

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

Mansour Khorasani<sup>1✉</sup>, Mohsen Hasani Barzi<sup>2</sup>, Bahman Derakhshan<sup>3</sup>

<sup>1</sup>Associated Professor of Oral and Maxillofacial Surgery Department, School of Dentistry, Qazvin University of Medical Sciences, Qazvin, Iran

<sup>2</sup>Assistant Professor of Neurosurgery Department, School of Medicine, Qazvin University of Medical Sciences, Qazvin, Iran

<sup>3</sup>Assistant Professor of Oral and Maxillofacial Surgery Department, School of Dentistry, Qazvin University of Medical Sciences, Qazvin, Iran

## Abstract

Plagiocephaly (oblique skull) is premature fusion of one of the coronal sutures. Frontal plagiocephaly is a rare congenital deformity in the skull that is the most complicated form of craniosynostosis to treat. Examination of all sutures is necessary for diagnosis of craniosynostosis

In this article, a 10-month-old, healthy girl with deformity of the right forehead and orbit that is caused by frontal plagiocephaly and coronal unilateral synostosis is presented.

This abnormality was corrected by frontal craniotomy and fronto-orbital complex advancement under general anesthesia.

An exact clinical and radiographic (Multislice CT scan with 3- dimensional three dimensional reconstruction) examination with the suitable time of surgery is necessary for diagnosis of skull abnormality and prevention of treatment delay. Furthermore, 3D stereo-photo-grammetry is a radiation-free, non invasive method for evaluating the growth pattern of children in long term.

**Key Words:** Plagiocephaly; Craniotomy; Craniosynostoses; X-Ray Computed Tomography

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✉ Corresponding author:  
V. Khorasani, Department of Oral and Maxillofacial Surgery, School of Dentistry, Qazvin University of Medical Sciences, Qazvin, Iran

vkhorasani1342@yahoo.com

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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



**Fig 1.** Patient's Photograph

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

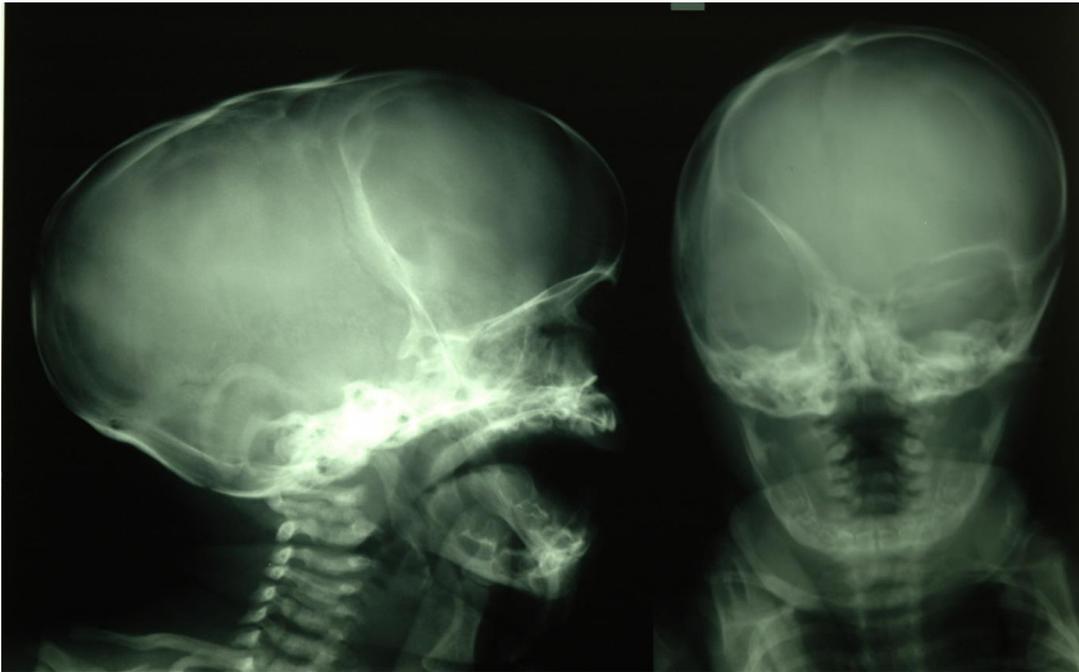
Based on the possible cause of plagiocephaly (closure of the right fronto sphenoidal suture), bifrontal craniotomy with right-sided advancement of a fronto-orbital complex was planned.

