Trigonocephaly case series

2021

A retrospective chart review of all patients with a diagnosis of a metopic ridge or metopic craniosynostosis was performed from 2000 to 2015 at Rady Children's Hospital. Patients were grouped based on computed tomographic scans consistent with metopic craniosynostosis versus metopic ridge.

Results: Data were available for 15 metopic ridge patients, 74 metopic craniosynostosis patients, and 213 normal patients. Mean metopic ridge ICV was greater than mean metopic craniosynostosis ICV at 4 to 6 months and 7 to 12 months. Controlling for age and sex, the difference in ICV associated with metopic ridging was 197.484 cm3 and 137.770 cm3 at 4 to 6 and 7 to 12 months, respectively. Similarly, mean metopic index was significantly greater in metopic ridge patients compared to mean metopic craniosynostosis at 4 to 6 months and at 7 to 12 months.

Conclusions: Our study provides volumetric and anthropometric data to support the hypothesis that isolated metopic ridge is an intermediate phenotype between metopic craniosynostosis and normal cranial anatomy. We hope that characterizing the spectrum of disease involving premature closure of the metopic suture with regard to ICV and metopic index will aid physicians in their management of patients with isolated metopic ridge [1)].

2020

A retrospective review of all coronal synostosis and metopic craniosynostosis patients undergoing cranial vault remodeling (CVR) with fronto-orbital advancement (FOA) from March 2010 to June 2019 was performed. Before 2014 (“Control group”), all patients received blood transfusion at the start of surgery. In 2014, a protocol of preoperative EPO and ferrous sulfate with perioperative TXA and non-automatic transfusion was instituted (“Study group”). Patient demographics and anthropometrics, perioperative hemoglobin (Hb) levels, and transfusion details were collected and compared.

Thirty-six patients met inclusion criteria. Twenty-one patients were in the control group, and 15 in the Study group. Nineteen patients had metopic synostosis, 11 had unicoronal synostosis, and 6 had bicoronal synostosis. There were no significant differences between groups in demographics, operative time, intraoperative crystalloid volume, craniofacial syndromes, or sutures affected. The Study group had higher preoperative Hb (13.9 ± 1.0 vs. 12.6 ± 0.8 g/dL, $p < 0.001$), lower intraoperative Hb nadir (7.4 ± 1.8 vs. 9.2 ± 1.2 g/dL) lower intraoperative transfusion rate (66.7% vs. 100%, $p = 0.008$), lower postoperative transfusion rate (0% vs 28.6%, $p = 0.03$), and exposure to fewer unique units of packed red blood cells (0.7 ± 0.6 vs. 1.5 ± 0.9 units).

The protocol resulted in decreased transfusion needs. These results add valuable information to the growing body of work on transfusion reduction in craniosynostosis surgery [2)].

2019

30 consecutive cases of metopic synostosis treated over a 14-year period in the Plastic Surgery Department in Polanica-Zdrój, Poland. The data was evaluated using the patients' clinical records, and preoperative and postoperative photographs. The patients showed up on a follow-up visit at a median age of 9 years and were examined by an ophthalmologist and a neurologist. The surgical outcomes were evaluated according to the Whitaker classification. In 23 patients, remodeling and the advancement of fronto-orbital skull segments was performed at a median age of 18 months and in 7 milder cases, simple suturectomy or burring of the metopic ridge was sufficient.

According to the Whitaker classification, results were considered good to excellent (category I and II). Only 1 patient was included into category III. None of the examined cases were included into category IV, which would require a major craniofacial procedure, duplicating or exceeding the original operation. Neurological abnormalities were found in 12 cases and vision defects in 15 cases.

Trigonocephaly is currently the 2nd most common type of isolated synostosis. Surgical treatment based on Tessier's and Marchac's modified methods provides good results in patients at the age of about 12 months and prevents the consequences of ICP increase. Primary neurological and behavioral disorders may occur, despite corrective surgery [3)].

