



Disponible en ligne sur

ScienceDirect
www.sciencedirect.com

Elsevier Masson France

EM|consulte
www.em-consulte.com



Craniosynostosis: State of the Art 2019

Deformational plagiocephaly: State of the art and review of the literature



1. Introduction

Several types of head deformation can be seen in children, including plagiocephaly and brachycephaly. These terms derive from the Greek “*plagios*” meaning oblique, “*brachy*” meaning short and “*kephale*” meaning head. The etymologies globally refer to an abnormal head shape without any specific etiology, and can refer to anterior or posterior deformity [1]. Anterior plagiocephaly is often due to premature fusion of a single coronal suture, resulting in unilateral flattening of the forehead with abnormal orbital roof shape, while anterior brachycephaly is often due to premature fusion of both coronal sutures, resulting in bilateral flattening of the forehead. In posterior deformities, the mechanism usually does not concern the synostosis but rather a prolonged pre- and/or post-natal mechanical force applied on the head, resulting in what is called positional or deformational plagiocephaly (DP) when the deformation is asymmetrical or deformational brachycephaly (DB) when it is symmetrical. This positional mechanism can also be involved in anterior deformations, but very uncommonly. Conversely, premature fusion of one or both lambdoid sutures may be found, resulting in true craniosynostosis with posterior deformation, but this is more rare.

This article is dedicated to positional or deformational plagiocephaly and brachycephaly, describing the history, diagnosis, treatment and controversies and our experience in its management.

2. History

The history of artificial cranial deformity (ACD) is extremely old. The very first evidence of ACD dates back the middle Paleolithic period, but the most reliable evidence dates back to the Inca world in Peru and ancient Egypt. In ancient Peru, wood planks were applied to the front and back of children’s heads, and progressively tightened by bandages. In Europe, the same kind of deformities were found up to the nineteenth century, especially in France where the custom was to put a tight bandage around the skull of the newborn until adolescence, resulting in the typical “Toulousaine deformation”. Different types of head shape were found by archeologists, suggesting that this practice had several different meanings. Anthropologists have explained ACD as a method of defining the membership of social or ethnic groups [2]. For example, in the Navajo custom, warriors’ cranial deformation promoted aggressive behavior during war and instilled fear in enemies.

In the modern era, these customs have disappeared, but skull deformities may still occur. A sudden rapid increase in the incidence of DP was observed in the early 1990s. The reason for this was a

1992 recommendation by the American Academy of Paediatrics, in the so-called “back to sleep campaign” [3], to position children on their back to sleep. Previously, infants had often been placed in prone position to sleep. Since the campaign, the incidence of sudden infant death syndrome (SIDS) dropped by 50% down to 0.54 per 1,000 live births [4], but the incidence of DP increased [5–8]. In 1974, DP was found in 1 per 300 live births in prone-sleeping infants [9]: in 1994, it was estimated at 1 per 60 [5] and now is as high as 48% [10,11]. Interestingly, before 1992, anterior DP was more frequently seen than posterior DP, but its incidence dropped with the supine sleeping position [6,12,13]. This increasing incidence of posterior DP led practitioners to be more diligent in diagnosing head deformities, resulting in earlier treatment initiation [4].

3. Diagnosis

3.1. Natural history of skull growth

The neurocranium is composed of two parts: the base, which is formed by endochondral ossification, and the vault, which is formed by membranous ossification. The vault or flat bones of the skull (frontal, parietals, squamous part of the temporal and part of the occipital bones) are formed by a membranous system. These membranous bones, making up the calvaria of the skull, are each derived from the primary ossification center, from which bone formation spreads outward. However, the individual plates do not fuse during prenatal development, and newborn babies have unclosed sutures and fontanelles. These temporary discontinuities between the bones of the calvaria allow easier passage of the head through the birth canal at birth and an increase in skull size to match brain growth after birth. Cranial growth is directly linked to brain growth, and is regulated by a complex system in which the bone forms perpendicularly to the suture. This involves osteoclasts that induce bone disintegration on the inner surface of the calvaria, and osteoblasts that thicken the bone on the outer surface. These two mechanisms allow adaptation of bone thickness to obtain homogeneous curvature of the calvaria. Complete fusion between the various bones normally occurs after the age of 3, is almost complete at 8 years of age, and finishes at the end of sexual maturity, except for the metopic suture. This suture, between the two frontal bones, begins to fuse in the first year of life. Sometimes the skull fails to close completely, usually at the level of some basal sutures.

In contrast to what happens in an unfused skull, in craniosynostosis the direction of growth that is stopped is perpendicular to the closed suture and growth becomes parallel to the suture with abnormal skull growth to compensate for the lack of development of the affected part. Moreover, as bone growth is directly linked to brain growth, closure of one or more sutures leads to abnormal brain growth, so that the modification of shape is the result of both compensatory growth and also possible alteration of the

brain, especially when the other sutures begin their physiological closure process.

3.2. Epidemiology, physiopathology and risk factors

DP is far more frequent than synostotic deformation. Incidence of lambdoid synostotic plagiocephaly is approximately 3 per 100,000 births (0.003%) [14]; this is the rarest form of craniosynostosis, while a certain degree of DP can presently be found in almost half of infants under 3 months of age [11]. DP is far more frequent in boys, and the right side affected twice as often as the left [15]. DP may also be present immediately at birth, due simply to mechanical stress during the end of pregnancy passage through the birth canal [11].

During the first months of life, the bone is extremely malleable and subject to deformation. It is, therefore, easy to understand why DP is strongly associated with head position and the ability to move the head spontaneously: cervical muscle strength is variable at the beginning of life and may be not sufficient [16]. Consequently, the child spends most of the time on the back with the head sideward, putting the occipital bone under pressure [15]. Flattening may thus occur, as expansion and growth of the cranial bone is consistently resisted by an external force [14]. When the infant is positioned prone, the head rests on a surface, exerting a counterforce force on the skull, according to Newton's law, equal to the weight of the head multiplied by the force of gravity. It is this counterforce that resists cranial growth. Progressively, the area of contact ceases to grow and the brain, which continues to grow, will be displaced to areas of less resistance, creating contralateral deformation (occipital bossing) and ipsilateral flattening [17]. As the child grows older, he/she becomes able to hold the head independently, and then to sit up. Time spent lying down decreases and the progression of the deformation slows down or may even regress [16].

Another factor that affects the high prevalence of DP at a young age is that the younger the child is the faster the skull grows [18], and DP will appear or worsens faster. This explains why DP is recognized between 3 and 4 months of age [15,19], when prevalence peaks [20].

