



## Diagnosis and treatment of plagiocephaly: clinical case presentation and clinical review

### *Diagnóstico y tratamiento de plagiocefalia: presentación de caso clínico y revisión de literatura*

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#### ABSTRACT

Plagiocephaly can be defined as the premature closing of the frontal-parietal suture or unilateral coronal suture which is caused by a frontal-parietal (unicoronal) synostosis, and/or fusions along the coronal suture. Plagiocephaly is characterized by facial asymmetry in the frontal region, inclination of the head towards the affected side as well as deviation of the chin towards the non-affected side. We herein present the case of a five month old female infant with plagiocephaly, facial asymmetry with right side predominance, posterior and superior displacement of the eyebrow and supra-orbital ridge. The case was treated with ipsilateral frontal-orbital advancement, at the Regional Hospital «Lic. Adolfo Lopez Mateos», ISSSTE, Mexico City.

**Key words:** Plagiocephaly, frontal-orbital advancement, stereolithography.

**Palabras clave:** Plagiocefalia, avance frontoorbitario, estereolitografía.

#### RESUMEN

La plagiocefalia se define como el cierre prematuro de la sutura frontoparietal o coronal unilateral, la cual es causada por una sinostosis frontoparietal (unicoronal) y/o fusiones a lo largo de la sutura coronal, se caracteriza por asimetría facial en la región frontal, inclinación de la cabeza hacia el lado afectado y desviación del mentón hacia el lado no afectado. Se presenta el caso de un infante de género femenino de cinco meses de edad con plagiocefalia, asimetría facial de predominio derecho, el reborde supraorbitario y ceja están desplazados posterior y superior, tratado mediante avance frontoorbitario ipsilateral, llevado a cabo en el Hospital Regional «Lic. Adolfo López Mateos» ISSSTE.

#### INTRODUCTION

Approximately 200 years ago, Dr. Sommering reported the first scientific research on cranial deformities. Dr. Otto, in 1830 suggested that premature suture closure caused deformities. Based on that, Dr. Virchow published his research on deformities which stemmed from scientific studies. Drs. Apert and Cruzon described the syndromes later named after them. Drs Lane and Lannelongue reported the first modern correction of skull deformities which were the result of a premature closing of sutures. Frontal synostotic plagiocephaly is commonly caused by the premature closure of the unilateral frontal-parietal suture. Synostosis of other sutures along the coronal suture (sphenotmoidal, frontal-sphenoid) is the result of a frontal asymmetry, which is clinically difficult to differentiate from a frontal-parietal synostosis.<sup>1-3</sup>

Clinically, a retrusion of the frontal-orbital region is observed. From the radiographic and tomographic perspective, a typical harlequin-like orbit can be observed. Equally observed are deviations of the

*crista galli* apophyses, asymmetry in the frontal region, as well as calcification of the frontal-parietal suture. The aforementioned situations were tomographically confirmed with a 3-D reconstruction. It is important to differentiate a true craniosynostosis, requiring surgical treatment, from a positional deformity.<sup>4-6</sup>

Plagiocephaly represents 85% of all cranial deformity cases, it is found more in males than in females (2:1), and affects predominantly the right side (2.7:1 in males, 1:1 in females) when compared to the left side.<sup>7-10</sup>

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**CASE REPORT**

The case herein presented is that of a five month old female, born at full term, who had received a previous diagnosis of plagiocephaly (*Figure 1*) at 10 days of age. The patient exhibited hyperbilirubinemia, which had been assessed when she was 4 months old. With respect to contour and facial structures, plagiocephaly diagnosis was confirmed with the help of diagnosis adjuvants such as stereolithography and CT with 3D reconstruction (*Figures 2 to 4*). Physical examination revealed retrusion and asymmetry at the ipsilateral frontal-orbital region (*Figure 5*).



**Figure 1.** Extraoral view.

Parameters to analyze cranial discrepancy were based on studies on cranial cephalometry described by Dr. Jeffrey C. Posnick based on computerized tomography.

