www.jkns.or.kr 10.3340/jkns.2011.49.3.194
Print ISSN 2005-3711 On-line ISSN 1598-7876
J Korean Neurosurg Soc 49 : 194-199, 2011

Technical Note

Immediate Spontaneous Shape Correction Using Expantile Zigzag Craniectomy in Infantile Scaphocephaly

-Is There an Improvement in the Developmental Quotient following Surgery?-

Sang-Dae Kim, M.D., Ph.D.,[1] Shizuo Oi, M.D., Ph.D.[2]
Department of Neurosurgery,[1] Korea University College of Medicine, Seoul, Korea
Division of Pediatric Neurosurgery,[2] Jikei Women's and Children's Medical Center, Jikei University School of Medicine, Tokyo, Japan

There is still debate over which method of the surgery is the most appropriate for the treatment of scaphocephalic infants. In addition, change in psychomotor development following these procedures is a very complex issue that has not yet been resolved. In this paper, the authors describe a surgical technique for immediate spontaneous shape correction of infantile scaphocephaly. There were significant differences between pre- and postoperative cephalic index. We also describe an improvement in the developmental quotient following surgery. Therefore, this expantile zigzag craniectomy should be recommended to correct for isolated sagittal craniosynostosis in infants.

Key Words : Scaphocephaly · Cephalic index · Developmental quotient · Expantile zigzag craniectomy.

INTRODUCTION

Scaphocephaly, which is the most common type of craniosynostosis, accounts for approximately 40 to 60% of the isolated suture synostosis, and occurs in 1 in 5,000 children at birth[2,5,8,18,26,27)].

Numerous techniques have been used to correct scaphocephaly in infants including bilateral strip craniectomy, wide-strip craniectomy, the π procedure, total vertex craniectomy, endoscopic craniectomy and cranial vault remodeling with parietal flap cranioplasties[3,4,6,7,10,11,13-15,17,18,20,22,23,25-30)]. However, there is still debate over which method of the surgery is most appropriate for the treatment of scaphocephalic infants. In addition, change in psychomotor development following these procedures is a very complex issue that has not yet been resolved. In spite of this, only few studies regarding the correlation between immediate calvarial shape correction and perioperative psychomotor development in infants with isolated sagittal craniosynostosis have been conducted to date[4,5,10,15,16,19,31)].

Therefore, this study was conducted to document the immediate head shape correction using by expantile zigzag craniectomy and to discuss the developmental improvement following the use of our surgical technique to treat scaphocephalic infants.

CLINICAL MATERIALS AND METHODS

Patient population

The authors retrospectively reviewed the medical records of our hospital to identify infants who had undergone expantile zigzag craniectomy to treat sagittal craniosynostosis (scaphocephaly) between January, 2003 and February, 2008. Only patients that were less than 12 months of age and only diagnosed with isolated sagittal suture synostosis were included in this study. In addition, all patients with associated hydrocephalus or premature synostosis of multiple sutures were excluded from this study. Diagnosis was based on clinical examinations and radiological findings, especially of three-dimensional reconstruction computed tomography (3D CT).

Radiological assessment of scaphocephaly

3D CT scans were obtained prior to surgery (Fig. 1A, 2A, 3A). And within 1 week after surgery, so called immediately postoperative 3D CT scan, we rechecked CT scans (Fig. 1B, 2B, 3B). In addition, all patients underwent 3D CT scans at our hospital during

---

• Received : December 7, 2010 • Revised : December 29, 2010
• Accepted : February 6, 2011
• Address for reprints : Shizuo Oi, M.D., Ph.D.
Division of Pediatric Neurosurgery, Jikei Women's and Children's Medical Center, The Jikei University School of Medicine, 3-25-8 Nishi-Shinbashi, Minato-ku, Tokyo 105-8461, Japan
Tel : +81-3-3433-1111, Fax : +81-3-3438-1161
E-mail : shizuo.prof.oi@jikei.ac.jp

follow-up period (Fig. 1C, 2C, 3C). These scans were then compared with preoperative CT scans to compare the change in the cephalic index (CI). The cranial CI was obtained by dividing the maximum measurable bitemporal or biparietal diameter by the maximum measurable midline anterior-posterior diameter and then multiplying by 100. All CIs were measured using 3D CT scans.

