Intracranial pressure in craniostenosis

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In this study, intracranial pressure (ICP) was recorded with an epidural sensor for periods of 12 to 24 hours in 92 cases of craniostenosis. Pre- and postoperative recordings were performed in 23 patients, and 55 children underwent preoperative psychometric testing. The ICP was found to be normal in one-third of the cases, was obviously elevated in one-third, and was borderline in one-third. Waves of increased ICP were recorded during rapid eye movement (REM) sleep. After surgery, ICP decreased progressively and returned to normal in several weeks. A significant statistical relationship was found between the patients' ICP and their mental level: the higher the ICP the lower the mental level. The regression curve of ICP as a function of age shows that ICP is maximal at the age of 6 years and decreases later. The significance of these results is discussed. The authors recommend that ICP be recorded in cases of craniostenosis since it is of some help in deciding whether patients should undergo surgery.

KEY WORDS • craniofacial dysostosis • craniostenosis • intracranial pressure • epidural pressure sensor

The need for surgery is obvious in many cases of craniostenosis, either because the patient's family is motivated by the aesthetic problem or because the child presents clinical signs of intracranial hypertension. In other cases, especially when only one cranial suture is involved or when the child is over 3 or 4 years old, it is more difficult to make a decision whether to operate, because it is then usually impossible to infer increased intracranial pressure (ICP) from the clinical features alone. Moreover, in craniostenosis, true papilledema is uncommon; exaggerated digital markings occur very late and they do not always correlate with increased ICP at the time of examination. Furthermore, since any increase of ICP would be moderate, if it were found, it may or may not be related to the functional long-term result.

Few studies deal with the measurement of ICP in craniostenosis. Most of these reports are of small series of patients, in whom very often ICP was monitored for only short periods of time.

In the present study, ICP was monitored for more than 12 hours in 92 cases of craniostenosis. Our objective was to answer two questions: 1) in cases of untreated craniostenosis, are the neuropsychological disorders related in any way to a chronic moderate increase of ICP, and 2) is ICP modified after surgery? Indeed these questions must be answered before ICP monitoring can be considered helpful in making a decision regarding surgery in craniostenosis.

Clinical Material and Methods

This series includes 92 children, ranging in age from 6 weeks to 15 years, who had the premature fusion of one or several cranial sutures. The age distribution in this series is given in Fig. 1 and types of craniosynostosis in Table 1. These children were all admitted to our institution between July, 1979, and March, 1981. Outpatients have been excluded.

Intracranial pressure was monitored in 75 children; 58 of these were operated on. In most cases, craniofacial techniques were used. In 23 patients, ICP was measured postoperatively, once 15 days after the operation and a second time 3 to 6 months later. In 17 other children, who had previously been operated on for craniosynostosis, ICP was recorded several years later because the recurrence of intracranial hypertension was suspected. The classical surgical techniques (free flaps, extensive craniectomies) were used in most of these cases.
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The ICP was recorded by the method described by de Rougemont. This method uses an epidural sensor filled with saline which is connected through the scalp by a catheter to an extracranial 800 Bentley Trantec transducer or an HP 1280 Hewlett-Packard transducer. These recordings lasted more than 12 hours, and always through the entire night, since the highest and lowest values of ICP are recorded during the sleep-waking cycle.

Whenever possible, the mental level of the child was tested by intelligence quotient (IQ) or development quotient (DQ). The development Brunet-Lézine scale was used for children aged less than 2 years. For those aged between 2 and 3 years, the complementary tests and the nonverbal scale of Brunet-Lézine were used. Children over 3 years old were studied with the NEMI (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 (WISC) was used.

