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 [1)].

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 [2)].

Pathogenesis

Nonsyndromic Craniosynostosis Pathogenesis.

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 [3].

Diagnosis

Nonsyndromic Craniosynostosis Diagnosis.

Treatment

Nonsyndromic craniosynostosis treatment.

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 [4].

Case series

A retrospective chart review was performed for patients with nonsyndromic craniosynostosis who underwent minimally-invasive nonendoscopic suturectomy between 2019 and 2020.

Thirteen patients (11 males; 2 females) were operated including 5 Metopic, 5 Sagittal, 2 coronal, and 1 lambdoid craniosynostosis. The average age at surgery was 4.35 months. The average length of surgery was 71 minutes. Averaged intraoperative estimated blood loss was 31.54 mL. Eleven patients received a blood transfusion (most before performing the skin incision) with a mean amount of 94.62 mL of blood transfused during surgery. The mean hemoglobin at discharge was 10.38 mg/dL. There was only 1 intraoperative mild complication. The mean intrahospital length of stay was 1.77 days with no postoperative complications noted. All patients initiated remodeling orthotic treatment following surgery. Long-term follow-up scans were available for 8 patients (5 metopic, 2 sagittal, and 1 lambdoid) with an average follow-up of 9 months. In all cases, there was a significant improvement in

the skull width at the synostosis location as well as in the skull proportions and symmetry. The above outcomes are similar to those published in the literature for endoscope-assisted strip-craniectomies.

Suturectomies assisted with cranial orthosis remodeling for the treatment of all types of nonsyndromic craniosynostosis can be performed without an endoscope while maintaining minimal-invasiveness, good surgical results, and low complication rates [5)].

Unclassified

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