Correction of Maxillofacial Deformities in a Patient with Unilateral Coronal Craniosynostosis (Plagiocephaly): A Case Report and a Review of Literatures

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INTRODUCTION

Craniosynostosis is premature fusion of one or several sutures in the skull bone [1]. Premature fusion of one of the coronal sutures results in plagiocephaly (oblique skull) occurring in 1 in 10000 live births [2]. Frontal plagiocephaly is caused by deformational or synostotic forces [2-4].

The signs of unilateral coronal synostosis include a flat frontal bone, retardation of the supra and lateral orbital rim, elevation of the sphenoid wing producing a harlequin appearance on plain radiography or CT scan and deviation of the root of the nose toward the affected side. Compensative changes consist of bossing of the left frontal bone, inferior...
replacement of the supra and lateral orbital rim and deviation of the tip of the nose toward the left side [1,3,5,6]. In this article, we introduce a patient with non-deformational unilateral plagiocephaly.

CASE REPORT
A 10-month-old infant with right-sided plagiocephaly referred to a neurosurgeon. Examination revealed a healthy girl infant who was normal apart from the craniofacial deformity. The mother had a normal pregnancy and delivery and she did not smoke, drink or use any medication. Physical examination showed an oblique head. The right ipsilateral forehead and parietal was flat and retruded. The right temporal bone was depressed. The right supra orbital ridge and lateral orbital rim were depressed and displaced posteriorly. The nasal tip was slightly deviated to the left side and the root of the nose was constricted and deviated to the affected side. The left side revealed frontal and parietal bossing (Fig 1). Plain radiographs revealed a unilateral coronal synostosis (harlequin appearance) (Fig2). A CT scan of the craniofacial skeleton was performed in both axial and coronal planes, with the axial slices reformatted for 3D reconstruction.

The craniofacial asymmetry described clinically was also appreciated on radiographic examination. In addition, the right sphenoid wing was elevated superiorly (harlequin appearance) (Fig 3).

PROCEDURE
Based on the possible cause of plagiocephaly (closure of the right fronto sphenoidal suture), bifrontal craniotomy with right-sided advancement of a fronto-orbital complex was planned. After the bicornoral incision, the coronal flap was elevated anteriorly in the subperiosteal plane. Temporalis muscles were dissected and the flap was 180-degrees rotated. Periorbital dissection was followed by releasing the lateral canthi and careful maintenance of the integrity of the medial canthi and the naso-lacrimal apparatus. After bifrontal craniotomy with retraction of the frontal and temporal lobes by the neurosurgeon, the first osteotomy was performed approximately 1 cm above the supra orbital rim and extended toward the temporal bone and lateral wall of the orbit (Fig 4). After fronto-orbital advancement, the bone parts were fixed with titanium screw and miniplates by the maxillofacial surgeon (Fig 5). Osteotomy and the additional bone cuts in the frontotemporal region created enough space for expansion of the brain. The fragments of the bone were inserted between the globe and the brain. The right and left frontal bones were switched thus, creating a more normal frontal head shape (Fig 6). Lateral canthopexy was completed, then the coronal incision was closed in layers and suction drains were placed. Standard follow-up visits to the neurosurgeon, maxillofacial surgeon and ophthalmologist occurred at one week, 4 weeks, 8 weeks, 6 months and 2 years (Fig 7).

DISCUSSION
Synostotic frontal plagiocephaly is most commonly caused by fronto-parietal synostosis.
Fig 2. PA and lateral view of the skull

Fig 3(a). Coronal view of CT scan
Fig 3 (b). Axial view of CT scan

Fig 3 (c). Three dimensional reconstruction
But it may occur by premature fusion along the coronal hemiring [7,8]. Unilateral Coronal synostosis is the main synostotic cause of frontal plagiocephaly[2,5]. Frontal plagiocephaly is caused by synostotic or deformational forces [2,4]. Differentiation between them is possible based on physical examination and radiographic evaluation (3D CT scan and skull x-ray) [2,9]. The difference between deformational and synostotic frontal plagiocephaly is insignificant. Sometimes visible deformities are misleading and result in delay in treatment. Meanwhile, examination of all sutures is necessary for recognition of craniosynostosis and prevention of incorrect diagnosis [9]. Clinically in positional plagiocephaly, the skull has a rhomboid form and in synostotic plagiocephaly, it is trapezoidal [10].