Assessment of psychomotor development

Evaluation of psychomotor development was performed both pre- and postoperatively using the developmental quotient (DQ, Tsumori-Inamo method) in conjunction with a K test from the Kyoto Scale of Psychological Development[12]. The DQs were divided into three scales that measure important aspects of infant and toddler development : performance, recognition and language. These DQs were then corrected for age (mean 100) and transformed into a total developmental quotient (mean 100).

Surgical technique

To perform this operation, the patient is positioned supine upon the operating table with their necks slightly flexed. Next, the patient's head is placed on a small horse-shoe headrest and then fixed in place using a bandage adhesive tape. A large S-shape skin incision is then made as shown in Fig. 4A, with the anterior margin of the incision being just below the coronal suture line including the anterior fontanelle, and the posterior margin being below the posterior fontanelle. When the dissection using the Raimondi clump is performed between the galea and osteal fascia, the scalp flap can be reflected bilaterally (Fig. 4B). Four or 5 burr holes are then placed 1-1.5 cm lateral to midline on both side of a large area of exposed skull that includes the bregma, lamda and synostosed suture lines. Next, craniotomy lines in a zigzag shape as large as possible are created after which multiple burr holes are created and expantile zigzag craniectomy is performed in the parietal bone from midline to the squamoid suture and from the coronal to lambdoid sutures bilaterally. Epidural dissection is

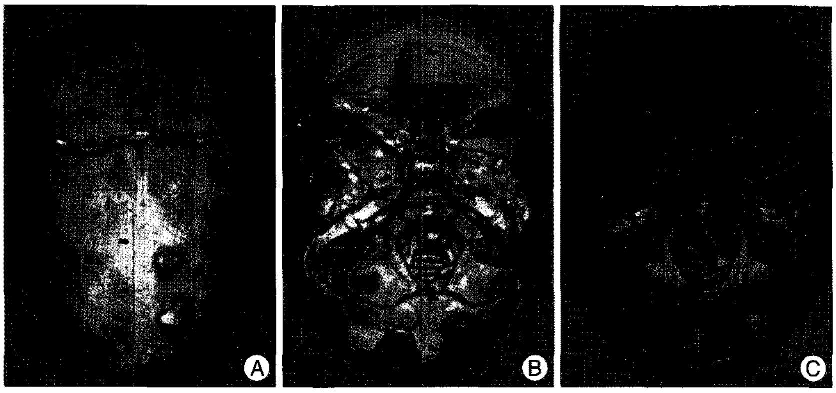

Fig. 1. 3D CT scans, vertex view. A : Preoperative. B : Immediate postoperative. C : Follow-up postoperative.

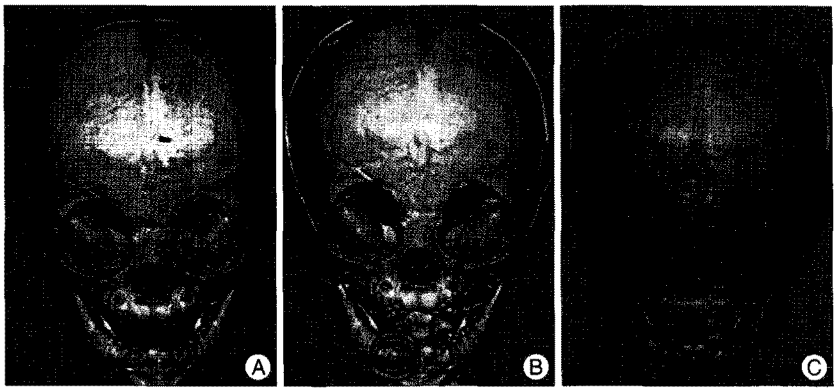

Fig. 2. 3D CT scans, anterior view. A : Preoperative. B : Immediate postoperative. C : Follow-up postoperative.

Fig. 3. 3D CT scans, left lateral view. A : Preoperative. B : Immediate postoperative. C : Follow-up postoperative.

