Pediatric Issue

Minimally Invasive Suturectomy and Postoperative Helmet Therapy : Advantages and Limitations

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Various operative techniques are available for the treatment of craniosynostosis. The patient's age at presentation is one of the most important factors in the determination of the surgical modality. Minimally invasive suturectomy and postoperative helmet therapy may be performed for relatively young infants, whose age is younger than 6 months. It relies upon the potential for rapid brain growth in this age group. Its minimal invasiveness is also advantageous. In this article, we review the advantages and limitations of minimally invasive suturectomy followed by helmet therapy for the treatment of craniosynostosis.

Key Words : Craniosynostosis · Suturectomy · Helmet therapy.

INTRODUCTION

Most infants with craniosynostosis require surgery for its correction. For the past few decades, various surgical methods have been proposed for the treatment of craniosynostosis, from open strip craniectomy to extensive cranial vault remodeling[3,16,31,35,38]. New techniques have also been applied to the surgical treatment of craniosynostosis such as distraction devices, absorbable plates and endoscopes[2,4,6,21,36].

Open strip craniectomy was first performed in 1890 for the treatment of microcephaly[22], and it has been used for the treatment of craniosynostosis since 1927[8]. Then, it was widely used to release fused sutures and to correct head contour in craniosynostosis. However, advancements in open strip craniectomy have led to arguments against its use due to dissatisfying and inconsistent esthetic outcomes. The late timing of surgery, the incomplete release of the involved sutures and an insufficient maintenance of this release are considered to be the causes of these problems[35].

With the development of intraoperative management and anesthesia in pediatric patients, more extensive surgery such as extensive cranial vault remodeling became possible. Extensive cranial vault remodeling methods have enabled effective cranial volume expansion and cephalic index (CI) correction via the removal and division of the skull bones into many parts to reshape bony contours, followed by rigid fixation[11]. These approaches became popular conventional methods for treating craniosynostosis, replacing open strip suturectomy[25,26]. However, extensive cranial vault remodeling has also raised concerns among many surgeons due to the potential for blood loss and the required operation time, even though efforts to reduce the need for blood transfusion have been made[3,7,9,10,17,20]. Aggravation of the deformation while patients age sufficiently to endure this extensive operation is another problem[14,29]. The necessity of stable fixation of the skull also restricts the use of extensive cranial remodeling to relatively early ages.

These concerns related to extensive cranial vault remodeling have raised a need for surgical methods with minimal invasiveness. Minimally invasive suturectomy is one method that began to appear in tandem with this concept. This method reappeared with modifications of the previous open strip suturectomy in 1998, from Jimenez and Barone[15]. They reported their experiences of endoscopic suturectomy for sagittal craniosynostosis. Using an endoscope during dissection made this procedure minimally invasive, and the use of an orthotic helmet applied after the operation assisted in reshaping the head contour. Suturectomy itself is not different from open strip craniectomy, but its minimal invasiveness enabled the operation to be used with early infants. Comparable esthetic outcomes were also reported in the literature, and this technique was acceptable to many sur-

geons[12,14,15,27,34,40]. Smaller incisions, less blood loss and shorter operative times and lengths of hospital stay were achieved with minimally invasive suturectomy[13,19,31,34,40]. Minimally invasive suturectomy has been performed since 2011 at our institution. In our experience, the magnification and illumination of the endoscope can be substituted with a loupe magnifier and a suction tip with a fiber optic cable and light source to visualize the operative field. Therefore, we will use the term 'minimally invasive suturectomy' and 'endoscopic-assisted suturectomy' interchangeably, although two terms do not mean precisely the same thing.

PATIENT SELECTION AND PREPARATION FOR THE TREATMENT

Patient age is the most important factor in deciding whether a surgical candidate is appropriate for minimally invasive suturectomy. Because taking advantage of a period of rapid brain growth phase as much as possible is the main point in this procedure, patients under the age of 6 months are good surgical candidates, and ages closer to 3 months of age are more ideal. Patients between 6 to 9 months of age may also be considered as surgical candidates for minimally invasive suturectomy if the deformity is minimal[16].

The diagnosis of craniosynostosis is made by clinical examination of the deformed head shape and confirmed by plain-skull radiographs and craniofacial computed tomography (CT) scans with 3-dimensional (3D) reconstruction. Gross photographs and anthropometric data are required to obtain preoperative morphologic information and to determine trends for postoperative

correction.

