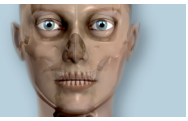
Orthodontics & Craniofacial Research



ORIGINAL ARTICLE

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Craniofacial cephalometric morphology in 8-year-old children with operated sagittal synostosis

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Date:
Accepted 30 August 2014

DOI: 10.1111/ocr.12056

Heliövaara A., Vuola P., Hukki J. Craniofacial cephalometric morphology in 8-year-old children with operated sagittal synostosis
Orthod Craniofac Res 2015; **18**: 27–32. © 2014 John Wiley & Sons A/S. Published by John Wiley & Sons Ltd

Structured Abstract

Objectives – To evaluate cephalometrically craniofacial morphology in children with operated sagittal synostosis and to compare the findings with age- and sex-matched controls.

Setting and Sample Population – Forty-two children (37 boys) with operated primary sagittal synostosis were compared retrospectively with age- and sex-matched controls from lateral cephalograms taken at a mean age of 8.1 (range 7.0–8.9) years.

Material and Methods – The operations had been performed between the ages of 2 months and 6.3 years at three Finnish hospitals. The surgical methods included strip craniectomy, pi-plasty and cranial vault expansion. A paired Student's *t*-test and Pearson's correlation analysis were used in the statistical analyses.

Results – Children with operated sagittal synostosis had wide cranial base angles and their mandibles were retrognathic with labially inclined lower incisors relative to the controls. Age at craniosynostosis operation did not correlate with the cranial base angle.

Conclusion – This study suggests that children with operated sagittal synostosis have minor distinctive morphological features in the cranial base and mandible. Orthodontic evaluation of craniofacial growth is recommended.

Key words: cephalometrics; cranioplasty; craniosynostosis; sagittal suture; scaphocephaly

Introduction

Premature fusion of the sagittal suture, scaphocephaly, is the most common type of non-syndromic isolated craniosynostosis, with an estimated prevalence between 1:2000 and 1:5000 live births (1). Most of the cases are sporadic, although familial

occurrence has been reported (1, 2). Boys are more likely to be affected than girls (1).

The first sign of sagittal synostosis is usually abnormal head shape, with elongation of the cranial vault, prominent forehead and occiput, variable ridging of the sagittal suture and increased head circumference. Thompson et al. (3) reported borderline or elevated intracranial pressure, ≥10 mmHg, in six (24%) of 25 patients with sagittal synostosis. Untreated progressive sagittal craniosynostosis may be associated with an increased risk of learning disabilities (4) and language impairment (5, 6). The goal of surgical correction of craniosynostosis is to ensure normal function of the brain and to correct the abnormal shape of the head. Several surgical techniques have been developed for treatment. There is no consensus regarding the optimal surgical technique. Although strip craniectomies are still used, concerns regarding inadequate correction of intracranial compartment volume have led to more extensive calvarial reshaping operations such as pi-plasty (7) and comprehensive calvarial vault reconstruction (8). Corrective surgery is preferentially performed within the first year of life to capitalize on the malleability of the cranial bone and the rapid growth of the infant's brain (9, 10).

Previous studies have described the head shape of children with scaphocephaly both pre- and post-operatively (11, 12). Secondary to premature synostosis of the sagittal suture, the calvaria becomes elongated and narrowed, with an increase in head circumference and a reduction in the cephalic index, CI (head width/head length × 100). After craniosynostosis operations, the CI increases. Computed tomography has been used to assess intracranial volume before surgery and to evaluate intracranial volumetric changes achieved by surgery (12, 13). However, no long-term data exist of cephalometric craniofacial growth in children with operated sagittal synostosis.

Craniosynostosis and subsequent corrective operations might influence the growth of the cranial base regions, thus also affecting the relationship of the maxilla and the mandible. The cranial base forms the floor of the cranial vault. It extends anteriorly from the foramen caecum to the basio-occipital bone posteriorly. The maxilla is attached to the anterior segment of the cranial base, and the mandible is attached to the posterior segment. The aim of this study was to evaluate cephalometrically the craniofacial morphology in children with operated sagittal synostosis and to compare the findings with age- and sex-matched controls.

