THE CUTTING EDGE OF PEDIATRICS

Texas Children's Hospital®

Large Heads, Flat Heads, Small Heads, Abnormally Shaped Heads

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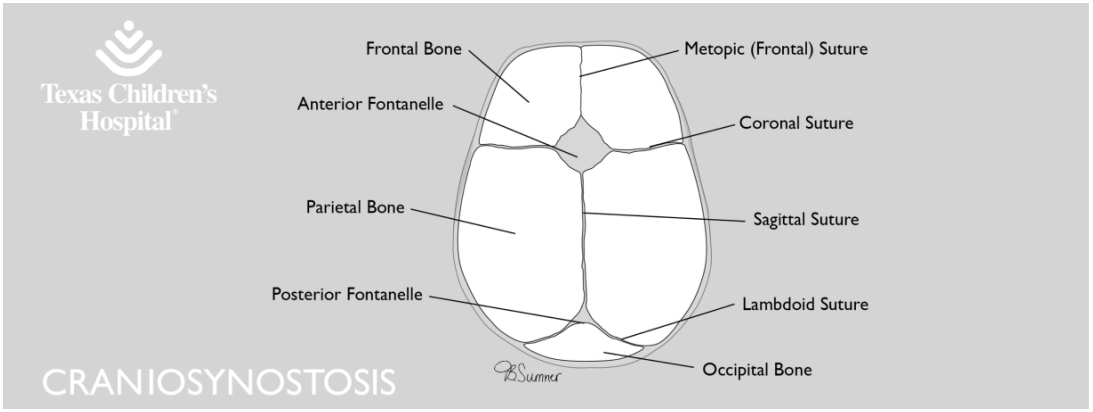
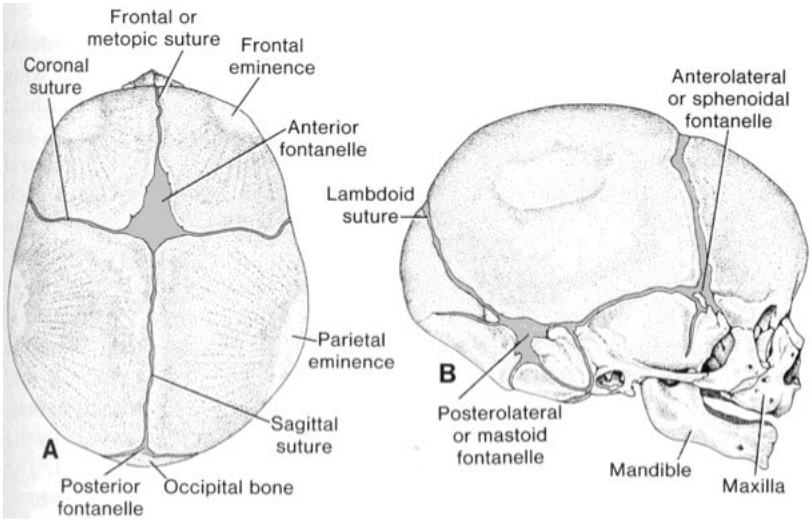
Abnormally Shaped Heads



Major Sutures Lines and Fontanels



Craniofacial Development

- Sutures line located at the junction of two bones
  - Corresponds to major dural reflections
- Major suture lines:
  - **Coronal**
  - **Sagittal**: Between the two parietal bones
  - **Squamosal**: Between the parietal bone and squamous part of the temporal bone
  - **Lambdoid**: Between the paired parietal bones and occipital bone
  - **Metopic**: Between the frontal bones
- Fontanels represent unossified regions of adjacent bone
  - 6 fontanels: Anterior, Posterior, Sphenoid and Mastoid

Craniofacial Development

- Sutures: fibrous tissues that unite the bones of the skull
  - Play a critical role in bone formation
- Sutures function as bone growth sites
  - Supply bone to the growing bone edge
  - Requires extrinsic signaling/bone formation
    - Brain expands → Stress on dura → Signaling to the suture → Bone formation
- **Sutures must be patent** for the skull to develop
  - Patency is maintained by molecular mechanisms
  - Errors in these mechanisms may cause premature fusion of the sutures → Craniosynostosis