Many other DP risk factors have been identified [21–30]: intra-uterine restriction such as oligoamnios, prematurity, primiparity (due greater birth canal constraint on the firstborn), prolonged labor, assisted delivery, multiple birth, or congenital muscular torticollis. Moreover, ophthalmological abnormalities and abnormal auditory processing and motor development are also associated with greater incidence of DP [31].

3.3. Diagnosis

Diagnosis is clinical. Clinical examination should include close observation of the baby's head from all points of view (anterior, lateral, posterior and superior). It is very important to differentiate positional deformity of the skull from true craniosynostosis: prognosis, natural history and management are very different. Positional deformity is rarely seen at birth and appears rather in the first months of life. On the other hand, true lambdoid craniosynostosis is visible at birth and worsens over time. It is thus very important to question the parents and even ask for picture of the baby at birth. At birth the head should have a symmetrical round shape. Not only is chronology different, but the head deformity is also very different (Fig. 1). In unilateral lambdoid craniosynostosis, the head has a trapezoid shape, the ear is posteriorly and interiorly displaced on the side of the posterior flattening, and there is often a palpable bony ridge along the fused lambdoid suture.

In DP, the head has a parallelogram shape, the ear is displaced anteriorly on the side of the posterior flattening, and there is contralateral occipital bossing. In severe forms, ipsilateral frontal and

temporal bossing can also be present. It is very important to examine the face, because some degree of facial deformation can occur (cheekbone prominence, deviation of the chin to the contralateral side of the occipital flattening and mandibular dysmorphia). In DB, there is bilateral parietal widening to compensate for the bilateral posterior flattening; there is then no rounding on the back, the occipital external protuberance is almost invisible, and the head is disproportionately wide in its transverse diameter and the maximal anterior diameter is too short. There is often a turriccephalic aspect, with a taller vertex than the anterior part of the skull. Sometimes the bilateral posterior flattening is not completely symmetrical, one side being more affected than the other. One important feature to assess on clinical examination is neck movement, to screen for torticollis [32], which is very frequent in neonates, the most frequent being congenital muscular torticollis (CMT), in up to 30% of neonates [33]. This can be explained by fibrosis of the sternocleidomastoid muscle, impairing neck range of motion. Roger et al. found that 90% of infants that developed DP already had CMT [34]. Diagnosis is based on the rotating chair or stool test, to test rotation amplitude on both sides. In DP, torticollis limits head rotation toward the side contralateral to the posterior flattening. Clinically, this leads to an abnormal head position, tilted toward the affected side and turned toward the opposite side. Sometime facial asymmetry and problems of gross motor development can be linked to torticollis.

3.4. Anthropometric assessment and classification

Clinical assessment of anthropometric variables is essential for diagnosis, severity assessment and follow up of the deformation related to cranial growth [35]. Argenta was the first to describe a 5-type clinical classification based on physical appearance and the degree of clinical deformation for both DP and DB [36].

Precise measurements can be made using a classical measuring tape or anthropometric cranial caliper. Some authors described more modern techniques: portable or stationary laser scanner, 2D and/or 3D digital photography are the most widely used [37]. A recent study showed that the anthropometric cranial caliper was the most widely used tool in clinical practice; besides being probably the cheapest, it was shown to provide highly precise and reliable measurement with low variability [38]. However, laser scanners were more often used exclusively for large centered volumes, due to the high cost of the device [37]. Several centers have developed custom-made techniques. Hutchinson et al. used 2D and 3D digital photography, with a small stocking cap on the infant's head to hold the hair down and a soft elastic headband around the maximal occipitofrontal circumference; measurement used a custom-made computer program [39]. Van Vlimmeren et al. used a thermoplastic material to mold the outline of the infant's skull; the skull shape was then reproduced on paper for anthropometric measurement; this method, called plagiocephalometry, proved inexpensive, reliable and accurate [40]. 3D digital photography is also used to assess facial dysmorphia [41].

Several parameters should be recorded:

- head circumference: measured at the glabella, passing by the posterior occipital protuberance (opisthocranion);
- cranial length: from glabella to opisthocranion;
- cranial width: maximal bi-parietal length;
- maximal and minimal cranial diagonal: respectively, from the tip of the parieto-occipital bone on the unaffected side to the tip of the contralateral frontal bossing, and from the middle of the flattening to the tip of the contralateral frontal bone. In DP the maximal diagonal should be on the unaffected side and the minimal diagonal on the side of the flattening.

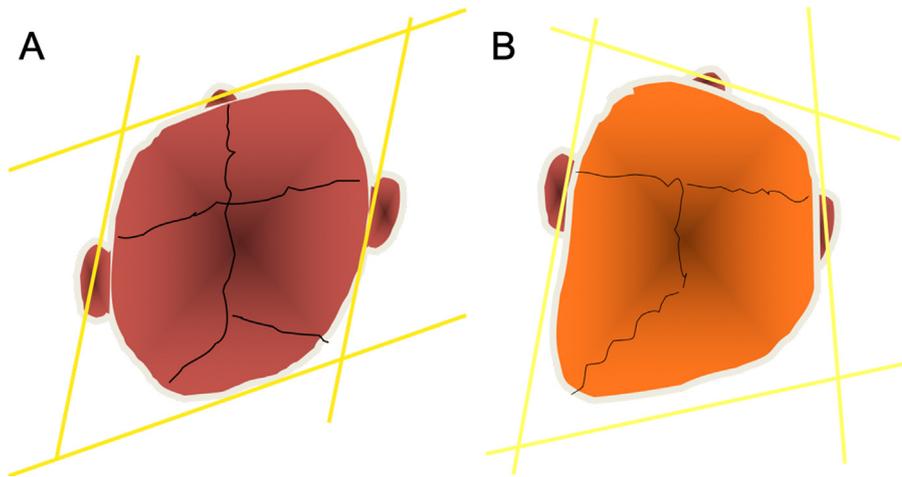


Fig. 1. Drawing representing the differences between position plagiocephaly (A) and true lambdoid craniosynostosis.

Once all these parameters have been measured, several indices should be calculated:

- cranial index (CI): ratio of head width to head length, multiplied by 100 ($CI = (\text{width}/\text{length}) \times 100$);
- cranial vault asymmetry index (CVAI): ratio of oblique measurements ($CVAI = [(\text{diagonal of the unaffected side} - \text{diagonal of the affected side}) / \text{diagonal of the unaffected side}] \times 100$);
- oblique cranial length ratio (OCLR): ratio of longest to shortest oblique cranial diameter;
- transcranial diagonal difference (TD): diagonal of the unaffected side minus diagonal of the affected side.

