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Learning objectives

1. Know what imaging options are available
2. Know normal developmental stages
3. Identify calvarial anatomy: bones, boundaries, synchondroses & sutures
4. Identify common & syndromic craniosynostoses
5. Understand potential complications of craniosynostosis (pre- & postoperative)

Imaging options include:

- Radiographs (still ordered by pediatricians)
- Low dose (20-30 mAs) CT/3D CT
- MRI (black bone' technique)
- Ultrasound evaluation of single suture patency (not skull base)
- 3D photogrammetric analysis
  - Can measure
    - Cranial index
    - Anterior skull volume
    - Symmetry ratio

CT dose is lowered to evaluate

- Skull & skin only:
  - Very low dose 20-30 mAs axial images
  - Helically scanned
  - Without tilting table gantry (to avoid 'stacked coin' on 3D)

- Late postoperative leaks or unusual skull base defects may require:
  - 15 mAs direct coronal (0.08mSv)
  - Contrast chalciagram
  - CTV

If your center uses CT: ALARA

It is what it is... so make sure your doses are ultra-low

- Published 'low-dose' CT at 120 mAs NOT really low-dose'
- Again: Images obtained in this talk used 20-30 mAs
Black bone’ 3D MRI, not yet as robust as CT, but software packages improving

Fetal ‘black bone’ & 3D MRI possible
- Skin surface phenotyping can be done
  - FISP 3D data set
    - Currently time consuming
    - Requires a ‘still’ fetus
- ‘Black bone’ source imaging
  - SWI

Finally: Ultrasound & sutures
- US can assess suture patency in single-suture synostosis, by evaluating:
  - Suture width, bone thickness, bone overlap, partial or complete synostosis, but NOT skull base
  - Normal: beveled or hypoechogenic gap of at least 0.5 mm

Next, calvarial development: First - Membranous
- Membranous (vault) derived from:
  - Neural crest (frontal)
  - Paraxial mesoderm (parietal)
- Mesodermal cells form membranous sheets:
  - Which differentiate directly into osteoblasts & osteocytes
- Membranous bones include:
  - Frontal, parietal, temporal squamous, supraoccipital
  - Nasal, lacrimal bones

Enchondral bone formation: is more complicated
- Basal condensation (4 wks GA)
  - Neural crest cells & paraxial mesoderm consolidate to form desmocranium
- Chondrification (8 wks GA)
  - Chondrocranium replaces mesenchyme of desmocranium
- Ossification of cartilage centers follows chondrification
  - Ossification also determined by presence of sensory neural penetration
- Cranial synchondroses
  - Develop within chondrocranium as cartilaginous junctions between cartilage centres

On to synchondroses & sutures
- The bones of the cranial vault are joined by sutures (a type of synarthrosis)
- The bones of the cranial base are joined by synchondroses (cartilaginous joints)
- Up-regulation of transforming growth factors/receptors signal sutural fusion
For example

- Calvarial bones arise in ossification centers (beginning around 2 mo GA)
- Spicules of bone grow towards each other
- Sutures form at the site of meeting bone fronts, sutural fate is determined by the underlying (regionally differentiated) dura mater
- Interdigiting fingers of bone project into sutures
- Osteoblastic & osteoclastic activity contribute to fusion activity (note FGFR expression on leading edges of bone)

Cribiform ossification, for example, is not identified until late infancy

1 day old
10 months old

Same infant

And now centrally...

The occipital bone is comprised of 6 bones

- Basi-occipital
- Exoccipital x 2
- Supraoccipital
- Squamous portion of the occipital bone
- Kerckring’s sssicle

* All but the squamous portion are contiguous with foramen magnum

This process is ongoing & unossified cartilage simulates bony defects during early life

20 weeks gestation
31 weeks gestation

Lets remind ourselves of anatomy: starting peripherally & here is basi-sphenoid

Here are mastoid...
...And pyramidal portions of petrous bone

Here is the basion (mid-point of anterior margin of foramen magnum)

It belongs to the basi-occipital bone

Here are paired exoccipital bones
Finally, the opisthion (midpoint of posterior margin of foramen magnum, which develops from Kerpizir's ossicle)

And the ventral chondal &...