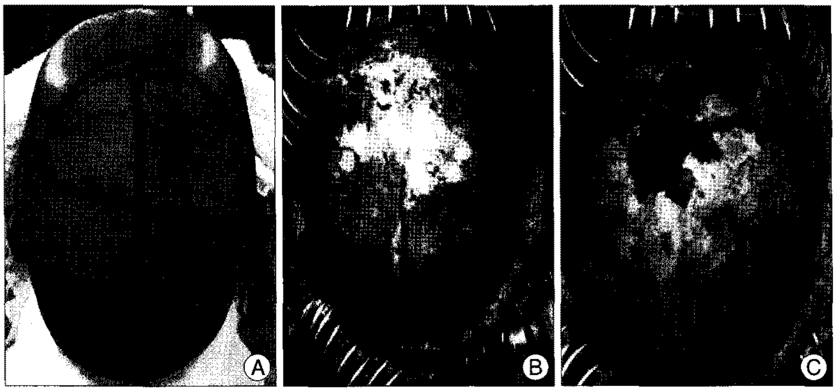

Fig. 4. Intraoperative findings. A : S-shape skin incision, vertex view. B : Vertex view after skin flap reflected was done bilaterally. C : Vertec view following expantile zigzag craniectomy. Noted widening bitemporal distance.

Table 1. Patient characteristics and perioperative data of the cephalic indices*

Patient No.	Age at surgery (months)	Sex	Preop CI (C_0)	Imm-postop CI (C_1)	CI change* (%)	Follow-up CI (C_2)	CI change‡ (%)	Follow-up period (months)
1	5	M	63.5	80.6	+26.9	82.4	+29.8	59
2	6	M	64.8	70.9	+9.4	80.0	+23.5	12
3	4	F	78.5	87.5	+11.5	92.3	+17.6	17
4	4	F	67.7	71.8	+6.1	83.2	+22.9	19
5	3	M	67.9	74.4	+9.6	90.1	+32.7	13
6	5	M	71.7	73.8	+2.9	79.8	+11.3	12
7	9	M	63.9	78.1	+22.2	79.5	+24.4	11
8	4	F	66.4	77.7	+17.0	78.9	+18.8	2
Mean	5		68.1	76.9	+13.2†	83.3	+22.6§	18.1

*CI change (%) : $(C_1-C_0)/C_0\times100$, †$p<0.005$ (signed-rank test). ‡CI change (%) : $(C_2-C_0)/C_0\times100$, §$p<0.005$ (signed-rank test). CI : cephalic index, F : female, Imm : immediate, M : male

Table 2. Pre- and postoperative developmental quotients*

Subscales	Preoperative DQ			Postoperative DQ			Mean change (%)
	Mean	SD	Range	Mean	SD	Range	
Performance	93.1	9.1	80-109	112.9	14.7	91-129	+21.3
Recognition	94.0	12.8	76-116	109.0	12.4	83-121	+16.0
Language	103.9	17.0	80-127	115.8	19.0	94-158	+11.5
Total	95.4	10.7	78-111	109.9	10.9	91-124	+15.2*

*p=0.0078 (signed-rank test). DQ : developmental quotients (Tsumori-Inamo method), SD : standard deviation

concurrently performed, while taking care to avoid dural injury. Finally, sagittal suturectomy is performed, which leaves all edges of the zigzag craniectomy floating (Fig. 4C). Hardwares such as screws and plates for fixation are not needed, and the transposition of free bone flaps is not necessary in this procedure. The operating time, estimated blood loss and amount of blood transfusion (if necessary) were also evaluated in this study.

Statistical analysis

All statistical analyses were performed using SAS (version 9.1, Cary, NC) statistical software. A Wilcoxon signed-rank test was used for the univariate analysis. The results were considered significant when a p-value less than 0.05 was obtained.

