

# Craniosynostosis and Positional Plagiocephaly

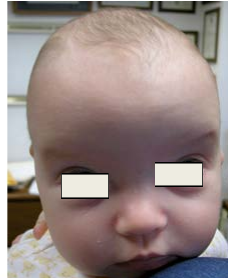
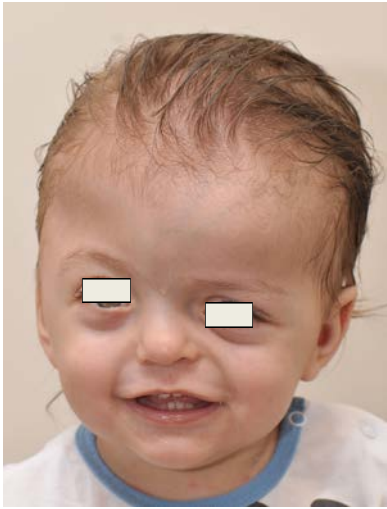
Stephanie Gurevich, PA-C  
Jessica Wisniewski, MSN, CPNP  
Department of Neurosurgery

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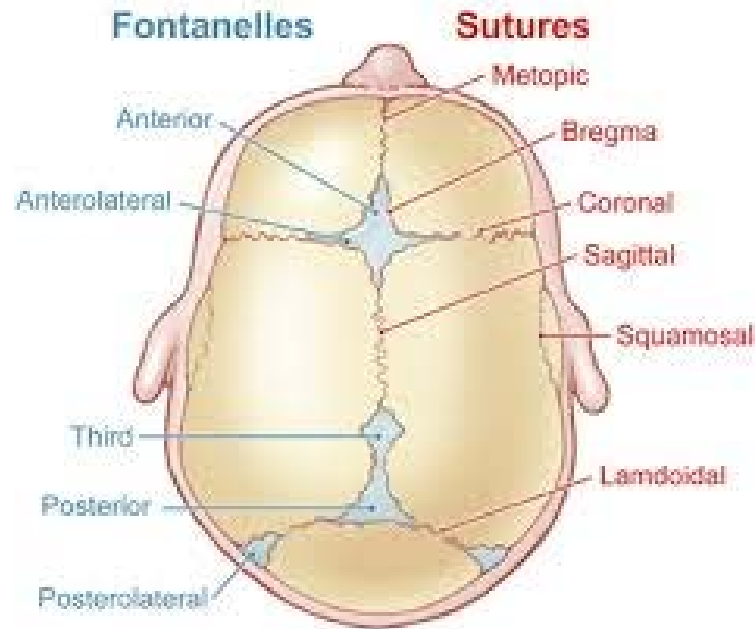
# Objectives

- 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 Uniconic 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

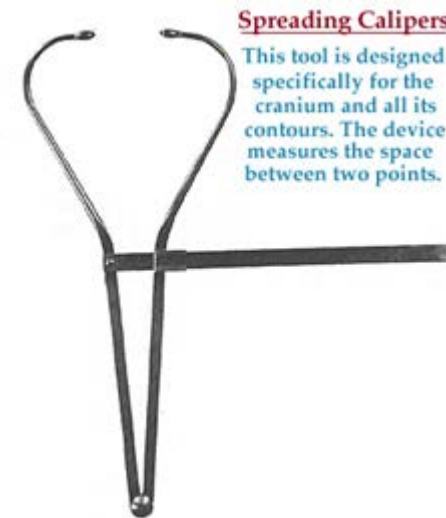
- Physical

- Head circumference
    - Calipers

- CT (low radiation/3D recon)

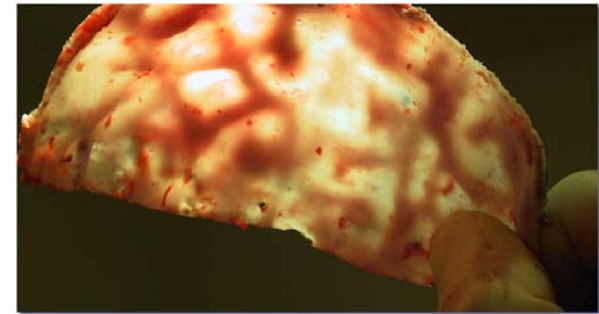
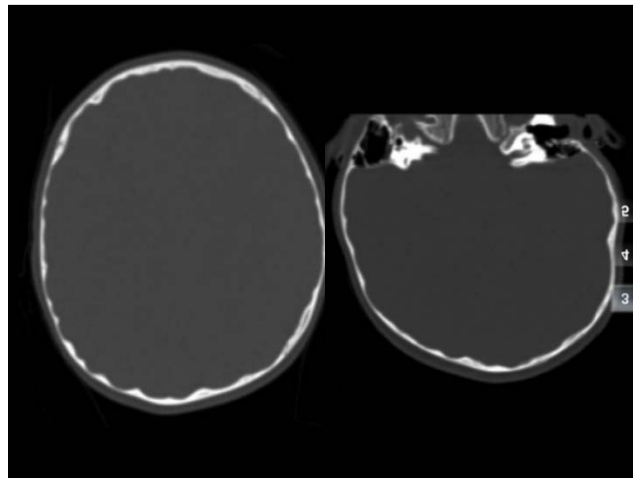
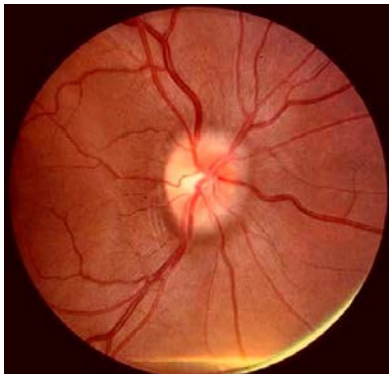
- Genetics (FGFR3-->crouzon,apert)