Results

Slow-Wave and REM Sleep Recordings

Two ICP values can be defined on sleep recordings: the ICP value during slow-wave (SW) sleep, and the waves of increased ICP recorded during rapid eye movement (REM) sleep. In most cases of craniosynostosis, when the SW sleep ICP is elevated, a sustained wave of increased ICP is recorded during each period of REM sleep. Sometimes, a plateau wave with a sudden onset and an abrupt ending is recorded (Fig. 2 lower). In other cases, the wave has a fast onset, immediately followed by a slow return of ICP to the baseline (Fig. 2 center). In these two types, phasic variations of ICP are superimposed on the sustained wave. A third type of recording was found, usually when SW sleep ICP was normal. In this case, only the phasic variations of ICP were recorded; the sustained wave was missing or of a small amplitude (Fig. 2 upper). This last type is considered normal because SW sleep ICP is not elevated, a finding that correlates well with studies in normal animals. The mean duration of the sustained wave was 11 minutes and that of the phasic variations of ICP about 1 minute. The average time between two REM sleep periods was 70 minutes; the number of REM sleep waves of increased ICP varied from four to eight during the night. The relationship of these waves of increased ICP to REM sleep, already proven in hydrocephalus, has been confirmed in craniosynostosis by polygraphic recordings (electromyography, electro-oculography, electroencephalography, and ICP monitoring) in eight patients (Fig. 3).

Preoperative ICP Recordings

Preoperative recordings were grouped into three categories (Table 2). In 23 patients, the ICP was obviously increased. The ICP recorded during SW sleep was above 15 mm Hg, with a mean ICP of 21 mm Hg. The mean ICP at the peak of REM sleep waves was 48 mm Hg. These waves of increased ICP

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* Epidural sensor, Enfants Malades 8624-10, distributed by Plastimed, 95320 Saint-Leu La Forêt, France.
† Bentley Trantec 800 transducer manufactured by Kontron S.A., 78194 Trappes Cédex, France; HP1280 transducer manufactured by Hewlett-Packard, 91401 Orsay Cédex, France.

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TABLE 1

Types of craniostenosis in 92 patients

<table>
<thead>
<tr>
<th>Type</th>
<th>No. of Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>scaphocephaly</td>
<td>25</td>
</tr>
<tr>
<td>plagiocephaly</td>
<td>10</td>
</tr>
<tr>
<td>trigonocephaly</td>
<td>5</td>
</tr>
<tr>
<td>brachycephaly</td>
<td>9</td>
</tr>
<tr>
<td>oxycephaly</td>
<td>27</td>
</tr>
<tr>
<td>Apert's syndrome</td>
<td>12</td>
</tr>
<tr>
<td>Crouzon's disease</td>
<td>4</td>
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</tbody>
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TABLE 2

Preoperative ICP in 75 cases of craniosynostosis

<table>
<thead>
<tr>
<th>Preop ICP Groups</th>
<th>Mean ICP (mm Hg)</th>
<th>REM Sleep (no. of cases)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>SW Sleep</td>
<td>REM Sleep</td>
</tr>
<tr>
<td>increased ICP (23 cases)</td>
<td>21</td>
<td>48</td>
</tr>
<tr>
<td>borderline ICP (22 cases)</td>
<td>14</td>
<td>35</td>
</tr>
<tr>
<td>normal ICP (30 cases)</td>
<td>9</td>
<td>27</td>
</tr>
</tbody>
</table>

* ICP = intracranial pressure; SW = slow wave; REM = rapid eye movement.
FIG. 2. Intracranial pressure recordings in three patients illustrating three different patterns during sleep. For a description see text.

