Child's Nervous System (2019) 35:1351–1356
https://doi.org/10.1007/s00381-019-04246-5

ORIGINAL ARTICLE

The unseen third dimension: a novel approach for assessing head shape severity in infants with isolated sagittal synostosis

Rosalinda Calandrelli[1] · Fabio Pilato[2] · Luca Massimi[2] · Marco Panfili[1] · Concezio Di Rocco[3] · Cesare Colosimo[1]

Received: 8 May 2019 / Accepted: 30 May 2019 / Published online: 12 June 2019

Abstract

Purpose This study aimed to develop a novel approach to assess the severity of skull dysmorphology in infants with isolated sagittal synostosis (ISS) and its relationship with the surgical results.

Methods We divided 66 infants with ISS into three groups by combining the scaphocephalic (SSI-A) and platycephalic (VLI) indices as descriptors of the relation between length, width, and height. We evaluated each skull for morphology as hyperdolichocephalic (< 66%) versus dolichocephalic (66–77%) and as hyperplatycephalic (< 78%) versus platycephalic skull (78–85%). A score system was developed as follows: 2 points for values < 66% and < 78% and 1 point for values between 66 and 77% and 78 and 85% in SSI-A and VLI, respectively. The overall score was calculated and it was used to classify our patients on a 4-point ordinal scale, according to the severity of head shape (2 = mild, 3 = moderate, 4 = severe).

Results Thirty-two infants resulted in mild group, 17 in moderate group, and 17 in severe group. SSI-A and VLI were reduced according to the severity of ISS. We demonstrated a positive correlation between SSA-A and VLI in mild subgroup of patients while we found a negative correlation between SSA-A and VLI in moderate and in severe subgroups. Moreover, a positive correlation was found between severe subgroup and Sloan III class of surgical results.

Conclusion This study describes a simple tool to better classify infants with ISS, considering the three-dimensional morphology of the skull, because it evaluates both the dolichocephalic and platycephalic component.

Keywords Isolated sagittal synostosis · Scaphocephalic index · Platycephalic index · Computer tomography · Skull morphology

Introduction

Closure of the sagittal suture normally begins in the third or fourth decade of life. In utero, closure of this suture is the most common form of isolated, non-syndromic synostosis [1]. This occurs at a rate of one in 2000 to 5000 live births and is more common in boys than in girls [2–4]. This premature fusion along with brain's development directs the cranium growing in the anteroposterior dimension, causing the scaphocephalic head shape. Distinctive findings of this deformity include frontal bossing, temporal protrusion, coronal constriction and occipital protuberance [5].

Scaphocephaly is a descriptive term for a typical head shape observed in children with premature synostosis of the sagittal suture. However, among patients with isolated sagittal synostosis (ISS), the head shape varies considerably in relation to the severity of the abnormality. Previous studies quantified the degree of severity of scaphocephalic skull shape measuring a set of cranial indices using different planes and internal landmarks [6] but the differences in methodologies limited the possibility to compare results among different studies.

Moreover, all the described cephalic indices cannot be considered reliable measures for severity evaluation because this ratio is not able to capture the amount of correction of the

✉ Rosalinda Calandrelli
rosalinda.calandrelli@policlinicogemelli.it

[1] Area diagnostica per immagini, Fondazione Policlinico Universitario Agostino Gemelli IRCCS, Università Cattolica del Sacro Cuore, Largo Francesco Vito 1, 00168 Rome, Italy

[2] Area neuroscienze, Fondazione Policlinico Universitario Agostino Gemelli IRCCS, Rome, Italy

[3] International Neuroscience Institute, Hannover, Germany

reduced posterior skull height associated with sagittal craniosynostosis [7, 8]. In particular, these cephalic indices described the two-dimensional ratio of maximum breadth and maximum length, only addressing the dolichocephalic component of the skull, but the vertical component accompanying several presentations of scaphocephaly was not taken into account.

It may explain the lack of relationship between the scaphocephaly severity indices and the measurements of intracranial volume as previously reported [9–13].