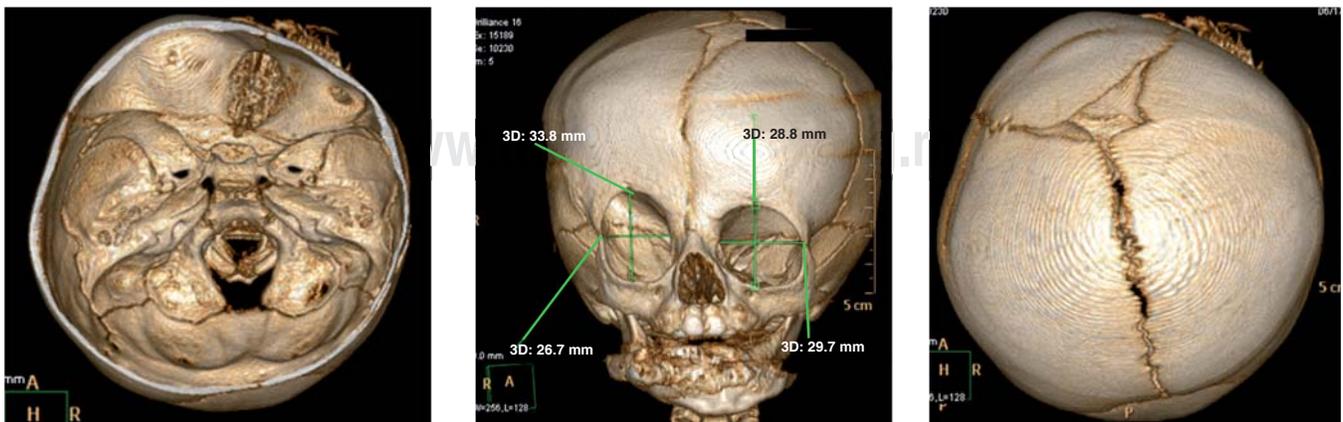
**Surgical technique**

A retro-auricular bi-coronal approach (SCALP) was performed. The anterior portion of the approach was raised along the temporal muscle in a sub-periosteal plane (*Figures 6 and 7*). A bilateral orbital circumferential dissection was then performed with respect to the lateral edge, preserving at all time the medial edge and the naso-lacrimal apparatus (*Figure 8*).

Subperiosteal dissection was continued under the lateral and infra-orbital portion of the orbital rim; it encompassed the superior and anterior portion of the zygoma and jaw (not the zygomatic arch). The SCALP posterior portion was dissected in the periosteal plane up to the middle portion located between coronal and lambdoidal sutures. The neurosurgeon performed a bi-frontal craniotomy. Following principles of craniofacial surgery, extradural dissection was performed in order to preserve the encephalon (*Figure 9*).

Osteotomy of three quarters of the orbit was completed through the floor of each of them, above the medial wall of each orbit, orbit lateral wall and in lateral aspects in the floor of the orbit, underneath the orbital fissure. Skull base osteotomy remains at an anterior position with respect to the respiratory bulb. Lateral aspects of each infra-orbital and lateral rim, upper and upper aspects of the medial orbital rim, include extension of the osteotomy which involves the canthal ligament.

In the present case, osteotomy was performed in a region located over the frontal-zygomatic suture



**Figures 2, 3 and 4.** CT with 3D reconstruction where orbit measures and cranial structures variations can be observed.



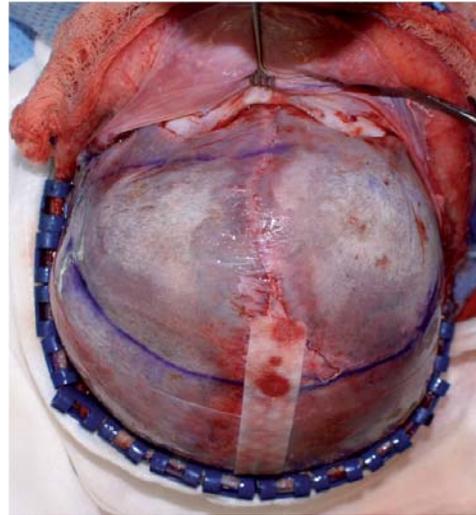
**Figure 5.** Asymmetry at frontal-orbital region.



