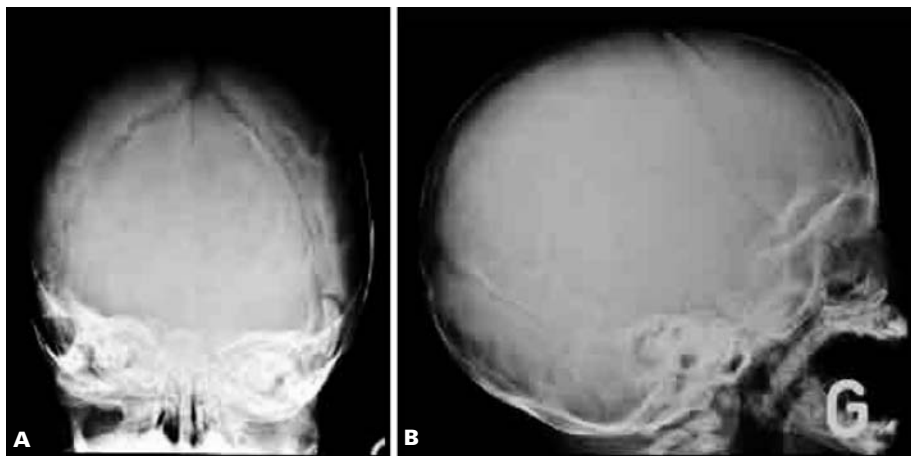
Discussion

Etymologically, the term “plagiocephaly” has Greek roots, “plagios” meaning “oblique” and “kephale” head. One cause of plagiocephaly is craniosynostosis, which is premature fusion of the cranial suture(s). The skull shape can be predicted from the suture(s) involved. Early fusion of the

lambdoid suture(s), which separate the parietal from the occipital bones, causes posterior plagiocephaly. However, premature lambdoidal fusion is extremely rare: whereas the overall incidence of craniosynostosis is 6 per 10,000 live births, plagiocephaly secondary to isolated lambdoid premature

Figure 5

Skull x-rays of a 5-month-old baby with left posterior positional plagiocephaly. **A.** AP view showing patent radiolucent lambdoid sutures. Note the sclerotic margin (arrows) parallel to the left lambdoid suture, on the side of the occipital flattening. **B.** Lateral view. The sclerotic margin is also visible (arrows).



fusion occurs in only 3 cases per 100,000 births [11–13]. In contrast, over the past decade there has been an increasing incidence of PPP [14, 15] which has been reported in up to 48% of live births [11, 16]. It is important to distinguish lambdoid craniosynostosis from PPP, because the course and the management of these two conditions are clearly different. True synostosis is habitually present from birth, and is progressive. It never improves spontaneously and carries the risk of intracranial hypertension, although this occurs in less than 10% of cases when only a single suture is involved [17]. In contrast, PPP is usually absent at birth, develops during the first months of life, and carries no risk of increased intracranial pressure.

Regarding the pathophysiological mechanism of PPP, it is accepted that external forces applied consistently to a specific region of the infant's head deform the skull. This explains why the increasing incidence of babies presenting with occipital flattening has coincided with current advice to position infants to sleep in the supine position to prevent sudden infant death syndrome [12, 14, 15]. Other pre- and postnatal factors, though rarer, may favour the development of PPP by external constraint. They include multiple births, decreased amniotic fluid, macrocephaly, malformed uterus, congenital torticollis, cervical spine anomaly or brain injury with asymmetrical spasticity [7, 11, 15, 16, 18–20]. In these situations the cranial deformity is usually present from birth and worsens over the following weeks.

Simple clinical examination is usually sufficient to diagnose postural posterior plagiocephaly [11]. In vertex view, patients presenting with PPP exhibit a parallelogram-shaped head: there is a flattening of one side of the posterior cranium, along with contralateral parieto-occipital and ipsilateral frontal bossing, and the ipsilateral ear is displaced anteriorly. In contrast, in unilateral lambdoid synostosis, the vertex view reveals a trapezium-shaped head: there is unilateral occipitoparietal flattening associated with contralateral frontal bossing. In addition, the area of the fused lambdoid suture presents as a bony ridge, with a bony prominence in the mastoid region behind the ear, which is displaced posteriorly and inferiorly [21]. Normally

there is no need for complementary radiological investigations. In this study, neither CT scans nor plain x-rays were done routinely. However, several patients referred to our clinic had already had skull x-rays done elsewhere. In this situation skull x-rays reveal a sclerotic margin parallel to a patent lambdoid suture on the side of the occipital flattening (Fig. 5). This aspect, sometimes called “lazy lambdoid”, must not be confused with a true lambdoid synostosis where the suture is no longer visible [22].

This study confirms some characteristic features of PPP: three quarters of the patients were boys, and there were a majority of infants with right-sided plagiocephaly. A male preponderance in up to 73% of cases and a right/left preference in a proportion of 2:1 are also reported in the literature [4, 9, 10, 14, 16, 19, 22–25].

The natural history of PPP is difficult to establish in the absence of reliable and reproducible data which would allow grading of its severity. It has however been reported that over 70% of cases improve spontaneously [4]. This improvement may be encouraged by regular changes in sleeping position and/or physiotherapy [4, 25, 26]. Severe positional skull deformations may not always correct satisfactorily [6, 15, 27]. Even though there is little information about the true risks of leaving this condition untreated [11], PPP is probably essentially a cosmetic problem of no significant neurological consequence [4, 15, 27]. The role of the various therapeutic modalities must therefore be determined. In the nineties, when this condition was emerging, surgical management was carried to epidemic proportions [3, 4] until cranial remodelling helmets were developed [8, 9]. This treatment by orthotics was inspired by ancestral ethnic practices once aimed at intentionally deforming children's skulls [28].

This study shows that cranial remodelling orthosis is a valuable treatment option for infants whose PPP is not satisfactorily corrected by physiotherapy. The earlier the helmet is applied, the more rapid and complete will be the correction [9, 11, 14, 24]. In practice, the ideal period for initiating this treatment is from 4 to 6 months of age. At this age, in our series, the treatment duration av-

eraged 3 months. Before 4 months of age, experience has shown that the infants usually do not have enough cervical muscle strength to tolerate the helmet. After 1 year of age, because cranial growth lessens and the skull thickens, correction by orthosis is longer and less impressive. In addition, older children tend to remove their helmets themselves as they do not tolerate them as well as younger ones. In practice, we do not usually propose treatment after 1 year of age except as an ultimate attempt to avoid surgical correction. Convincing results in such rare cases have however been reported in a small series [29].

Conclusion

This study shows that treatment by cranial remodelling orthosis is effective, well tolerated, and has zero morbidity. It can be recommended in in-

fants with PPP whose skull deformity is not satisfactorily corrected by physiotherapy. This therapeutic option should always be taken up before surgery is considered for patients with recognised PPP in the first year of life, bearing in mind that this type of surgery would be a cosmetic procedure frequently requiring blood transfusion and not without risk in view of the proximity of posterior dural sinuses.

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