We used platycephalic term to refer to the vertical growth of the head favored by a minor resistance at the level of the anterior fontanel resulting, in some patients, in a large and more developed frontal region compared with the two posterior thirds of the skull.

The purpose of our study was (1) developing a novel approach to better classify the severity of ISS considering the three dimensions (length, width, and height) by combining two indices as descriptors of their relationship and (2) assessing surgical results in relation to the severity of the skull deformity.

Material and method

Patients

Preoperative cranial computed tomographic (CT) scans of 66 infants with isolated sagittal synostosis, consecutively admitted to our hospital between 2015 and 2017 (13 females and 53 males; mean age 124 days, range 76–194 days) were reviewed.

Sixty-six age-matched healthy subjects (mean age 125 days, range 94–192 days) who underwent CT examinations for craniofacial trauma were enrolled as controls. The study was approved by the local Institutional Review Board.

Skull shape quantification

CT images implemented by three conventional orthogonal reformatted viewports of the skull were used to compute the "traditional" cephalic index at the skull vault (CI), three distinct scaphocephaly severity indices (SSIs) in the A, F, and M planes in order to assess the plane with the best diagnostic accuracy value to describe the dolichocephalic component of the skull.

Traditional cephalic index represents the ratio of maximum cranial width to maximum cranial length × 100 [14].

SSIs in the A , F, and M planes With a lateral view of a 3D reformation of the skull, a skull base plane was determined by using the frontal nasal suture anteriorly and the opisthion (the middle point on the posterior margin of the foramen magnum of the occipital bone of the skull) posteriorly. This plane was shifted superiorly until positioned (1) just above the top of the lateral ventricles (A plane), (2) at the foramina of Monroe (F plane), and (3) at the level of the maximal dimension of the fourth ventricle (M plane). CT slices taken at the level of these planes were used to calculate three ratios (cranial width/the cranial length × 100), which defined the three indices SSI-A, SSI-F, and SSI-M, respectively [6].

Vertical-longitudinal index (VLI) was calculated to describe the platycephalic component of the skull and it represents the ratio between cranial height and cranial length × 100. The cranial height represents the distance between the basion and the bregma (Fig. 1).

All measurements were repeated twice and performed independently by two investigators. Moreover, all measurements were compared with those of normal controls.

Concerning the scaphocephalic index, we divided skull morphology into hyperdolichocephalic (< 66%) and dolichocephalic (66–77%) according to previously published data [6, 15]. About the VLI, because previous data are lacking, we collected the distribution of values ranging from the lower value of the controls to the lower value of the affected children and as cut-off value the mean-2SD of the patient group values. As a result, infants were divided into hyperplatycephalic (< 78%) and platycephalic skull (78–85%).

A score system was developed assigning 1 point or 2 points as the following: 2 points for values < 66% and < 78% and 1 point for values between 66 and 77% and 78 and 85% in SSI-A and VLI, respectively. The overall score was calculated by the sum and it was used to classify our patients according to the severity of head shape on a 4-point ordinal scale (2 = mild, 3 = moderate, 4 = severe).

Surgical result after reconstruction for craniosynostosis

The cosmetic results were evaluated according to the outcome classification proposed by Sloan et al. [16]. In this system, classes 1 through 4 represent excellent to good overall correction of the deformity, but with varying degrees of minor visible and/or palpable irregularities: none in class 1, palpable but not visible irregularities in class 2, visible irregularities in class 3, and requiring reoperation in class 4. Classes 5 through 7 represent patients with significantly compromised correction: not requiring further surgery in class 5, requiring further surgery in class 6, and believed by the surgeon to require further surgery but with the family declining further surgery in class 7 [16].

We compared surgical results with the severity of skull deformity in our groups of patients.

