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Clinical evaluation of non-syndromic scaphocephaly surgically corrected with the procedure of total vertex craniectomy

Matthias Kreppel*, Martin Kauke, Ali-Farid Safi, Andrea Grandoch, Nina Pocek-Behn, Hans-Joachim Nickenig, Joachim Zöller

Department for Oral and Craniomaxillofacial Plastic Surgery, University of Cologne, Germany

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ABSTRACT

The present investigation constitutes a retrospective evaluation of the outcome in children who received surgical correction of a scaphocephalic phenotype by median total vertex craniectomy. Between September 2009 and September 2015, a total of 35 infants with non-syndromic scaphocephaly were treated according to the same standardized operative technique of total vertex craniectomy by a single surgeon approach. At the time of surgery, the patients were between 3 and 12 months of age, with a median of 5 months. The mean duration of the procedure was 94 min. The duration of postoperative follow-up was a mean of 24 months (range 6–49 months). A total of 34 (97%) patients were successfully treated by total vertex craniectomy and were thus classified as category I according to the Whitaker score. Only one individual was assigned to category IV, necessitating secondary major craniofacial corrective surgery. Aesthetic outcomes were excellent in 34 cases and poor in one case. No major complication occurred. The reoperation rate was 3%. The surgical method we present herein is a wide median craniectomy which can be applied in young individuals with non-syndromic single-suture scaphocephaly.

1. Introduction

Craniosynostosis is defined by the premature closure of one or multiple cranial sutures, showing an annual incidence of approximately 1 in 2500 births (Persing, 2008; Safi et al., 2018a,b). An interplay of multiple causative factors (e.g., genetics, environment and teratogenics) are held responsible for the development of this disease (Wilkie et al., 2017). In approximately 15%, craniosynostotic deformities are associated with a genetic defect and a consecutive syndromic phenotype (termed syndromic craniosynostosis). However, nonsyndromic cases are much more frequent and constitute about 85% of all craniosynostosis cases (Abraham et al., 2018).

Scaphocephaly, the premature closure of the sagittal suture, is the most frequent single suture nonsyndromic craniosynostosis, contributing 50–60% of all cases (Simpson et al., 2017). The incidence is about 1 in 5000 children, with a male to female ratio of 4 to 1 (Lajeunie et al., 1996; Kolar, 2011; Simpson et al., 2017). Considering the pathophysiology as propagated by Rudolph Virchow in 1851, sutural premature closure entails growth restriction perpendicular to the prematurely closed suture and compensatory growth at the remaining patent sutures (Virchow, 1851; Al-Shaqsi et al., 2018). Therefore in the case of sagittal synostosis, one can observe compensatory growth from the coronal and lambdoid sutural complex (Virchow, 1851; Simpson et al., 2017). This pathological growth pattern results in the typical scaphocephalic phenotype: a dysmorphic cranial vault, appearing boat-shaped due to an enlargement of the anterior-posterior diameter with a varying degree of temporal hollowing (narrowed transverse dimension) (Marsh and Vannier, 1986; Mühling, 1986; Zöller, 2003; Simpson et al., 2017). Consequently, the head is rather long and small, frequently showing frontal bossing and occipital coning (Persing, 2008; Simpson et al., 2017; Al-Shaqsi et al., 2018). As the sagittal suture is closed and osseous overgrowth occurs, an osseous prominence can frequently be palpated in the median plane (Al-Shaqsi et al., 2018).

Craniofacial anomalies involving the sutural system are very complex entities of disease. Depending on the number and combination of premature suture obliterations, the repercussions are

* Corresponding author. Department for Oral and Craniomaxillofacial and Plastic Surgery, University of Cologne, Kerpener Straße 62, 50931, Cologne, Germany. Fax: +49 221 478 7360.
E-mail address: mattheskreppel@yahoo.de (M. Kreppel).

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manifold. Most importantly, pathological restrictive growth patterns of the cranial vault may negatively impact the integrity of the brain. As the cranial vault represents an osseous cage specially designed to protect the brain, restricted expansion and elevated rigidity poses a hindrance to the brain's expansion, potentially resulting in an entrapment syndrome with neurological impairment (Zöller, 2003; Safi et al., 2018a,b). Thus, in cases of clinical manifestation of intracranial hypertension, immediate surgical intervention is warranted (Stein and Schut, 1977; Zöller, 2003; Safi et al., 2018a,b).