- Ophthalmology



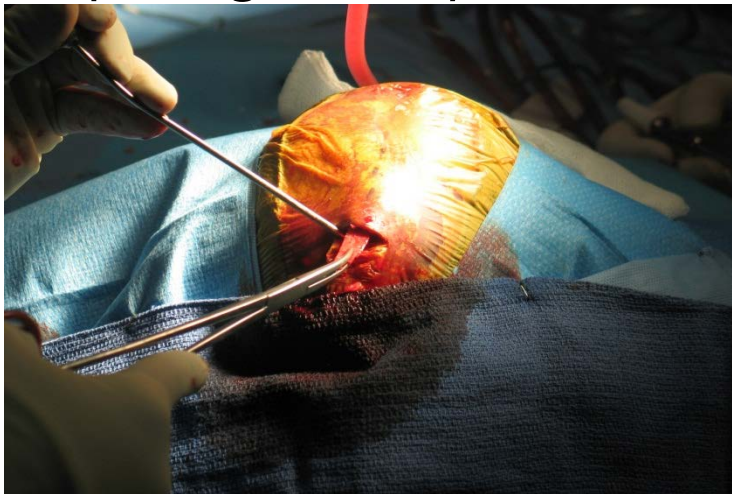
# Craniosynostosis- Treatment

- 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



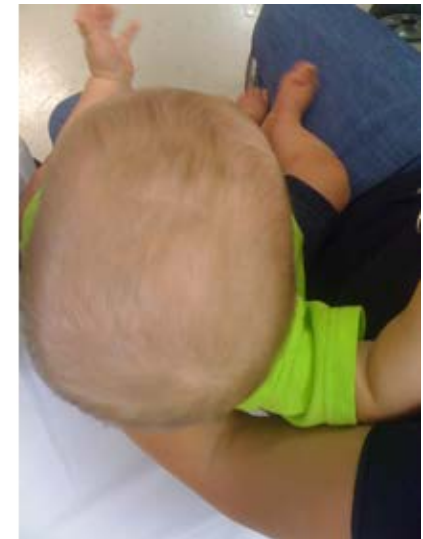
# Surgical Options

- 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



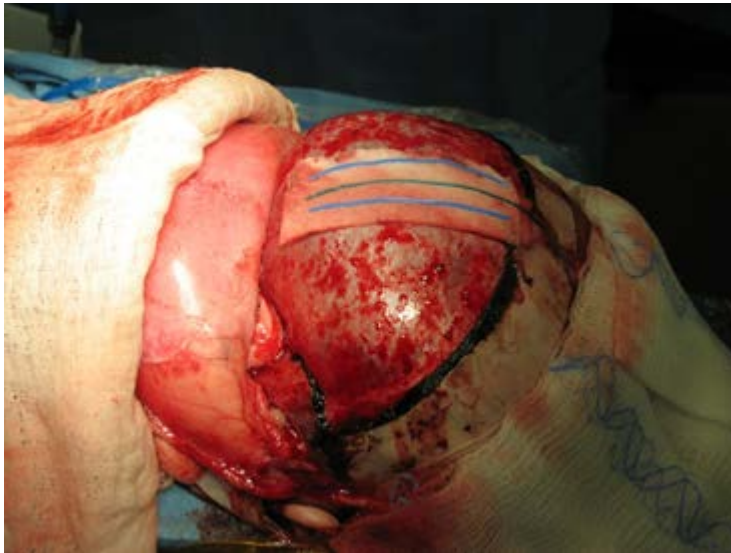
## Surgical Options (continued)

- Pi Procedure
  - Typically done at 3-9 months of age
  - Sagittal craniosynostosis
  - Removal of Pi-shaped piece of bone



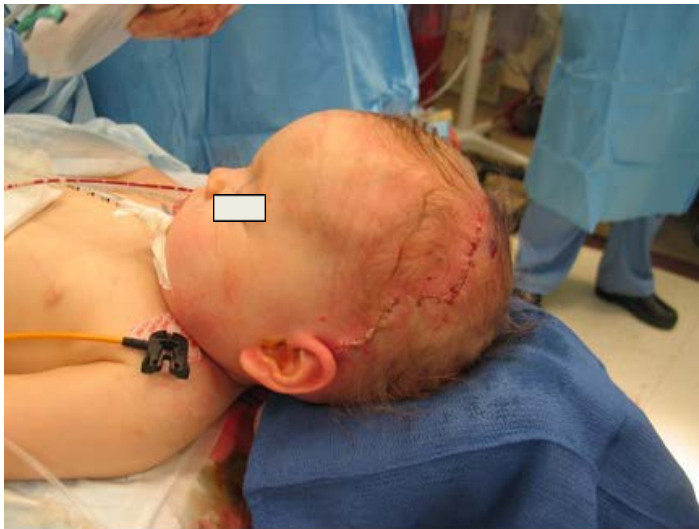
## Surgical Options (continued)

- 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

# References

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Questions?