Nonsynostotic scaphocephaly: the so-called sticky sagittal suture

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Object. Scaphocephaly is a common craniofacial abnormality that results from craniosynostosis of the sagittal suture. The authors have treated a group of infants who presented with nonsynostotic scaphocephaly, or a so-called sticky sagittal suture. The purpose of this study was to describe these patients with nonsynostotic scaphocephaly, the natural history of the disease, and its treatment.

Methods. At the University of Texas–Houston Medical School between 1996 and 2002, nine patients presented with nonsynostotic scaphocephaly. When the abnormality in patients was diagnosed prior to 12 months of age, the majority (seven of eight) were successfully managed by molding helmet therapy. The only child in this group in whom this therapy failed to correct the deformity was noted on repeated computerized tomography scanning to have true sagittal synostosis, which required surgical correction. One child with nonsynostotic scaphocephaly, who presented after 1 year of age, required surgical correction followed by postoperative molding helmet therapy.

Conclusions. Patients with nonsynostotic scaphocephaly appear to have sagittal synostosis. If treated early (at < 12 months of age), head shape in these patients can be normalized by molding helmet therapy. Surgery is reserved for older patients (> 12 months of age) or those with sagittal synostosis.

Key Words • craniosynostosis • dolichocephaly • sagittal suture • pediatric neurosurgery

Clinical Material and Methods

We conducted a retrospective chart review of 48 children with scaphocephaly referred to The University of Texas–Houston Cleft and Craniofacial Clinic between 1996 and 2002. Nine patients with nonsynostotic scaphocephaly were identified (Table 1). These patients were seen both by a pediatric neurosurgeon and a craniofacial surgeon. A detailed history was obtained and a physical examination was performed; CT scanning of the head was performed to assess the patency of the sagittal suture. These children ranged in age from 2 months to 1 year. In this group, eight children underwent molding helmet therapy (DOC; Cranial Technologies, Inc., Phoenix, AZ). Two patients required surgery prior to helmet therapy. Anthropomorphic measurements were taken using calibrated calipers. The cephalic index was the primary measurement used to assess each patient’s treatment resulting from helmet therapy and surgery. This value represents the ratio of the widest point on the head above the ears to the distance measured from the
So-called sticky sagittal suture

Illustrative Cases

Case 7

This girl was born at 37 weeks’ gestation by cesarean section secondary to breech presentation. She presented at 3.5 months of age with an abnormal head shape (Fig. 1a and c). A CT scan revealed scaphocephaly with all sutures open and no evidence of craniosynostosis (Fig. 1e and f). At 4 months of age, she was placed in a DOC band and her head shape normalized within 2 months. At 5 years of age, her cephalic index remained normal (76.7) (Fig. 1b).

Results

The median age at the time of diagnosis was 4.5 months (range 3–12.5 months); the median age at commencement of helmet therapy was 5.75 months (range 4–12 months), and the median duration of treatment was 2.75 months (range 2–4.5 months). Of the nine children with nonsynostotic scaphocephaly, eight underwent helmet therapy. Of these, head shapes normalized clinically in seven children by the end of therapy. Early posttreatment cephalic index findings indicated that results in six children were within the range of one SD of the mean, whereas those in the remaining child were within the range of two SDs of the mean. Long-term cephalic index results were obtained in only one patient; all other patients were lost to follow up. The long-term results in this child (Case 7) indicated a persistent normalized cephalic index (76.7 [normal 76.8]). Two children required surgery—one after helmet therapy failed and another in whom repeated CT scanning revealed sagittal synostosis. A child who presented at 12.5 months of age underwent cranial vault reconstruction and postoperative helmet therapy.

Discussion

Sagittal synostosis is the most common of all types of craniosynostosis; however, in some children who present with the stigmata of sagittal synostosis the corresponding suture fusion may not exist. Developmentally, children with nonsynostotic scaphocephaly appear to have a so-called sticky sagittal suture that results in functional craniosynostosis. The term “sticky” suture has historically been used in conjunction with the lambdoidal suture in children with positional plagiocephaly. It was used to

Table 1

Demographic and treatment data obtained in nine patients with nonsynostotic scaphocephaly*