Shimoji and Taira, performed surgery on 15 patients (ten girls and five boys) who were all diagnosed as microcephalic during infancy. All patients presented clinical symptoms and evidence of short stature. Symptoms included mental retardation, language delay, hyperactivity, motor dysfunction, and self-mutilation (head banging). Head circumferences were > 2 standard deviations below the normal range for their sex and age at the time of surgery. All patients were diagnosed with mild trigonocephaly based on three-dimensional computed tomography (3D-CT). No abnormal findings could be observed in the brain of 14 patients, as assessed by magnetic resonance imaging (MRI). One patient showed brain atrophy. 3D-CT showed marked digital markings in all. Intracranial pressure (ICP) was measured under normocapnia. Increased ICP could be observed in 13 patients. Decompressive cranioplasty was performed in all patients. After surgery, evidence of enlargement of the head circumference could be observed in six patients. Some degree of intelligence problems remained. However, every patient made some improvement in at least one of the other symptoms.

They suggest that decompressive cranioplasty may be indicated in patients with mild trigonocephaly associated with microcephaly, if pre-surgical evaluation shows high ICP and no abnormal brain findings can be identified on MRI [4)].

2018

A total of 141 patients with single-suture metopic nonsyndromic craniosynostosis sutures were treated between 1998 and 2017 by endoscopically resecting the synostosed bone followed by postoperative custom cranial orthosis use. All data used in the case series were collected prospectively and stored in a secure database. A comprehensive literature review was performed that included all previous case series reporting common surgical performance measures. A statistical comparison of traditional open methods versus MIS techniques was performed with regard to age,

length of hospital stay (LOS), surgical time, estimated blood loss (EBL), and transfusion rate.

RESULTS: The mean age at the time of surgery in the current series was 4.1 months. The mean EBL was 33 ml (range 5-250 ml). One patient underwent an intraoperative blood transfusion and 5 underwent postoperative blood transfusion for a total transfusion rate of 4.3%. The mean operating time was 56 minutes. Ninety-eight percent of patients were discharged on the 1st postoperative day. The median size of the removed synostosed bone was 0.6 cm × 10 cm. The primary goal of achieving correction of the forehead deformity was obtained in 94% of the patients. One hundred eight patients presented with hypotelorism (76.6%). Those with a minimum 1-year follow-up achieved 99% correction (n = 97). Six patients younger than 1 year had not achieved correction at the time of follow-up (6%). There were no intra- or postoperative deaths. One patient had a temporary contact dermatitis to the helmet materials and 2 patients developed pseudomeningoceles, which were successfully treated with a lumbar drain and/or spinal tap. No patient required nor underwent a second surgical procedure. Regarding the previously published literature through 2014, the reported EBL in patients who underwent MIS versus traditional open methods was 54.7 ml versus 224 ml, respectively. The reported average age for patients undergoing MIS versus traditional open methods was 3.8 months versus 11.5 months. The average LOS for patients undergoing MIS versus traditional open methods was 1.7 days versus 3.7 days. The average reported surgical time for those undergoing MIS versus traditional open methods was 66.7 minutes versus 223.7 minutes. The transfusion rate for patients undergoing MIS versus traditional open methods was 22% versus 77%. All of the above differences demonstrated statistical significance.

CONCLUSIONS: The authors' team has safely and effectively performed 141 metopic craniosynostosis corrections over the past 19 years, with excellent outcomes. Literature review comparing metrics such as LOS, EBL, operating time, and transfusion rate demonstrates a statistically significant improvement in all commonly reported measurements. MIS techniques are safe and effective and should be offered to parents and patients as an option at craniofacial centers treating this condition [5)].

The specific aim of a study was to determine if the senior author's method of “hypercorrection” decreases relapse and the need for subsequent revisional surgery.