Materials and methods

The patients comprised 42 Finnish children with operated sagittal synostosis who at the age of 8 years had attended follow-up at the Cleft Palate and Craniofacial Center, Department of Plastic Surgery, Helsinki University Central Hospital. The children were born between 1996 and 2003. Their mean age when the lateral cephalograms were taken was 8.1 (range 7–8.9) years. Most of the children (n = 37, 88%) were boys. Patients with syndromes were excluded.

The craniosynostosis operations had been performed between the ages of 2 months and 6.3 years (mean age 1.3 years) at three Finnish hospitals. The surgical methods were strip craniectomy (n = 8), pi-plasty (n = 19) and cranial vault expansion including modified Barrel stave technique (n = 15). Six children had had secondary operations before the 8-year cephalogram. Two re-operations were performed after strip craniectomy, two after pi-plasty and two after cranial vault expansion. The re-operations were performed by modified Barrel stave (n = 5) and pi-plasty (n = 1) techniques. The age at re-operation varied from 1.2 to 8.6 (mean 4.6) years. None of the children had undergone orthodontic treatment.

Standard lateral cephalometric radiographs, taken with the head positioned according to the Frankfort horizontal plane with molar teeth occluded and lips in repose, were used in the cephalometric analysis. The cephalograms were traced twice by the same orthodontist by a computer-connected digitizer. The computer was programmed to calculate the mean of the two digitalizations, which were determined to an

accuracy of 1 mm. The reference points and landmarks are shown in Fig. 1.

Cephalometric values of the children with operated sagittal synostosis were compared with the values of 42 healthy age- and sex-matched controls collected from the normative Finnish database of the University of Helsinki, Department of Orthodontics. The difference in age between each of the comparative pairs was <6 months. The mean age of the controls was 8 (range 6.8–8.6) years. A Student's paired *t*-test was used to compare the children with operated sagittal synostosis with age- and sex-matched controls.

The research protocol was approved by the Helsinki University Central Hospital.

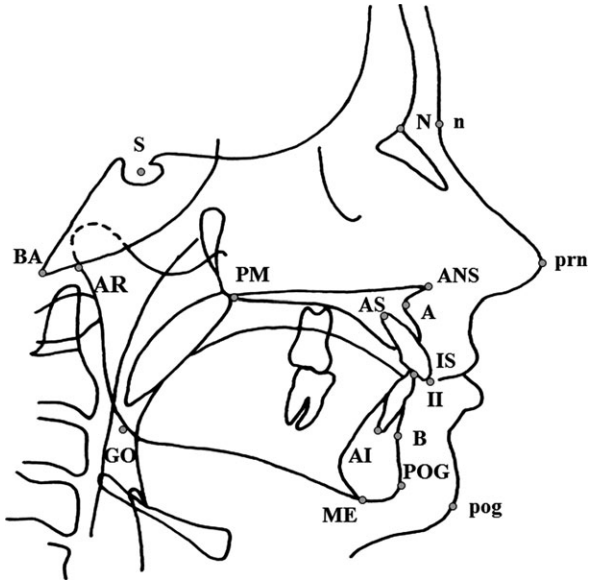

Fig. 1. Cephalometric landmarks. Abbreviations, full names and definitions. A (Point A): deepest point on the anterior contour of the maxillary alveolar arch; AI (Apex inferior): apex of the root of the average mandibular central incisor; ANS (Anterior nasal spine): tip of anterior nasal spine; AR (Articulare): intersection between the external contour of the cranial base and the dorsal contour of the mandible; AS (Apex superior): apex of the root of the average maxillary central incisor; B (Point B): deepest point on the anterior contour of the mandibular alveolar arch; BA (Basion): most inferior point of the clivus of the occipital bone; GO (Gonion): intersection between the external contour of the mandible and the bisector of the angle between the ramus line and mandibular line; II (Incisive inferior): incisal edge of the average mandibular central incisor; IS (Incisive superior): incisal edge of the average maxillary central incisor; ME (Menton): most inferior point on mandibular symphysis; N (Nasion): most anterior point on the nasofrontal suture; n (soft tissue nasion): intersection between NSL and soft profile contour; PM (Pterygomaxillare): intersection between nasal floor and the posterior contour of maxilla; POG (Pogonion): most prominent point of the bony chin; pog (soft tissue pogonion): most anterior point of soft tissue chin; prn (pronasale): most prominent point of apex nasi; S (Sella): centre of sella turcica; ML (mandibular line): tangent to the lower border of mandible through points ME and GO; NL: (nasal line): line through points ANS and PM; NSL (Nasion-Sella line): line through points N and S; RL (ramus line): tangent to the mandibular ramus through AR.