Craniosynostosis: Background

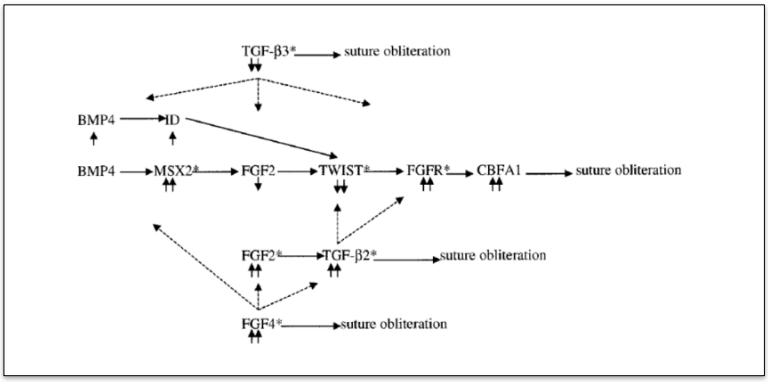
Premature ossification of a cranial suture – likely results from failure of the mechanisms that maintain suture patency

Epidemiology

- 1:2500 live births
- No gender predilection: Bilateral coronal, metopic, lambdoida
- Sagittal – M:F / 4:1
- Unilateral coronal – F:M / 3:2

Classifications

- Single vs. multiple sutures
- Syndromic vs. nonsyndromic
  - **Syndromic:** dysmorphisms of the face, trunk, limbs
  - **Nonsyndromic:** typically single suture. Multiple or pansuture craniosynostosis is described but rare



Syndrome	Gene	Major Clinical Manifestations	Craniosynostosis
Apert	FGFR2	Symphalangism, syndactyly, radiohumeral fusion, learning disabilities, craniofacial deformity	Bicoronal synositosis
Crouzon	FGFR2	Crouzonoid facies, no syndactyly	Bicoronal/Pansynostosis
Pfeiffer	FGFR1	Broad, laterally deviated first digits, cutaneous syndactyly	Kleeblatschädel, milder variations
Muenke	FGFR3	Macrocephaly, sensorineural deafness, thimble-shaped phalanges, carpal and tarsal fusion	Uni/bicoronal synostosis, cloverleaf skull
Saethre-Chotzen	TWIST1	Broad laterally deviated first digits, characteristic facies, craniofacial and vertebral defects	Uni/bicoronal synostosis

Clinical Consequences

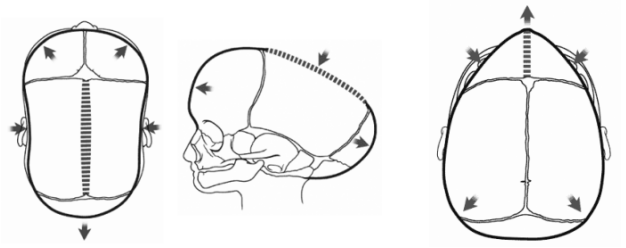
- Generally, intracranial volume is approximately equal to that of “normal” cranial vaults
- Slight increased risk of increased ICP in some studies
- Generally, no significant changes in IQ in non-syndromic craniosynostosis
- Overwhelmingly an issue of cosmesis

Craniosynostosis Diagnosis

- Typically made by characteristic headshape with subsequent imaging to confirm ossification of the suture
  - CT w/ 3D reconstruction is modality of choice

Craniosynostosis

Virchow's principle:
Growth restricted perpendicular to suture: Compensatory growth along other patent sutures