RESULTS

The results of data from data of 8 infants comprised of 5 males and 3 females, aged 3 to 9 months (mean 5 months) were reviewed. All cases met the above criteria. The results of the radiological assessment are presented in Table 1 and Fig. 5. The preoperative CI (%) was available for all patients and ranged from 63.5 to 78.5 (mean 68.1, SD 4.7). The mean CI of the scans taken immediately after surgery was 76.9 (SD 5.1, range 70.9-87.5). The CI of the CT scans was increased by an average of 13.2% (SD 7.7, p<0.005). And, mean CI of the scans taken after several months was 83.3 (SD 4.8). Of the cases evaluated, 7 (88%) were followed up for longer than 11 months. When the CI of postoperative scans of patients who underwent follow-up were compared with the preoperative CI, the CI of the follow-up CT was 22.6% greater than that of the preoperative CI (SD 6.4, p < 0.005) (Table 1).

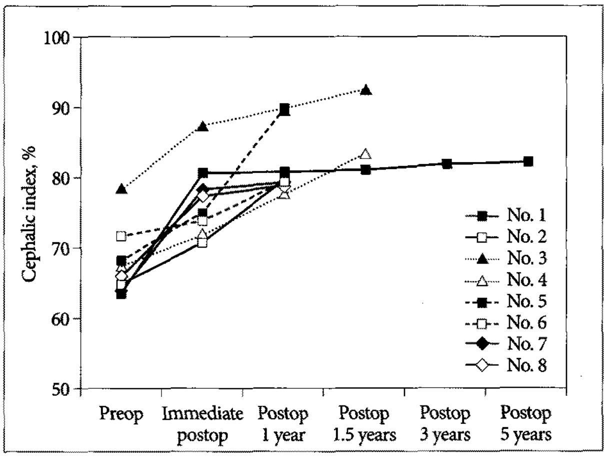

Fig. 5. Graphic showing the significant expansion in cephalic index following surgery.

The mean preoperative total DQ of all 8 infants, was 95.4 (SD 10.7, range 78-111) (Table 2). However, the mean postoperative total DQ, which was determined over an average of 24.1 months (range 2-60 months), was 109.9 (SD 10.9, range 91-124), indicating an overall postoperative improvement in DQ of 15.2% (Fig. 6). Evaluation of the mean total DQ of all patients using a signed-rank test indicated that there were significant differences between the pre- and postoperative scores (p=0.0078) (Fig. 7). However, when the language subscale was evaluated, no significant differences between the pre- and postoperative scores were observed (p=0.0742).

The mean operation time for each procedure was 3.3 hours (SD 0.5, range 2.5-4.2) (Table 3). Furthermore, the procedure resulted in a mean blood loss of 113 mL (SD 63.2, range 46-249), a mean blood transfusion of 132 mL (SD 55.5, range 50-210) and a transfusion rate of 75 % (6 of 8 patients).

No fatalities or incidents of intracranial complications were observed among the 8 infants who underwent this operation. In addition, no further surgery was required as a result of cosmetic worsening or any other complications. In all cases, the parents and surgeons were satisfied with the improvement in cranial shape.

DISCUSSION

Optimal timing of surgical correction

Many papers have shown that the optimal age period for surgical correction of scaphocephaly is between birth and 12 months of age[3-6,9,11,18,23-25,27,29]. As the skull grows, the degree of stiffness increases markedly with age due to an increase in the thickness of the two tables of skull[11,21,25]. Furthermore, most surgeons agree that thicker and less ductile skull bones require more invasive surgical procedures, which results in a higher risk of complications, as well as longer operation times[18]. The modern craniofacial surgical technique has improved greatly; therefore the mortality and morbidity associated with this procedure have decreased[5,21,18]. Some authors suggest that surgical correction after the first 6 months of life produces less favorable results than surgery performed during the first 6 months[29], which indicates that it is very important that surgery to correct scaphocephaly be conducted at as early an age as possible.

However, simple synostectomy conducted within the second month of life appears to be complicated by an increased rate of re-craniostenosis, which occurs due to the rapid growth and the high reossification power of the skull within the first few months of life[6,18,23]. For this reason, Boop et al.[6] advocated waiting until patients were three months of age to perform the operation. Although it is essential to correct the skull early in scaphocephalic infants because of further expansion of the brain, it is also important to consider the relationship between the age of the patient and the flexibility of the skull[25].