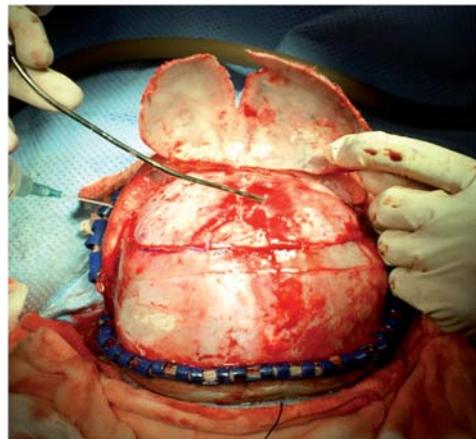
**Figure 6.** Surgical approach.



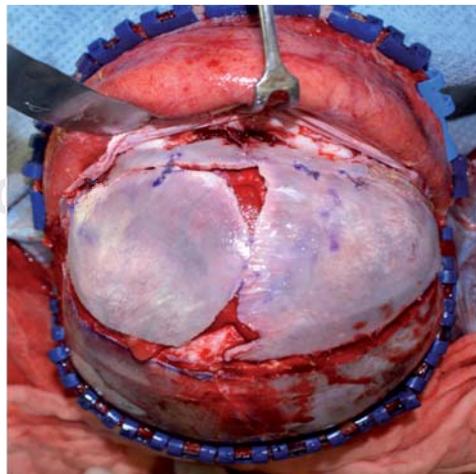
**Figure 7.** Sub-periosteal dissection.



**Figure 8.** Bilateral orbital circumferential dissection.



**Figure 9.** Extradural dissection.



**Figure 10.** Placement of bone fragments.



**Figures 11, 12 and 13.** Fixation with plate and resorbable screws.

in order to avoid fracture of the mask at that level, additionally achieving suitable preservation of canthal ligament in lateral and medial direction. Once the osteotomy was completed, the frontal bone fragments were placed, interchanging them in order to correct the deformity (*Figure 10*). In the mask, jagged fracture was performed to shape it and achieve symmetry in the patient. The fragment of the mask was fixated with plate and resorbable screws (*Figures 11 and 12*) and the rest of the segments were fixated with vicryl 3-0 (*Figure 13*). The flap was then re-positioned observing present asymmetry correction, after placing a drenovac, deep planes were sutured with polyglycolic acid and skin, using simple sutures of 3-0 nylon (*Figures 14 and 15*). The patient was then admitted at the pediatric intensive care unit to be closely monitored.

## DISCUSSION

Synostoses involve nearby structures of the coronal suture. Frontal synostotic plagiocephaly is the result of a fusion of the coronal suture components. Unilateral frontal-parietal synostosis is the most common synostosis, occurring in one out of every 10,000 live births.

Authors such as Dr. Bartlet, report that it is logical to think that neurological damage previous to the development of plagiocephaly is an important and predisposing factor for the later occurrence of a positional plagiocephaly, among other reasons due to the lesser mobility that these children might be afflicted with.

Several authors have proposed a common pathophysiological origin for both entities. According to Dr. Dias et al, mechanical forces that cause the primitive deformation can originate, when very persistent, pathological changes in the lambdoid sutures and skull base at the end of the process, transforming this into true craniosynostoses.

It must be borne in mind that factors which imply immobilization and compression of similar sutures are implicated in order to explain some forms of



**Figure 14.** Flap repositioning.

craniosynostoses, such as sagittal and methodic craniosynostoses.

The most severe cranial deformities are problems which affect brain function, ocular alterations or psychosocial disorders. Nevertheless, in recent years, there have been published studies stating that in plagiocephaly afflicted newborn there are delays (retardations) in the psychomotor or cognitive development, or affectation of auditory potentials. In 2000, Drs. Miller and Clarren<sup>11</sup> published a study where they compared long-term neurological development of newborn patients diagnosed with plagiocephaly. In it they found that children afflicted with plagiocephaly presented a significant increase of mild brain dysfunctions (39.7% of afflicted children), language and learning disorders, behavioral problems as well as attention deficit.

## CONCLUSIONS

For centuries, cranial anomalies have represented a great challenge, since sometimes, selecting the



**Figure 15.** Suture with simple stitches.



**Figure 17.** One week after surgery.



**Figure 16.** Pre-surgical image.

ideal treatment can be a subject of controversy. Some prefer performance of unilateral frontal craniotomy, others consider bi-frontal craniotomy the best option. The case herein presented supports the second trend, since results obtained have been excellent, satisfactory for the patient as well as the surgeon (Figures 16 and 17).

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