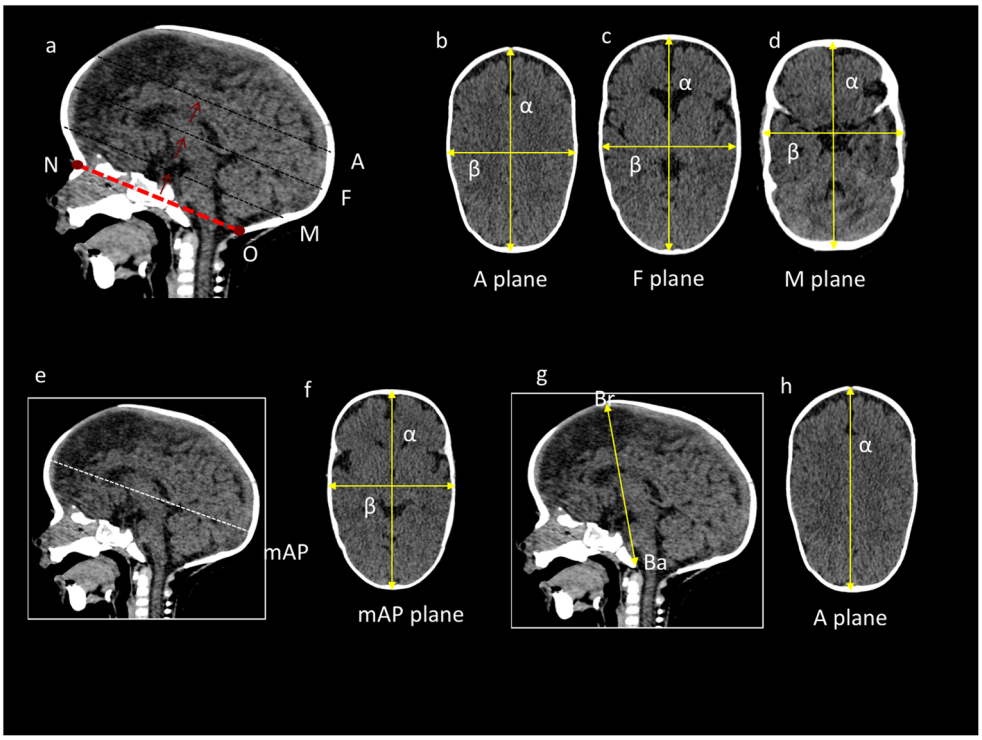

Fig. 1 Skull shape severity quantification using traditional cephalic index (CI), scaphocephaly severity indices (SSIs) in the A, F, and M planes, and vertico-longitudinal index (VLI). SSIs in the A, F, and M planes (**a**–**d**). With a lateral view of a 3D reformation of the skull, a skull base plane was determined using the frontal nasal suture anteriorly and the opisthion posteriorly (red dot line in **a**). This plane was shifted superiorly until positioned (1) just above the top of the lateral ventricles (A plane in **a**), (2) at the foramina of Monroe (F plane in **a**), and (3) at the level of the maximal dimension of the fourth ventricle (M plane in **a**). Three ratios (cranial width/cranial length × 100) define SSI-A (**b**), SSI-F (**c**), and SSI-M (**d**), respectively. Traditional CI (**e**, **f**) represents the ratio of maximum cranial width to maximum cranial length × 100. VLI in plane A (**g**, **h**) represents the ratio of cranial length to cranial height × 100. The cranial height represents the distance between the basion (Ba) and the bregma (Br)

Statistical analysis

Descriptive statistics were expressed as the means ± SD for continuous variables if not otherwise specified.

Receiver operating characteristic (ROC) analysis was used to evaluate the accuracy, specificity, and sensibility. Diagnostic accuracy of all SSIs and CI was calculated and reported: area under the ROC curve (AUC) with 95% confidence intervals (95% CI), sensitivity, and 1-specificity.

One-way ANOVA was used to compare the more accurate CI, VLI, and age in the three subgroups. Pearson correlation coefficients were used to compare the more accurate CI versus VLI in all subjects and for each group (controls, mild, moderate, severe).

Pearson correlation was used to compare age and the more accurate CI or VLI in the population. χ^2 was used to compare surgical results and head deformity severity.

Statistical significance was set at $p < 0.05$. All statistical comparisons were corrected for multiple comparisons.