These measurements should be compared first to age-related norms, and secondly they should be taken regularly for a given patient to compare individual progression [21]. For children with thick hair, measurement error can be reduced by using a caliper with its tips pressed firmly against the skin [42].

The indices to be used depend on the type of deformity (DP or DB). DP is more accurately monitored by TDD and DB by CI. All the parameters aim to contribute to diagnosis, to assess the severity of deformation and to monitor progression of head shape during treatment.

Using these anthropometric parameters, several classifications of positional head deformity have been proposed. Hutchison et al. proposed a DP classification in 3 stages according to TDD: mild for TDD 3–10 mm, moderate for 10–12 mm and severe for > 12 mm [39]. DP is also defined by $OCLR \geq 106\%$ [39]. DB can be classified in 3 stages according to CI: mild for CI 82–90%, moderate for 90–100% and severe for > 100% [39]. Wilbrand et al. used CVAI in a 3-level (mild/moderate/severe) severity categorization of DP and DB according to age and gender, based on curves of normal cranial vault growth percentile during the first year of life: mild for CVAI 75th to 90th percentile, moderate for 90th to 97th, and severe for > 97th [35].

3.5. Complementary imaging

There is no need for skull imaging for diagnosis of typical DP, as analysis of the skull deformity coupled with interview is in most cases sufficient. Systematic skull computed tomography scan (CT) is thus not recommended in typical cases [15] (Fig. 2). The pediatric population is vulnerable, and CT entails radiation and its consequences [43,44]. The vulnerability of children comes from the fact that they have a long remaining life expectancy and a thinner, less

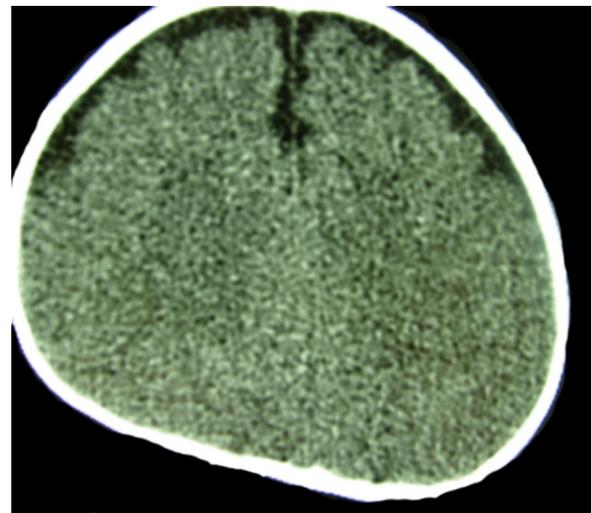


Fig. 2. Axial CT slice showing position plagiocephaly. It is no longer recommended to use CT.

dense skull that protects the brain less against radiation. Moreover, it may be necessary to sedate the baby, which can also lead to complications. If a CT scan is needed in case of doubtful diagnosis, 3D reconstruction may be useful [45]. To reduce radiation levels for children, some teams use low-dose CT that does not impair image quality while maintaining the diagnostic contribution [46]. Plain skull X-rays are also commonly used to confirm diagnosis when needed [47]; but X-ray is an irradiating examination, although far less than CT. More recently, cranial ultrasound (CUS) was described, showing a clear hypoechoic gap in case of patent suture and obliteration of the gap in case of synostosis [48]. It has thus become more and more widely used to confirm lambdoid suture patency in DP [49,50]. In addition to confirming diagnosis of DP, CUS can be used to explore the severity of the deformation. Kim et al. used an ultrasound method to measure the occipital angle ratio (angle between the lines projected along the lambdoid sutures of the skull), which they reported to correlated positively with the classic anthropometric variables [51].

3.6. Potential complications

Because neurodevelopment may be retarded in children with single suture synostosis [52,53], health care providers that manage

DP began to be interested in the potential implications of DP for neurodevelopment (mental retardation, retarded motor function, language, learning, etc.). In the past two decades, studies reported retarded neurodevelopment in children with DP [16,54–56]. Miller and Clarren found out that children with DP were more likely to require special education [16]. Kordestani et al. and Pancha et al. found significant differences in psychomotor development and mental developmental indices between a DP and an aged-matched population [54,55]. Speltz et al. reported DP to be associated with early neurodevelopmental disadvantage, most clearly in motor function [56]. The main mechanisms for these findings were reviewed by Collett et al. [57], who distinguished 3 possible pathways. The first is mechanical: the deformation of the skull leads to deformation of the brain and impaired function. This pathway is an extrapolation of the findings of cortical and subcortical anomalies in synostotic plagiocephaly [58]. However, no evidence could be found in the literature for DP, and the authors also showed that the cortical and subcortical anomalies found in craniosynostosis were not obviously related to the shape of the skull itself [58]. No evidence of raised intracranial pressure in DP children has been found [7,15,19]. The second pathway is environmental: if the child's movement is too frequently restricted (over-use of car seats or baby swings, or unvarying sleep positioning), this inexorably leads to delay or deficits in the development of “motor-driven” cognitive functions. The motor development of babies was shown to be related to sleep position and the amount of “tummy time” [59]. On this hypothesis, the developmental delay is a consequence of excessive constraint rather than being caused by DP. The last pathway is developmental: central nervous system (CNS) pathologies may lead to tonus disorder and DP. Whether deficiency is the cause or the consequence of the DP remains in doubt, but it is more likely that developmental delay is related to the CNS pathology itself [57]. It should be added that the retardation can be aggravated by impairment of motor control, as in the second pathway. One very important point, made by Paquereau, is that no studies have explored the effect of treatment on neurocognitive and motor behavior [60]. Because there is no evidence of DP directly causing neurodevelopmental delay, some authors consider DP as a purely an aesthetic issue [61,62].

Neurodevelopment is not the only issue in DP, and other potential complications have also been studied.

Ocular motion abnormalities are well known in craniosynostosis, with horizontal and vertical strabismus [63]. Very few studies have reported ophthalmologic findings in DP. Gupta et al. found that the prevalence of strabismus and astigmatism in DP was comparable to that in the general population [64]. In a very recent study [65], none of the infants with DP had anisometropic astigmatism or occlusion amblyopia, with 93.7% having some pseudoptosis. These findings differ from those of Gupta et al. regarding the rate of strabismus, affecting 15.6% of patients, compared to 1% for Gupta, thus considerably higher than the 2–5% prevalence reported in the general population [66]. However, the authors considered that this “may represent referral bias due to the nature of our sub-specialty clinic or sampling error due to the size of our study population”, and concluded that the frequent ocular asymmetry found in DP had no clinical significance.