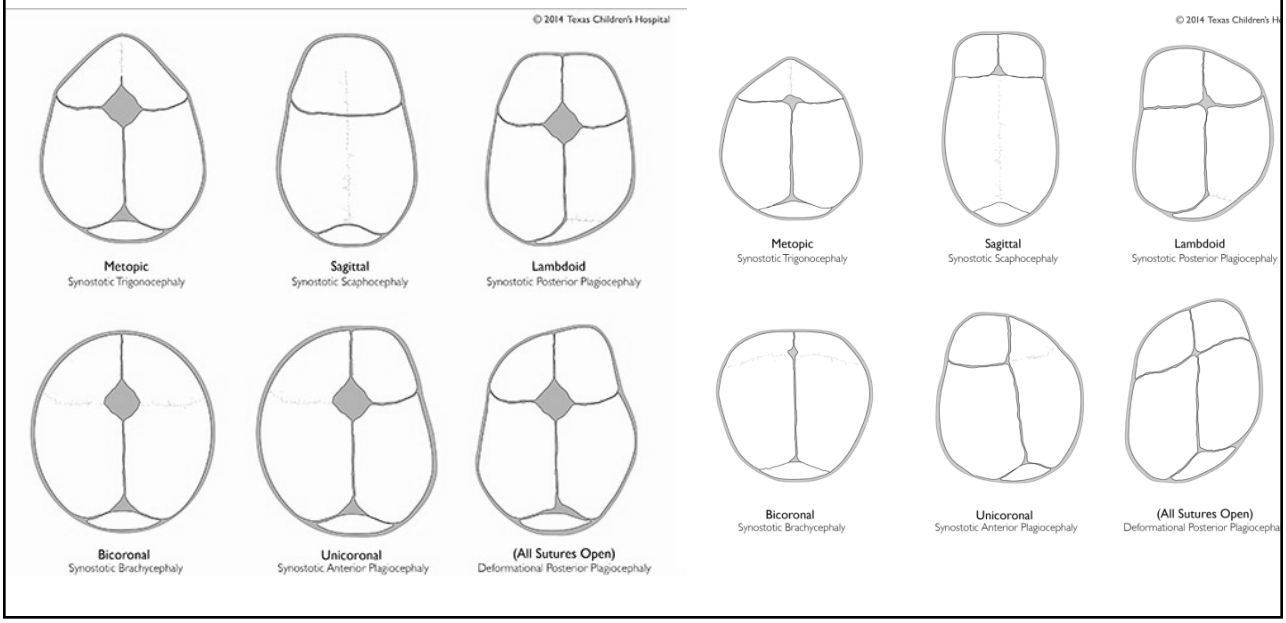


Morphology is determined by 4 principles:

- Calvarial bones adjacent to an ossified suture act as a single bone with limited growth potential
- Asymmetrical bone deposition at the sutures along the perimeter of the bone plate with increased deposition at the outer margin
- Nonperimeter sutures in line with the fused suture deposit bone symmetrically at their sutural edges
- Perimeter sutures adjacent to the fused suture compensate to a greater degree than the other distant sutures

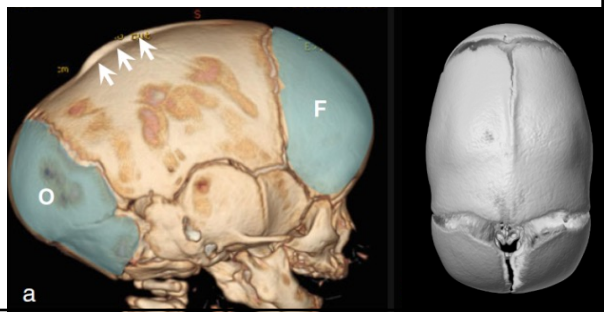
Fusion of Sutures Produce Certain Head Shapes

Synostosis	Deformity	Prevalence (M:F)
Sagittal	Scaphocephaly	50% (4:1)
Metopic	Trigonocephaly	20% (3:1)
Unicoronal	Anterior plagiocephaly	20% (2:3)
Bicoronal	Brachycephaly	<10%
Unilambdoid	Posterior plagiocephaly	<10%
Bilambdoid	Turricephaly – posterior tall cranium	<10%
Bilambdoid + sagittal	"Mercedes Benz" synostosis	<10%
Sagittal + bicoronal +/- metopic	Oxycephaly – anterior tall cranium	<10%
Sagittal + bicoronal +/- bilambdoid	Kleeblatschädel: cloverleaf cranium	<10%



