Craniosynostosis and Positional Plagiocephaly

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Objectives

• Identify common features of positional plagiocephaly and craniosynostosis
• How to monitor changes in head shape
• Treatment options for positional plagiocephaly/craniosynostosis
• Know when to consult NSG
• What to send with your patient before their NSG appointment
• Current department research
Do you know what the diagnosis is?
What is a ‘normal’ head shape?
Case Study-Positional Plagiocephaly

- 4 month old male presents with R. occipital flattening worsening over the last two months
- He prefers to lay/look to the right side
- He cries and does not tolerate tummy time
- No unexplained vomiting, lethargy, irritability or downward gaze

Physical Exam
- Right ear looks more anterior than left
- Right frontal bossing
Right Occipital Positional Plagiocephaly

How to evaluate:

• Head circumference
• Suture ridging
• C-spine range of motion
• Facial symmetry
• Cranial Index (CI)
  – AP/L
• Cranial Vault Asymmetry
  – RO-LF, LO-RF
• Plagio vs lambdoid synostosis
Positional Plagiocephaly

• Treatment Options
  – Repositioning
  – Physical Therapy
    • Need to treat underlying torticollis
  – Cranial Remolding Helmet
  – When to refer
Case Study- Sagittal Craniosynostosis

• 1 month old female presents with elongated head and occipital prominence more defined since birth

• Repositioning has not changed head shape

• No unexplained vomiting, lethargy, irritability or downward gaze

Physical Exam

• Frontal bossing
• Sagittal suture ridge
• Face is symmetric
Sagittal Craniosynostosis

- Most common occurrence (50%)
- Scaphocephalic head shape
- Widened forehead, narrow occiput
- Ridging over sagittal suture
- CI <0.70
Case Study- Coronal Craniosynostosis

- 6 month old male presents with asymmetric eyes
- Appears to have left eye ptosis
- Left occipital flattening that has improved over two month period
- No unexplained vomiting, lethargy, irritability or downward gaze

Physical Exam
- Right frontal flattening and left frontal bossing
- Right coronal suture ridging
- Left ear mildly more anterior than right
- Anterior fontanelle open, soft and flat
Right Unicoronal Craniosynostosis
Coronal Craniosynostosis

- Second most prevalent type
- May involve one or both sutures
- Affected eye looks elevated (harlequin eye)
- Forehead is flat on affected side
- Bilateral may indicate syndromic process
Craniosynostosis

• Definition
  – sagittal 53%, unicoronal 20%, metopic 13.5%, bicoronal 6%, lambdoid 1%, multiple 4.5%

• Diagnosis
  – History
    • Prenatal/birth
    • Time of deformity onset
    • Deformity changes
  – Physical
    • Head circumference
    • Calipers
  – CT (low radiation/3D recon)
  – Genetics (FGFR3-->crouzon, apert)
  – Ophthalmology
Craniosynostosis- Treatment

• Surgery vs no surgery
  • Cosmesis (helmet, hat, bullying)
  • Possible Increased ICP
    – 50% of kids presenting >15 months have ICP>20mmHg
    – Seruya, Oh, Boyajian, Posnick, Keating et al

• Papilledema

• Possible Motor/Cognitive Delays
  – Chieffo, Tamburrini, DiRocco et al, J Nsurg Ped
  – Magge SN et al, J Craniofacial Surgery
Surgical Options

• Minimally invasive endoscopic strip craniectomy
  – Typically done prior to 3 months old, but can be done up to 6 months old
  – Any suture craniosynostosis
  – One to two small incisions
  – Cut out affected suture (suturectomy)
  – Remodeling helmet must be worn after surgery 23hrs/day for 10 months to 1 year
  – Helps to gradually correct child’s head shape
Pre op vs 9 month post op
Surgical Options (continued)

- Pi Procedure
  - Typically done at 3-9 months of age
  - Sagittal craniosynostosis
  - Removal of Pi-shaped piece of bone
Surgical Options (continued)

- **Bifrontal Orbital Advancement (BFOA)**
  - > 6 months of age
  - Metopic and coronal craniosynostosis
  - Forehead and orbital bones remodeled
  - Done in conjunction with Plastic Surgery
Surgical Options (continued)

• Calvarial Reconstruction
  – > 6 months of age
  – Sagittal Craniosynostosis
  – Total remodeling of the skull
  – Done in conjunction with Plastic Surgery
After Surgery

• Post Op Care
  – Transfer to the PICU for 24 hours (except endoscopic)
  – Maintain head wrap and drain in place
  – Manage immediate pain

• Complications
  – Infection
  – Wound breakdown
  – CSF leak

• Follow Up
Research Productivity in Our Department on Plagio/Cranio

• Clinical outcomes studies
  – Endoscopic vs open Pi surgery (Magge, Bartolozzi et al., Under review; Seruya, Oh et al., 2011)
  – Re-do percentage (Wood, Oh et al., 2015; Rogers, Greene et al., 2015; Keating, in prep.)
    • 5%
  – Hospital stay
    • ICU? 4.7%
  – Transfusion rate (Reddy, Swink et al., 2016)
    • Endo 23% vs Pi 40%
  – Optimal age
  – Helmet (Wood, Ahn et al., 2017)

• Volumetrics (Jha, Barnawi et al., 2016; Jha, Quigley et al., in prep.)
• 3D imaging (Tu, Porras et al., 2017; Porras, Paniagua et al., 2017; Wood, Mendoza et al., 2016)
• Non invasive ICP monitoring
  – Vittamed w/ Boston Neurosciences
• Positional Plagiocephaly (Wisniewski, future)
What can you do as our Primary Providers?

• Consistent head circumference measurements
  – same person measuring HC
• When you refer a patients, send a copy of HC chart
• If you suspect craniosynostosis send them earlier than later so parents have option of strip craniectomy
• Always evaluate for increased ICP
New guidelines review evidence on PT, helmets for positional plagiocephaly.
http://www.aappublications.org/news/2016/10/27/Plagiocephaly102016

Plagiocephaly - Hampshire Orthotics
Hampshire Orthotics

Muenke Syndrome | Hellenic Craniofacial Center
craniofacial.org

Magge SN et al, J Craniofacial Surgery, 2002
http://www.craniofacial.org/en/content/craniosynostoses

http://www.myhealthyfeeling.com/papilledema-symptoms-causes-treatment-pictures/papilledema-eyes/
http://www.3dmd.com/wp-content/uploads/2012/03/Lily-head-individual-shot.jpg

Seruya, Roagers, Myseros et al, Pediatric Craniofacial, 4:582-588
Questions?