Results

The measurements of the children with operated sagittal synostosis and the controls, and the comparisons using the Student's paired *t*-test are shown in Table 1. The children with operated sagittal synostosis had obtuse cranial base angles. No difference in the maxillary position in relation to cranial base was observed between the two groups, but the mandibles of the children with operated sagittal synostosis were retrognathic and the lower incisors labially inclined. Also their soft tissue profile was more convex relative to the controls. Age at craniosy-

Table 1. **Means, standard deviations and *p*-values of the cephalometric variables in paired Student's *t*-test between children with operated sagittal synostosis and controls. Angles are reported in degrees**

	Sagittal synostosis	SD	Controls	SD	*p*-Value
N-S-BA	134.2	6.1	131.4	4.3	0.02*
SNA	81	4	81.7	3.2	0.354^{ns}
SNB	75.5	3.9	77.8	3	0.004**
ANB	5.5	1.8	3.9	1.9	0.001**
SNPOG	76.2	4	77.9	3	0.041*
NSL/ML	34.1	5	34.6	4.2	0.598^{ns}
NSL/NL	6.9	4	6	2.8	0.231^{ns}
ML/ RL	130.2	4.8	131.1	5.2	0.445^{ns}
ANS/ME/N-ME × 100	58	2.1	58.2	2	0.743^{ns}
S-GO/N-ME × 100	63.8	4.3	64.6	3.6	0.412^{ns}
IS-AS/NSL	109.4	8.7	107.2	7.7	0.306^{ns}
II-AI/ML	99.8	8.3	93.2	7.2	0.001***
n-prn-pog	132.6	4.8	135.3	3.4	0.025*

*$p < 0.05$
**$p < 0.01$
***$p < 0.001$

nostosis operation did not correlate with the cranial base angle $r = 0.072$, ns.

Discussion

The goal of the surgical correction of sagittal craniosynostosis is to remodel the cranial vault to allow unimpeded brain growth and to avoid elevated intracranial pressure. In addition, the goal is a long-lasting correction of the abnormal head shape deformity so that further craniofacial growth is not disturbed. The major findings in the children with operated sagittal synostosis at 8 years of age were obtuse cranial base angles and retrognathic mandibles. Cranial surgery can cause morphological alterations during growth, but evaluation of the roles of intrinsic growth deficiency associated with craniosynostosis, genetic factors, functional adaptations and growth disturbance caused by surgery is complicated.

The cranial base angle can be altered by posterior flexion or by changes in anterior facial height. In this study, no differences in vertical measurements were found, but the cranial base angle was more obtuse (134°) in children with sagittal synostosis than in controls (131°). Guimareaes-Ferreira et al. (9) studied 82 children with scaphocephaly who had been operated on by modified pi-plasty. The cranial base angles did not change significantly from the pre-operative value (132°) at 7 months of age to the post-operative value at 5 years of age. The cranial base angle is approximately 142° at birth, diminishing to 130° at 5 years of age (14). From 5 to 15 years of age, the cranial base angle is relatively stable (14). However, variation is relatively large, with a standard deviation of 5 or more (15).

Findings of the relationship between cranial base angulation and malocclusion in healthy children are contradictory. Whereas some researchers have found no association between cranial base angle and malocclusion (16–19), other have reported an increased cranial base angle and a posterior position of the mandible in Class II patients (20, 21). Baccetti et al. (22) concluded that the glenoid fossa is more posteriorly positioned in Class II than in Class III subjects. According to Björk (23), the glenoid fossa follows the displacement of temporal bone in association with an obtuse cranial base angle during growth. This has been also simulated experimentally in rabbits. The increased distal displacement of the glenoid fossa was caused by artificial premature cranial synostosis (24). Interestingly, no differences in our study were noted in the maxillary position in relation to the cranial base. Enlow (25) has shown that the growth of the maxilla is under the influence of the cranial base, which in turn is influenced by growth of the brain. The mandible, by virtue of its remoteness from the region, acts in a more independent way, although its articulation at the glenoid fossa provides the potential for influence from the cranial base (25). While the anterior cranial base influences the growth of the maxilla, the middle and posterior cranial bases may influence mandibular position because of the location of the glenoid fossa. The cranial base anterior to sella stabilizes by 7 years of age through ossification of the spheno-ethmoidal and intersphenoidal synchondroses (23). Among cranial base synchondroses, the spheno-occipital synchondrosis is the most important site for sagittal and vertical growth. The spheno-occipital synchondrosis ossifies by 17 years of age until which time the clivus grows in length (26).