Orthodontic problems such as malocclusion are a major point of interest in pediatric dentistry. Early malocclusion can lead to orthodontic problems in the permanent dentition. DP was studied in this population, as it causes facial asymmetry, and higher prevalence than normal is reported in unilateral coronal craniosynostosis [67]. In DP, the most recent study, by Kluba et al., in 2016 [68], reported higher prevalence of orthodontic abnormalities in children with previous positional plagiocephaly compared to a control group of normal children, although none of the differences were

significant [68]. There is thus still no scientific evidence for the involvement of DP in orthodontic problems.

4. Management

The debate over the management of DP is still ongoing. Different conceptions of the natural history of DP underlie the different therapeutic options. Some authors believe that DP corrects itself spontaneously [69], while others disagree [70].

However, because the deformation of the skull is not present or is minimal at birth and appears during the first months of life, the most important aspect of management is prevention, to avoid the deformation occurring in the first place. Once the deformation is present, it is unanimously agreed that the earlier the treatment, is the better the result, independently of type of deformation, because of the remodelling capacity of the growing skull [60,71].

In this section, we review the different techniques of prevention, and then the different types of management proposed in the literature.

4.1. Prevention

The main aspect of prevention is teaching the parents good positioning and handling and how to organize the baby's environment so as to avoid prolonged uneven external forces on the skull. Two studies specifically proved the usefulness of prevention in DP [72,73]. The first was a non-randomized trial conducted in France [72] in which the prevalence of DP at 4 months of age was analyzed. One group received classic positioning advice to prevent SIDS, without specific education about DP, and the second group received the classic SIDS advice plus specific oral recommendations by a trained pediatrician for the prevention of DP and an information booklet on DP. This early, simple and cheap preventive measure significantly lowered the incidence of DP in the intervention group. A few years later, a randomized trial confirmed these findings and added that the intervention also reduced the severity of the deformation when present [73]. These studies are of paramount importance because they proved that a simple inexpensive educational program could favorably impact the incidence of DP. Prevention should be started by pediatricians during the stay in maternity. The first step is to draw pediatricians' attention to this pathology and to the preventive messages to be delivered to the parents. The written information booklet is also essential, to help the parents adhere to the preventive measures [74].

These preventive measures are aimed at reducing the time the infant spends in supine position and at promoting an unhindered environment for the child to develop spontaneous and symmetrical motor function. The main preventive instructions are summarized in Table 1.

4.2. Active counter-positioning, physiotherapy and osteopathy

Even with early prevention, deformation may occur [73]. In that case, several therapeutic options are available. The first is active counter-positioning. The purpose is to actively prevent the baby leaning on their affected side. To achieve this, thorough parental guidance is needed. The parents' adherence to the program is essential, because they need to implement active repositioning (with the unaffected side against the mattress) whenever the infant is lying down (in sleep or in play), and to change the position of the crib in the room or alternate which end of the crib the baby is placed at to sleep, so that the view of the room the baby prefers will be found via the opposite position; the parents should do the same when using the changing table, and likewise, instead of putting toys on

Table 1
Preventive instructions to reduce the incidence and severity of DP.

Situation	Instructions
Sleeping	Supine position (to prevent SIDS) Alternate the position of head from left to right Switch the side of the bed where you put the head if something always catches his/her attention on one side Size of the bed should large enough to able your baby to move/turn freely Stimuli should be placed evenly on both sides of the bed
Feeding	Alternate the position while holding the baby during breast-feeding (alternate breasts) or bottle-feeding
Tummy time	Always under adult supervision From the first day of life Gradually increase the time from just 1 or 2 minute At least 30–60 minutes a day
Travel	Minimize time in car seat and carrier Alternate head position while in car seat or carrier
Day time	No prolonged restriction of movement Put stimuli on both side while playing Be careful to switch sides while playing with the child

both sides, as usually recommended for preventive measures, toys should be placed only on the unaffected side. During feeding, parents should put the unaffected side against their chest. “Tummy time” should be initiated, or increased if already practiced.

Apart from active counter-positioning, physiotherapy (PT) can help to correct positional preference and thus DP, acting mainly on the associated torticollis. PT also contributes to the parents’ education, teaching stretching exercises that will loosen the sternocleidomastoid and the trapezius muscle [75,76]. There used to be no strong scientific evidence for the effectiveness of PT; however, a randomized trial by Van Vlimmeren et al. proved its efficacy [77]. They concluded that PT and parental counseling on the different preventive and counter-positioning measures were more effective than parental education alone. However, it is to be noted that the PT program was specifically developed for this trial and was performed by 6 experienced pediatric physical therapists specially trained in the program. It was implemented between 7 weeks and 6 months of age, with 8 sessions. Thus, the trial did not reflect usual PT, which is generally performed initially by the parents themselves. Of course, if the torticollis or DP worsens, the parents need to be referred to a specialist [62,74]. It is recommended to practice the PT during each change of diapers: placing one hand on the child’s upper chest and moving the head with the other hand; the first exercise consists of chin-to-shoulder rotation and the second of ear-to-shoulder tilting. These movements should be performed on both sides and repeated several times. Steinberg et al. analyzed risk factors for failure of conservative treatment (counter-positioning alone or associated with PT); deformity severity (as assessed by the anthropometric variables), persistence of torticollis beyond 6 months of age, and neuromuscular developmental delay were risk factors, in addition to age and parental compliance [78].

The last active maneuver to deal with DP is osteopathic manipulative treatment (OMT). Osteopaths are trained to use palpation to identify abnormal suture mobility during the respiratory cycle, shortening of tissue (fascia and cervical muscles) and abnormal position of the spine, pelvis and lower limbs. OMT aims to mobilize all the impaired structures by gentle maneuvers, to restore the normal balance of the body. A randomized trial comparing OMT



Fig. 3. Intra-operative view of a two-year-old boy with severe plagiocephaly that was not treated with a helmet.