Sagittal Synostosis

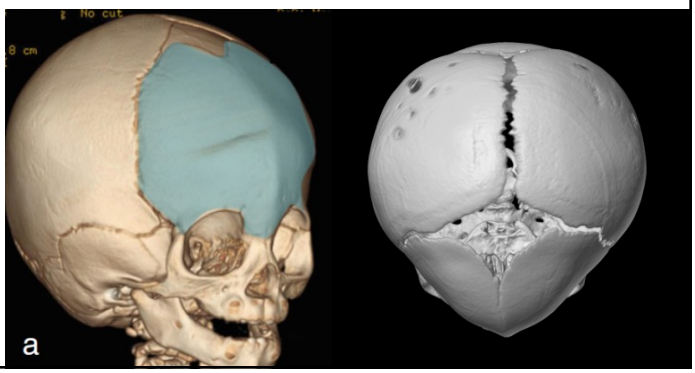
- Most common
- Scaphocephaly
  - "Inverted boat with a keel"
- Decreased biparietal diameter
- Frontal and/or occipital bossing



Metopic Synostosis

Trigonocephaly

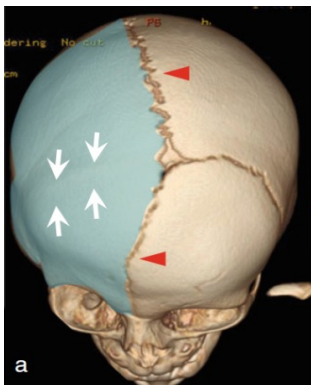
- Triangular, pointed forehead
- Frontal and bitemporal narrowing – narrow anterior fossa
- Parietoccipital bossing
- Hypotelorism

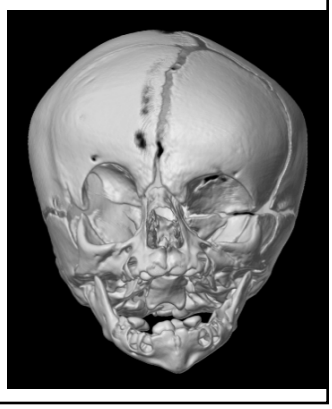


Unicoronal Synostosis

Anterior plagiocephaly – "oblique cranium"

- Ipsilateral frontal flattening and contralateral bossing
- Widened biparietal dimension
- Rotated midface – nose towards, chin away
- "Harlequin eye"

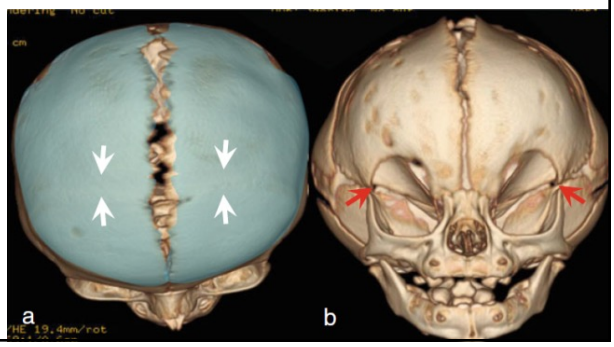




Bicoronal Synostosis

Brachycephaly – "short cranium"

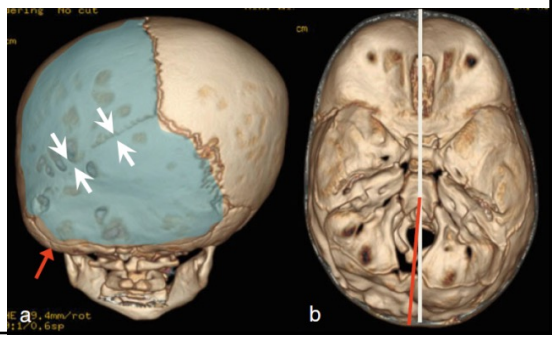
- Commonly seen in syndromic synostosis
- Shortened AP dimension – frontal/occipital flattening
- Biparietal widening
- Bilateral "harlequin eyes"



Unilamboid Synostosis

Posterior plagiocephaly

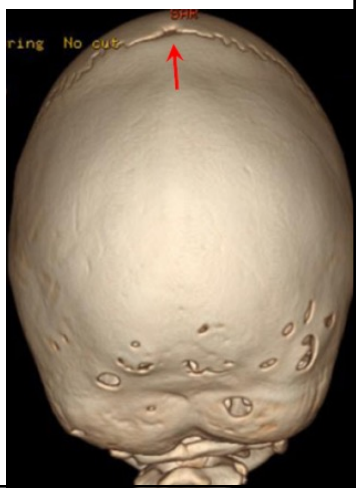
- Ipsilateral occipitoparietal flattening
- Contralateral occipitoparietal and frontal bossing
- Ipsilateral ear posteriorly displaced
- Trapezius shaped – posterior fossa deformed toward affected side