Based on these findings, we recommend that surgical expansion to treat scaphocephaly should be conducted during the fourth to sixth month of life due to the flexibility of the skull and the minimal risk of early reossification and restenosis. The mean patient age of our data is 5 months.

Concepts and advantages of expantile zigzag craniectomy

In this paper, the authors describe a surgical technique for immediate spontaneous shape correction of infantile scaphocephaly. The S-shape skin incision allowed for sufficient operative fields. The plasticity of the infant cranial vault is an important factor in this surgical technique[2]. The authors previously suggested that infantile calvarial normalization using a non-fixed procedure is

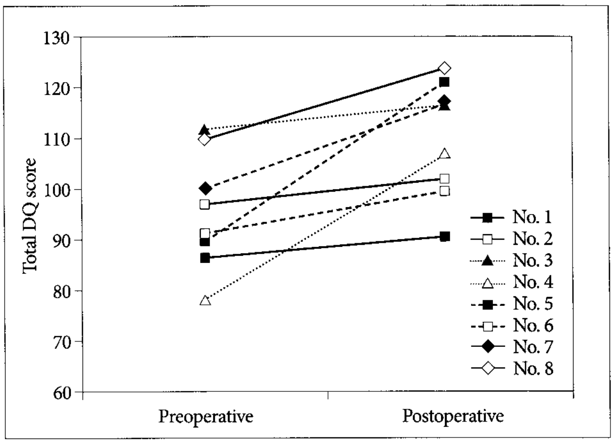

Fig. 6. Preoperative versus postoperative total DQ. Note consistent improvement. The mean improvement defined was 15.2% (singed-rank test comparison, *p*=0.0078).

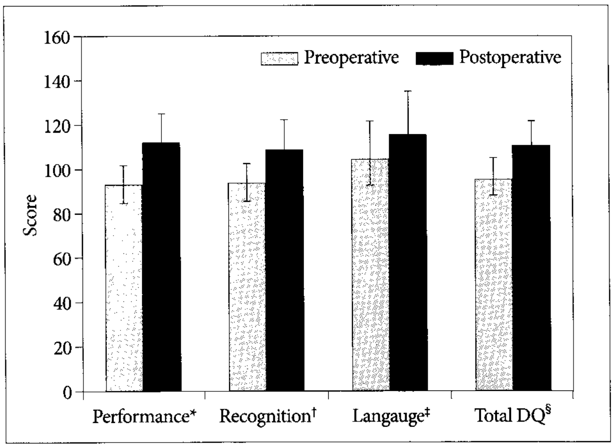

Fig. 7. Preoperative versus postoperative each subscales DQ. There were a significant differences between pre- and postoperative DQs except language subscales (signed-rank test comparison, **p*=0.0156, †*p*=0.0039, ‡*p*=0.0742, §*p*=0.0078).

Table 3. Patient body weights and intraoperative data in eight infants

Patient No.	Body weight (g)	Blood loss (mL)	Transfusion (PRBC, mL)	Op time (hours)
1	8,560	72	86	4.2
2	6,200	72	180	3.5
3	7,700	78	50	3.6
4	7,300	46	-	3.0
5	5,800	160	210	4.0
6	8,020	80	-	3.0
7	8,010	146	110	2.5
8	7,200	249	160	3.1
Mean	7,350	113	132	3.3

PRBC : packed red blood cell

essential and necessary for treatment of patients that are in the neonatal early infancy stages of life[20]. According to above concepts, we designed the zigzag craniectomy. When this craniectomy is performed in bilateral parietal bone, the edges of the craniectomy remain floating, which results in less of an obstacle to transverse brain growth and improved cosmesis. In addition, this method allows widening toward the bitemporal and biparietal directions immediately following the operation. The results of this study showed that the CI improved significantly immediately after surgery and persisted until the follow-up period, at which point the mean improvement was 13.2% and 22.6% when compared to the preoperative CI. Agrawal et al. suggested that the maximum improvement occurs in the early postoperative period due to the removal of restrictions previously placed on skull growth by the synostotic sagittal suture[1]. The immediate correction of skull shape is also of great importance in the parent-child relationship[13]: therefore, surgical management of scaphocepaly should also ensure that good cosmetic results are obtained[18]. The technique described here allows easy reshaping of the immature skull bone, enabling the skull contour to be corrected immediately. Futhermore, the intraoperative blood loss was less than that reported for other techniques.