SURGICAL TECHNIQUES

Surgical techniques for minimally invasive suturectomy in previous articles are all similar in that they were all aim to release a fused suture with small exposure[12,15,27,31]. Procedures in our center are both similar and different from previous articles introduced as follows.

In sagittal craniosynostosis, the patient is prepared with the head extended in a prone position. Skin preparation is done with povidone-iodine. Two transverse incisions are made of 3−4 cm length at 1 cm behind the anterior fontanelle and 1 cm in front of the lambdoid suture. An additional incision may be needed between the two sites to manipulate safely in a patient with a longer head. A subperiosteal dissection is made along the desired craniectomy site. Burr holes are placed over the fused suture at both incision sites. The dura is dissected and carefully detached from the fused bone. During these procedures, a fiber optic suction tip or endoscope is used for the safe and accurate manipulation of the compromising space. Strip craniectomy is performed using curved Mayo scissors, sternal scissors and straight rongeurs. The fused bone is removed from the anterior fontanelle anteriorly to the lambda posteriorly. The width of the craniectomy site is targeted to be between 3 cm to 4 cm. After the strip craniectomy, additional lateral wedge osteotomies or barrel stave osteotomies might be conducted according to the surgeon's preference. Bleeding from the diploic space is controlled by bone wax and monopolar electrocautery. With the in-
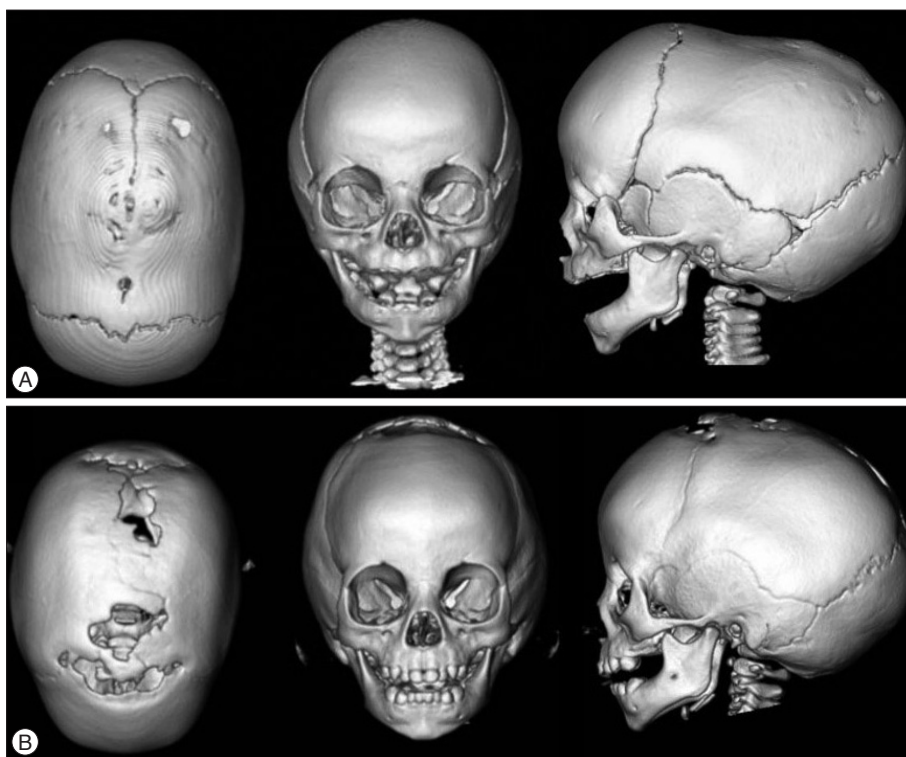

Fig. 1. Preoperative (A) and 1-year postoperative (B) 3D reconstructed CT views of a patient with sagittal craniosynostosis treated with minimally invasive suturectomy and postoperatihelmet therapy (left : superior view, middle : anterior view, right : lateral view). Cephalic index is 68% preoperatively and is improved to 75% 1-year after the operation.

sertion of a drain, the wound is closed layer by layer (Fig. 1).