Bilamboid Synostosis

Turricephaly (acrocephaly/oxycephaly)

- Turricephaly - “tall cranium”
- Bilateral occpitoparietal flattening
- Compensatory growth in cranio-caudal direction



Deformational Plagiocephaly

- Mimics unilateral lamboid synostosis
  - Occipital flattening from lying on one side of skull
- Deformational plagiocephaly has become more common since the initiation of the “Back to Sleep” Campaign in 1992
  - Incidence as high as ~46.6%
- Treated conservatively

Positioning and SIDS

AAP Task Force on Infant Positioning and SIDS

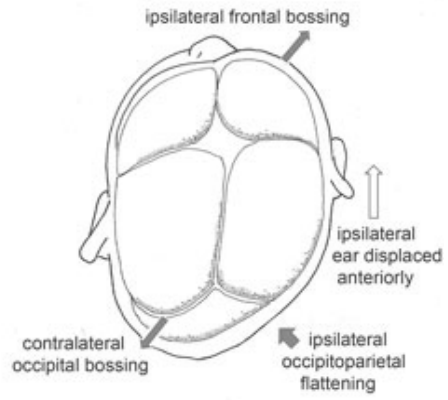
SUMMARY STATEMENT

Based on careful evaluation of existing data indicating an association between Sudden Infant Death Syndrome (SIDS) and prone sleeping position for infants, the Academy recommends that healthy infants, when being put down for sleep, be positioned on their side or back. The most common position currently used in the United States is prone.

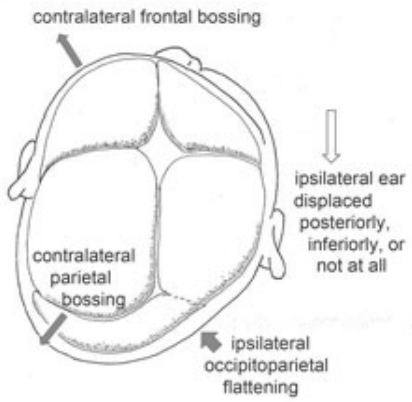
Common Sleep Positions

The predominant infant sleeping position appears to vary considerably from country to country. In the United States, most infants are placed in their beds prone (Hoffman H. National Institute of Child Health and Human Development: Cooperative Epidemiological Study of Sudden Infant Death Syndrome Risk Factors. Personal communication, 1992). Reasons

Positional Plagiocephaly



Unilat. Coronal/ Lambdoid Synostosis

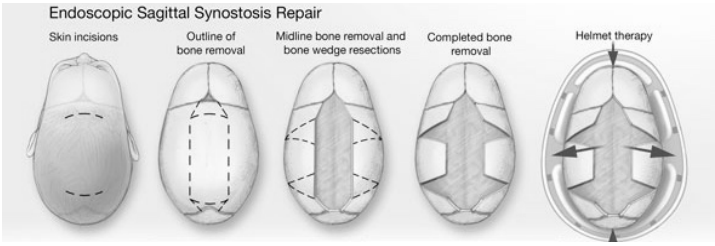


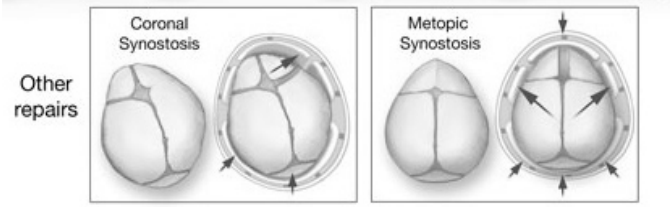
Positional Plagiocephaly

- Recommend repositioning off “flat” side as early as possible
  - Tummy time and sitting up off the back of the occiput
- Start physical therapy as soon as possible for torticollis or preferential head tilt
- Avoid Rock n Play Sleepers or swings for sleeping
  - They mold the back of the head
- 0-4 months = reposition and therapy in hopes of following natural history
- 5-7 months = “helmet heaven”
- 9 + months = unable to helmet