Statistical analyses were conducted using Statistical Package for the Social Sciences (SPSS) for Windows version 21.0 (SPSS Inc., Chicago, IL, USA).

Results

The SSI-A resulted the more accurate index if compared to standard CI and SSI-F and SSI-M (Table 1). According to SSI-A and corresponding VLI, our patients were subdivided into three clinical severity subgroups.

In particular, 32 infants were attributed to the mild group (mean age 127 days, range 90–194 days; score = 2); 17 infants were attributed to the moderate group (mean age 121 days,

Table 1 AUC, confidence intervals, sensitivity, and 1-specificity for traditional CI and SSIs in planes A, F, and M

Index	AUC	95% confidence intervals	Sensitivity	1-specificity
CI	0.999	0.997–1	0.955	0.015
SSI-A	1	0.999–1	0.985	0.015
SSI-F	0.980	0.952–1	0.970	0.045
SSI-M	0.994	0.985–1	0.985	0.182

AUC, area under the curve; *CI*, cephalic index at the skull vault; *SSIs*, scaphocephaly severity indices

range 76–176 days; score = 3) and 17 infants were attributed to the severe group (mean age 125 days, range 88–172 days; score = 4) (Table 2).

One-way ANOVA showed significant differences in SSI-A and VLI values among the three subgroups ($p < 0.001$). We found a positive correlation between the SSA-A and VLI in the control group (r^2 0.380, $p < 0.001$) and in mild subgroup of patients (r^2 0.123, $p < 0.05$) while we found an inverse correlation between the SSA-A and VLI in the moderate (r^2 0.358, $p < 0.05$) and in the severe subgroups (r^2 0.250, $p < 0.05$) (Figs. 2 and 3).

All infants belong to the first three classes of the Sloan classification (I class, 51 children; II class, 12 children; III class, 3 children) (Table 2). A significant difference was found between surgical results and the severity of the head deformity ($p < 0.05$) in the three subgroups but a positive correlation was found only between severe subgroup and Sloan III surgical results (r^2 0.137, $p < 0.05$).

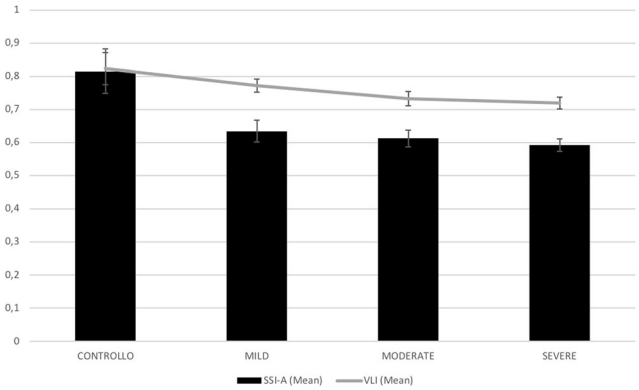

Fig. 2 Trend of SSI-A and VLI in the healthy control group and in the three severity subgroups of isolated sagittal synostosis (ISS). SSI-A and VLI are reduced according to the severity of ISS; the reduction of SSI-A is greater compared with that of VLI. SSI-A is shown with plots while VLI is shown as a gray line

Discussion

The degree of suture fusion, severity of head shape deformity, and additional changes such as frontal and occipital bossing and biparietal narrowing vary significantly among children with ISS [6]. The degree of severity of ISS is commonly quantified by measuring the CI [17]. Previous reports published new SSIs using CT scan bone windows to create specific two-dimensional planes from which to measure skull length and width [6, 18, 19]. When compared with the standard CI, these studies found that the SSIs had a higher specificity and sensitivity than CI for quantifying sagittal synostosis because, in the traditional CI, the points of maximal skull length change dramatically due to variable degrees of frontal and occipital bossing [6]. However, although these indices are reproducible objective measurements, they underestimated the severity of scaphocephaly because they describe the relation only between two morphological measurements (length and width) and they do not consider the overall abnormal head shape because height is not taken into account. The reduction in posterior skull height accompanying the various presentations of scaphocephaly has never been assessed in this condition.