**TABLE 3**

<table>
<thead>
<tr>
<th>Intracranial pressure (ICP) in different types of craniosynostosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type</td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td>scaphocephaly</td>
</tr>
<tr>
<td>plagiocephaly</td>
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<tr>
<td>trigonocephaly</td>
</tr>
<tr>
<td>brachycephaly</td>
</tr>
<tr>
<td>oxycephaly</td>
</tr>
<tr>
<td>Apert's syndrome</td>
</tr>
<tr>
<td>Crouzon's disease</td>
</tr>
<tr>
<td>total</td>
</tr>
</tbody>
</table>

**TABLE 4**

<table>
<thead>
<tr>
<th>Progressive decrease in mean ICP after surgery in 23 patients*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Time of Measurement</td>
</tr>
<tr>
<td>mean preop ICP (mm Hg)</td>
</tr>
<tr>
<td>mean postop ICP (mm Hg)</td>
</tr>
<tr>
<td>15th day</td>
</tr>
<tr>
<td>3rd to 6th month</td>
</tr>
</tbody>
</table>

* ICP = intracranial pressure.

were sustained in 17 cases (among these, plateau waves were found in 12 patients). In six cases, only phasic waves were recorded, in spite of the fact that SW sleep ICP was increased.

Thirty patients had normal ICP, with recordings during SW sleep below 10 mm Hg (mean 9 mm Hg). In these cases, the mean ICP at the peak of REM sleep waves was 27 mm Hg. In 25 of these 30 cases, REM sleep waves of increased ICP were of the phasic type. In the 22 patients with borderline recordings, ICP values during SW sleep were between 10 and 15 mm Hg (mean 14 mm Hg). The mean ICP at the peak of REM sleep waves was 35 mm Hg. These waves were sustained in seven cases and phasic in 15.

The ICP recordings have been found to vary in different types of craniosynostosis (Table 3), and depending upon the number of cranial sutures involved (Fig. 4). The ICP recordings were abnormal (15 mm Hg or greater during SW sleep) in 14% of cases involving one suture and in 47% of cases involving several sutures.

**Postoperative Recordings**

The ICP was recorded in 23 unselected children both before and after the operation. Recordings were...
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FIG. 3. Intracranial pressure (ICP) during REM sleep in craniosynostosis. Upper: Mean number of eye movements per 10 seconds. Center: Mean electroencephalographic (EEG) frequency (arbitrary units). Lower: Mean ICP recordings in mm Hg. The means were computed every 10 seconds. On the middle curve during the 1st hour, EEG frequency is high and the subject is first in light sleep (Stage 1 or 2, with no eye movements) then in REM sleep. Afterward, EEG frequency gets slower, indicating that the subject enters deep sleep (Stage 3 or 4).

made after surgery whenever possible. Of these patients, eight had oxycephaly, four scaphocephaly, four plagiocephaly, two brachycephaly, two trigonocephaly, two Apert’s syndrome, and one Crouzon’s disease. The postoperative recordings showed that the mean ICP was lower after surgery. Table 4 and Fig. 5 show the decrease of the mean ICP during SW sleep and REM sleep in the 23 children recorded before and after surgery. On the recordings taken 15 days after the operation, the mean decrease of ICP during SW sleep was 3 mm Hg ($p < 0.002$), and the waves of increased ICP during REM sleep were reduced by an average of 11 mm Hg ($p < 0.001$). However, this slight decrease was amplified during the following 6 months. Thus, on the recordings taken 6 months after surgery the final mean decrease from preoperative levels of ICP during SW sleep was 6 mm Hg ($p < 0.001$) and that of waves of increased ICP during REM sleep was 16 mm Hg ($p < 0.001$). Postoperatively, ICP during SW sleep dropped below 15 mm Hg in all cases (Table 5). The proportion of cases in which abnormal sustained waves of increased ICP were found decreased from 57% before the operation to 26% after surgery. Seventeen children, previously operated on for craniosynostosis, were hospitalized because the recurrence of intracranial hypertension was suspected from clinical evaluation. In these children, the mean ICP during SW sleep was 17 mm Hg and the mean REM

![Fig. 4. Distribution of intracranial pressure (ICP) values according to the number of fused sutures. White blocks = less than or equal to 10 mm Hg; crossed blocks = 10 to 15 mm Hg; double-crossed blocks = more than 15 mm Hg.](image)

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sleep ICP recorded at the peak of the wave was 40 mm Hg. In these cases, the preoperative ICP was unknown. However, the mean ICP was comparable to that found in the 75 children recorded either before surgery or with no surgery. It can thus be inferred that, in these cases, ICP had returned to its preoperative value and that a surgical procedure was needed again.

