CRANIOFACIAL ABSTRACTS

Intracranial Hypertension and Cortical Thickness in Syndromic Craniosynostosis

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INTRODUCTION: Intracranial hypertension (ICH) is a frequent indication for surgical intervention in syndromic craniosynostosis. Various clinical risk factors have been described, but potential effects on underlying brain morphology have not been investigated. This study seeks to evaluate the impact of ICH risk factors on cerebral cortex thickness in syndromic craniosynostosis.

METHODS: Patient records and imaging were reviewed for ICH risk factors and demographic data including papilledema, hydrocephalus, moderate to severe obstructive sleep apnea, cerebellar tonsillar position, occipitofrontal circumference curve deflection, age at the time of scan, and sex in 107 syndromic (Apert, Crouzon, Pfeiffer, Muenke, Saethre-Chotzen) craniosynostosis patients. One hundred seventy-one magnetic resonance imaging scans of these patients were then analyzed. Average cortical thickness estimates were obtained via an auto-segmentation/auto-parcellation image processing software (FreeSurfer) and exported for statistical analysis. A linear mixed-effect model accounting for repeated measurements, age, gender, and syndrome influences was developed to determine impact of ICH risk factors on cerebral cortex thickness changes (significance P < 0.05).

RESULTS: Average cortical thickness in this cohort was 2.78 ± 0.17 mm with an average age of 8.88 years (range, 1.15–34.03) at the time of scan. Cortical thickness did not vary significantly by sex (P = 0.534) or syndrome (P = 0.896) as independent predictors. A history of papilledema (P = 0.036) or hydrocephalus (P = 0.007) before scan date was associated with thinner cortices than those without. Average cortical thickness was also shown to significantly vary with the age of the patient at the time of magnetic resonance imaging (P < 0.001), with older patients having thinner cortices. History of moderate to severe obstructive sleep apnea (oAHI > 5) (P = 0.464), cerebellar tonsil position (P = 0.682), or history of occipitofrontal circumference curve deflection (P = 0.375) before scan date did not result in significant cortical thickness changes.

CONCLUSIONS: Our results indicate that a history of hydrocephalus or papilledema results in a thinner cerebral cortex on average in syndromic craniosynostosis patients. This suggests structural consequences from the development of ICH and may support early intervention to avoid such effects. Further investigation is needed to evaluate the link between these findings, timing of intervention, and neuropsychological development.

Increasing Incidence of Craniosynostosis in the United States: Is Folic Acid Supplementation Responsible?

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PURPOSE: Craniosynostosis, the premature fusion of cranial sutures, has increased in both prevalence and incidence as reported by international studies.1,2 To our knowledge, no recent studies have evaluated increasing incidence in the United States; therefore, we sought to evaluate if there was a significant increase in our national incidence of craniosynostosis. Methotrexate, a folic acid antagonist, has been associated with an increase in craniosynostosis.3 There has been a decrease in the incidence of cleft anomalies following the implementation of the folic acid supplementation program in 1998 within the United States. Both of these anomalies seem affected by folate. We hypothesize that there is a reciprocal relationship between cleft and craniosynostosis and seek to investigate the theory that as folate supplementation penetrates the population, we see a gradual increase in the incidence of craniosynostosis.

METHODS AND MATERIALS: The National Inpatient Sample Database was consulted to identify infants born with craniosynostosis between 2004 and 2013. Data were collected from the US Center for Disease Control and Prevention, including incidence of influenza virus infection according to year and month. Using multivariable logistic regression, we examined the relationship between craniosynostosis and the independent variables month and year. We then utilized mixed-effects logistic regression to estimate the odds ratio of occurrence of craniosynostosis in
relation to previous months’ flu incidence. E values were calculated to evaluate for unmeasured confounders.

RESULTS: In 2004, there were 4,110 infants born with craniosynostosis, which increased to 6,155 infants in 2013. A statistically significant increase in the incidence of craniosynostosis within the United States was found (odds ratio of 1.57 in 2013; \( P < 0.001 \)). Mixed-effects logistic regression revealed a lower incidence of craniosynostosis associated with an increased incidence of influenza infection. E values for national incidence of craniosynostosis and association with influenza incidence were 2.51 and 11.6, respectively.

CONCLUSIONS: To our knowledge, this is the first study demonstrating a significant increase in the national incidence of craniosynostosis in the United States, which we believe may be a result of folic acid supplementation penetrating the population. We also report for the first time a decreased incidence of craniosynostosis in association with influenza incidence, which support our hypothesis of a possible inverse relationship with cleft, as maternal influenza during pregnancy demonstrates increased incidence of cleft anomalies. We are further investigating the relationship between cleft and craniosynostosis at this time to uncover a mechanism that might explain this relationship.

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Correcting Orbital Hypertelorism With Supraorbital Bipartition Osteotomy: Technique and Advantages

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INTRODUCTION: Orbital hypertelorism can exist in a variety of craniofacial anomalies such as midline anterior encephaloceles, frontonasal dysplasia, and syndromic bicoronal craniosynostosis. Facial bipartition corrects hypertelorism and benefits patients with a narrow, V-shaped maxilla. However, in young children with hypertelorism, there is a higher risk of injury to dental follicles. The supraorbital bipartition allows for correction hypertelorism in this younger population of patients undergoing frontal craniotomy without the need for osteotomies extending into tooth-bearing segments of the maxilla.

MATERIALS AND METHODS: The supraorbital bipartition technique was performed in 15 patients with hypertelorism. Of these, 3 patients had associated meningoencephaloceles, 5 patients had facial clefting, and 7 patients had hypertelorism associated with Crouzon or Apert syndrome. All patients underwent preoperative evaluation by neurosurgery, ophthalmology, and pediatrics. Neuropsychiatric testing and preoperative computerized tomography scans were performed. The technique, advantages, and complications are described.

RESULTS: The patient age ranged from 8 months to 8 years old, with a mean of 40 months. Seven patients were female, and 8 were male. All cases were uneventful. The interorbital distance was normalized for age in 11 cases. The remainder 4 cases had dramatic improvement in interorbital distance. Blood loss ranged from 250 to 600 ml, with mean EBL of 350 ml. Blood transfusion was required in 12 patients. No major complications occurred. In 4 cases, unilateral detachment of the medial canthal ligament occurred. In one case, bilateral detachment of the medial canthal ligament occurred. In all cases, these detachments were repaired intraoperatively. Two cases had minor wound dehiscence that healed with local wound care.

CONCLUSIONS: The classical techniques for management of hypertelorism entail either complete bilateral orbital osteotomies and translocation or facial bipartition. These techniques are not suitable for younger patients given the presence of tooth buds before the eruption of permanent dentition. In the proposed technique, the infraorbital osteotomy was avoided, thus sparing the developing tooth buds. The rate of complication of the present technique is lower than in the other techniques, with no major complications. The improvement in interorbital distance is comparable to that obtained with classical techniques.

Minimally Invasive Migraine Surgery: Our 9-year Experience

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