Taken together, these considerations might explain why all these cephalic indices have been shown to poorly correlate with measurements of intracranial volume [9–13].

The first goal of our study was to introduce a new approach to better classify the severity of ISS by combining two indices as descriptors of the relationship of three morphological measurements (length, width, and height). To correctly measure the dolichocephalic component of the skull, the accuracy of the traditional cephalic index and of the three SSIs in the A, F, and M planes of the skull base was tested. As previously described, the SSI-A resulted the most accurate index [6]. This plane captured both the frontal bossing and the inferior projection, or bossing of the occiput. Consequently, plane A was used also to calculate the VLI, evaluating the platycephalic component of the skull and we divided our children in three degrees of skull dysmorphology using a score severity scale combining the SSI-A index and the corresponding VLI.

Our study demonstrates that both SSI-A and VLI are reduced according to the severity of ISS even if the dolichocephalic component is more pronounced with respect to

Table 2 Classification of patients according to SSI-A and VLI

Standard	Mild	Moderate		Severe
SSI-A	66–76% (point 1)	66–76% (point 1)	< 66% (points 2)	< 66% (points 2)
VLI	78–85% (point 1)	< 78% (points 2)	78–85% (point 1)	< 78% (points 2)
Total score	***2***	***3***	*3*	***4***
Patients (*n*)	32	9	8	17
Total patients (*n*)	32	17		17
Sloan classification	I/II	I/II		I/II/III

SSI-A, scaphocephalic severity index in A plane; *VLI*, vertico-longitudinal index

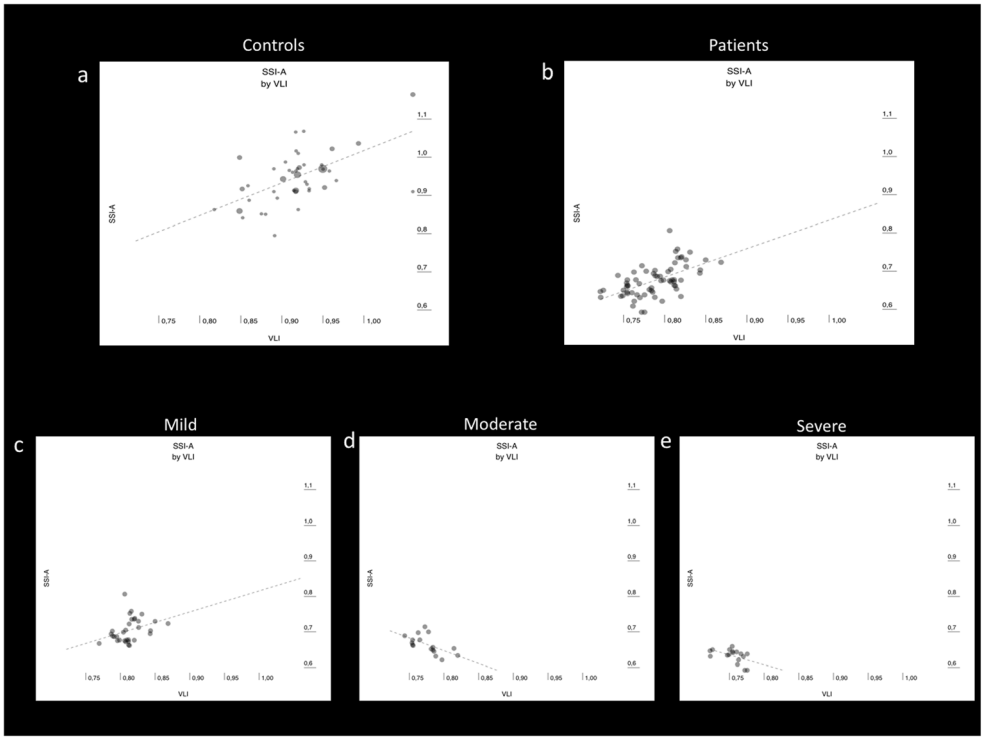

Fig. 3 Correlation between the scaphocephaly index (SSI-A) and platycephalic index (VLI) for the healthy control group (**a**), all patients (**b**), and for the three severity subgroups of isolated sagittal synostosis (**c**–**e**). In the control group (**a**) and in the mild subgroup (**c**) a positive correlation between the SSA-A and VLI is represented while in the moderate (**d**) and in the severe subgroups (**e**) an inverse correlation is showed