**REM Sleep ICP Related to SW Sleep ICP**

Previous studies have shown that the wave of increased ICP during REM sleep is due to an increase in cerebral blood volume and that the amplitude of...

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**FIG. 5.** Intracranial pressure (ICP) recording in a patient with scaphocephaly, preoperatively (*upper*), and 15 days (*center*) and 4.5 months postoperatively (*lower*). There is a progressive return to a normal ICP level.

**FIG. 6.** Relationship between intracranial pressure (ICP) levels in slow-wave (SW) and REM sleep. The ICP given for REM sleep is the value recorded at the peak of the wave of increased ICP.

**FIG. 7.** Pre- and postoperative relationship between intracranial pressure (ICP) in slow-wave (SW) and REM sleep.
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TABLE 5

<table>
<thead>
<tr>
<th>Time of Recording</th>
<th>Slow-Wave Sleep ICP</th>
<th>REM Sleep</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>&lt; 10 mm Hg</td>
<td>10-15 mm Hg</td>
</tr>
<tr>
<td>preop</td>
<td>5</td>
<td>5</td>
</tr>
<tr>
<td>postop</td>
<td>15</td>
<td>8</td>
</tr>
</tbody>
</table>

* ICP = intracranial pressure; REM = rapid eye movement.

this wave should be related to the cerebral compliance. Therefore, the ratio between maximum ICP values during SW sleep and during REM sleep (Figs. 6 and 7) was studied in order indirectly to evaluate this cerebral compliance. The mean ratio in the 75 preoperative recordings was 2.7:1, and did not differ significantly regardless of whether the craniosynostosis involved one suture (2.7:1) or several (2.72:1). However, this mean ratio was not sensitive to variations from one case to another; indeed, it varied from 1.7:1 to 6:1. This ratio was also studied in the 23 patients recorded pre- and postoperatively. It did not decrease significantly after surgery (preoperative mean value 2.69:1, postoperative mean value 2.52:1).

ICP Related to Mental Level

The IQ levels and ICP were compared in order to determine if a relationship could be found between these different parameters. This comparison was carried out only before the operation; the follow-up period in this series was far too short for useful postoperative study of the mental level. Among the 75 children whose ICP was recorded preoperatively, 55 underwent psychometric testing. It can be seen in Fig. 8 that there was a relationship between both SW sleep ICP and REM sleep ICP and the IQ level. The IQ level decreased slightly when ICP increased. This relationship was statistically significant (SW sleep ICP/IQ: p < 0.01; REM sleep ICP/IQ: p < 0.05).

Intracranial Pressure Related to Age

When the regression curve of ICP as a function of age is plotted, it becomes apparent that the preoperative ICP was maximum at the age of about 6 years in this series (Fig. 9). After this age, ICP decreased. The relationship was statistically significant and was true for both SW sleep ICP (p < 10^-6) and REM sleep ICP (p < 10^-5).

Discussion

The purpose of this study was to establish the consequences of the discrepancy between brain and cranial volume in craniosynostosis, and the possible effect of this discrepancy on ICP.

The definition of "normal" and "abnormal" ICP recordings in children raises an initial problem. Indeed, normal ICP is different in infants and adults. With regard to infants, most studies agree with our results, and consider mean ICP values between 7 and 8 mm Hg to be normal. Normal ICP was evaluated in adults by Gilland, et al., and was found to be 11 ± 2.7 mm Hg. However, normal ICP is not precisely defined for patients in these age groups. In our study, we considered ICP to be normal when it was below 10 mm Hg, abnormal above 15 mm Hg, and borderline in between.