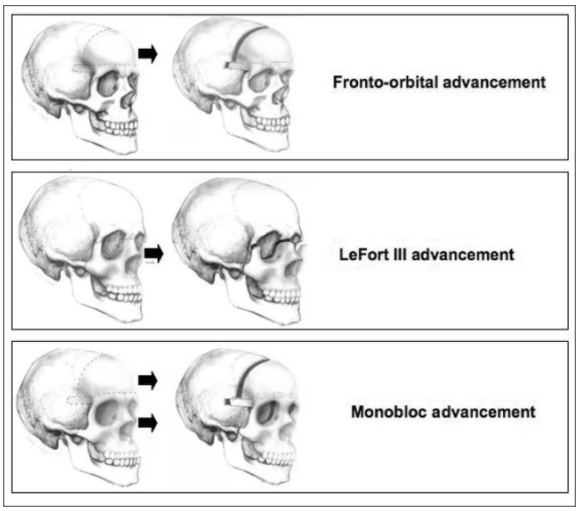
If any concerns for craniosynostosis, please place referral immediately
Call 832.822.3950

Surgery: Endoscopic Suturectomy, 1-3 Months

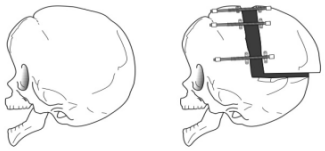




Surgery, Open, Vault Expansion, Age 9-11 Months



Posterior Cranial Vault Expansion with Distraction Osteogenesis



Take Home Points

Positional Plagiocephaly:

- Recommend repositioning off “flat” side as early as possible
  - Tummy time and sitting up off the back of the occiput
- Start physical therapy as soon as possible for torticollis or preferential head tilt
- Helmet between 5-7 months (If needed)

Craniosynostosis:

- Most kids are nonsyndromic with normal IQ
- Surgery for craneosynostosis is for cosmesis

THE CUTTING EDGE OF PEDIATRICS

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Pediatric Traumatic Brain Injury