platycephalic component. In the subgroups analysis, we demonstrated a positive correlation between the SSA-A and VLI in mild subgroup of patients while we found an inverse correlation between the SSA-A and VLI in the moderate and in the severe subgroups. Then, in the mild group, the dolichocephalic skull component is more noticeable with respect to the platycephalic skull dysmorphology while in the moderate and severe group, the dolichocephalic morphology is associated with evident platycephalic skull morphology. It suggests that the degree of overall deformity is lower in the mild group with respect to the moderate and severe groups because the effects of the compensatory changes accompanying the sagittal suture synostosis cause a different compliance of the skull. It is probably that in the mild group, the compensatory process is still active because of a later closure of the sagittal suture while in the moderate and severe groups, there is an earlier closure of sagittal suture causing a ceiling effect of compensatory mechanisms over the time determining a more evident dolichocephalic and platycephalic deformity of the skull.

Finally, we found a difference between surgical results and the severity of the head deformity. In particular, although all our infants belong to the first three classes of the Sloan classification, only patients of the severe subgroups were in class III.

This data strengthens the idea that, although the surgical management of sagittal synostosis results in good cosmetic results, a part of the infants belonging to the severe group of skull dysmorphology may have worse cosmetic results than of the infants in the mild and moderate subgroups.

Conclusion

This study develops a new approach to better quantify the severity of skull dysmorphology in infants with ISS by combining two indices as descriptors of the relationship of three morphological measurements (length, width, and height). This approach may be considered a simple and useful tool to better classify infants with isolated sagittal synostosis because it evaluates both the dolichocephalic and platycephalic component. Further studies, involving a larger sample, are required to validate these results.

Compliance with ethical standards

Conflict of interest Rosalinda Calandrelli declares that she has no conflict of interest.

Fabio Pilato declares that he has no conflict of interest.

Luca Massimi declares that he has no conflict of interest.

Marco Panfili declares that he has no conflict of interest.

Concezio Di Rocco declares that he has no conflict of interest.

Cesare Colosimo declares that he is scientific consultant for Bracco Diagnostics Inc. and Bayer HealthCare.

Research involving human participants Ethical approval: We declare that all procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. For this type of study, formal consent is not required.

Informed consent Informed consent was obtained from all individual participants included in the study.