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age at Diagnosis (mos)</th>
<th>Sex</th>
<th>Age at Helmet Therapy (mos)</th>
<th>Treatment Duration (mos)</th>
<th>Cephalic Index Values</th>
<th>FU†</th>
<th>Normal</th>
<th>Op Required</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>3.5, M</td>
<td>5</td>
<td>2.5</td>
<td>69.8</td>
<td>73.7 ± 5.0</td>
<td>73.6 ‡</td>
<td>78.0 ± 6.6</td>
<td>no</td>
</tr>
<tr>
<td>2</td>
<td>4.5, F</td>
<td>7</td>
<td>2</td>
<td>66.5</td>
<td>78.0 ± 6.6</td>
<td>68.5 §</td>
<td>78.0 ± 6.6</td>
<td>no</td>
</tr>
<tr>
<td>3</td>
<td>7.0, F</td>
<td>12</td>
<td>4.5</td>
<td>69.8</td>
<td>78.0 ± 6.6</td>
<td>73.8 ‡</td>
<td>76.7 ± 4.9</td>
<td>no</td>
</tr>
<tr>
<td>4</td>
<td>2.5, M</td>
<td>4</td>
<td>2.5</td>
<td>69.9</td>
<td>73.7 ± 5.0</td>
<td>71.4 ‡</td>
<td>78.0 ± 6.6</td>
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</tr>
<tr>
<td>5</td>
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<td>6.5</td>
<td>4</td>
<td>76.0</td>
<td>78.0 ± 6.6</td>
<td>78.0 ‡</td>
<td>78.0 ± 6.6</td>
<td>no</td>
</tr>
<tr>
<td>6</td>
<td>4.5, M</td>
<td>7</td>
<td>3</td>
<td>72.9</td>
<td>78.0 ± 6.6</td>
<td>73.1 §</td>
<td>78.0 ± 6.6</td>
<td>no</td>
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<tr>
<td>7</td>
<td>3.5, F</td>
<td>4</td>
<td>2</td>
<td>71.4</td>
<td>73.7 ± 5.0</td>
<td>74.2 ‡</td>
<td>73.7 ± 5.0</td>
<td>no</td>
</tr>
<tr>
<td>8</td>
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<td>4</td>
<td>67.6</td>
<td>73.7 ± 5.0</td>
<td>67.5 §</td>
<td>78.0 ± 6.6</td>
<td>yes</td>
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<tr>
<td>9</td>
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<td>NA</td>
<td>59.9</td>
<td>78.0 ± 6.6</td>
<td>NA</td>
<td>NA</td>
<td>yes</td>
</tr>
</tbody>
</table>

* FU = follow up; NA = not applicable.
† The follow-up cephalic index represents the value determined at completion of helmet therapy.
‡ Within the range of one SD of the normal mean value.
§ Within the range of two SDs of the normal mean value.
explain the cranial vault asymmetry in cases of positional deformities in which the pathogenesis of the deformities was not clearly understood. Therefore, the term sticky suture is associated with a pseudodisease entity.1

The physiological mechanisms for aberrant suture growth remain poorly understood. Recent research into cranial suture phenomena has demonstrated that the cranial vault develops as a result of a tissue interaction between the brain, dura mater, sutures, and cranial bones.2,7 In the murine model, regionally derived growth...
factors such as transforming growth factor–β and the family of fibroblast growth factors have been noted to guide calvarial development. Fibroblast growth factor mutation is associated with syndromic craniosynostosis.8 In our series, the sticky suture affected cranial growth resulting in scaphocephaly. It has often been hypothesized that partial synostosis causes cranial changes seen in synostosis.3 To date, most suture imaging studies have failed to identify the presence of partial synostosis.1 The natural history of an untreated sticky suture is illustrated by the outcome in our Case 9. The child presented with prominent scaphocephaly because a delay in diagnosis resulted in alteration of normal head growth. When craniosynostosis is not present, the origin of abnormal head growth in these children remains unclear. None of the patients’ families had a history of a naturally occurring scaphocephalic skull. There was no history of lateral head positioning or prematurity; however, positioning appears to be the most probable cause of the deformity. To date, there is no evidence, radiologically or histologically, of the so-called sticky sagittal suture.

Treatment of nonsynostotic scaphocephaly is based on an early and accurate diagnosis. Initially CT scanning is undertaken to rule out the craniosynostosis of the sagittal suture. There is a group of infants with histories of prematurity and treatment in the neonatal intensive care unit who present with nonsynostotic scaphocephaly. This cranial deformity results from lateral head positioning. Its natural history is one of resolution once the infant assumes normal posturing and head control. In our patients with nonsynostotic scaphocephaly, there was no history of prematurity or lateral head positioning. Because scaphocephaly in these children is progressive, early intervention reduces the extent of their deformity and the amount of correction required. Moreover, for helmet therapy to be successful, the potential for adequate cranial growth must be present. In this series, helmet therapy was successful in all patients with nonsynostotic scaphocephaly in whom the diagnosis was made prior to 6 months of age. When the diagnosis is delayed beyond 1 year (as in Case 9), molding helmet therapy is of limited use. Surgical intervention followed by helmet therapy becomes the treatment of choice. If helmet therapy fails to correct the deformity, CT scanning should be repeated to rule out undiagnosed craniosynostosis (as in Case 8).

Clinically all patients with nonsynostotic scaphocephaly treated with helmet therapy exhibited normal head shape in time. Cephalic index results were improved in all but one patient (Case 8) who required surgery. The use of the cephalic index as a marker of scaphocephaly, however, has always been controversial because it treats the skull as a two-dimensional planar object. The cephalic index provides no information on vertical height or the anterior cranial vault, both of which are critical in determining a clinically acceptable head shape. Opisthion is a relative landmark, but because the skull is always increasing in length, it does not cause a problem with cephalic index. Maximum cranial breadth (euryon–euryon) is difficult to determine because it may be high on the parietal bones or low on the temporal bones. Clinically, although this is not a problem, if the malposition is indicated, it does hinder statistical analysis. As an end point, cephalic index is not ideal, but as a measure of treatment progress it is useful, particularly in cases in which helmet therapy fails to correct the deformity.6 Patients then require repeated CT scanning to rule out true craniosynostosis.

Conclusions

Scaphocephaly can exist without corresponding sagittal synostosis. When diagnosed early, this condition is amenable to nonsurgical therapy involving molding helmets. When the diagnosis is delayed, however, the deformity is often severe and may require surgical correction.
All children with scaphocephaly should undergo CT scanning of the head to determine the cause of the abnormality. For patients with a so-called sticky suture, we recommend molding helmet therapy.

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Disclaimer
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References

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