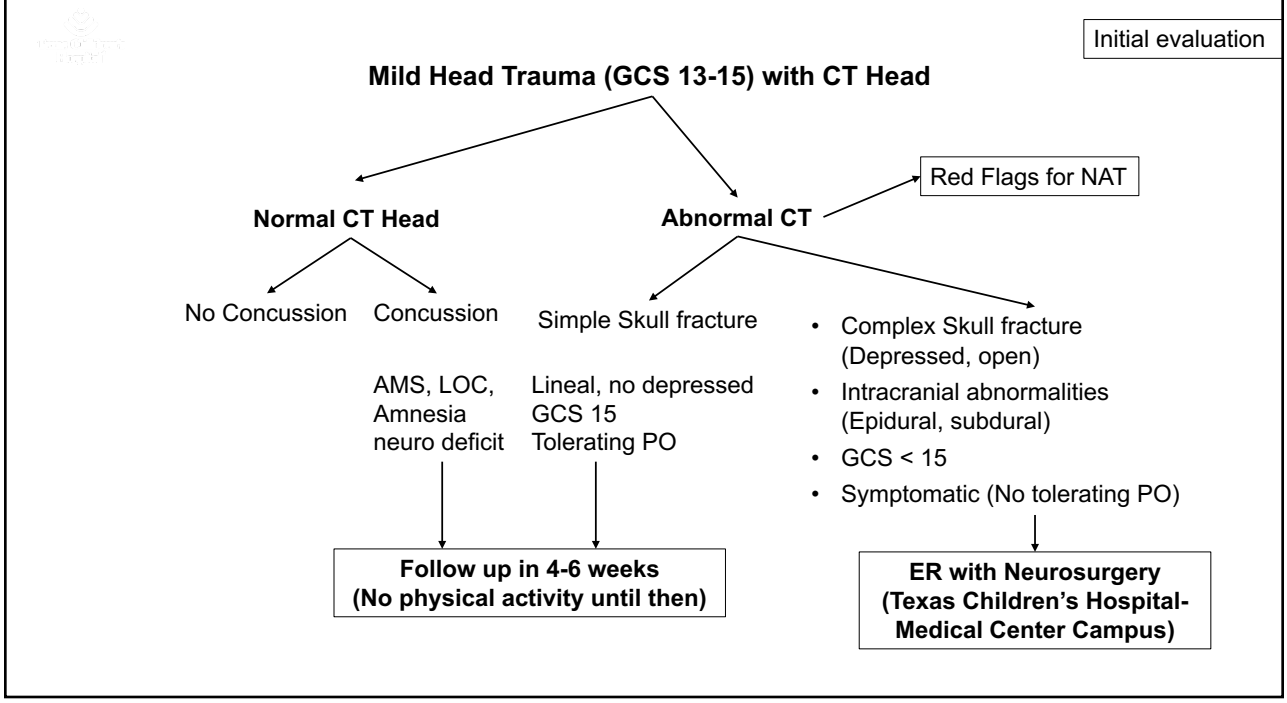
Pediatric TBI Epidemiology

- 435,000 ER visits
- 37,000 Hospitalizations
- 2,500 deaths
- 637 pages to neurosurgery junior residents

Walker, 2009 J Trauma

Sources of TBI

- Falls
- Transportation related
  - MVC
  - Bicycle
  - Peds v auto
- Non-accidental trauma (NAT)

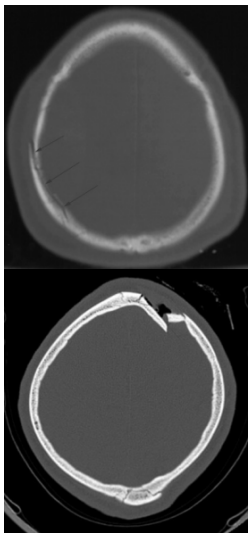


Every single patient with
head trauma & abnormal CT
(except from simple lineal skull fracture)
NEEDS
a neurosurgery evaluation

Skull Fractures

Linear
- +/- ICH
- Typically do not require intervention

Depressed
- Open vs closed
- Surgery
  - Depression > thickness of calvaria
  - Dural laceration / CSF leak
  - Neurologic deficit
  - Cosmesis



Initial evaluation

Mild Head Trauma (GCS 13-15) with CT Head

Normal CT Head

No Concussion

Concussion

AMS, LOC, Amnesia neuro deficit

Lineal, no depressed GCS 15 Tolerating PO

Follow up in 4-6 weeks (No physical activity until then)

Abnormal CT

Red Flags for NAT

Simple Skull fracture

- Complex Skull fracture (Depressed, open)
- Intracranial abnormalities (Epidural, subdural)
- GCS < 15
- Symptomatic (No tolerating PO)

ER with Neurosurgery (Texas Children's Hospital-Medical Center Campus)

Concussion: A Mild Form of TBI

Concussion is a traumatically induced physiological disruption of brain function where the Glasgow Coma Score range is 13 to 15, and has at least ONE of the following manifestations

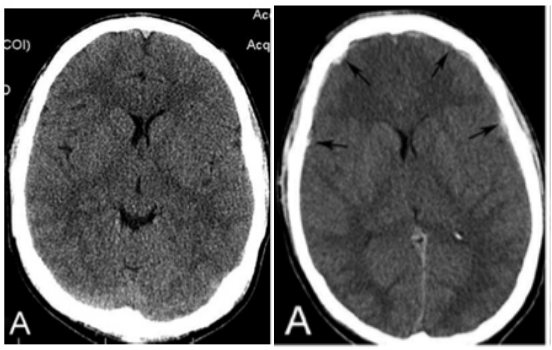
- Any alteration in mental status at the time of the accident (example: feeling dazed, disoriented, or confused
- Any loss of memory for events immediately before or after the accident, where the amnesia is less than 24 hours
- Any loss of consciousness less than 30 minutes
- Focal neuroligical deficits that may or may not be transient

Or "a complex pathophysiological process affecting the brain, induced by traumatic biomechanical forces"

Does not require loss of consciousness
Imaging studies will usually be normal

Second Impact Syndrome

- Second head injury while symptomatic from previous injury
- Impaired cerebral autoregulation
  - Hyperemia
  - Malignant cerebral edema
- Mortality 50-100%



After initial impact
CGS 15

After second impact
CGS 7T

Weinstein, 2013 JNS peds

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Mild Head Trauma (GCS 13-15) with CT Head

Normal CT Head

No Concussion

Concussion

AMS, LOC, Amnesia neuro deficit

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Lineal, no depressed
GCS 15
Tolerating PO

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ER with Neurosurgery (Texas Children's Hospital-Medical Center Campus)

Severity of NAT

Table 1 Comparison of the demographics of non-accidental trauma (NAT) to accidental trauma (AT) patients from 2007–2011.