... posterior squamous (or membranous) portions of Supraoccipital bone

A reminder, the ventral chondal...

... posterior squamous portions of Supraoccipital bone

Are ventral & posterior to the Mendosal sutures

The superior tip of the squamous supraoccipital bone is called the Interparietal portion

And here is the interparietal OR bi-interparietal suture AKA median occipital fissure

Sutura mendosa = transverse suture...at level of transverse sinus

Mendosal sutures connecting in the middle = transverse occipital suture (subjacent transverse sinuses/torcular at risk during birth)

Here are some examples of:

- The upper fragment is the interparietal bone OR inca bone
- Failure to fuse the occipital centers leads to persistence of accessory sutures
- Posterior fontanel bones (like these) are larger than intrasutural & smaller than inca bones

Accessory ossicles: here, the lateral intrasutural sutures connect with Mendosal sutures, parallel to the lambdoid sutures

Intrasutural ossicles: AKA Wormian bones are within the sutures
Excessive Wormian bones can be clues to a diagnosis

They are also present in cases of delayed sutural ossification

In Menke disease

As in this pre-teen with delayed & delayed dentition, abnormal CR & hard radiograph in keeping with Clidocranial Dysplasia

Here are examples of accessory frontal & parietal bones versus anterior fontanel bone

The accessory frontal bone is abnormal

- Has been called duplication of metopic suture, but is more than that
- Usually seen in midline deficiency disorders:
  - Agenesis of corpus callosum
  - Frontonasal dysplasia
  - Goldenhar
  - Chiari 2
  - Etc.

While the anterior fontanel bone is a normal variant usually referred because of pseudo-early fontanel closure

On the synchondroses, the occipital bone has 6 pairs (mostly named by adjacent bones)

- Petro-occipital
  - Between petrous portion of temporal bone & basioccipital bone
- Sphen-occipital
  - Between sphenoidal body & basal portion of occipital bone
- Keckring-supra-occipital
  - Between Keckring ossicle & supra-occipital bone
- Petro-mastoïd named as expected
- Anterior intra-occipital
  - Between basis-occipital & exoccipital
- Posterior intra-occipital (AKA innominate suture)
  - Between exoccipital portion of occipital bone & petrous portion of temporal bone

Let's start peripherally:

Mastoid or postero-lateral fontanels: site of asterion (junction of parieto-mastoïd, occipito-mastoïd & lambdoid sutures)
And move centrally:

- Sphenoparietal synchondrosis
- Anterior intracerebral synchondrosis
- Posterior intracerebral synchondrosis AKA basal cranial synchondrosis AKA inonminate sutures

Finally: Kerckring’s ossicle (or center)

- Last of the occipital centers
- Appears ~16 weeks GA
- Fuses with supraventricular portion of occipital bone in 1st month (or so)
- Forms the posterior margin (opisthion) of foramen magnum

Kerckring’s ossicle rapidly incorporates

- 30 wks GA
- 36 wks GA
- 5 wks old
- 8 mo
- 12 mo
- 4 yrs

Asymmetric fusion of Kerckring-supraventricular synchondrosis → misshapen foramen magnum

Kleeblattschild
Plagiocephaly

Hypertrophied Kerckring’s ossicle & over-ambitious fusion leads to:

- Triangular shaped foramen magnum & ossified inferior internal occipital crest

Foramen magnum in normal 11 day old
X-linked chondrotyplasia punctata in 13 day old

Accessory bifid ossification center replacing Kerckring’s ossicle:

Will lead also lead to a malformed foramen magnum

Foramen magnum in normal 11 day old
In 8 wks old
Incidental bifid cpiestion
Absent ossification of Kerckring’s ossicle leads to:

- An oval foramen magnum (absent bony opisthion)
- Median cerebellar suture (in pre-term with acelidocranial dysostosis)

While pan-fusion intraoccipital synchondroses leads to: Coarced foramen magnum

- Foramen magnum in normal 11 day old
- Kleeblattschadel in 7 day old

Factors affecting sutural fusion include:

- Defective dural-mesenchymal signaling issues
  - Thought to be idiopathic
- Syndromes
  - Remember to assess extremity plain films
- Metabolic disorders
  - Hypothyroidism
  - Hypophosphatemic vitamin D-resistant rickets
  - Inborn errors of metabolism
    - Mucopolysaccharidoses / mucolipidoses / PBD (Zellweger)

Influence of intrauterine position is controversial

- However, synostoses more common with multiple births & uterine constraint

Onward to Craniosynostoses

- 1:2,500, heterogeneous group of disorders with premature osseous collateration of cranial sutures
- Leading to calvarial (and facial) distortion
- Single suture, non-syndromic fusion = most common
  - Still may have genetic etiology

Sagittal synostosis: most common

- Growth at patent metopic, coronal, squamous, lambdoid sutures ⇒ scaphocephaly
- Chiari 1 not uncommon
- Other malformations rare
**Beaking, bridging & endosteal notch**

- Restricted occipital growth & continued growth at open sutures

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**Metopic suture next most commonly stenosed**

- Frontal squama, separate at birth, fuse between 3 & 9 months of age along the metopic (AKA frontal) suture
- The metopic suture closes from nasion to anterior fontanel

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**Brain in metopic synostosis may be normal or small**

- Trigonoccephaly
- Microcephaly vera
- 19 wks GA
- Due to deficient brain growth, as in MCPH vera & HPE

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**Typical orbit features in metopic synostosis are:**

- Hypotelorism & “quizzical” orbits

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**‘Beak & notch’ in metopic synostosis**

- Ectocranial beak or ridge seen in both normally & prematurely fusing metopic sutures
- Endocranial notch only seen in synostoses cases (controversial)

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**Next: Unilateral lambdoid synostosis**

- Least likely to be treated (or imaged)
- Note typical ipsilateral mastoid “bump”
- This child has Muir-Torr syndrome with additional ipsilateral coronal synostosis
- If > 1 synostosis, think syndromal
Unilateral lambdoid synostosis:
- Leads to mild contralateral frontal bossing
- Note that the ipsilateral ear is pushed forward

Unilateral lambdoid synostosis differs from skew
- Lambdoid synostosis
  - Contralateral frontal bossing
  - Trapezoid configuration
- Positional plagiocephaly
  - Ipsilateral frontal bossing
  - Parallelogram configuration

Bilateral lambdoid synostosis is rare...
Leads to a flat occiput & can be seen in progressive stenoses

Mesenchymal thinning (holes) confirm sutural fusion in this 4 month old with Apert

Before coronal synostoses, back to anatomy
- coronal
- fronto-sphenoidal
- zygomatico-frontal
- squamosal
- lambdoid suture
- parieto-frontal
- parieto-lateral fontanel
- spheno-squamosal suture

Sutures lateral view, 1 month

A word about the fontanels
- Pterion is seen at the junction of frontal, temporal, sphenoid & parietal bone (pterion fontanel seen in prem)
- Bregma point = anterior fontanel
- Lambdoid point = posterior
- Asterion is the postero-lateral fontanel (closes at 1 month)
- Delayed fusion of fontanels from any etiology often associated with larger numbers of sutural bones

Imaging in unilateral coronal synostosis reveals
- Progressive plagiocephaly & angling of sagittal suture
- Nasal tilt due to involved naso-maxillary suture
- Contralateral forehead bulging
It is more than just coronal synostosis
Here is an obliterated coronal suture
... and nearly obliterated sphenoparietal & sphenofrontal sutures

With coronal synostosis: the hemi-ring (uniconoral) or entire cranial coronal ring (biconoral) is involved