Patients who underwent operative correction of metopic craniosynostosis between 1988 and 2011 were reviewed. All patients underwent the “hypercorrection” technique performed by the senior author. Hypercorrection consisted of a fronto-orbital advancement of 2.5 to 3.5 cm and a concomitant hyperexpansion of bitemporal projection. Split cranial bone grafting ensured adequate coverage of the significantly expanded cranial vault. Only patients who had at least 5 years of follow-up were included for review of outcomes. Relapse was defined as recurrence of bitemporal constriction or lateral orbital retrusion, requiring surgical correction.

Fifty-eight patients met criteria. Mean age at the time of surgery was 11 months. Mean follow-up was 9.0 years. During this time, 2 patients exhibited relapse requiring camouflage procedures. Cranial bone defects were found in 4 patients (7%), 3 of whom underwent cranial bone grafting, while 1 underwent methylmethacrylate placement at an outside institution. One patient underwent fat grafting for areas of soft tissue irregularity. No patients exhibited persistent sequelae of hypercorrection significant enough to require repeat fronto-orbital advancement.

Surgical hypercorrection of trigonocephaly seems to minimize relapse and the need for revision in long-term follow-up and is therefore an important technique to consider [6)].

2017

Kalantar Hormozi et al. retrospectively evaluated the outcomes of the simplified horse-shoe technique with previous procedures performed on 169 children severe trigonocephaly during 1996 to 2015 at Mofid Hospital, Tehran, Iran. Demographic data of the patients and the surgical outcomes and complications and the need for reoperation were recorded and scored using the Whitaker system. The male-to-female ratio was 2.75:1. The mean ± standard deviation age of all the patients at the time of surgery was 7.09 ± 7.5, 9.95 ± 7.71, 10.53 ± 6.57, and 10.59 ± 7.96 months for the traditional, horse-shoe, and simplified horse-shoe techniques, respectively. The total reoperation rate was 6.5% (4.7% in the traditional technique, 1.2% in the horse-shoe, and 0.6% in the simplified horse-shoe technique) ($P < 0.001$). The main reason for reoperation was hardware removal (in the traditional technique). Whitaker scoring showed no patients of class IV in any of the groups and no patients of class III and IV in simplified horse-shoe technique, but the difference in the Whitaker scores among the 3 groups was not statistically significant ($P = 1.176$). The new surgical technique is easier and simpler with fewer complications than the traditional technique and is suggested to be recommended for surgical treatment in children with trigonocephaly, especially in younger children [7].

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72 patients with trigonocephaly who were operated on. Postoperatively, intelligence was assessed prospectively. The two independent variables, secondary mechanical and primary brain developmental mechanisms, were evaluated retrospectively. Computed tomographic imaging was used to assess skull volume and severity of the frontal stenosis (secondary mechanical factors), the width of the central part of the lateral ventricles, and other structural brain anomalies (primary brain developmental factors). Extracranial congenital anomalies were also taken into account.

No association was found between secondary mechanical factors and postoperative IQ score. Width of the central part of the lateral ventricles, and an interaction effect between this width and additional extracranial anomalies, showed a significant negative association with postoperative IQ.

Primary brain developmental disorders seem to play an important role in the development of cognitive problems in trigonocephaly. Assessment of width of the central part of the lateral ventricle scores and additional extracranial congenital anomalies for the early prediction of cognitive problems in patients with trigonocephaly could be clinically valuable and can be performed using routinely available tools [8].

During a period of 20 years (from 1996 to 2015) at the Pediatric department of the Clinic for Neurosurgery in Skopje, we observed 18 babies with trigonocephaly, including one with Carpenter syndrome and trigonocephaly, 14 males and 4 females. All children had simple trigonocephaly, one had syndromic trigonocephaly (Carpenter's syndrome). According to Oi and Matsumoto classification done in 19865 severe trigonocephaly is observed in 11 cases and, moderate trigonocephaly in 7 cases. Our method: Our treatment consisted of slightly modified Di Rocco's3 surgical procedure named “shell” operation, adding transposition of the “bone flap”.