For the treatment of anterior plagiocephaly caused by unilateral coronal craniosynostosis, the patient is positioned supine with the head rotated to the contralateral side. A single incision on the stephanion is used by several surgeons, but we use two incisions[13,37]. Two incisions, approximately 2 cm in length, are made at both ends of the fused coronal suture, just lateral to the anterior fontanelle and over the pterion. We prefer two incisions rather than a single incision for the feasibility of manipulation, the direct visualization of the fused ends and to accomplish complete release of the suture. A strip craniectomy is performed 1 cm in width subperiosteally. The craniectomy should run from the anterior fontanelle to the squamosal suture. An additional anterior directed wedge osteotomy might also be performed in cases of accompanied frontosphenoidal craniosynostosis at the level of the squamosal suture (Fig. 2).

In infants with unilateral lambdoid craniosynostosis presenting as posterior plagiocephaly, the operation is performed with the patient in the prone position. As mentioned for the anterior plagiocephaly, one incision on the fused suture is possible, but we use two incisions, one just lateral to lambda and the mastoid

fontanelle along the fused bone. Dissection is conducted in a similar pattern, and the width of subperiosteal craniectomy is also 1 cm (Fig. 3).

POSTOPERATIVE HELMET THERAPY

After the subgaleal swelling is absorbed and all stitches are removed, a cranial helmet is prescribed to redirect balanced growth of the head. The helmet is fitted by orthotists. We recommend wearing it until patients are 12 to 18 months old, during which period rapid brain growth occurs. The helmet allows for three dimensional growth and individual adjustments to its course of correction[30]. As the patient's head grows, one or two additional orthoses may be required until sufficient correction is achieved. In our experiences, one helmet is sufficient for most cases of sagittal craniosynostosis, and two helmets might be required in cases with plagiocephaly similar to previous reports[14,31].

TREATMENT OUTCOMES

In sagittal craniosynostosis, correction of the scaphocephalic shape can be measured by changes in CI postoperatively. Jimenez et al.[19] reported that 87% of sagittal craniosynostosis patients who were treated with minimally invasive suturectomy and postoperative helmet therapy showed excellent results (CI >75%),
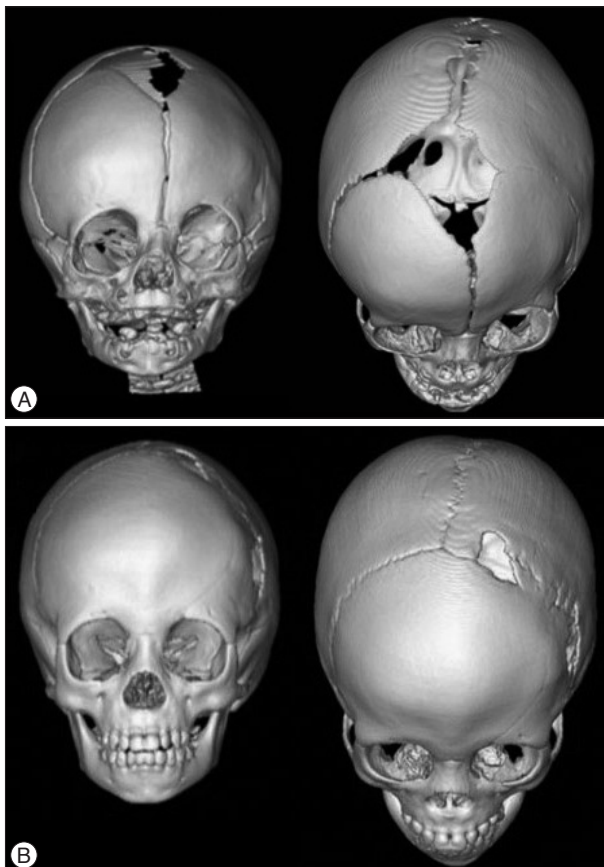

