Prognosis for mental function in scaphocephaly

ERIC ARNAUD, M.D., DOMINIQUE RENIER, M.D., AND DANIEL MARCHAC, M.D.

Unit of Craniofacial Surgery, Department of Neurosurgery, Hôpital Necker-Enfants Malades, Paris, France

... Three hundred ninety-six children with scaphocephalies were prospectively studied to analyze the correlation between age, intracranial pressure (ICP), and mental function outcome. The ICP measurements and the early and late psychometric assessments were compared. The influence of surgery, when performed, was analyzed. In most cases, the mental function outcome of the patients was good whether or not they had undergone surgery. The mental level and the frequency of increased ICP both correlated with patient age. A correlation was found between the early and late psychometric assessments in all patients. Thus, the main predictive factor of mental function outcome appears to be the initial developmental level.

KEY WORDS • craniosynostosis • scaphocephaly • craniofacial surgery • children

SCAPHOCEPHALY, or premature closure of the sagittal suture, is rarely associated with increased intracranial pressure (ICP) or mental retardation, as are most of the single craniosynostoses. For the past decade, surgery has been performed on scaphocephalies for both functional and morphological reasons. Because of the tremendous decrease in the mortality and morbidity associated with modern craniofacial surgery techniques, remodeling of the scaphocephalic skull can be performed early in infancy in almost all cases, with minimal risks. The indication for surgery for scaphocephaly remains controversial. Some authors discuss a possible deterioration of mental function if surgery is withheld, whereas others describe the absence of deterioration when nonsurgical care is provided.

Scaphocephalies are the most common (42%) of the craniosynostoses seen in our unit. Our observation that several nonsurgically treated scaphocephalics (some with increased ICP) showed excellent mental development led us to question whether ICP is an important prognostic factor of mental function outcome, as we have previously considered for this defect. Therefore, the purpose of this study was to evaluate the relationship between intelligence development and ICP in surgically and nonsurgically treated scaphocephalic children.

Clinical Material and Methods

Patient Selection

Between 1976 and 1991, 396 scaphocephalies were prospectively studied. The following data were considered: ICP and development quotient (DQ) prior to any treatment, and final intelligence quotient (FIQ) assessment in both surgically and nonsurgically treated patients later in the follow-up period.

The ICP measurements were systematically performed in the first half of the series. They were obtained from a 12-hour continuous recording, according to the method described by De Rougemont. This method uses a saline-filled epidural sensor that is connected through the scalp via a catheter to an extracranial transducer (Bentley Transtec model 800, Kontron S. A., Trappes, France, or model 1280; Hewlett-Packard, Orsay, France). Recordings were made for 12 hours or more, encompassing the entire night, because the highest and lowest values of ICP are known to be recorded during the sleep-waking cycle. Intracranial pressure was recorded as abnormally high when its baseline value (corresponding to slow-wave sleep) was greater than or equal to 15 mm Hg for at least 12 hours. When the baseline ICP was elevated, a sustained wave of increased ICP was recorded during each period of rapid eye movement sleep in almost all cases.

The level of mental functioning in each child was assessed using IQ or DQ tests. The developmental Brunet–Lézine scale was used for children less than 2 1/2 years of age. For those between 2 1/2 and 3 years old, the complementary tests and the nonverbal scale of Brunet–Lézine were used. Children more than 3 years old were studied with the Nouvelle Échelle Métrique de l’Intelligence, a revision by Zazzo, et al., of the Binet–Simon test. When there were language difficulties, the performance test of the Wechsler Intelligence Scale for Children was used. The DQ and FIQ assessments were conducted by the same pediatric psychology team. These
Mental function prognosis in scaphocephaly

TABLE 1

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<tr>
<th>Age</th>
<th>DQ†</th>
<th>ICP†</th>
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<tr>
<td>&lt;1 yr</td>
<td>&gt;1 yr</td>
<td>&lt;90</td>
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<tr>
<td>≥15</td>
<td>7</td>
<td>12</td>
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</tbody>
</table>

* DQ = development quotient; ICP = intracranial pressure.
† For the correlation between DQ and ICP, chi square = 10.033; p = 0.0015 (Pearson’s chi square test). For the correlation between DQ and ICP, p = 0.166 (Fisher’s exact test).