After the bicoronal 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 nasolacrimal 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).

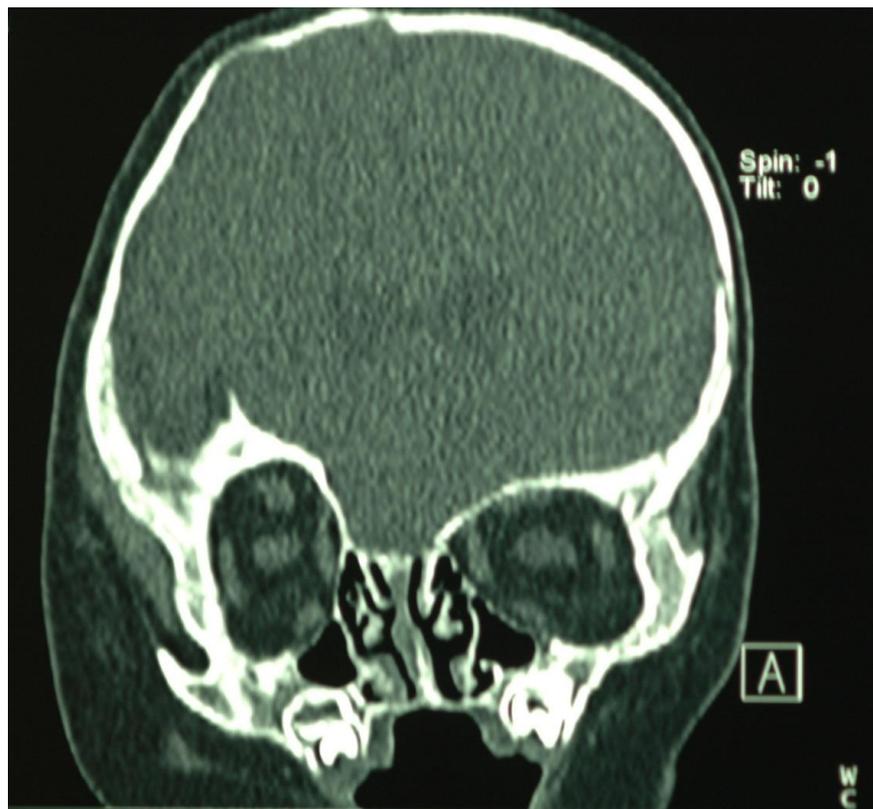
Osteotomy and the additional bone cuts in the frontotemporal region created enough space for expansion of the brain. The fragments of the bone were inserted between the globe and the brain. The right and left frontal bones were switched thus, creating a more normal frontal head shape (Fig 6). Lateral 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