The postoperative period was uneventful except for the expected forehead swelling. The babies were discharged from the hospital on average at the 8th postoperative day. At the three months control after the surgery, the head had excellent aesthetic appearance, with regular psychomotor development according to the age of the patient (Fig 3a and 3b). We had no serious complications

except the expected postoperative swelling of the forehead. All operated children had excellent “long term” aesthetic effect and normal psychomotor development.

The early recognition of these anomalies including all craniosynostoses, the deformities of the newborn and infant's head and the preventive operative reconstruction would prevent abnormal disturbance of the psychomotor development during the child's growth. The multidisciplinary approach can prevent new disabled individuals in the society. Our technique allows shortening the entire surgical procedure, especially in the departments where blood saving devices are not available [9)].

The aim of a study was to quantitatively assess aesthetic outcomes of FOR by capturing 3D forehead scans of metopic patients pre- and post-operatively and comparing them with controls. Ten single-suture metopic patients undergoing FOR and 15 age-matched non-craniosynostotic controls were recruited at Great Ormond Street Hospital for Children (UK). Scans were acquired with a three-dimensional (3D) handheld camera and post-processed combining 3D imaging software. 3D scans were first used for cephalometric measurements. Statistical shape modelling was then used to compute the 3D mean head shapes of the three groups (FOR pre-op, post-op and controls). Head shape variations were described via principal component analysis (PCA). Cephalometric measurements showed that FOR significantly increased the forehead volume and improved trigonocephaly. This improvement was supported visually by pre- and post-operative computed mean 3D shapes and numerically by PCA ($p < 0.001$). Compared with controls, post-operative scans showed flatter foreheads ($p < 0.001$). In conclusion, 3D scanning followed by 3D statistical shape modelling enabled the 3D comparison of forehead shapes of metopic patients and non-craniosynostotic controls, and demonstrated that the adopted FOR technique was successful in correcting bitemporal narrowing but overcorrected the rounding of the forehead [10)]

2016

Patients with anterior plagiocephaly or trigonocephaly were included. All had undergone coronal or metopic suturectomy with bilateral orbitofrontal bandeau resection between 1997 and 2005. The primary endpoint was the Whitaker classification. The secondary endpoints were anthropometric measurement, assessments of the bone defects on computed tomography scan, and the comments of patients and their relatives concerning the final skull outcome. Patients with anterior plagiocephaly also attended an ophthalmological consultation.

Seventeen patients were included in the study: 8 with anterior plagiocephaly and 9 with trigonocephaly. Mean age at the time of surgery was 6.91 months. Mean age at the time of craniofacial consultation was 14 years. Fourteen patients (82%) were classified as Whitaker Class III and IV, corresponding to poor esthetic results and persistent bone defects. Seven patients requested further surgery.

This study shows that suturectomy seems to yield poor esthetic results in the long term and patients should be followed up throughout adolescence to correct any craniofacial deformities [11)].

2014

A retrospective chart review of patients with metopic synostosis who underwent fronto-orbital advancement with pre- and postoperative STARscanner imaging at Texas Children's Hospital was performed. Two patients were identified and evaluation and discussion of the data produced by the STARscanner was undertaken. A novel symmetry index created by the authors, called the anterior-posterior volume ratio (APVR), was discussed for use in metopic synostosis patients. The postoperative growth metrics demonstrated an interval increase compared with the preoperative data. The anterior symmetry ratio, posterior symmetry ratio, overall symmetry ratio, cranial vault volumes, cranial vault asymmetry index, and cephalic ratio were not found to be useful in evaluating resolution of dysmorphology after fronto-orbital advancement in metopic synostosis. The APVR does not characterize dysmorphology, but may help show degree of expansion of the anterior cranial vault after fronto-orbital advancement. The STARscanner imaging device does not appear to have significant utility in characterizing head shape for surgical outcomes assessment in metopic synostosis. The minor utility of this device may be that it is a safe and fast way to derive growth parameters for both short-term and long-term follow-up of cranial vault remodeling [12].

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