Retrospective studies have shown that the basilar coronal ring is involved in one-third of the patients with unilateral coronal craniosynostosis [11,12]. Frontosphenoidal synostosis is a rare cause of frontal plagiocephaly [7]. Fracèle et al., 1955; Rogers et al., 2002; Dun-dulis et al., 2004; and Ribaupierre et al., 2007; have reported only 10 cases of plagiocephaly caused by an isolated stenosis of the frontosphenoidal suture [2,4,5,11]. Sometimes synostosis of the fronto-sphenoidal suture is misdiagnosed as coronal unilateral synostosis; therefore, an exact radiologic and clinical examination to recognize the abnormalities in the length of the coronal hemi-ring is necessary [7]. Main morphologic differences exist between the unilateral coronal craniosynostosis and fronto-sphenoidal craniosynostosis suture.
Fig 5(a). Osteotomy and advancement of the fronto-orbital segment

Fig 5(b). Rigid fixation of the fronto-orbital segment
In the first group, the limited expansion of the frontal and parietal bones is compensated by extra growth at the ipsilateral fronto-sphenoidal and spheno-ethmoidal sutures that lead to deviation of the nose to the contralateral side and an elevated position of the ipsilateral eye socket (Harlequin appearance). In the second group, growth is probably immediately restricted at the basilar coronal ring. This leads to ipsilateral deviation of the nose and a downward retracted position of the ipsilateral orbit [12]. Dundulis et al. (2004) stated in the first group of patients that had synostotic fronto-parietal suture and patent fronto-sphenoidal suture, the ipsilateral to contralateral vertical orbit dimension (1.11) was more than the second group patients in whom both sutures of the fronto-sphenoidal and fronto-parietal were synostotic (1.04). The horizontal orbit dimension did not show a significant difference between the two groups [11].

Another study reported that the dynamic behavior of the orbit in response to intracranial pressure (ICP) mentioned a significant difference between the two groups.

In a study conducted by Nagasao et al., the orbit displacement was higher in group 1 compared to group 2, because premature fusion of the fronto-sphenoidal suture disturbs orbit growth in response to ICP. In fronto-sphenoidal synostosis, quick releasing of the fusion at an early stage improves the appearance of the orbit [13].

Conventional cephalogram and CT scan do not have inherent accuracy for determining whether the minor skeletal sutures such as the spheno etmoidal and fronto-sphenoidal suture are open [12]. Therefore, multi-slice CT scan with 3-dimension reconstruction is necessary for the diagnosis of skull abnormality and prevention of delay in treatment [9,12,14].
Meanwhile, assessment of the minor and major sutures is advised to confirm or exclude craniosynostosis as a cause of skull deformity [12]. Although three dimensional CT scan is accepted as the diagnostic tool for craniosynostosis, it is not considered for long term follow-up due to the radiation dose and needs for anesthesia [14,15]. It is proved that 3D stereo photo grammetry is suitable for children as a non invasive method and devoid of ray for evaluating the growth pattern in long term [9,15]. According to different studies, it is recommended to release the fusion at an early stage of cranial growth (before one year) to improve the appearance of the orbital region [1-3,12,13]. In this case, we released a frontoparietal suture at 10-months of age.

Similar to the case presented, Ecklet et al. (2007), used elegant titanium plates instead of a resorbable plate because of the large step and major advancement between the fronto-orbital part and frontal bone and to determine constancy [13].

Existence of non resorbable plates does not cause any restriction for growth [17]. Other studies have reported coronal suturectomy through minimal incisions under endoscopy with less morbidity and mortality in patients with plagiocephaly due to coronal synostosis [18,19].

CONCLUSION
Synostotic frontal plagiocephaly is most commonly caused by fronto-parietal synostosis, but it can be caused by premature fusion of other sutures in the length of the coronal ring. Exact physical and radio graphic examination (multislice CT scan with 3D reconstruction) result in accurate diagnosis of skull abnormality and prevention of treatment delay.

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