Behavioral, developmental, and educational problems in children with nonsyndromic trigonocephaly

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Object. The neurobehavioral morbidity of nonsyndromic trigonocephaly is incompletely understood. The purpose of this study was twofold: first, to assess the degree of developmental, educational, and behavioral problems in patients with nonsyndromic trigonocephaly and second, to establish whether patients with mild degrees of trigonocephaly had a lower frequency of such problems.

Methods. The authors performed an observational study of the frequency of developmental, educational, and behavioral problems in 63 children with trigonocephaly at the National Craniofacial Centre in the Republic of Ireland between 1989 and 2004. The parents of the children completed a follow-up questionnaire. Thirty percent of patients had a mild form of trigonocephaly and were treated conservatively. The remainder underwent surgical correction. Speech and/or language delay was reported in 34% of the children. Thirty-three percent of the children needed to be assessed by a school psychologist, and 47% were receiving remedial or resource hours within the school system. Twenty percent of children required a special needs classroom assistant because of behavioral issues, and 37% of parents expressed concerns about their child’s behavior. There were no statistically significant differences between children treated with surgery and those who had a mild deformity and were treated conservatively.

Conclusions. Nonsyndromic trigonocephaly is associated with a high frequency of developmental, educational, and behavioral problems. The frequency of these problems is not related to the severity of the trigonocephaly.

KEY WORDS • craniosynostosis • developmental delay • pediatric neurosurgery

MONOSUTURAL metopic synostosis constitutes 7 to 23% of all craniosynostotic disorders, and its cause and pathogenesis are poorly understood. The phenotypic features of trigonocephaly vary from mild, with slight prominence of the metopic ridge, to severe, with gross distortion of the forehead, supraorbital bar, and orbits. Surgery is often required for cosmetic correction of the resultant deformity.

The majority of the literature on trigonocephaly contains descriptions of surgical techniques or cosmetic outcomes. Over the last 10 years, there has been increasing interest in the associated cognitive, behavioral, and developmental problems that often occur in association with craniosynostosis. It has been suggested that early surgical release of craniosynostosis might increase mental abilities. However, a recent critical review of the literature did not show any evidence to support the theory that surgery prevented or reduced the long-term risk of neurobehavioral impairment.

There is now an increasing body of evidence suggesting that children with nonsyndromic trigonocephaly might have a higher risk of cerebral impairment. Bottero, et al., reported that 31.6% of the patients in their study had some degree of impairment, although the study included children with nonsyndromic trigonocephaly as well as those with associated anomalies. In similar studies, 50% of children with single suture sagittal craniosynostosis were found to have reading or learning disabilities. It is estimated that isolated craniosynostosis is associated with a three- to fivefold increase in the risk of cognitive deficits or learning and language difficulties. Although the causal relation between this condition and neurodevelopment is unknown, it would appear that these types of calvarial abnormalities are at the very least a visible and easily diagnosable marker for an increased risk of neurodevelopmental problems.

Clinical Material and Methods

The National Craniofacial Surgical Unit at The Children’s University Hospital Dublin is a tertiary referral center for all patients with craniofacial abnormalities in the Republic of Ireland and serves a catchment population of approximately 3.5 million. All patients with nonsyndromic trigonocephaly who were assessed and treated by the senior authors between 1989 and 2004 were included in this study. The charts of patients were reviewed, and patients and/or
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their parents were asked to complete a follow-up questionnaire. The questionnaire included items pertaining to developmental milestones of speech and walking; assessment of the degree of help the child needed in the school setting (Table 1); and what behavioral issues, if any, were apparent.

Examination of the charts indicated that each patient had undergone a comprehensive, multidisciplinary evaluation by a craniofacial team. Follow-up assessments were multidisciplinary and occurred annually for the first few years, and biannually thereafter. Since 2000, a craniofacial coordinator has acted as a liaison with the families and has provided a periodic telephone follow-up service.

Statistical analysis was performed using a standard statistical package (JMP; SAS Institute, Cary, NC). The Pearson nonparametric test was used to evaluate the hypothesis that children with mild degrees of trigonocephaly had educational and behavioral problems similar to those of children with more severe deformities.