Fig. 2. Preoperative (A) and postoperative follow up 3D reconstructed CT images at 2 year (B) after the minimally invasive suturectomy in a patient with left coronal craniosynostosis (left : anterior view, right : superoanterior view). Note that supraorbital asymmetry is improved in 2-year follow up images compared with the preoperative image.
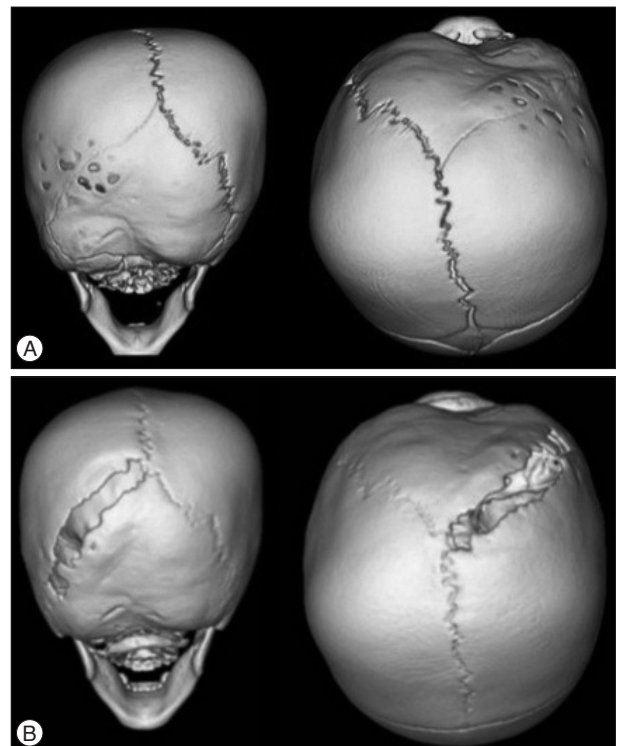

Fig. 3. Preoperative (A) and postoperative 1-year follow up (B) 3D reconstructed CT images of a patient with left lambdoid craniosynostosis (left : posterior view, right : superoposterior view). Deformation at contralateral parietal bone and cranial base is improved at the postoperative 1-year follow up images.

and 8.7% of patients showed good results (CI 70−75%). Similar supporting data have been presented by other groups, and comparisons of the CI between the minimally invasive suturectomy with helmet group and the extensive cranial vault remodeling group in a single center revealed equivalent outcomes[12,34]. These results support the promise of minimally invasive suturectomy and postoperative helmet therapy and suggest it to be different from open strip craniectomy, which showed inferior outcomes compared with extensive cranial vault remodeling[28]. The feasibility of the operation in the early ages due to its minimal invasiveness might be important in this difference because manipulating or releasing the fused suture is important and similar in both open strip craniectomy and minimally invasive suturectomy. Correction of the CI occurs mostly within 2 months of the operation and improves until 6 months[1,19]. After that period, correction begins to decrease and reaches a plateau[1]. Suppressing relapse of the scaphocephaly is one of the expected roles of the postoperative helmet[16,33]. However, we do not suggest that postoperative helmet therapy should be applied in all cases; improvements in CI could be achieved just by suturectomy, and there are few issues of asymmetry of laterality in sagittal craniosynostosis. More studies are needed to identify the effects of the postoperative helmet in sagittal craniosynostosis patients treated by minimally invasive suturectomy.

In view of its safety and minimal invasiveness, the outcomes of minimally invasive suturectomy are remarkable. Transfusion rates in minimally invasive suturectomy during the perioperative period range from 3.3% to 25%, while almost patients need transfusion in extensive cranial vault remodeling[13,19,27,31,40]. The operating time (45−100 minutes) is also shorter than in extensive cranial vault remodeling (4−8 hours)[4,19,31,34]. These advantages make the patient less prone to morbidity and require shorter periods of hospital stay.

