Nonsyndromic craniosynostosis

Nonsyndromic craniosynostosis (NCS) is the premature ossification of cranial sutures, without associated clinical features.

It can be familial in which more than one of the family members are involved.

**Epidemiology**

Nonsyndromic craniosynostosis is significantly more common than syndromic craniosynostosis, affecting the sagittal, coronal, metopic, and lambdoid sutures in decreasing order of frequency. Nonsyndromic craniosynostosis is most frequently associated with only 1 fused suture, creating a predictable head shape.

In a prospective study which is carried out from April 2015 to January 2018 in 2 academic hospitals. Those patients who had nonsyndromic craniosynostosis and completed medical follow-up were included in the study as well as their 1st degree relatives. Age of patients, gender, existing consanguineous marriage, type of deliveries, type of pregnancy (assisted reproductive technologies [ART] versus sexual intercourse), severity and type of craniosynostosis were gathered.

Ninety-four (46.0%), 58 (28.4%), 28 (13.7%), 16 (7.8%), and 8 (3.9%) of patients had trigonocephaly, scaphocephaly, anterior plagiocephaly, complex, and brachycephaly, respectively. A total number of 204 patients were included in the study. Of all 204 families which were included, 30 (14.7%) families had positive familial history. Familial patients were determined in 10, 15, 8, 1, and 5 patients with scaphocephaly, trigonocephaly, anterior plagiocephaly, rachycephaly, and mixed type. Male to female ratio was 2:1, 1.9:1, 1.3:1, 1:1, and 1:1 for scaphocephaly, trigonocephaly, anterior plagiocephaly, brachycephaly, and mixed craniosynostosis. Twelve (5.9%) women had applied ART.

The study reveals that metopic suture is the most frequent craniosynostosis within nonsyndromic types. All the types of nonsyndromic craniosynostosis had male prevalence but for complex one which was equal in both gender. Nonsyndromic craniosynostosis in about 14.7% of patients was familial.

**Pathogenesis**

BBS9 gene in nonsyndromic craniosynostosis

**Clinical features**

The ocular and systemic abnormalities of nonsyndromic craniosynostosis are often considered to be less severe than those of syndromic craniosynostosis and are less well described. The nature of ophthalmic abnormalities in children treated for nonsyndromic craniosynostosis by expansion cranioplasty in a retrospective review identified 88 consecutive children with nonsyndromic...
craniosynostosis who underwent expansion cranioplasty with distraction osteogenesis. Assessment of presence and type of strabismus, refractive error, and amblyopia before and 6 months after surgery was recorded. Children with a mean age of 24.4 months were treated for nonsyndromic craniosynostosis (27 with coronal and 61 with sagittal and/or lambdoid). One-fourth of the patients had a fixation preference. Significant refractive errors were found in 45 (51%) of the 88 patients: hyperopia in 27%, myopia in 5%, and astigmatism in 35%. Anisometropia was present in 20%. Of the 85 patients who completed orthoptic examination, 48 (56%) had strabismus: exodeviation in 26%, esodeviation in 14%, and vertical deviation in 5%. Fourteen patients (16%) had abnormal head posture. Significant refractive error and strabismus were more likely to occur in cases with coronal synostosis. The procedures used for cranial vault expansion improved the abnormal head posture but did not affect the refractive error or ocular misalignment. Of children with nonsyndromic craniosynostosis who need neurosurgical correction, more than half were found to have significant refractive error and strabismus. These findings support the importance of ophthalmic evaluation in these children.

Diagnosis

Preoperative computed tomography (CT) evaluation of patients with nonsyndromic craniosynostosis (NSC) has focused on the bony cranial vault while ignoring the surrounding soft tissues.

CT-derived temporal muscle and temporal fat pad morphomics (tissue thickness, area, and volume) can be used to calculate temporal morphomic indices (TMIs), which are unique to each NSC subtype (metopic, coronal, and sagittal) and divergent from normal individuals.

High-throughput image analysis was used to reconstruct the 3-dimensional anatomy and quantify a TMI. These steps were completed in a semiautomated method using algorithms programmed in MATLAB v13.0. Differences in TMI across various craniosynostosis subtypes were assessed using Wilcoxon nonparametric tests for both patients with NSC and a control cohort of patients with trauma. Using preoperative CT images, the evaluation of 117 children with NSC from the University of Michigan Health System and 50 age-matched control patients between 1999 and 2011, indicate significant differences in TMI among the normal and NSC groups, with normal patients having significantly higher TMI values than patients with metopic, sagittal, and coronal synostosis. In addition, significant differences were found to exist between each craniosynostosis category.

Patients with craniosynostosis demonstrate diminished temporalis muscle and overlying fat pad volume and thickness compared with control patients. The unique changes in temporal morphomics demonstrate not only that the bony calvaria is affected by craniosynostosis but also that there exist quantifiable aberrations in the temporalis muscle and temporal fat pad. The methodologies described offer a novel methodology to use pre-existing CT scans to glean additional preoperative information on the soft tissue characteristics of patients with craniosynostosis.

Treatment

Repair of craniosynostosis is recommended to avoid potential neurodevelopmental delay. Early intervention at 3 to 4 months of age allows minimally invasive approaches, but requires postoperative molding helmet therapy and good family compliance. Open techniques are deferred until the child is
older to better tolerate the associated surgical stress. Cranial vault remodeling is generally well-tolerated with a low rate of complications 5).

Healthy patients with NSC undergoing cranial vault surgery can potentially be safely managed without routine admission to the ICU postoperatively. Key elements are proper preoperative screening, access to ICU should an adverse intraoperative event occur and necessary postoperative surgical care. The authors hope that this experience will encourage other craniofacial surgeons to reconsider the dogma of routine ICU admission for this patient population 6).

Outcome

Public insurance and nonwhite race/Hispanic ethnicity were statistically significant predictors for older age at surgery, adjusted for sex, zip code median family income, year, and hospital factors such as size, type, region, and teaching status. Further research into these disparities is warranted 7).

Unclassified


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