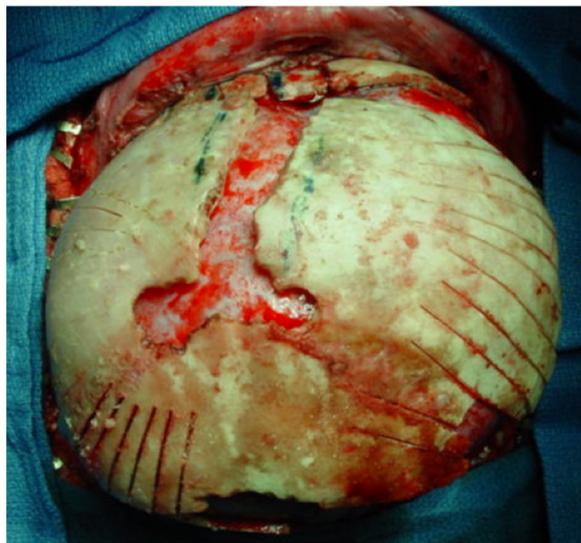


Fig. 4. Intra-operative view of the Lyon technique, which consists in posterior vault remodeling in prone position.

versus sham treatment showed significant improvement in head asymmetry and concluded that OMT is effective in the management of DP [79]. OMT should be performed by trained specialist as a complement or when active counter-positioning and PT have failed [80].

4.3. Helmet molding therapy

The helmet uses the forces exerted by the growing brain so as to alleviate skull deformation. This treatment seems to be effective, although efficacy remains to be scientifically demonstrated.

4.4. Surgery

When the child is referred late with a severe deformation or when he treatment was not effective, surgical correction may be proposed, as reported by Di Rocco et al., [81] and by Czorny (not published) (Fig. 3). The indications are very strict, as this surgical

procedure is associated with possible life-threatening complications [82]. Generally, surgical correction is proposed after 2 years of age [81]. Many surgical techniques have been described. Our technique in Lyon is a modification of the one described by Czorny. It consists in posterior vault remodeling with the patient in prone position (Fig. 4). A parieto-occipital bone flap is performed, then, an osseous arc is elevated at the vertex, positioned in the occipital region and fixed with absorbable plates or sutures. The occipital bone flap is then cut and molded into two symmetrical parietal bossings to build a more symmetrical posterior cranial vault.

Results are immediate and do not require molding therapy. The disadvantages are the need for surgery, and especially the risk of dural tear, persisting bone defects and severe blood loss [81] such as described in craniosynostosis correction. In experienced hands, the risk of lesion of the sagittal sinus or of the torcular is low. Caution is also needed to control bleeding from the Pacchioni granulations, which can be induced by ventral positioning. Any hemorrhage should be controlled using Surgicel® and Cottonoid® or by ligature of the dura matter. Surgeons should avoid aspiration, and rather use compression with surgical cottonoid.

5. Conclusions

DP is presently a major issue in the pediatric community. Prevention is the keystone, to decrease incidence. Primary health-care providers, who are at the forefront of parental education, must educate the parents from the outset.

Diagnosis of DP is clinical. Skull CT scan should be reserved for complex cases when diagnosis is not certain and craniosynostosis is suspected.

Once DP is present, there are several effective means to deal with it. Active counter-positioning, PT and OMT can be recommended. The last two options need to be performed by well-trained specialists. Close follow-up is mandatory, to help the parents and to improve treatment adherence. Pediatricians also need to know when to refer the child to a neurosurgeon if progression is not satisfactory. Helmet molding therapy is recommended for severe deformations and when the various non-instrumental therapies have failed; nevertheless, it should be started early, ideally before 6 months of age, to have maximum effectiveness. Conversely, surgery should be reserved for extremely deformed cases with severe aesthetic impact.

Disclosure of interest

The authors declare that they have no competing interest.