Waves of increased ICP during REM sleep were regarded as abnormal every time they were sustained.
whether they were plateau waves or not. In previous reports, these waves have been recorded only in pathological situations; in this study, they were associated with an obviously increased SW sleep ICP in about three cases out of four. Sustained waves of increased ICP during REM sleep are due to an increase in cerebral blood volume. They are found in cases of hydrocephalus, subdural hematomas, craniosynostosis, and space-occupying lesions; that is, in cases in which the space available for the cerebral parenchyma is reduced. Thus, it is very likely that they indicate a diminution of cerebral compliance. On the contrary, the phasic variations of ICP recorded during REM sleep with no or almost no sustained wave were associated with normal SW sleep ICP in about the same proportion of cases. Since they are also found in the normal cat, they should be considered as normal events during REM sleep in man.

Normality being thus defined, the present study shows that, in craniosynostosis, ICP is normal in roughly one-third of the cases, elevated in one-third, and borderline in one-third. These variations are in agreement with the findings of Goblet, et al., and van Effenterre, et al. In other studies of craniosynostosis, ICP was found to be normal or elevated. These, however, were only small series of patients.

The fact that increased ICP is most often found in cases of craniosynostosis involving several sutures is not surprising. Indeed, the cranial volume is probably reduced in this group. Moreover, clinical and radiological signs of intracranial hypertension are more often discovered in these patients than in those with only one suture involved. It should be stressed, however, that this finding is not constant, so that ICP levels cannot be predicted from the number of fused sutures.

The main problem in craniosynostosis is to determine if a moderately increased ICP can, in the long term, be responsible for a low IQ level. This study suggests such a relationship, but does not prove it definitively. A statistically significant relationship between IQ and ICP was found at the time of surgery; ICP was usually elevated in cases of craniosynostosis involving several sutures, and it is well known that in these cases IQ is often low. However, it could be argued that increased ICP and low IQ are two consequences of a third variable. Two other arguments can be cited in favor of a direct relationship between IQ and ICP. This study indicates that the longer the duration of the craniosynostosis and of its associated increased ICP, the lower the mental level of the patient (Table 6). Moreover, the decrease of ICP in children more than 6 years old possibly indicates the progressive development of a cerebral atrophy likely related to chronic intracranial hypertension. The demonstration of a relationship between ICP and IQ could be clarified by a comparative study of pre- and postoperative IQ levels. Such a study is not yet available in this series of patients, because the postoperative follow-up period is, at the moment, too short.

The lowering of ICP after surgery is proven in this work by comparative pre- and postoperative record-

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**TABLE 6**

<table>
<thead>
<tr>
<th>Age (yrs)</th>
<th>IQ &gt; 90</th>
<th>IQ &lt; 90</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt; 3</td>
<td>22</td>
<td>5</td>
</tr>
<tr>
<td>&gt; 3</td>
<td>9</td>
<td>19</td>
</tr>
</tbody>
</table>

*Significance: p < 0.001. IQ = intelligence quotient; this includes development quotient in children aged less than 2½ years. ICP = intracranial pressure.
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ings. However, the progressive nature of that decrease is, at first sight, surprising since an increase of the cranial volume occurs immediately after the surgical procedure. The resistance of the dura which is not opened at surgery, and the slow expansion of the brain in the dead space created by the surgical procedure, probably explain the progressive decrease of ICP. The demonstration of this hypothesis is, at the present time, the subject of another study, relating the progressive diminution of the dead space and the lowering of ICP.

Conclusions

In craniostenosis, ICP is obviously elevated in one-third of the cases, mainly when several sutures are involved. Moreover, it is likely that there is a relationship between ICP and mental level. After surgery, ICP decreases progressively. This study shows that, if ICP can be estimated easily and without danger, it should be recorded in cases of craniostenosis. These recordings could be one, but only one, of the elements that lead to the final therapeutic decision.

References


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