Strategic approach to squint surgery in craniofacial patients

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Introduction- What are Craniofacial disorders?

• Craniosynostosis is the premature fusion of one or more of the calvarial sutures.
• Single suture craniosynostoses (SSC) prevalence of
  • 1 in 1700-2500 live births
• Syndromic forms (hereditary forms with extracranial malformations)
  • 1 in 25,000
• Premature suture fusion results in abnormalities in calvarial shape due to the restriction of growth perpendicular to the fused suture
Craniosynostosis

sagittal

scaphocephaly

bicoronal

brachycephaly

metopic

trigonocephaly

unicoronal

plagiocephaly
Introduction-Craniofacial disorders identified

- Crouzon,
- Apert,
- Pfeiffer
- Jackson–Weiss,
- PMuenke,
- Crouzonodermoskeletal
- Beare–Stevenson,
- Antley–Bixler with or without genital abnormalities and disordered steroidogenesis

- Saethre–Chotzen
- Baller–Gerold, craniosynostosis Boston-type
- craniofrontonasal,
- Carpenter
- CRSDA (craniosynostosis and dental anomalies)
Introduction

• Children have multiple problems with
  • Speech and development
  • Multiple surgeries
  • Episodes of raised intracranial pressure

• Ophthalmology may be called in late due to the above pressures
• Patients are best managed by multidisciplinary teams where the ophthalmologist is a part.
• The ophthalmologists priority is the preservation and development of vision and not the squint.
Vision

- May be compromised by
  - Optic nerve atrophy due to raised ICP
  - Corneal exposure
  - Hypermetropia
  - Myopia
  - Astigmatism
  - Anisometropia secondary to orbital asymmetry
  - Amblyopia

- Colobomas
- Dry eye
- Ocular Dermoids
- Exophthalmos
- Hypertelorism
- Missing muscles
- Ptosis
- Strabismus
Preventable visual loss

- Astigmatism
- Anisometropia
- Corneal exposure
- Colobomas
- Amblyopia
- Dry Eye
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- Missing Muscles
- Optic atrophy
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- Strabismus
Visual Loss requiring rapid response

- Optic Nerve swelling from raised intracranial pressure
- Corneal Exposure
Preventable visual loss

- **Astigmatism**
  - Anisometropia
  - Corneal exposure
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- Exophthalmos
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- Missing muscles
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Astigmatism in Craniosynostosis

• Significant astigmatism (>1.00D) is found
  • 46% Apert’s
• 45% Pfeiffer’s
  • 24% Crouzon’s
  • 18% Uni-coronals
### Types and frequency of strabismus

<table>
<thead>
<tr>
<th>Type</th>
<th>Apert's (44)</th>
<th>Crouzon's (33)</th>
<th>Pfeiffer's (20)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Exotropia</td>
<td>18 (41%)</td>
<td>20 (61%)</td>
<td>15 (75%)</td>
</tr>
<tr>
<td>Esotropia</td>
<td>23 (52%)</td>
<td>8 (36%)</td>
<td>1 (5%)</td>
</tr>
<tr>
<td>Eso and Exo</td>
<td>6 (14%)</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Nil</td>
<td>2 (5%)</td>
<td>6</td>
<td>4 (20%)</td>
</tr>
</tbody>
</table>
Not all children with squint need surgery

Binocularity may exist in a small area even in children with what appears to be a constant manifest squint.
Strategy

• Understanding the reason for squint
  • guides the management
• Preparation with investigations before surgery
  • prevents nasty surprises
• Understand the syndromic abnormalities
  • Treat underlying treatable visual problems
  • Work with the rest of the team to investigate the orbits and muscles
  • CT scan and MRI scan
  • Consider squint surgery during same General anaesthetic as another procedure
  • Help the MDT to understand the importance of binocularity if attainable.
Abnormal muscles and abnormal orbits

• Muscles do not subtend the globe at the normal angles
  • A pattern
  • V pattern
  • X pattern

• Muscles may be missing
• May be different lengths in the 2 orbits leading to mechanical advantage and imbalance between the 2 eyes.
• Orbits may be shallow
• Hypertelorism
Plagiocephaly - example of orbit asymmetry
Plagiocephaly

The trochlear is displaced to the rear on the affected side. Vertical power of superior oblique is reduced whilst tortional effect is increased.
Plagiocephaly

Patient with unicoronal synostosis who has already lost binocular fusion and presents with a V pattern esotropia. In her case, SO underaction was greater than IO overaction (Panel A) Bi-medial recessions and bilateral SO tucks were performed (Panel B).
Apert syndrome with tilted orbits
Tilted/extorted orbits

• See-saw eye movements
• Looks like inferior oblique over action
• Weakening the I/O will not resolve the problem
• Horizontal recti need moving up/down
Missing muscles
Anatomical considerations

• Lids
• May require tarsorrhaphy
  • If squint only obvious in lateral gazes may not be important to do squint repair
Syndromic craniosynostosis

Pfeiffer’s Syndrome
orbits

• Shallow
• Assymmetric longer/shorter/ frontal prominance
• Tilted/ Extorted
• Wide IPD/ Hypertelorism
Importance of understanding the squint

• Expect the unexpected

• MRI scan of orbits to locate the muscles
  • Missing muscle at the time of surgery is disconcerting and preparation beforehand makes for good planning.

• CT scan of orbits (likely already requested by craniofacial surgeons) to ascertain shape of orbits.
  • Is there a difference in mechanical advantage?

• Consider mimicry from other muscles
  • eg horizontal recti acting vertically
Timing

• Preservation of binocular function is a reason to operate early
  • Consider piggy back onto a combined admission /GA episode

• Craniofacial surgical corrections do not affect the squint as key orbital landmarks are not changed

• Strabismus surgery may have to take lower place in line of multiple surgeries
Strategy- Integrate with the MDT

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