References

1. Chummun S, McLean NR, Flapper WJ, David DJ (2016) The management of nonsyndromic, isolated sagittal synostosis. J Craniofac Surg 27(2):299–304. https://doi.org/10.1097/SCS.0000000000002363
2. Hunter AG, Rudd NL (1976) Craniosynostosis. I. Sagittal synostosis: its genetics and associated clinical findings in 214 patients who lacked involvement of the coronal suture(s). Teratology 14(2):185–193. https://doi.org/10.1002/tera.1420140209
3. Wilkie AO, Wall SA (1996) Craniosynostosis: novel insights into pathogenesis and treatment. Curr Opin Neurol 9(2):146–152
4. Fearon JA, McLaughlin EB, Kolar JC (2006) Sagittal craniosynostosis: surgical outcomes and long-term growth. Plast Reconstr Surg 117(2):532–541. https://doi.org/10.1097/01.prs.0000200774.31311.09
5. Schmelzer RE, Perlyn CA, Kane AA, Pilgram TK, Govier D, Marsh JL (2007) Identifying reproducible patterns of calvarial dysmorphology in nonsyndromic sagittal craniosynostosis may affect operative intervention and outcomes assessment. Plast Reconstr Surg 119(5):1546–1552. https://doi.org/10.1097/01.prs.0000256067.42651.30
6. Ruiz-Correa S, Sze RW, Starr JR, Lin HT, Speltz ML, Cunningham ML, Hing AV (2006) New scaphocephaly severity indices of sagittal craniosynostosis: a comparative study with cranial index quantifications. Cleft Palate Craniofac J 43(2):211–221. https://doi.org/10.1597/04-208.1
7. Fearon JA, Ditthakasem K, Herbert M, Kolar J (2017) An appraisal of the cephalic index in sagittal craniosynostosis, and the unseen third dimension. Plast Reconstr Surg 140(1):138–145. https://doi.org/10.1097/PRS.0000000000003422
8. Fearon JA (2014) Evidence-based medicine: craniosynostosis. Plast Reconstr Surg 133(5):1261–1275. https://doi.org/10.1097/PRS.0000000000000093
9. Lee SS, Duncan CC, Knoll BI, Persing JA (2010) Intracranial compartment volume changes in sagittal craniosynostosis patients: influence of comprehensive cranioplasty. Plast Reconstr Surg 126(1):187–196. https://doi.org/10.1097/PRS.0b013e3181dab5be
10. Heller JB, Heller MM, Knoll B, Gabbay JS, Duncan C, Persing JA (2008) Intracranial volume and cephalic index outcomes for total calvarial reconstruction among nonsyndromic sagittal synostosis patients. Plast Reconstr Surg 121(1):187–195. https://doi.org/10.1097/01.prs.0000293762.71115.c5
11. Anderson PJ, Netherway DJ, McGlaughlin K, David DJ (2007) Intracranial volume measurement of sagittal craniosynostosis. J Clin Neurosci 14(5):455–458. https://doi.org/10.1016/j.jocn.2006.07.001
12. Netherway DJ, Abbott AH, Anderson PJ, David DJ (2005) Intracranial volume in patients with nonsyndromal craniosynostosis. J Neurosurg 103(2 Suppl):137–141. https://doi.org/10.3171/ped.2005.103.2.0137
13. Fischer S, Maltese G, Tarnow P, Wikberg E, Bernhardt P, Tovetjarn R, Kolby L (2015) Intracranial volume is normal in infants with sagittal synostosis. J Plast Surg Hand Surg 49(1):62–64. https://doi.org/10.3109/2000656X.2014.971804
14. Haas LL (1952) Roentgenological skull measurements and their diagnostic applications. Am J Roentgenol Radium Therapy, Nucl Med 67(2):197–209
15. Bendon CL, Sheerin FB, Wall SA, Johnson D (2014) The relationship between scaphocephaly at the skull vault and skull base in sagittal synostosis. J Craniomaxillofac Surg 42(3):245–249. https://doi.org/10.1016/j.jcms.2013.05.009
16. Sloan GM, Wells KC, Raffel C, McComb JG (1997) Surgical treatment of craniosynostosis: outcome analysis of 250 consecutive patients. Pediatrics 100(1):E2
17. Dvoracek LA, Skolnick GB, Nguyen DC, Naidoo SD, Smyth MD, Woo AS, Patel KB (2015) Comparison of traditional versus normative cephalic index in patients with sagittal synostosis: measure of scaphocephaly and postoperative outcome. Plast Reconstr Surg 136(3):541–548. https://doi.org/10.1097/PRS.0000000000001505
18. Ruiz-Correa S, Starr JR, Lin HJ, Kapp-Simon KA, Cunningham ML, Speltz ML (2007) Severity of skull malformation is unrelated to presurgery neurobehavioral status of infants with sagittal synostosis. Cleft Palate Craniofac J 44(5):548–554. https://doi.org/10.1597/06-190.1
19. Ruiz-Correa S, Gatica-Perez D, Lin HJ, Shapiro LG, Sze RW (2008) A Bayesian hierarchical model for classifying craniofacial malformations from CT imaging. Conf Proc IEEE Eng Med Biol Soc 2008:4063–4069. https://doi.org/10.1109/IEMBS.2008.4650102