Correlation Between ICP and DQ Before Any Treatment

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<tr>
<td>&lt;1 yr</td>
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Correlation Between ICP and DQ

One hundred forty-two patients had both ICP recording and DQ assessments prior to any treatment. The mean age at testing was 360 days (range 73 to 1069 days).

Correlation Between ICP and FIQ

Ninety-eight patients with ICP recording at first evaluation had an IQ assessment at their most recent review. One group of 51 patients included the nonsurgically treated scaphocephalies (mean age at recording, 9 months; mean age at review, 5.5 years). The other group of patients included surgically treated scaphocephalies (mean age at recording, 6.4 months; mean age at surgery, 10 months; mean age at review, 6.4 years).

Correlation Between DQ and FIQ

One hundred forty-two patients had both preliminary DQ and FIQ assessments. One group of 41 included the nonsurgically treated patients. The other group of 59 included the surgically treated cases. The mean age at first consultation was 9 months in both groups, and the age at last review was 6 and 6.4 years, respectively. Surgery was performed at a mean age of 11 months in the second group.

Results

Correlation Between ICP and FIQ in Surgically and Nonsurgically Treated Patients

The mean ICP in the nonsurgically treated patients was 10.3 mm Hg (standard deviation (SD) 3.3), ranging from 3 to 22, and in the surgically treated patients was 10.3 mm Hg (SD 2.8), ranging from 5 to 20. There was no statistical difference in ICP for the surgically and nonsurgically treated patients (Student’s unpaired t-test). The mean FIQ in the nonsurgically treated patients was 106 (SD 12), ranging from 76 to 135, and in surgically treated patients was 105 (SD 12), ranging from 77 to 141. In both groups, the ICP was not correlated to the FIQ assessments (Spearman’s test, and Pearson’s chi square, Table 2). Because of the small number of children with mental retardation or intracranial hypertension, the influence of age in this correlation could not be analyzed.

Correlation Between DQ and FIQ in Surgically and Nonsurgically Treated Patients

In the nonsurgically treated patients, the mean DQ was 105 (SD 8), ranging from 80 to 125, and the mean FIQ was 106 (SD 12), ranging from 76 to 136. In the surgically treated patients, the mean DQ was 101 (SD 12), ranging from 50 to 120, and the mean FIQ was 103, ranging from 44 to 145 (SD 17). In each group, there was a sig-

Statistical Analysis

Statistical significance was analyzed using commercially available software (SPSS for Windows, Version 5.0; SPSS Inc., Chicago, IL; and Systat for Windows, Version 5; Systat Inc., Evanston, IL). The Spearman test was used for correlation studies. The comparison between data was performed with the Student t-test (paired or unpaired), Pearson’s chi square and Fisher’s exact test were used when splitting the data within groups. The results were considered significant when probability was less than 0.05.
ICP showed abnormal clinical signs. Gordon et al. reported analyzing 18 cases of scaphocephalies, Hemple, et al., noted no mental impairment in a series of 23 scaphocephalies. Scaphocephaly have rarely been considered as having excessive resistance. These nonsurgically treated scaphocephalies of the surgical team's advice, mostly because of parental resistance. Nevertheless, some cases were not treated surgically regardless of the surgical team's advice, mostly because of parental resistance. These nonsurgically treated scaphocephalies subsequently had normal development in the majority of cases, but one may wonder whether absence of surgery might cause impaired psychological evolution. As with most of the single craniosynostoses, patients with scaphocephaly have rarely been considered as having an excessive ICP, and it has been reported previously that increased ICP occurred in 7% of scaphocephalies. In this study, we report a higher rate, approximately 13%.

In the literature, the conclusions vary between absolute necessity for surgery to absence of risk to mental function impairment. To evaluate the mental outcome in the scaphocephalic children, we analyzed the relationship between age, ICP, and intellectual development as measured by psychometric assessments. In our unit, surgery was routinely performed for the major deformities and for those patients with increased ICP. However, surgery was often performed for minor deformities as well. Nevertheless, some cases were not treated surgically regardless of the surgical team's advice, mostly because of parental resistance. These nonsurgically treated scaphocephalies subsequently had normal development in the majority of cases, but one may wonder whether absence of surgery might cause impaired psychological evolution. As with most of the single craniosynostoses, patients with scaphocephaly have rarely been considered as having an excessive ICP, and it has been reported previously that increased ICP occurred in 7% of scaphocephalies. In this study, we report a higher rate, approximately 13%.