Correlation of developmental quotients

In 1965, Anderson et al.[4] proposed that early surgical release of craniosynostosis allows normal brain growth while protecting the brain from irreversible damage caused by pressure. However, the results of recent studies suggest that the cranial release and reconstruction do not impact mental abilities during infancy[5,10,16,19,24,29,31]. Actually, the test of infant can also be problematic because instruments may not be sensitive to subtle changes in cognitive functions. In addition, many aspects of cognitive development do not emerge until a child is older and therefore cannot be measured at the preschool age[31]. Some authors stated that the likelihood of serious brain dysfunction as a result of a single-suture craniosynostosis was extremely low[24]. It seems that patients with simple craniosynostosis are not likely to experience increased intracranial pressure[16]. Nevertheless, there were significant changes observed between pre and postoperative DQ in our study except language subscale (p=0.0742).

Limitation of our study

The surgical technique evaluated here is limited because it only allows treatment of a single pathology among the many types of craniosynostosis. In addition, this study is limited by the relatively small number of cases and short follow-up period.

CONCLUSION

The results of this study indicate that expantile zigzag craniectomy is safe, simple and effective and therefore applicable to the scaphocephalic infants. However careful patient selection is important to ensure that better results are obtained. A multidisciplinary approach can minimize the risk of morbidity and mortality in the treatment of craniosynostosis, thereby leading to a satisfactory outcome[9].

And, it is also important to understand the psychomotor development of the patient as well as the natural course of skull growth prior to conducting surgery to correct skull shape. Once it is determined that surgery is appropriate, immediate correction of the skull shape can be obtained by conducting an early expantile zigzag craniectomy.

References

1. Agrawal D, Steinbok P, Cochrane DD : Long-term anthropometric outcomes following surgery for isolated sagittal craniosynostosis. **J Neurosurg 105** : 357-360, 2006
2. Alvarez-Garijo JA, Cavadas PC, Vila MM, Alvarez-Llanas A : Sagittal synostosis: results of surgical treatment in 210 patients. **Childs Nerv Syst 17** : 64-68, 2001
3. Amm CA, Denny AD : Correction of sagittal synostosis using foreshortening and lateral expansion of the cranium activated by gravity : surgical technique and postoperative evolution. **Plast Reconstr Surg 116** : 723-735, 2005
4. Anderson FM, Greiger L : Craniosynostosis : a survey of 204 cases. **J Neurosurg 22** : 229-240, 1965
5. Arnaud E, Renier D, Marchac D : Prognosis for mental function in scaphocephaly. **J Neurosurg 83** : 476-479, 1995
6. Boop FA, Shewmake K, Chadduck WM : Synostectomy versus complex cranioplasty for the treatment of sagittal synostosis. **Childs Nerv Syst 12** : 371-375, 1996
7. Erdinçler P, Kaya AH, Kafadar A, Canbaz B, Kuday C : Bilateral peninsula-shaped linear craniectomy for mild degrees of craniosynostosis: indication, technique and long-term results. **J Craniomaxillofac Surg 32** : 64-70, 2004
8. Fearon JA, McLaughlin EB, Kolar JC : Sagittal craniosynostosis: surgical outcomes and long-term growth. **Plast Reconstr Surg 117** : 532-541, 2006
9. Ferreira MP, Collares MV, Ferreira NP, Kraemer JL, Pereira Filho Ade A, Pereira Filho Gde A : Early surgical treatment of nonsyndromic craniosynostosis. **Surg Neurol 65 Suppl 1** : S1:22-S1:26; discussion S1:26, 2006
10. Gewalli F, Guimarães-Ferreira JP, Sahlin P, Emanuelsson I, Horneman G, Stephensen H, et al. : Mental development after modified pi procedure: dynamic cranioplasty for sagittal synostosis. **Ann Plast Surg 46** : 415-420, 2001
11. Hendel PM, Nadell JM : Projection geometry and stress-reduction technique in craniofacial surgery. **Plast Reconst Surg 83** : 217-227, 1989
12. Ikuzawa M : **Kyoto Scale of Psychomotor Development,** ed 2. Kyoto, Japan : Nakanishiya Shuppan, 1989
13. Jane JA, Edgerton MT, Futrell JW, Park TS : Immediate correction of sagittal synostosis. **J Neurosurg 49** : 705-710, 1978
14. Jimenez DF, Barone CM : Endoscopic craniectomy for early surgical correction of sagittal craniosynostosis. **J Neurosurg 88** : 77-81, 1998
15. Kaiser G : Sagittal synostosis--its clinical significance and the results of three different methods of craniectomy. **Childs Nerv Syst 4** : 223-230, 1988
16. Kapp-Simon KA, Figueroa A, Jocher CA, Schafer M : Longitudinal assessment of mental development in infants with nonsyndromic craniosynostosis with and without cranial release and reconstruction. **Plast Reconstr Surg 92** : 831-839; discussion 840-841, 1993
17. Kim SW, Shim KW, Plesnila N, Kim YO, Choi JU, Kim DS : Distraction vs remodeling surgery for craniosynostosis. **Childs Nerv Syst 23** : 201-206, 2007
18. Massimi L, Tamburrini G, Caldarelli M, Di Rocco C : Effectiveness of a