Patient Demographics	NAT (n = 267) Median (range)	AT (n = 4781) Median (range)	P-value
Age in months	7 (0.4 - 122)	72 (0.03 - 228)	<0.001
Male, sex	61%	64%	0.327
Private Insurance	19%	40%	<0.001
Race			
Hispanic	36.0%	34%	0.507
African American	34%	15%	<0.001
White	25.8%	36.3%	<0.001
ISS	13 (1–50)	9 (1–59)	<0.001
Length of stay, days	3, (1–63)	1 (1–106)	<0.001
% pt admitted to the ICU	34%	9%*	<0.001
Mortality	7%	0.3%	<0.001

Red Flags for Nonaccidental Trauma

1. CT Findings consistent with moderate or high energy on the impact:
   - Fracture crossing sutures
   - Bilateral Fractures
   - Intracranial abnormalities (Subdural, epidural)
2. Fracture without history of Trauma (“Feel a bump on his/her head”)
3. Inconsistency of the history (2 or more versions of what happened)
4. Incongruence of the history (baby of 1-3 month old does not roll over)

Follow up Visit (4-6 weeks)

If symptoms or edema above the fracture persist refer to Neurosurgery
Neurosurgery Head Injury Clinic (832-822-3950)

Concussion at now asymptomatic
Resume physical activity progressively. (If he/she has symptoms stop for 24h)

Skull fracture recommendations:
1. No activity greater than walking for 3 months
2. No routine imaging done if asymptomatic and physical exam normal

Signs and Symptoms of a Concussion

Physical	Cognitive	Emotional	Sleep
Headache	Feeling mentally ”foggy”	Irritability	Drowsiness
Nausea	Feeling slowed down	Sadness	Sleeping more than usual
Vomiting	Difficulty concentrating	More emotional	Sleeping less than usual
Balance problems	Difficulty remembering	Nervousness	Difficulty falling asleep
Visual problems	Forgetful of recent information		
Fatigue	Confused about recent events		
Sensitivity to light	Answers questions slowly		
Sensitivity to noise	Repeats questions		
Dazed			
Stunned			

Return to Play

Based on clinical symptoms – can progress to next level if no symptoms

Rehabilitation Stage	Functional Exercise
1. No activity	Complete physical and cognitive rest
2. Light aerobic activity	Walking, swimming, stationary cycling at 70% maximum heart rate, no resistance exercises
3. Sport-specific exercise	Sport specific related drills but no head impact
4. Noncontact training drills	More complex drills, may start light resistance training
5. Full contact practice	After medical clearance participate in normal training
6. Return to play	Normal game play

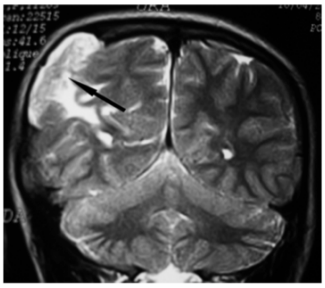
Each stage should lass no less than 24 hours, with a minimum of 5 days required to consider a full return to competition. If symptoms recur during the rehabilitation program, the athlete should stop immediately. Once asymptomatic after at least another 24 hours, the athlete should resume at the previous asymptomatic level and try to progress again. Contact healthcare provider if symptoms return.

Follow up

- Neurosurgery Head Injury Clinic (832-822-3950)
  Follow up if needed in 4-6 weeks
- Screens for post-concussive symptoms & surgical intervention
- Referrals made to Physical Medicine and Rehabilitation Clinic for post-concussive symptom management, if screening positive
- Surgical intervention if growing skull fractures

Growing Skull Fractures

- Post traumatic leptomeningeal cyst
  - 1% of all skull fractures
- Skull fracture w/ dural laceration
  - Entrapment of arachnoid in fracture
  - Herniation of brain through laceration
  - Enlarging pulsitile scalp mass



Repair requires closure of dural defect