Differences in size, position and form of the cranial base and jaws are important when analyzing craniofacial growth. Varrela (17) studied 5-year-old Finnish children and found no differences in the cranial base, including the base flexure, between Class 1 and Class II groups. Class II malocclusion in deciduous dentition was characterized by a short mandibular corpus and lower facial height, a large gonial angle and retruded dentoalveolar position of the mandible. In this study, angular and proportional measurements were used to eliminate possible differences in body growth and head size. It should be remembered, however, that besides the sagittal synostosis and its corrective surgery, such factors as mastication, mode of breathing, head posture and habits can be related to the aetiol-

ogy of Class II malocclusion and the inclination of incisors.

Our sample size was relatively small, and only four girls were included. The low number of girls reflects the greater incidence of sagittal synostoses in boys (1). The variations in type and timing of surgery, skills of the surgeon and number of corrective operations introduce uncontrolled variables to this study. Although it would have been interesting to compare the results of different surgical methods on post-operative growth, the material was too small for further analysis. The choice of the operative procedure depends on patient's age at the time of operation, and the degree and location of the deformity. Possibly, the more expansive operative methods result in increased intracranial volume, immediate correction of the abnormal skull shape and more favourable craniofacial growth than the strip craniectomy. Potentially the post-operative growth and treatment outcome may also vary according to the initial severity and location of the synostosis. In Finnish material, the typical features in sagittal synostosis are frontal bossing, posterior narrowing and posterior ridging of the sagittal suture even if there is a total synostosis. The saddle-shaped heads are less frequent.

The age of the craniosynostosis operation may be important in later growth. In our small sample, the age at surgery did not correlate with the cranial base angle. In animal experiments, premature fusion of sutures has been induced by several methods, including gluing bones across sutures with cyanoacrylate. The cyanoacrylate bridge, causing premature union of coronal bones, resulted in cranial deformity in rabbits (27). Surgical removal of growth restriction by linear craniectomy before completion of skull growth was followed by growth, making up for earlier restriction, and by spontaneous correction of the skull deformity during the remaining period. The degree of spontaneous correction was found to be dependent on age at release (27). In children, by the age of 2 years, intracranial volume has reached 77%, and, by 5 years, 90% of the volume observed at the age of 15 years (28). On the other hand, the cephalograms had been taken at a mean age of 8.1 years, before the pubertal growth spurt of the mandible.

No previous cephalometric studies of sagittal synostosis are available. Frontal cephalometric radiographs were not available for this study. It would have been interesting to evaluate also the transverse dimensions of the 8-year-old children with operated sagittal synostosis. In addition to frontal bossing, occipital lengthening, and sagittal ridging, the classical characteristics of scaphocephaly include biparietal narrowing.

Children with syndromatic and non-syndromatic craniosynostosis have been recommended to be treated at a craniofacial centre and followed by a multidisciplinary team throughout the growth period (29). Dental and orthodontic recommendations also exist (30). According to our study, the role of the orthodontist is in the craniofacial team is emphasized.

Conclusion

In conclusion, this study suggests that the children with operated sagittal synostosis have minor distinctive morphological features in the cranial base and mandible. Orthodontic evaluation of craniofacial growth is recommended.

Clinical relevance

Craniosynostosis and subsequent corrective operations in early childhood might influence the growth of the cranial base regions, thus also affecting the relationship of the maxilla and the mandible**.** However, no cephalometric data exists of children with operated sagittal synostosis although children with syndromatic and non-syndromatic craniosynostosis have been recommended to be treated at a craniofacial centre and followed by a multidisciplinary team throughout the growth period. Based on the results of this study continuous evaluation of craniofacial growth is recommended, and the role of the orthodontist is in the craniofacial team is emphasized.

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