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# **Craniosynostosis and Other Congenital Deformities of Skull, Face, and Jaw**

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# Objectives for Today's Discussion

## **Craniosynostosis and deformations of skull, face, and jaw:**

1. Introduce the clinical background
2. Describe experiences with clinical classification of the current ICD codes
3. Propose our hopes to optimize the clinical classification of the codes

# Acknowledgements

## **Contributing societies and organizations**

- American Society of Pediatric Neurosurgeons
- American Society of Craniofacial Surgeons
- American Academy of Pediatrics
- Children's Hospital Association
- Children and Youth with Special Healthcare Needs Research Network

# Craniosynostosis

## Clinical Background

- **Definition**

- Premature closure of one or more sutures that separate the bones of the cranium

- **Impact**

- Abnormally shaped head
- Risk for increased intracranial pressure, which can damage the eyes and brain

- **Epidemiology**

- 1 in 2000 births

# Craniosynostosis

## Clinical Background

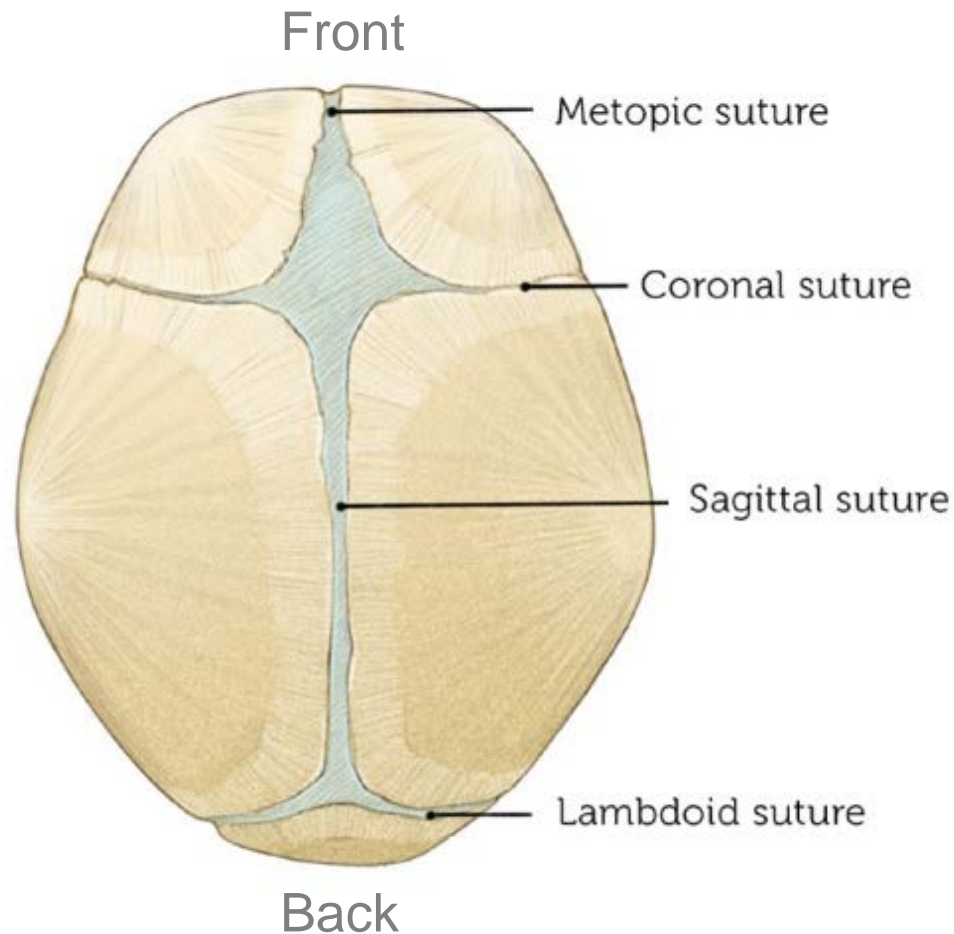
- **Clinically classified according to the involved suture(s)**
  - Sagittal (60%)
  - Coronal\* (25%)
  - Metopic (15%)
  - Lambdoid\* (2%)
  - Other (<1%)