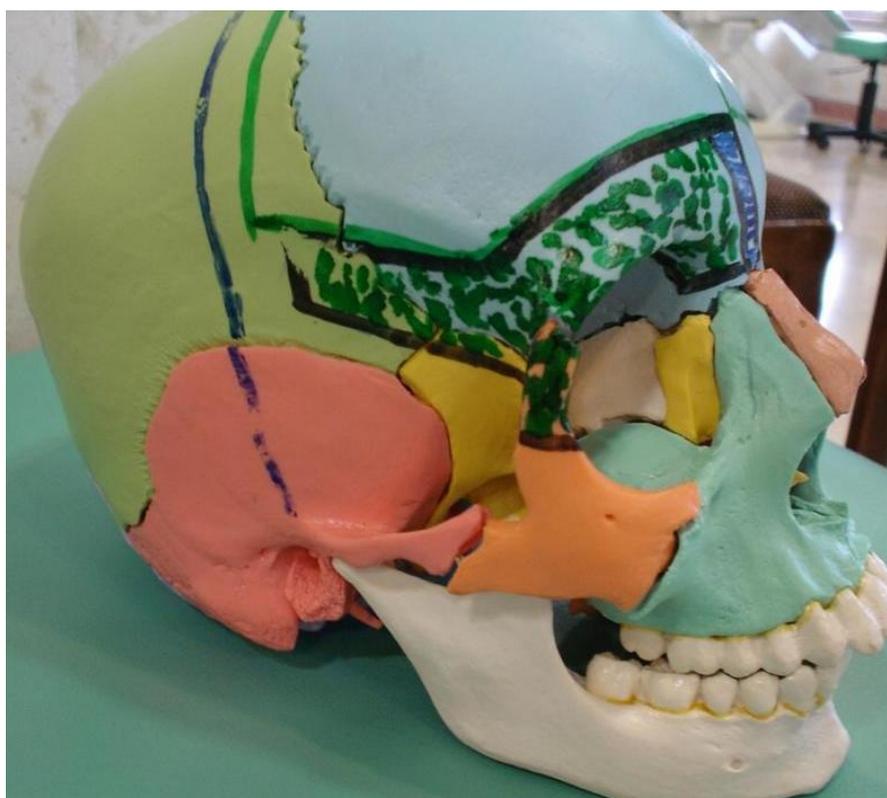
**Fig 3(a).** Coronal view of CT scan



Fig 3 (b). Axial view of CT scan



Fig 3 (c). Three dimensional reconstruction



**Fig 4.** Plan of osteotomy on the model

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].

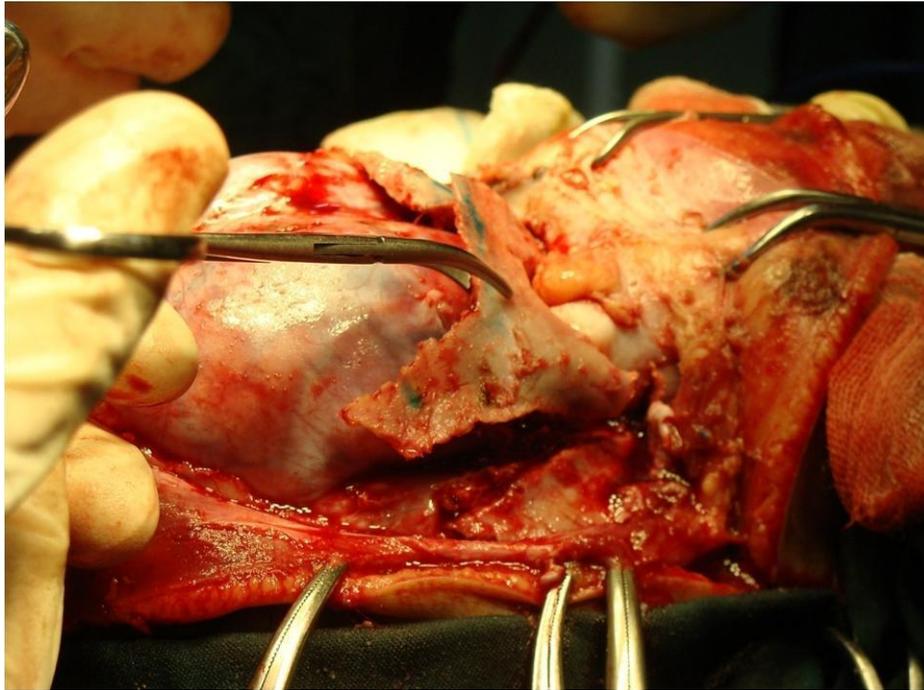
Differentiation between them is possible based on physical examination and radiographic evaluation (3D CT scan and skull x-ray) [2,9].

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].

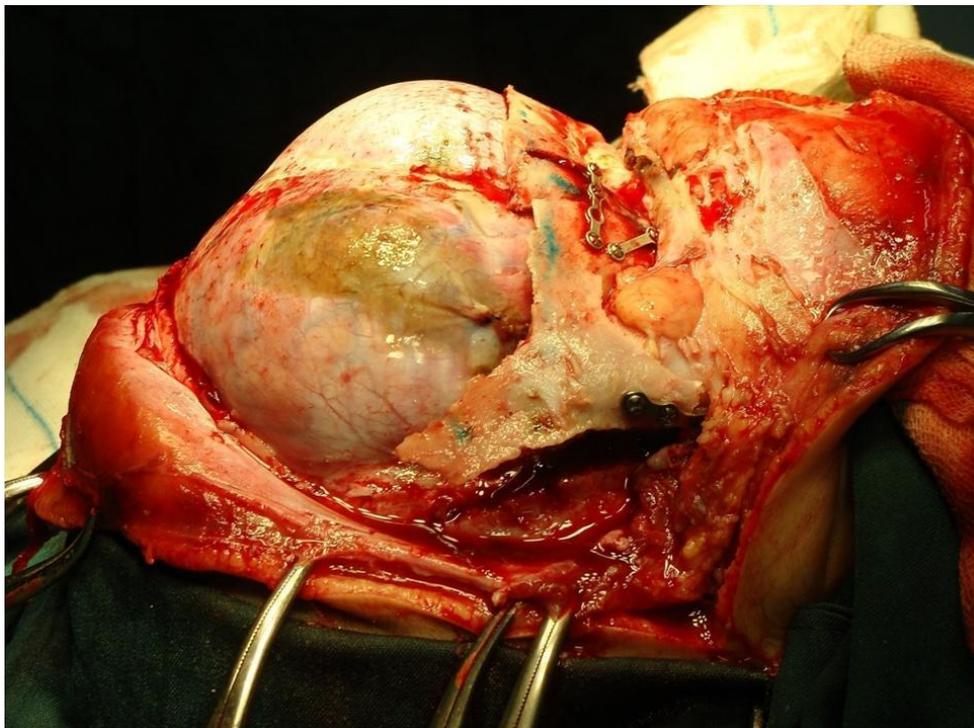
Clinically in positional plagiocephaly, the skull has a rhomboid form and in synostotic plagiocephaly, it is trapezoidal [10].