References

- Governale LS. Craniosynostosis. *Pediatr Neurol* 2015;53(5):394–401. <http://dx.doi.org/10.1016/j.pediatrneurol.2015.07.006>.
- Ayer A, Campbell A, Appelboom G, et al. The sociopolitical history and physiological underpinnings of skull deformation. *Neurosurg Focus* 2010;29(6):E1. <http://dx.doi.org/10.3171/2010.9.FOCUS10202>.
- American Academy of Pediatrics. AAP Task Force on Infant Positioning and SIDS: Positioning and SIDS. *Pediatrics* 1992;89(6 Pt 1):1120–6.
- Branch LG, Kesty K, Krebs E, Wright L, Leger S, David LR. Deformational plagiocephaly and craniosynostosis: trends in diagnosis and treatment after the “back to sleep” campaign. *J Craniofac Surg* 2015;26(1):147–50. <http://dx.doi.org/10.1097/SCS.0000000000001401>.
- Argenta LC, David LR, Wilson JA, Bell WO. An increase in infant cranial deformity with supine sleeping position. *J Craniofac Surg* 1996;7(1):5–11.
- Turk AE, McCarthy JG, Thorne CH, Wisoff JH. The “back to sleep campaign” and deformational plagiocephaly: is there cause for concern? *J Craniofac Surg* 1996;7(1):12–8.
- Kane AA, Mitchell LE, Craven KP, Marsh JL. Observations on a recent increase in plagiocephaly without synostosis. *Pediatrics* 1996;97(6 Pt 1):877–85.
- McKinney CM, Cunningham ML, Holt VL, Leroux B, Starr JR. Characteristics of 2733 cases diagnosed with deformational plagiocephaly and changes in risk factors over time. *Cleft Palate-Craniofacial J Off Publ Am Cleft Palate-Craniofacial Assoc* 2008;45(2):208–16. <http://dx.doi.org/10.1597/06-227.1>.
- Graham Jr JM, Gomez M, Halberg A, et al. Management of deformational plagiocephaly: Repositioning versus orthotic therapy. *J Pediatr* 2005;146(2):258–62. <http://dx.doi.org/10.1016/j.jpeds.2004.10.016>.
- Rogers GF, Miller J, Mulliken JB. Comparison of a modifiable cranial cup versus repositioning and cervical stretching for the early correction of deformational posterior plagiocephaly. *Plast Reconstr Surg* 2008;121(3):941–7. <http://dx.doi.org/10.1097/01.prs.0000299938.00229.3e>.
- Mawji A, Vollman AR, Hatfield J, McNeil DA, Sauv e R. The incidence of positional plagiocephaly: a cohort study. *Pediatrics* 2013;132(2):298–304. <http://dx.doi.org/10.1542/peds.2012-3438>.
- Peitsch WK, Keefer CH, LaBrie RA, Mulliken JB. Incidence of cranial asymmetry in healthy newborns. *Pediatrics* 2002;110(6):e72.
- Spermon J, Spermon-Marijnen R, Scholten-Peeters W. Clinical classification of deformational plagiocephaly according to Argentina: a reliability study. *J Craniofac Surg* 2008;19(3):664–8. <http://dx.doi.org/10.1097/SCS.0b013e31816ae3ec>.
- Rekate HL. Occipital plagiocephaly: a critical review of the literature. *J Neurosurg* 1998;89(1):24–30. <http://dx.doi.org/10.3171/jns.1998.89.1.0024>.
- Vernet O, de Ribaupierre S, Cavin B, Rilliet B. Traitement des plagiocephalies post rieures d’origine positionnelle. *Arch P diatrie* 2008;15(12):1829–33. <http://dx.doi.org/10.1016/j.arcped.2008.09.007>.
- Miller RI, Clarren SK. Long-term developmental outcomes in patients with deformational plagiocephaly. *Pediatrics* 2000;105(2):E26.
- Rogers GF. Deformational plagiocephaly, brachycephaly, and scaphocephaly. Part I: terminology, diagnosis, and etiopathogenesis. *J Craniofac Surg* 2011;22(1):9–16. <http://dx.doi.org/10.1097/SCS.0b013e3181f6c313>.
- Guo S, Roche AF, Moore WM. Reference data for head circumference and 1-month increments from 1 to 12 months of age. *J Pediatr* 1988;113(3):490–4.
- Kluba S, Lypke J, Kraut W, Krimmel M, Haas-Lude K, Reinert S. Preclinical pathways to treatment in infants with positional cranial deformity. *Int J Oral Maxillofac Surg* 2014;43(10):1171–5. <http://dx.doi.org/10.1016/j.ijom.2014.05.011>.
- Boere-Boonekamp MM, van der Linden-Kuiper LT. Positional preference: prevalence in infants and follow-up after two years. *Pediatrics* 2001;107(2):339–43.
- Looman WS, Flannery ABK. Evidence-based care of the child with deformational plagiocephaly, Part I: assessment and diagnosis. *J Pediatr Health Care Off Publ Natl Assoc Pediatr Nurse Assoc Pract* 2012;26(4):242–50. <http://dx.doi.org/10.1016/j.pedhc.2011.10.003> [quiz 251–253].
- Hutchison BL, Stewart AW, Mitchell EA. Characteristics, head shape measurements and developmental delay in 287 consecutive infants attending a plagiocephaly clinic. *Acta Paediatr Oslo Nor* 1992 2009;98(9):1494–9. <http://dx.doi.org/10.1111/j.1651-2227.2009.01356.x>.
- Rubio AS, Griffet JR, Caci H, B rard E, El Hayek T, Boutt  P. The moulded baby syndrome: incidence and risk factors regarding 1,001 neonates. *Eur J Pediatr* 2009;188(5):605–11. <http://dx.doi.org/10.1007/s00431-008-0806-y>.
- McKinney CM, Cunningham ML, Holt VL, Leroux B, Starr JR. A case-control study of infant, maternal and perinatal characteristics associated with deformational plagiocephaly. *Paediatr Perinat Epidemiol* 2009;23(4):332–45. <http://dx.doi.org/10.1111/j.1365-3016.2009.01038.x>.
- Oh AK, Hoy EA, Rogers GF. Predictors of severity in deformational plagiocephaly. *J Craniofac Surg* 2009;20(Suppl 1):685–9. <http://dx.doi.org/10.1097/SCS.0b013e318193d6e5>.
- Robinson S, Proctor M. Diagnosis and management of deformational plagiocephaly. *J Neurosurg Pediatr* 2009;3(4):284–95. <http://dx.doi.org/10.3171/2009.1.PEDS08330>.
- Mulliken JB, Vander Woude DL, Hansen M, LaBrie RA, Scott RM. Analysis of posterior plagiocephaly: deformational versus synostotic. *Plast Reconstr Surg* 1999;103(2):371–80.
- Dias MS, Klein DM. Occipital plagiocephaly: deformation or lambdoid synostosis? II. A unifying theory regarding pathogenesis. *Pediatr Neurosurg* 1996;24(2):69–73.
- Littlefield TR, Kelly KM, Pomatto JK, Beals SP. Multiple-birth infants at higher risk for development of deformational plagiocephaly. *Pediatrics* 1999;103(3):565–9.
- de Chala n TMB, Park S. Torticollis associated with positional plagiocephaly: a growing epidemic. *J Craniofac Surg* 2005;16(3):411–8.
- Shweikeh F, Nu o M, Danielpour M, Krieger MD, Drazin D. Positional plagiocephaly: an analysis of the literature on the effectiveness of current guidelines. *Neurosurg Focus* 2013;35(4):E1. <http://dx.doi.org/10.3171/2013.8.FOCUS13261>.
- Kuo AA, Tritasavit S, Graham JM. Congenital muscular torticollis and positional plagiocephaly. *Pediatr Rev Am Acad Pediatr* 2014;35(2):79–87. <http://dx.doi.org/10.1542/pir.35-2-79> [quiz 87].
- Stellwagen L, Hubbard E, Chambers C, Jones KL. Torticollis, facial asymmetry and plagiocephaly in normal newborns. *Arch Dis Child* 2008;93(10):827–31. <http://dx.doi.org/10.1136/adc.2007.124123>.
- Rogers GF, Oh AK, Mulliken JB. The role of congenital muscular torticollis in the development of deformational plagiocephaly. *Plast Reconstr Surg* 2009;123(2):643–52. <http://dx.doi.org/10.1097/PRS.0b013e318196b9be>.
- Wilbrand J-F, Schmidtberg K, Bierther U, et al. Clinical classification of infant nonsynostotic cranial deformity. *J Pediatr* 2012;161(6):1120–5. <http://dx.doi.org/10.1016/j.jpeds.2012.05.023> [e1].
- Argenta L, David L, Thompson J. Clinical classification of positional plagiocephaly. *J Craniofac Surg* 2004;15(3):368–72.