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In our study, we used well-known psychometric tests with standard development scores, as well as ICP recordings. When dividing the groups by categories of age, we chose 1 year as a limit because the craniosynostosis was congenital and therefore, 1 year was the age of maximum conflict between cerebral growth and skull. When comparing ICP and mental level (DQ) prior to any treatment, little correlation was noted. Mental retardation was found in 16% of the patients with increased ICP, and in 6% of the group with normal pressure. Although the difference is not significant (p = 0.17), this may be considered an important trend. Mental development seemed to be correlated with the age at which the patient presented for initial consultation. This could be a bias: the parents of the patients who consulted late (after 1 year of age) were generally concerned by the child’s altered developmental evolution rather than by the morphological deformity, which they had always considered normal. Only when developmental or behavioral problems appeared did they seek consultation. However, there was more abnormally high ICP in the group of children older than 1 year (20% vs. 2% before 1 year). The vast majority of scaphocephalic children in this series have normal mental function (92%) and normal ICP (87%). However, the children with delays were significantly older and had increased ICP significantly more often than the normal ones. Therefore, it must be concluded that scaphocephaly is a condition not completely without functional consequences.

Analyses correlating ICP and FIQ, and DQ and FIQ confirmed the good mental function outcome of nonsurgically treated scaphocephalies and of those that were surgically treated. The influence of surgery on psychological development was studied through comparisons between early and late mental function level in nonsurgically and surgically treated patients. The Student paired t-test showed no improvement between DQ and FIQ in surgically or nonsurgically treated patients. A correlation was found between the preliminary and the FIQ assessments for both surgically and nonsurgically treated patients. This correlation would tend to indicate that the preliminary evaluation of mental function could be the main predictive factor for outcome, especially in surgically treated cases (p < 0.001, r = 0.64). In fact, a greater alteration in initial DQ necessitated surgical intervention, which could bias the significance of our results. In these cases, the surgery did not improve the FIQ, as one might expect if ICP had been the main concern. Even surgery by itself as a stimulating factor on the mental function outcome, by its additional reinforcement of the relationship between parent and child, did not improve the prognosis.

In the nonsurgically treated group, there were four patients with increased ICP recordings who experienced very good mental development. On the other hand, two patients with normal ICP showed impaired evolution. These two children had neonatal hypoxia or difficult deliveries, as in the series published by Hemple, et al., and Hunter. These two special groups are obviously too small to have statistical relevance.

**Conclusions**

Because the mental function level was worse in the old-

<table>
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<tr>
<th>ICP (mm Hg)</th>
<th>Non Surgically Treated</th>
<th>Surgically Treated</th>
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<tbody>
<tr>
<td>&lt;15</td>
<td>FIQ &lt;90 45</td>
<td>FIQ &lt;90 38</td>
</tr>
<tr>
<td>≥15</td>
<td>0 4</td>
<td>0 4</td>
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*ICP = intracranial pressure; FIQ = final intelligence quotient. Spearman and Pearson chi square tests revealed no significance.
pler children, and because the surgery does not influence the functional outcome when the initial DQ is low, we can conclude that the indication to perform surgery in scaphocephaly is sometimes not only to correct a cosmetic problem.

On the other hand, one must consider the psychic trauma of children with deformed heads.2,7 Barritt, et al.,2 have extensively discussed this point in an excellent paper, pointing out that increased ICP and restriction of brain growth are not the only way for scaphocephaly to impair mental development. Pediatricians and neurosurgeons should take into account potential emotional disturbances when discussing the surgical indication with parents.

References
8. Hunter AGW, Rudd NL: Craniosynostosis. I. Sagittal synostosis; its genetics and associated clinical findings in 214 patients who lacked involvement of the coronal suture(s). Teratology 14:185–194, 1976