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].

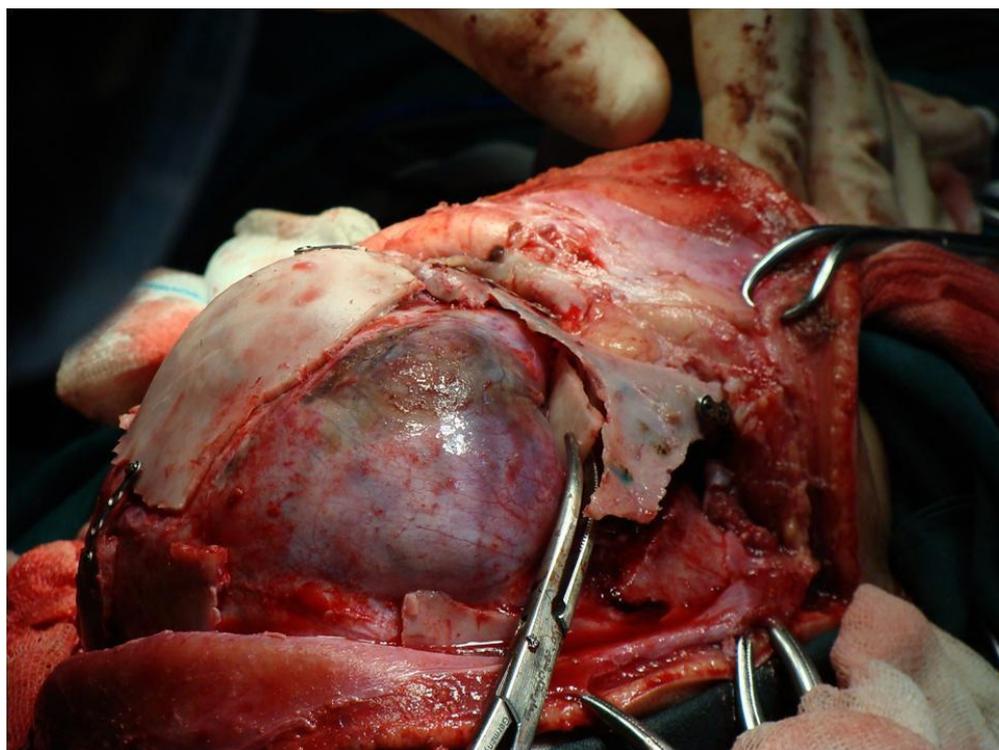
Francle et al., 1955; Rogers et al., 2002; Dundulis et al., 2004; and Ribaupierre et al., 2007; have reported only 10 cases of plagiocephaly caused by an isolated stenosis of the fronto-sphenoidal suture [2,4,5,11]. Sometimes synostosis of the fronto-sphenoidal suture is misdiagnosed as coronal unilateral 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



**Fig 6.** Insertion of bone between the globe and the brain and switching of the right and left frontal bone

In the first group, the limited expansion of the frontal and parietal bones is compensated by extra growth at the ipsilateral fronto-sphenoidal and speno-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 speno ethmoidal 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].



Fig 7. Patient's photograph two years after surgery

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.