- [37] Purnell CA, Benz AW, Gosain AK. Assessment of Head Shape by Craniofacial Teams: Structuring Practice Parameters to Optimize Efficiency. *J Craniofac Surg* 2015;26(6):1808–11, <http://dx.doi.org/10.1097/SCS.0000000000001948>.
- [38] Wilbrand J-F, Wilbrand M, Pons-Kuehnemann J, et al. Value and reliability of anthropometric measurements of cranial deformity in early childhood. *J Cranio-Maxillo-fac Surg Off Publ Eur Assoc Cranio-Maxillo-fac Surg* 2011;39(1):24–9, <http://dx.doi.org/10.1016/j.jcms.2010.03.010>.
- [39] Hutchison BL, Hutchison LAD, Thompson JMD, Mitchell EA. Quantification of plagiocephaly and brachycephaly in infants using a digital photographic technique. *Cleft Palate-Craniofacial J Off Publ Am Cleft Palate-Craniofacial Assoc* 2005;42(5):539–47, <http://dx.doi.org/10.1597/04-059r.1>.
- [40] van Vlimmeren LA, Takken T, van Adrichem LNA, van der Graaf Y, Helders PJM, Engelbert RHH. Plagiocephalometry: A non-invasive method to quantify asymmetry of the skull; a reliability study. *Eur J Pediatr* 2006;165(3):149–57, <http://dx.doi.org/10.1007/s00431-005-0011-1>.
- [41] Othman SA, Ahmad R, Mercant AF, Jamaludin M. Reproducibility of facial soft tissue landmarks on facial images captured on a 3D camera. *Aust Orthod J* 2013;29(1):58–65.
- [42] Hall JG, Allanson JE, Gripp KW, Slavotinek AM. *Handbook of Physical Measurements*. Oxford University Press; 2007.
- [43] Brenner D, Elliston C, Hall E, Berdon W. Estimated risks of radiation-induced fatal cancer from pediatric CT. *AJR Am J Roentgenol* 2001;176(2):289–96, <http://dx.doi.org/10.2214/ajr.176.2.1760289>.
- [44] Pearce MS, Salotti JA, Little MP, et al. Radiation exposure from CT scans in childhood and subsequent risk of leukaemia and brain tumours: a retrospective cohort study. *Lancet Lond Engl* 2012;380(9840):499–505, [http://dx.doi.org/10.1016/S0140-6736\(12\)60815-0](http://dx.doi.org/10.1016/S0140-6736(12)60815-0).
- [45] Mottolese C, Szathmari A, Ricci AC, Ginguene C, Simon E, Paulus C. Positional plagiocephaly: the place of cranial orthotics. *Neurochirurgie* 2006;52(2–3 Pt 2):184–94.
- [46] Morton RP, Reynolds RM, Ramakrishna R, et al. Low-dose head computed tomography in children: a single institutional experience in pediatric radiation risk reduction: clinical article. *J Neurosurg Pediatr* 2013;12(4):406–10, <http://dx.doi.org/10.3171/2013.7.PEDS12631>.
- [47] Linz C, Collmann H, Meyer-Marcotty P, et al. Occipital plagiocephaly: unilateral lambdoid synostosis versus positional plagiocephaly. *Arch Dis Child* 2015;100(2):152–7, <http://dx.doi.org/10.1136/archdischild-2014-305944>.
- [48] Sze RW, Parisi MT, Sidhu M, et al. Ultrasound screening of the lambdoid suture in the child with posterior plagiocephaly. *Pediatr Radiol* 2003;33(9):630–6, <http://dx.doi.org/10.1007/s00247-003-1009-3>.
- [49] Krimmel M, Will B, Wolff M, et al. Value of high-resolution ultrasound in the differential diagnosis of scaphocephaly and occipital plagiocephaly. *Int J Oral Maxillofac Surg* 2012;41(7):797–800, <http://dx.doi.org/10.1016/j.ijom.2012.02.022>.
- [50] Rozovsky K, Udjus K, Wilson N, Barrowman NJ, Simanovsky N, Miller E. Cranial ultrasound as a first-line imaging examination for craniosynostosis. *Pediatrics* 2016;137(2):1–9, <http://dx.doi.org/10.1542/peds.2015-2230>.
- [51] Kim JK, Kwon DR, Park G-Y. A new ultrasound method for assessment of head shape change in infants with plagiocephaly. *Ann Rehabil Med* 2014;38(4):541–7, <http://dx.doi.org/10.5535/arm.2014.38.4.541>.
- [52] Magge SN, Westerveld M, Pruzinsky T, Persing JA. Long-term neuropsychological effects of sagittal craniosynostosis on child development. *J Craniofac Surg* 2002;13(1):99–104.
- [53] Shipster C, Hearst D, Somerville A, Stackhouse J, Hayward R, Wade A. Speech, language, and cognitive development in children with isolated sagittal synostosis. *Dev Med Child Neurol* 2003;45(1):34–43.
- [54] Panchal J, Amirshaybani H, Gurwitch R, et al. Neurodevelopment in children with single-suture craniosynostosis and plagiocephaly without synostosis. *Plast Reconstr Surg* 2001;108(6):1492–8 [Discussion 1499–1500].
- [55] Kordestani RK, Patel S, Bard DE, Gurwitch R, Panchal J. Neurodevelopmental delays in children with deformational plagiocephaly. *Plast Reconstr Surg* 2006;117(1):207–18 [Discussion 219–220].
- [56] Speltz ML, Collett BR, Stott-Miller M, et al. Case-control study of neurodevelopment in deformational plagiocephaly. *Pediatrics* 2010;125(3):537–42, <http://dx.doi.org/10.1542/peds.2009-0052>.
- [57] Collett B, Breiger D, King D, Cunningham M, Speltz M. Neurodevelopmental implications of “deformational” plagiocephaly. *J Dev Behav Pediatr JDBP* 2005;26(5):379–89.
- [58] Aldridge K, Marsh JL, Govier D, Richtsmeier JT. Central nervous system phenotypes in craniosynostosis. *J Anat* 2002;201(1):31–9.
- [59] Salls JS, Silverman LN, Gatty CM. The relationship of infant sleep and play positioning to motor milestone achievement. *Am J Occup Ther Off Publ Am Occup Ther Assoc* 2002;56(5):577–80.
- [60] Paquereau J. Non-surgical management of posterior positional plagiocephaly: orthotics versus repositioning. *Ann Phys Rehabil Med* 2013;56(3):231–49, <http://dx.doi.org/10.1016/j.rehab.2012.12.005>.
- [61] Gautschi OP, Riilliet B, Schaller K, Jenny B. Positional plagiocephaly in infancy: diagnosis and management. *Praxis* 2013;102(25):1537–42, <http://dx.doi.org/10.1024/1661-8157/a001506>.