Results

Sixty-three patients with nonsyndromic trigonocephaly (15 girls and 48 boys) were referred to the unit. Two families were lost to follow-up and did not complete a follow-up questionnaire. A family history of craniosynostosis was noted in 9% of the children. Thirty percent of the patients had a mild form of trigonocephaly and were treated conservatively. The remainder underwent surgical correction. The median age at surgery was 12 months (range 4–144 months). Speech and/or language delay was reported in 34% of the children.

Thirty-three percent of the parents reported a developmental delay in their child. Thirty-seven percent of parents expressed concerns about their child’s behavior, reporting attention deficit disorder, autism, and hyperactivity.

Of the 63 children, 42 were attending school at the time of this study, and thus only this number was available for assessment of school progress. The majority (90%) attended mainstream schools; the remaining 10% attended special needs schools or were taught in separate classrooms. Nevertheless, 10% of children who attended mainstream schools had been required to repeat at least 1 year. Forty percent of the parents expressed concerns about their child’s learning ability. Thirty-seven percent of the children had specific needs with regard to reading and writing, and 33% had been assessed by a school psychologist. Forty-seven percent of the children were receiving remedial or resource hours within the school system. Twenty percent required the presence of a special needs classroom assistant because of behavioral issues. Preoperative computed tomography imaging did not reveal any evidence of frontal lobe abnormalities in this cohort.

There was no statistically significant difference in developmental, educational, or behavioral domains between patients who underwent surgery (70%) and those who had a mild deformity and were treated conservatively (30%).

Discussion

Until the last 10 years, nonsyndromic craniosynostosis was considered a morphological disorder devoid of functional morbidity. This was largely because infants rarely exhibit signs or symptoms of neurological impairment before or shortly after craniosynostosis surgery.4 Earlier studies were limited because they focused on “mental retardation” as evidence of primary neurological impairment.4 The authors of these studies ignored the more subtle nuances of brain dysfunction, such as learning difficulties or behavioral issues. Authors who previously tried to find a linkage between synostosis and neurobehavioral functioning usually assumed a direct pathway in which suture fusion leads to brain deformation and consequent neuropsychological impairment.27 However, there are insufficient data to support a hypothesis regarding a particular causal pathway.23

In the general pediatric population, developmental delay is estimated to be present in approximately 10% of children.6 This figure is significantly less than that seen in the present cohort and in previous studies.4,7,23 Cognitive and/or behavioral abnormalities have also been found in a large number of patients with trigonocephaly.4,22 These impairments are often not fully appreciable until the children reach school age.22

In the present study, children with nonsyndromic trigonocephaly were found to be two-and-a-half times more likely to need remedial or resource hours than healthy children (47 and 19%, respectively), and four times more likely to be referred to a psychologist (33 and 7%, respectively).10 This cohort also exhibited a fourfold increase beyond the 5 to 10% incidence of attention deficit disorder and other behavioral disorders reported in the general pediatric population.6

With few exceptions, the neurobehavioral assessment of craniosynostosis has been focused on a single index of global functioning.21 Global indexes are less likely to detect aberrant development than the assessment of multiple domains of functioning.21 In assessing the present cohort, we chose to perform an observational study of behavioral and educational needs described by the children’s parents, as opposed to subjecting them to a battery of psychometric or language tests. We believe that the results obtained in this way are more tangible and easier to explain to the family of a patient in whom trigonocephaly has recently been diagnosed, as they seek information on the possible problems their child might face. Overall, the results of our study are in concurrence with those of earlier published studies that have suggested that a large proportion of children with nonsyndromic trigonocephaly experience learning disorders and behavioral problems as they become older.2,4,7,8,14,17,19,22

Because a large portion of this patient population experiences learning and behavioral difficulties, it seems prudent to screen these children proactively as part of a multidisci-
plinary approach, rather than allow time to elapse until it be-
comes obvious that the children need help and have already
fallen behind.

Some authors have suggested that more severe forms of
trigonocephaly might be associated with higher levels of
cognitive dysfunction. 7 Bottero, et al., found developmen-
tal delay in 31.6% of patients who required surgery (syn-
dromic and nonsyndromic), compared with 23% in those
who had a mild form of trigonocephaly and did not require
surgery. 7 In our study, there was no statistical difference be-
tween those patients who underwent surgery and those who
were treated conservatively.

Conclusions

A significant number of children with trigonocephaly ex-
perience developmental, educational, and behavioral prob-
lems as they become older. The incidence of such problems
within this population appears to be independent of the se-
verity of the cosmetic deformity.

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