## REFERENCES

- 1- Bell WH. Modern practice in orthognatic and reconstructive surgery. 1st ed. Dallas: WB Saunders Co; 1992. p. 1845-54.
- 2- Francel PC, Park TS, Marsh JL, Kaufman BA. Frontal plagiocephly secondary to synop-sis of the frontosphenoideal suture. Case report. J Neurosurg. 1995 Oct;83(4):733-6.
- 3- Bruneteau RJ, Mulliken JB. Frontal plagiocephaly: synostotic, compensational, or deformational. Plast Reconstr Surg. 1992 Jan;89(1):21-31.
- 4- de Ribaupierre S, Czorny A, Pittet B, Jacques B, Rilliet B. Frontosphenoideal synostosis: a rare cause of unilateral anterior plagiocephaly. Childs Nerv Syst. 2007 Dec;23(12):1431-8.
- 5- Rogers GF, Proctor MR, Mulliken JB. Unilateral fusion of the frontosphenoideal suture: a rare cause of synostotic frontal plagiocephaly. Plast Reconstr Surg. 2002 Sep;110(4):1011-15.
- 6- Schmidek A, Sweet WH. Operative neurosurgical techniques, indications, methods and results. Philadelphia, 4th ed. WB Saunders Co; 2000. p. 126-8.

- 7- Marucci DD, Jones BM, Dunaway DJ, Hayward RD. Unilateral isolated fronto-sphenoidal craniosynostosis causing frontal plagiocephaly. *J Plast Reconstr Aesthet Surg.* 2009 Aug;62(8):e255-8.
- 8- Hansen M, Mulliken JB. Frontal plagiocephaly: Diagnosis and treatment. *Clin Plast Surg.* 1994 Oct;21(4):543-53.
- 9- Plooiij JM, Verhammey Y, Berge SJ, van Lindert EJ, Borstlap-Engels VM, Borstlap WA. Unilateral craniosynostosis of the fronto-sphenoidal suture: a case report and a review of literature. *J Craniomaxillofac Surg.* 2009 Apr;37(3):162-6.
- 10- Zoller JE, Kubler AC, Lorber WD, Muhling JFH. Kraniozynostosen. In: Zoller JE, Kubler AC (eds). *Kraniofaziale chirurgie.* Stuttgart: Thieme; 2002 (Chapter 2).
- 11- Dundulis JA, Becker DB, Govier DP, Marsh JL, Kane AA. Coronal ring involvement in patients treated for unilateral coronal craniosynostosis. *Plast Reconstr Surg.* 2004 Dec;114(7):1695-703.
- 12- Rogers GF, Mulliken JB. Involvement of the basilar coronal ring in unilateral coronal synostosis. *Plast Reconstr Surg.* 2005 Jun;115(7):1887-93.
- 13- Nagasao T, Miyamoto J, Uchikawa Y, Tamaki T, Yamada A, Kaneko T et al. A biomechanical study on the effect of premature fusion of the frontosphenoidal suture on orbit asymmetry in unilateral coronal synostosis. *Cleft Palate Craniofac J.* 2010 Jan;47(1):82-91.
- 14- Mathijssen IM, van der Meulen JJ, van Adrichem LN, Vannrager JM, van der Hulst RR, Lequin MH et al. The frontosphenoidal suture: fetal development and phenotype of its synostosis. *Pediatr Radiol.* 2008 Apr;38(4):431-7.
- 15- Littlefield TR, Cherney JC, Luiss JN, Beals SP, Kelly KM, Pomatto JK. Comparison of plaster casting with three-dimensional cranial imaging. *Cleft Palate Craniofac J.* 2005 Mar;42(2):157-64.
- 16- Eckelt U, Nitsche M, Muller A, Pilling E, Pinzer T, Roesner D. Ultrasound aided pin flaxation of biodegradable osteosynthetic materials in cranioplasty for infants with craniosynostosis. *J Craniomaxillofac Surg.* 2007 Jun-Jul;35(4-5):218-21.
- 17- Berryhill WE, Rimell FL, Ness J, Marentette L, Haines SJ. Fate of rigid fixation in pediatric craniofacial surgery. *Otolaryngol Head Neck Surg.* 1999 Sep;121(3):269-73.
- 18- Stelnicki E, Heger I, Brooks CJ, Ghersi MM, Stubbs CB, Bahuleyan B et al. Endoscopic release of unicoronal craniosynostosis. *J Craniofac Surg.* 2009 Jan;20(1):93-7.
- 19- Tellado MG, Lema A. Coronal suturectomy through minimal incisions and distractosteogenesis are enough without other craniotomies for the treatment of plagiocephaly due to coronal synostosis. *J Craniofac Surg.* 2009 Nov;20(6):1975-7.