Treatment of unilateral coronal craniosynostosis is somewhat more complex than sagittal craniosynostosis because asymmetries between the ipsilateral and contralateral side should be considered. Frontal, supraorbital and orbital asymmetries and nasal deviations are well known deformations related to unilateral coronal craniosynostosis. Fronto-orbital advancement is one of the extensive cranial vault remodeling methods for the treatment of unilateral coronal craniosynostosis, which can expand cranial volume and correct asymmetries. However, fronto-orbital advancement often does not modify facial or ocular asymmetries sufficiently modify facial or ocular asymmetries, which result in ophthalmologic problems such as strabismus, ocular torticollis and astigmatism[13,14,23]. These are problems with the age of the patient rather than the surgical methods because incidences are lowered when fronto-orbital advancement is performed at earlier ages[5,24]. Uncorrected facial asymmetries may persist or even worsen in some cases. Jimenez and Barone[13] reported that craniofacial scoliosis was corrected completely in 14% of unilateral coronal craniosynostosis patients and partially in 66%. Improvement of vertical dystopia was also achieved
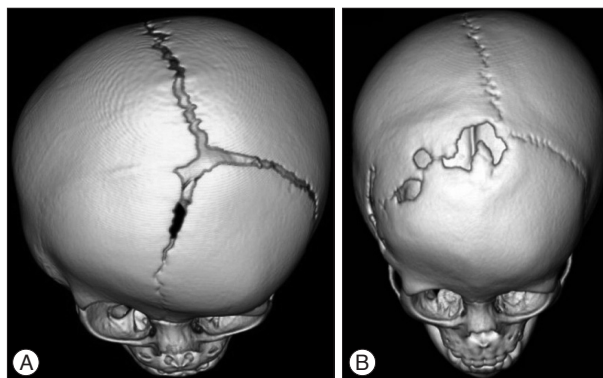

Fig. 4. Persistent supraorbital asymmetry of patients with right coronal craniosynostosis. Compared to preoperative (A) images, asymmetry is improved but still present at postoperative 1-year follow up (B).

completely in 14% and partially in 86% of patients. In comparison with fronto-orbital advancement, minimally invasive suturectomy and helmet therapy showed better outcomes for middle and lower facial asymmetries on comparison[37]. Contrary to the direct correction of forehead and supraorbital asymmetries in fronto-orbital advancement, changes in minimally invasive suturectomy and postoperative helmet therapy require time, and its final outcome is suboptimal in some cases (Fig. 4)[13]. However, its improvements in plagiocephaly may be persistent even following reossification of the suturectomy site, and further correction was noted when the frontal sinuses developed[14].

Unilateral lambdoid craniosynostosis has the feature of posterior plagiocephaly and compensatory contralateral parietal bossing. Compensatory and deformational growth also occurs at the cranial base level, which results in asymmetric external acoustic meatus and mastoid bulging. Extensive cranial vault remodeling for unilateral lambdoid craniosynostosis corrects the cranial vault shape but does not change asymmetries occurring in the cranial base directly. In one study of posterior cranial vault and base asymmetries after open and endoscopic operations, persistent cranial asymmetries were observed with both extensive cranial vault remodeling and minimally invasive suturectomy, and the treatment outcomes were equivocal[40]. Posterior asymmetries are more acceptable than anterior asymmetries because they can be concealed by hair. The simplicity of the procedure and the theoretical possibility of correction in the cranial base induced by rapid brain growth during the early infant period should make surgeons consider minimally invasive suturectomy favorably.

Multiple-suture craniosynostosis has many problems, including increased intracranial pressure, abnormal head shape depending on involved sutures, compensatory growth and asymmetric ophthalmologic findings. It is more complex than single suture craniosynostosis and differs case by case. Surgical strategies are also different depending upon the major problem. There are few studies of early minimally invasive suturectomy for multiple suture craniosynostosis, but good results have been presented in limited cases[18,32]. In cases with increased intracranial

pressure, distractor osteogenesis which is another minimal invasive technique would be more suitable for the sufficient volume expansion[39].

LIMITATIONS

The underlying concepts in minimally invasive suturectomy and postoperative helmet therapy are releasing the fused suture in the early period, before severe deformity occurs, and utilizing the potential for normal rapid brain growth during early infancy to reshape it. Therefore, this technique has age limitations, and it is unlikely there will be good outcomes in late infancy, when the driving force is much diminished. Patients over 9 months of age are not suitable for this technique, and extensive cranial vault remodeling should be considered[16].

Another limitation of this treatment is the necessity of wearing a helmet for up to 1 year. During that period, regular examinations and adjustments of the helmet are required. Compliance with helmet use can affect treatment outcomes.

CONCLUSION

Minimally invasive suturectomy and postoperative helmet therapy are safe methods to treat craniosynostosis in early infancy, and their esthetic results are comparable to conventional extensive cranial vault remodeling.