*\*extend laterally, left and right*

# Craniosynostosis, cont.

## Clinical Background

### Normal Cranium

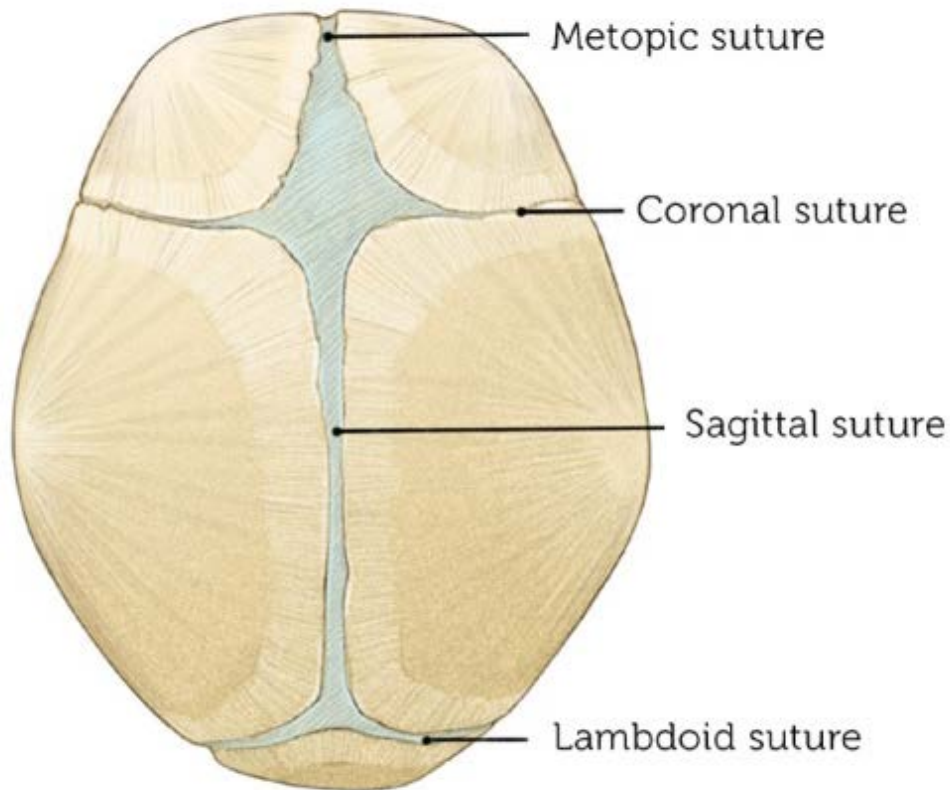


# Craniosynostosis. cont.

## Clinical Background

### Normal Cranium

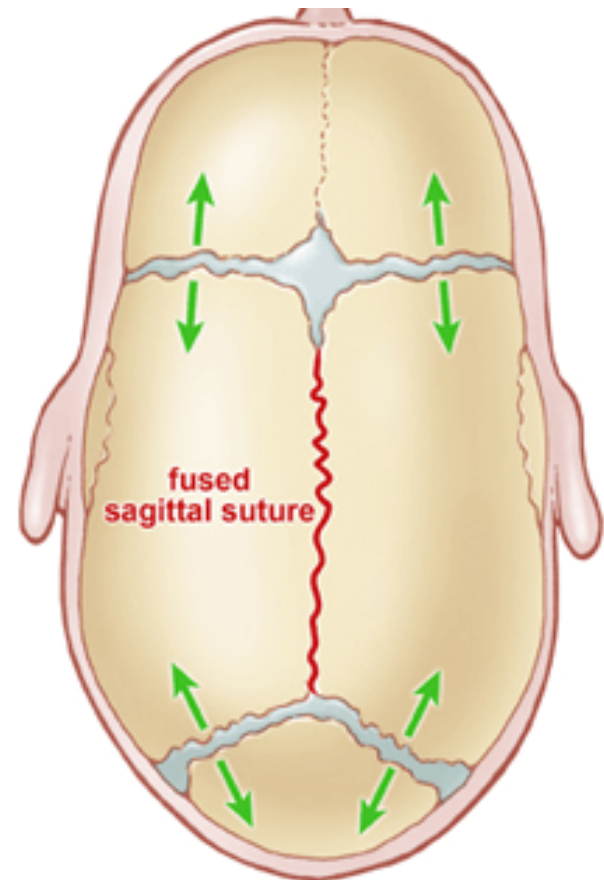
Front



Back

### Scaphocephaly

Sagittal craniosynostosis



# **Craniosynostosis,** Clinical Background

- **Clinical work-up**

- General pediatricians screen for it
- Referral made to craniosynostosis specialists
- Treatment and outcomes vary by craniosynostosis type

- **Non-synostosis head deformations**

- Not all deformations are craniosynostosis
- Metopic ridge, sagittal crest can be benign



# Craniosynostosis

Clinical Background **cont.**

## Benign Metopic Ridge



# Craniosynostosis

Current ICD Diagnosis Code

## ICD-10-CM Hierarchy

Q00-Q99 - Congenital malformations, deformations and chromosomal abnormalities

Q65-Q79 - Congenital malformations and deformations of the musculoskeletal system

Q75 - Other congenital malformations of skull and face bones

**Q75.0 – Craniosynostosis**

# Craniosynostosis

Current ICD Diagnosis Code **cont.**

## **Q75.0 – Craniosynostosis**

Acrocephaly, imperfect fusion of skull, oxycephaly, trigonocephaly

### ***Clinical Classification Challenges:***

- *Suture(s) and laterality are not distinguished*
- *Inclusion terms incomplete*

# Skull Deformities

Current ICD Diagnosis Code

## ICD-10-CM Hierarchy

Q00-Q99 - Congenital malformations, deformations and chromosomal abnormalities

Q65-Q79 - Congenital malformations and deformations of the musculoskeletal system

Q67 - Congenital musculoskeletal deformities of head, face, spine and chest

**Q67.4 – Other congenital deformities of skull, face, and jaw**

# Skull Deformities, cont.

Current ICD Diagnosis Code

## **Q67.4 – Other congenital deformities of skull, face, and jaw**

Congenital depressions in skull; Congenital hemifacial atrophy or hypertrophy; Deviation of nasal septum, congenital; Squashed or bent nose, congenital; Asymmetric crying face association; Asymmetric crying facies; Brow asymmetry, congenital; Congenital asymmetry of forehead; Congenital facial asymmetry

***Skull deformations mixed in with others***

***Skull deformation terms incomplete***

# References

Gonzalez SR, Han A, Golinko MS. Shifting epidemiology of single-suture craniosynostosis and the need for a more granular ICD classification system: a national survey of members from the American Society of Pediatric Neurosurgeons (ASPN) and the American Society of Craniofacial Surgeons (ASCFS). *Childs Nerv Syst.* 2019;35(9):1443-1444

Gonzalez SR, Light JG, Golinko MS. Assessment of Epidemiological Trends in Craniosynostosis: Limitations of the Current Classification System. *Plast Reconstr Surg Glob Open.* 2020;8(3):e2597

# References, cont.

- <https://www.ohsu.edu/doernbecher/pediatric-sagittal-synostosis>
- <https://babyandbump.momtastic.com/threads/metopic-ridge-vertical-ridge-on-forehead-anyone.664779/>

**Thank you!**