4.1 Classification of Craniosynostosis: Therapeutical implications.

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Craniosynostosis
Today’s Topics
(Single suture synostosis)

✓ Classification

✓ Surgical implication
Scaphocephaly

Elongation of the skull resulting from the premature fusion of the sagittal suture.

Most common type of synostosis.

40-60% (including non-syndromic & syndromic cases)

Male > Female (3.5:1)

Cephalic Index (CI)

\[ CI = \frac{a}{b} \times 100 \]

Cl under 75 : Scaphocephaly
Correct

Scaphocephaly

Surgical Indication: Cosmetic correction of the skull to normalize its form.

- Prevention of raised Intracranial pressure?
- Preservation of cognitive development?

Still in debate
Trigonocephaly

Premature fusion of the metopic suture.

Obvious ridge of the midline due to the ossification of the suture.

Restriction of the lateral growth of the frontal bone.

The number of this pathology is increasing.

2\textsuperscript{nd} single suture synostosis in terms of incidence.
Trigonocephaly

Indication

Obtain the normalization of the anterior skull fossa volume and to correct the abnormal position of the superior orbital ridges and the hypotelorysm.
Plagiocephaly
Plagiocephaly
Case Presentation

sagittal synostosis w/o scaphocephalic deformation
Compared with typical scaphocephaly

Cl over 75 : 19 cases
Non-scaphocephalic deformation: Group A

Cl under 75 : 15 cases
Typical Scaphocephaly: Group B (control group)
## Results

<table>
<thead>
<tr>
<th></th>
<th>CI Mean ± SD</th>
<th>Age of Diagnosis Mean ± SD ( range )</th>
<th>Developmental Delay</th>
</tr>
</thead>
<tbody>
<tr>
<td>A</td>
<td>80.3 ± 3.4</td>
<td>5.7 ± 3.9 y (0.4 – 15 y)</td>
<td>14 (73.6%)</td>
</tr>
<tr>
<td>B</td>
<td>65.4 ± 5.1</td>
<td>1.5 ± 1.2 y (0.6 – 5 y)</td>
<td>1 (6.7%)</td>
</tr>
</tbody>
</table>

A : Non-scaphocephalic group  
B : Typical scaphocephalic group

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Results

Neuroradiological analysis

<table>
<thead>
<tr>
<th></th>
<th>Sagittal Ridge</th>
<th>Parietal Foramina</th>
<th>Digital Marking in the frontal area</th>
<th>Metopic Ridge</th>
</tr>
</thead>
<tbody>
<tr>
<td>A</td>
<td>3 (15.7%)</td>
<td>12 (63.1%)</td>
<td>17 (89.4%)</td>
<td>14 (73.6%)</td>
</tr>
<tr>
<td>B</td>
<td>9 (60.0%)</td>
<td>1 (6.7%)</td>
<td>1 (6.7%)</td>
<td>9 (60.0%)</td>
</tr>
<tr>
<td>P value</td>
<td>p = 0.012</td>
<td>p = 0.001</td>
<td>p &lt; 0.0001</td>
<td>p = 0.63</td>
</tr>
</tbody>
</table>

A : Non-scaphocephalic group
B : Typical scaphocephalic group

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ICP monitoring

Raised intracranial pressure (ICP)

Overnight ICP monitoring

Increase ICP 76%

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In the literature

Absence of the sagittal suture does not result in scaphocephaly

Dipti Padmalayam • R. Shane Tubbs • Marios Loukas •
Aaron A. Cohen-Gadol

Childs Nerv Syst (2013) 29:673-677

3 / 400 (0.75%) osteological collection

No description of digital marking
And other information

In conclusion
Isolated sagittal suture absence does not result in a head configuration that is scaphocephalic.
In the literature

Management of Isolated Sagittal Synostosis in the Absence of Scaphocephaly: A Series of Eight Cases


8 / 193 (4.1%) Oxford craniofacial unit database

ICP measurement: 6 cases
B waves (+): 4 cases (67%)
Developmental. delay: 5 cases (62.5%)

In conclusion
It is important to recognize these patients because they are at a high risk of developing raised ICP.

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Hypotheses

Management of Isolated Sagittal Synostosis in the Absence of Scaphocephaly: A Series of Eight Cases


1. Late fusion of the suture after the majority of the skull growth had taken place.
2. The metopic suture have resulted compensatory.
3. Other sutures appeared normal but did not function normally.
Conclusion

• Cases of sagittal suture synostosis without scaphocephalic deformation are seen in a certain incidence.

• These children tend to

1. Have a developmental delay in some degree.
2. Detected later than normal scaphocephaly.
3. May have a higher incident of increased intracranial pressure.
Another cases
Mild trigonocephaly with clinical symptoms

Language delay, hyperactivity, autistic symptoms etc.
Surgery 手術
Surgery  手術
Surgery  手術
Surgery  手術
Surgery  手術
Surgery 手術
Materials and Method

2-4 years old
Developmental delay (+)
Bony ridge in the forehead
Normal head circumference
No morphological change in MR imaging.

Registration: 2012/11/1-2014/10/30
Follow up: 6 months

n=23  Primary end point : Improvement of DQ
Secondary end point: Improvement in other tests

3m pre Op
Informed Consent
Register: undergo 5 psychological examinations.

Pre Op
Decompressive Cranioplasty
psychological examinations

3m post Op
psychological examinations

6m post Op
psychological examinations
MRI/CT

2. National rehabilitation center Sign-Significance Test
3. Pervasive Developmental Disorders Autism Society
   Japan Rating Score (PARS)
4. Japanese Child Behavior Check list (CBCL)
5. Mother’s nurturing behavior question sheet

Natural history

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Results  DQ (Total)

- Natural History: 6/23 (26.1%)
- Post Op 3m: 8/23 (34.8%)
- Post Op 6m: 11/23 (47.8%)
Results

- Pre Op – Pre Op Pos Op 3m – Post Op 6m

- Natural History 6/23 (26.1%)
- Post Op 3m 10/23 (43.5%)
- Post Op 6m 10/23 (43.5%)
Results **DQ(total ) & PARS**

<table>
<thead>
<tr>
<th></th>
<th>Bot</th>
<th>DQ</th>
<th>PARS</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Natural history</td>
<td>2</td>
<td>4</td>
<td>4</td>
<td>10/23 (43%)</td>
</tr>
<tr>
<td>Post Op 3m</td>
<td>5</td>
<td>3</td>
<td>5</td>
<td>13/23 (56.5%)</td>
</tr>
<tr>
<td>Post Op 6m</td>
<td>4</td>
<td>7</td>
<td>6</td>
<td>17/23 (73.9%)</td>
</tr>
</tbody>
</table>

- Improvement of DQ: 43.5% (3,6m) 47.8% (6m)
- Improvement of PARS: 43.5%(3,6m)
- Improvement in some degree: 56.5% (3m) 73.9% (6m)

Adverse event: Overlap of the bone 1 case
Conclusion

Over 70% of these cases showed improvement in some degree.

So that this results leads us to believe surgery is providing a positive effect to these children.

There should be a structural disorder. So that continuous support is necessary. Surgery is improving only a part of this disease.
Conclusion

Surgical implication of single suture synostosis is mainly considered as cosmetic facts.

However, there might be some cases that increased ICP should be considered more.

This is not a issue that plastic surgeons will consider.
We (Neurosurgeons) should be aware of this issue.
Thank you for your attention