References

1. Agrawal D, Steinbok P, Cochrane DD : Long-term anthropometric outcomes following surgery for isolated sagittal craniosynostosis. **J Neurosurg 105 (5 Suppl)** : 357-360, 2006
2. Akai T, Iizuka H, Kawakami S : Treatment of craniosynostosis by distraction osteogenesis. **Pediatr Neurosurg 42** : 288-292, 2006
3. Boop FA, Chadduck WM, Shewmake K, Teo C : Outcome analysis of 85 patients undergoing the pi procedure for correction of sagittal synostosis. **J Neurosurg 85** : 50-55, 1996
4. Clayman MA, Murad GJ, Steele MH, Seagle MB, Pincus DW : History of craniosynostosis surgery and the evolution of minimally invasive endoscopic techniques : the University of Florida experience. **Ann Plast Surg 58** : 285-287, 2007
5. Denis D, Genitori L, Bolufer A, Lena G, Saracco JB, Choux M : Refractive error and ocular motility in plagiocephaly. **Childs Nerv Syst 10** : 210-216, 1994
6. Eppley BL, Sadove AM, Havlik RJ : Resorbable plate fixation in pediatric craniofacial surgery. **Plast Reconstr Surg 100** : 1-7; discussion 8-13, 1997
7. Esparza J, Hinojosa J : Complications in the surgical treatment of craniosynostosis and craniofacial syndromes : apropos of 306 transcranial procedures. **Childs Nerv Syst 24** : 1421-1430, 2008
8. Faber HK, Towne EB : Early craniectomy as a preventive measure in oxycephaly and allied conditions. **Am J Med Sci 173** : 701-711, 1927
9. Faberowski LW, Black S, Mickle JP : Blood loss and transfusion practice in the perioperative management of craniosynostosis repair. **J Neurosurg Anesthesiol 11** : 167-172, 1999
10. Fearon JA, Weinthal J : The use of recombinant erythropoietin in the reduction of blood transfusion rates in craniosynostosis repair in infants and children. **Plast Reconstr Surg 109** : 2190-2196, 2002
11. Heller JB, Heller MM, Knoll B, Gabbay JS, Duncan C, Persing JA : Intracranial volume and cephalic index outcomes for total calvarial reconstruction among nonsyndromic sagittal synostosis patients. **Plast Reconstr Surg 121** : 187-195, 2008
12. Hinojosa J, Esparza J, Muñoz MJ : Endoscopic-assisted osteotomies for the treatment of craniosynostosis. **Childs Nerv Syst 23** : 1421-1430, 2007
13. Jimenez DF, Barone CM : Early treatment of anterior calvarial craniosynostosis using endoscopic-assisted minimally invasive techniques. **Childs Nerv Syst 23** : 1411-1419, 2007
14. Jimenez DF, Barone CM : Early treatment of coronal synostosis with endoscopy-assisted craniectomy and postoperative cranial orthosis therapy : 16-year experience. **J Neurosurg Pediatr 12** : 207-219, 2013
15. Jimenez DF, Barone CM : Endoscopic craniectomy for early surgical correction of sagittal craniosynostosis. **J Neurosurg 88** : 77-81, 1998
16. Jimenez DF, Barone CM : Endoscopic technique for sagittal synostosis. **Childs Nerv Syst 28** : 1333-1339, 2012
17. Jimenez DF, Barone CM : Intraoperative autologous blood transfusion in the surgical correction of craniosynostosis. **Neurosurgery 37** : 1075-1079, 1995
18. Jimenez DF, Barone CM : Multiple-suture nonsyndromic craniosynostosis : early and effective management using endoscopic techniques. **J Neurosurg Pediatr 5** : 223-231, 2010
19. Jimenez DF, Barone CM, McGee ME, Cartwright CC, Baker CL : Endoscopy-assisted wide-vertex craniectomy, barrel stave osteotomies, and postoperative helmet molding therapy in the management of sagittal suture craniosynostosis. **J Neurosurg 100 (5 Suppl Pediatrics)** : 407-417, 2004
20. Kearney RA, Rosales JK, Howes WJ : Craniosynostosis : an assessment of blood loss and transfusion practices. **Can J Anaesth 36** : 473-477, 1989
21. Kumar AV, Staffenberg DA, Petronio JA, Wood RJ : Bioabsorbable plates and screws in pediatric craniofacial surgery : a review of 22 cases. **J Craniofac Surg 8** : 97-99, 1997
22. Lannelongue M : De la craniectomie dans la microcéphalie. **Compt Rend Seances Acad Sci 50** : 1382-1385, 1890
23. Macintosh C, Wall S, Leach C : Strabismus in unicoronal synostosis : ipsilateral or contralateral? **J Craniofac Surg 18** : 465-469, 2007
24. MacKinnon S, Rogers GF, Gregas M, Proctor MR, Mulliken JB, Dagi LR : Treatment of unilateral coronal synostosis by endoscopic strip craniectomy or fronto-orbital advancement : ophthalmologic findings. **J AAPOS 13** : 155-160, 2009
25. Marsh JL, Jenny A, Galic M, Picker S, Vannier MW : Surgical management of sagittal synostosis. A quantitative evaluation of two techniques. **Neurosurg Clin N Am 2** : 629-640, 1991
26. Maugans TA, McComb JG, Levy ML : Surgical management of sagittal synostosis : a comparative analysis of strip craniectomy and calvarial vault remodeling. **Pediatr Neurosurg 27** : 137-148, 1997
27. Murad GJ, Clayman M, Seagle MB, White S, Perkins LA, Pincus DW : Endoscopic-assisted repair of craniosynostosis. **Neurosurg Focus 19** : E6, 2005
28. Panchal J, Marsh JL, Park TS, Kaufman B, Pilgram T, Huang SH : Sagittal craniosynostosis outcome assessment for two methods and timings of intervention. **Plast Reconstr Surg 103** : 1574-1584, 1999
29. Persing J, Babler W, Winn HR, Jane J, Rodeheaver G : Age as a critical factor in the success of surgical correction of craniosynostosis. **J Neurosurg 54** : 601-606, 1981
30. Proctor MR : Endoscopic craniosynostosis repair. **Transl Pediatr 3** : 247-258, 2014