- [62] Flannery ABK, Looman WS, Kemper K. Evidence-based care of the child with deformational plagiocephaly, Part II: Management. *J Pediatr Health Care* 2012;26(5):320–31, <http://dx.doi.org/10.1016/j.pedhc.2011.10.002>.
- [63] Morax S. Oculo-motor disorders in craniofacial malformations. *J Maxillofac Surg* 1984;12(1):1–10.
- [64] Gupta PC, Foster J, Crowe S, Papay FA, Luciano M, Traboulsi EL. Ophthalmologic findings in patients with nonsyndromic plagiocephaly. *J Craniofac Surg* 2003;14(4):529–32.
- [65] Schweigert A, Merrill K, Mokhtarzadeh A, Harrison A. Periocular Asymmetry in Infants with Deformational Posterior Plagiocephaly. *J Binocul Vis Ocul Motil* 2019;69(1):18–23, <http://dx.doi.org/10.1080/2576117X.2019.1565275>.
- [66] Fieß A, Kölb-Keerl R, Schuster AK, et al. Prevalence and associated factors of strabismus in former preterm and full-term infants between 4 and 10 Years of age. *BMC Ophthalmol* 2017;17(1):228, <http://dx.doi.org/10.1186/s12886-017-0605-1>.
- [67] Pelo S, Marianetti TM, Cacucci L, et al. Occlusal alterations in unilateral coronal craniosynostosis. *Int J Oral Maxillofac Surg* 2011;40(8):805–9, <http://dx.doi.org/10.1016/j.ijom.2011.02.023>.
- [68] Kluba S, Rofkopp F, Kraut W, et al. Malocclusion in the primary dentition in children with and without deformational plagiocephaly. *Clin Oral Investig* 2016;20(9):2395–401, <http://dx.doi.org/10.1007/s00784-016-1716-4>.
- [69] Hutchison BL, Hutchison LAD, Thompson JMD, Mitchell EA. Plagiocephaly and brachycephaly in the first two years of life: a prospective cohort study. *Pediatrics* 2004;114(4):970–80, <http://dx.doi.org/10.1542/peds.2003-0668-F>.
- [70] Wilbrand J-F, Lautenbacher N, Pons-Kühnemann J, et al. Treated versus untreated positional head deformity. *J Craniofac Surg* 2016;27(1):13–8, <http://dx.doi.org/10.1097/SCS.0000000000002167>.
- [71] Pollack IF, Losken HW, Fasick P. Diagnosis and management of posterior plagiocephaly. *Pediatrics* 1997;99(2):180–5.
- [72] Cavalier A, Picot M-C, Artiaga C, et al. Prevention of deformational plagiocephaly in neonates. *Early Hum Dev* 2011;87(8):537–43, <http://dx.doi.org/10.1016/j.earlhumdev.2011.04.007>.
- [73] Aarnivala H, Vuollo V, Harila V, Heikkinen T, Pirttiniemi P, Valkama AM. Preventing deformational plagiocephaly through parent guidance: a randomized, controlled trial. *Eur J Pediatr* 2015;174(9):1197–208, <http://dx.doi.org/10.1007/s00431-015-2520-x>.
- [74] Laughlin J, Luerssen TG, Dias MS. Committee on practice and ambulatory medicine, section on neurological surgery. Prevention and management of positional skull deformities in infants. *Pediatrics* 2011;128(6):1236–41, <http://dx.doi.org/10.1542/peds.2011-2220>.
- [75] Biggs WS. Diagnosis and management of positional head deformity. *Am Fam Physician* 2003;67(9):1953–6.
- [76] Persing J, James H, Swanson J, Kattwinkel J. American Academy of Pediatrics Committee on Practice and Ambulatory Medicine, section on plastic surgery and section on neurological surgery. Prevention and management of positional skull deformities in infants. American Academy of Pediatrics Committee on Practice and Ambulatory Medicine, section on plastic surgery and section on neurological surgery. *Pediatrics* 2003;112(1 Pt 1):199–202.
- [77] van Vlimmeren LA, van der Graaf Y, Boere-Boonekamp MM, L’Hoir MP, Helders PJM, Engelbert RHH. Effect of pediatric physical therapy on deformational plagiocephaly in children with positional preference: A randomized controlled trial. *Arch Pediatr Adolesc Med* 2008;162(8):712–8, <http://dx.doi.org/10.1001/archpedi.162.8.712>.
- [78] Steinberg JP, Rawlani R, Humphries LS, Rawlani V, Vicari FA. Effectiveness of conservative therapy and helmet therapy for positional cranial deformation. *Plast Reconstr Surg* 2015;135(3):833–42, <http://dx.doi.org/10.1097/PRS.0000000000000955>.
- [79] Philippi H, Faldum A, Schleupen A, et al. Infantile postural asymmetry and osteopathic treatment: a randomized therapeutic trial. *Dev Med Child Neurol* 2006;48(1):5–9, <http://dx.doi.org/10.1017/S001216220600003X> [Discussion 4].
- [80] Amiel-Tison C, Soyez-Papiernik E. Place de l’ostéopathie dans la correction des déformations crâniennes du nouveau-né et du jeune enfant. *Arch Pédiatrie* 2008;15(Supplement 1):24–30, [http://dx.doi.org/10.1016/S0929-693X\(08\)73944-7](http://dx.doi.org/10.1016/S0929-693X(08)73944-7).
- [81] Di Rocco F, Marchac A, Duracher C, et al. Posterior remodeling flap for posterior plagiocephaly. *Childs Nerv Syst ChNS Off J Int Soc Pediatr Neurosurg* 2012;28(9):1395–7, <http://dx.doi.org/10.1007/s00381-012-1842-5>.
- [82] Marchac A, Arnaud E, Di Rocco F, Michienzi J, Renier D. Severe deformational plagiocephaly: long-term results of surgical treatment. *J Craniofac Surg* 2011;22(1):24–9, <http://dx.doi.org/10.1097/SCS.0b013e3181f7dd4a>.

P.-A. Beuriat^{a,b,c}

A. Szathmari^{a,b,c}

F. Di Rocco^{a,b,c}

C. Mottolese^{a,b,c,*}

^a Department of Pediatric Neurosurgery, hôpital Femme-Mère-Enfant, 32, avenue du Doyen-Jean-Lépine, 69677 Bron Cedex, France

^b *Claude-Bernard University Lyon 1, 8, avenue
Rockefeller, 69003 Lyon, France*

^c *Reference center for craniosynostosis, hôpital
Femme-Mère-Enfant, 32, avenue du
Doyen-Jean-Lépine, 69677 Bron Cedex, France*

* Corresponding author at: Department of Pediatric
Neurosurgery, hôpital Femme-Mère-Enfant, 32,
avenue du Doyen-Jean-Lépine, 69677 Bron Cedex,
France.

E-mail address: carmine.mottolese@chu-lyon.fr
(C. Mottolese)

Received 23 July 2019

Received in revised form 1st September 2019

Accepted 3 September 2019
Available online 25 September 2019