- Coronal ring/hemi-ring
  - Separates the frontal & sphenoid bones
  - Classically includes the contiguous frontoparietal (coronal), frontosphenoidal & frontothmoidal sutures (Rogers et al. Plast Reconstr Surg 2006;118:718-73)
  - Also includes sphenooccipital & sphenoparietal (Cattermull et al. Neuroradiology 2014;56:685-7)

Additionally, early sphenoccipital synchondrosis fusion in the syndromic biconoral synchondroses contributes to:
- Decreased AP skull base growth
- Angled skull base
- Progressive midface retrusion

The "Herlequin" eye is not the only ocular finding in the coronal synostoses

Additional ocular findings are: hypertropia, strabismus, malfunctioning or abnormal oblique muscles

"Herlequin" eyes are also seen in bilateral involvement

Apert infant, serial images:
Note the large fontanel (instead of metopic synostosis)

... And all of the biconoral synostoses can exhibit exorbitism
Syndromic bicoronal synostoses typically have FGF/R gene mutations & anomalies of the extremities

The “best known” is Apert
- Most severe syntactody in Apert is seen with Pro253Arg mutation in FGFR2

Other extremity anomalies in the FGF/R gene mutations are common

Classically:
- Pfeiffer: Broad thumbs
- Crouzon: Normal extremities
- Muenke: Tarsal/carpal coalitions

However, a wide range of syntactody, anomalies & coalitions can be seen in each

As in this child with Pfeiffer syndrome

The syndromic bicoronal synostoses often progress

Crouzon syndrome
- Bi-coronal & frequently metopic synostosis → turricphaly
- Malar / midface hypoplasia, long face, increased gonial angle

Progression to Kleebattschädel: clover leaf

- Pansynostosis & bulging temporal squamosa
- May be seen with multiple disorders, in particular:
  - Congenital & syndromic synostoses: Pfeiffer type 2, etc
- But also with:
  - Brain growth failure
  - Metabolic: I-cell disease

Temporal lobes ‘push out’ the temporal squama in severe kleebatschädel
**Kleebletschädel** often rapidly progressive
- 7 days: focal stenosis sagittal suture, large squamosal suture
- 8 weeks: loss of squamosal suture, early coronal synostosis, faster fusing

**Fused foramen magnum: the best clue**
- The "waist" & bulging temporal squamosa; were additional early clues to kleebletschädel (rather than sagittal synostosis)
- 7 days
- 8 weeks

**Universal synostosis**
- May also be acquired, due to inadequate brain growth
- 3 day old with HIE & diffuse diffusion restriction

Follow up at 3 months: Microcephaly & palpable overlapping sutures

**A quick word about pre- & postoperative complications**
- Complications are relatively rare, despite extensive calvarial surgery... Except in the syndromic bicornal synostoses
- Hydrocephalus
- Chiari I
- Coronalization of foramen magnum
- Basilar imnagination
- Venous compression or thrombosis
- Infection
- Segmentation & fusion anomalies
- Hydroxyasphyxia

Let's look at complications in a single disorder: for example, Crouzon

- Venous complications predater surgery (2 different children)
  - Jugular foramen stenosis
  - Venous collaterals

Pre-operative sinus thrombosis
- Another Crouzon child with straight sinus thrombosis (Chiari I): following stent placement, no craniofacial surgery yet
Collateral veins may also follow operative intervention

Due to skull base distortion or damage to dura

And arteries may become unusually positioned...

"Kissing" carotids, due to early transverse synchondrosis fusion in Crouzon patient

Finally, remember to assess the cervical spine

Chiari 1 in Crouzon

Syrinx & basilar invagination
Segmentation / fusion anomalies

Summary

1. Knowledge of basic skull anatomy
   Crucial
2. 3d reconstructions extremely useful
3. Remember associated anomalies
   CNS & extra-CNS
4. Look for complications that may complicate surgical approach
   Vascular, spinal, CNS

Thank-you for your attention!