31. Ridgway EB, Berry-Candelario J, Grondin RT, Rogers GF, Proctor MR : The management of sagittal synostosis using endoscopic suturectomy and postoperative helmet therapy. **J Neurosurg Pediatr 7** : 620-626, 2011

32. Rivero-Garvía M, Marquez-Rivas J, Rueda-Torres AB, Ollero-Ortiz A : Early endoscopy-assisted treatment of multiple-suture craniosynostosis. **Childs Nerv Syst 28** : 427-431, 2012

33. Seymour-Dempsey K, Baumgartner JE, Teichgraeber JF, Xia JJ, Waller AL, Gateno J : Molding helmet therapy in the management of sagittal synostosis. **J Craniofac Surg 13** : 631-635, 2002

34. Shah MN, Kane AA, Petersen JD, Woo AS, Naidoo SD, Smyth MD : Endoscopically assisted versus open repair of sagittal craniosynostosis : the St. Louis Children's Hospital experience. **J Neurosurg Pediatr 8** : 165-170, 2011

35. Shillito J Jr, Matson DD : Craniosynostosis : a review of 519 surgical patients. **Pediatrics 41** : 829-853, 1968

36. Sugawara Y, Hirabayashi S, Sakurai A, Harii K : Gradual cranial vault expansion for the treatment of craniofacial synostosis : a preliminary report. **Ann Plast Surg 40** : 554-565, 1998

37. Tan SP, Proctor MR, Mulliken JB, Rogers GF : Early frontofacial symmetry after correction of unilateral coronal synostosis : frontoorbital advancement vs endoscopic strip craniectomy and helmet therapy. **J Craniofac Surg 24** : 1190-1194, 2013

38. Whitaker LA, Bartlett SP, Schut L, Bruce D : Craniosynostosis : an analysis of the timing, treatment, and complications in 164 consecutive patients. **Plast Reconstr Surg 80** : 195-212, 1987

39. White N, Evans M, Dover MS, Noons P, Solanki G, Nishikawa H : Posterior calvarial vault expansion using distraction osteogenesis. **Childs Nerv Syst 25** : 231-236, 2009

40. Zubovic E, Woo AS, Skolnick GB, Naidoo SD, Smyth MD, Patel KB : Cranial base and posterior cranial vault asymmetry after open and endoscopic repair of isolated lambdoid craniosynostosis. **J Craniofac Surg 26** : 1568-1573, 2015