limited invasive scalp approach in the correction of sagittal craniosynostosis. **Childs Nerv Syst 23** : 1389-1401, 2007

19. Mathijssen I, Arnaud E, Lajeunie E, Marchac D, Renier D : Postoperative cognitive outcome for synostotic frontal plagiocephaly. **J Neurosurg 105** : 16-20, 2006
20. Nonaka Y, Oi S, Miyawaki T, Shinoda A, Kurihara K : Indication for and surgical outcomes of the distraction method in various types of craniosynostosis. Advantages, disadvantages, and current concepts for surgical strategy in the treatment of craniosynostosis. **Childs Nerv Syst 20** : 702-709, 2004
21. Oi S, Matsumoto S : Trigonocephaly (metopic synostosis). Clinical, surgical and anatomical concepts. **Childs Nerv Syst 3** : 259-265, 1987
22. Oi S, Okuda Y, Sasaki K, Yamada H, Ijichi A, Matsumoto S : [Analysis of intracranial pressure and operative procedures for craniosynostosis.] **Neurol Med Chir (Tokyo) 27** : 110-116, 1987
23. Olds MV, Storrs B, Walker ML : Surgical treatment of sagittal synostosis. **Neurosurgery 18** : 345-347, 1986
24. Persing JA : Immediate correction of sagittal synostosis. **J Neurosurg 107** : 426, 2007
25. Roddi R, Vaandrager JM, Gilbert PM, van der Meulen JC : Reshaping of the skull in the early surgical correction of scaphocephaly. **J Craniomaxillofac Surg 21** : 226-233, 1993
26. Sikorski CW, Iteld L, McKinnon M, Yamini B, Frim DM : Correction of sagittal craniosynostosis using a novel parietal bone fixation technique : results over a 10-year period. **Pediatr Neurosurg 43** : 19-24, 2007
27. Smyth MD, Tenenbaum MJ, Kaufman CB, Kane AA : The "clamshell" craniotomy technique in treating sagittal craniosynostosis in older children. **J Neurosurg 105** : 245-251, 2006
28. Venes JL, Sayers MP : Sagittal synostectomy. Technical note. **J Neurosurg 44** : 390-392, 1976
29. Vollmer DG, Jane JA, Park TS, Persing JA : Variants of sagittal synostosis: strategies for surgical correction. **J Neurosurg 61** : 557-562, 1984
30. Wagner W, Wiewrodt D : A simple technique for the correction of frontal bossing in synostotic scaphocephaly. **Childs Nerv Syst 24** : 373-377, 2008
31. Warschausky S, Angobaldo J, Kewman D, Buchman S, Muraszko KM, Azengart A : Early development of infants with untreated metopic craniosynostosis. **Plast Reconstr Surg 115** : 1518-1523, 2005