However, scaphocephaly is rarely associated with elevated intracranial pressure. Hence, surgical intervention generally aims to correct the cranial dysmorphia in an elective cosmetic surgical setting (Barritt et al., 1981; Greene and Winston, 1988; Al-Shaqsi et al., 2018). Due to our intrinsic sense of beauty, mainly governed by the fact that we judge aesthetics based on the absence or presence of specific proportions between craniofacial components, we are likely to negatively perceive individuals with clinically visible scaphocephaly. Hence, this malformation is likely to entail negative social pressure (Foltz and Loeser, 1975; Barritt et al., 1981). Here we evaluate a one-step surgical procedure that we routinely perform at our institution for correction of cranial dysmorphia due to premature sagittal synostosis, namely, the total vertex craniectomy.

2. Materials and methods

Subjects in the retrospective cohort study presented herein were all patients who were diagnosed with a craniosynostosis and were consecutively treated at the Department of Oral and Craniomaxillofacial Plastic Surgery at the University of Cologne between September 2009 and September 2015. For consideration, patients had to meet the following criteria: nonsyndromic premature closure of the sagittal suture (scaphocephaly), treatment naive, total vertex craniectomy for correction of the dysmorphic skull, and follow-up care at our department.

Predictor variables were age, sex, time point of surgical assessment, reason for initial presentation, postoperative complications, pre- and postoperative hemoglobin and duration of postsurgical follow-up. All patients who were referred to our clinic were clinically and radiographically evaluated by our head physician and surgeon. Accessory evaluation included standard preoperative hematologic diagnostics, ophthalmological examination and photographic documentation of the patient's dysmorphic craniofacial system. Postoperative evaluation included hematologic analysis and supervision by a pediatric specialist for 1 day in the intensive care unit (ICU) as well as for the remaining days of their postoperative hospitalization. In consideration of the as low as reasonably achievable (ALARA) principle, patients did not regularly receive pre- and/or postoperative x-ray–based imaging. Imaging was applied only in difficult diagnostic situations and whenever postoperative imaging was necessary (e.g. exclusion of complications). Additionally, following the surgical procedure, each patient was re-evaluated by our head physician and surgeon. After their stay, patients were advised to regularly check in for follow-up. The follow-up standardly consisted of an evaluation of the surgical outcome (physical appearance, skull shape) and an evaluation of the overall aesthetics by both the surgeon and the parents of the individual. The comments on the aesthetic assessment were used to judge the success of the surgical procedure presented below.

2.1. Surgical technique

All patients were treated by the same surgeon in cooperation with assisting surgeons from our local institutional neurosurgical department. Per standard procedure, every individual received general endotracheal anesthesia and perioperative care according to a standardized protocol. All individuals underwent to equal treatment for the correction of a single-suture sagittal craniosynostosis with scaphocephalic phenotype. We performed the procedure of total vertex craniectomy (approximately 9 cm wide) including the sagittal suture as well as the proximal coronal and lambdoid sutural complex. Figs. 1–6 present pre-, intra-, and postoperative pictures. With to this procedure, we intended to achieve unhindered brain expansion and consecutive autoinductive remodeling of the cranial vault, led by the expanding brain. Thus, this procedure does not involve active remodeling of the cranial vault.

A detailed description follows.

Prior to incision, a vasoconstrictive agent was administered intracutaneously following the course of the marked skin incision lines. The bicoronal incision was placed slightly posterior to the habitual bicoronal incision line. The superficial incision was followed by careful subgaleal dissection anteriorly and posteriorly, thereby exposing the cranial vertex (Figs. 4 and 5). In the median plane, the periost was divided. It is particularly important to visualize the coronal and lambdoid sutures. Multiple bilateral burr holes were made between the coronal and lambdoid sutures up to 4.5 cm lateral to the midline (Fig. 5). The osteotomy was carried out extending posteriorly of the lambdoid suture and anteriorly of the coronal suture (Fig. 5). A large portion of the occipital bone is removed. In cases in which the infant exceeded the fifth month of age, additional lateral osteotomies posterior to the lambdoid suture and at the coronal suture were performed. This is important, because the correction of the biparietal width becomes increasingly difficult as the malleability of the cranial bone decreases with the individual's age. Special care must be taken, as the additional osteotomies potentially increase the individual's blood loss. Importantly, an osseous fragment was left in situ for protection of the sagittal sinus (Fig. 5). In the end, after removal of the cranial vertex, tight wound closure was achieved. Two surgical drains were placed in order to allow for postoperative drainage (Fig. 6).

2.2. Evaluation of surgical outcome

We evaluated the surgical outcome according to the classification of Whitaker et al. as recently modified and applied by Engel et al. and Safi et al. following fronto-orbital advancement, the subjective view of the patient's parents and the surgeon, as well as the rate of postoperative complications (Tables 1 and 2) (Whitaker et al., 1987; Engel et al., 2012; Safi et al., 2018a,b). If the surgeon as

Fig. 1. Preoperative view of an individual affected by scaphocephaly.

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