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

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Objectives for Today's Discussion

Craniosynostosis :

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

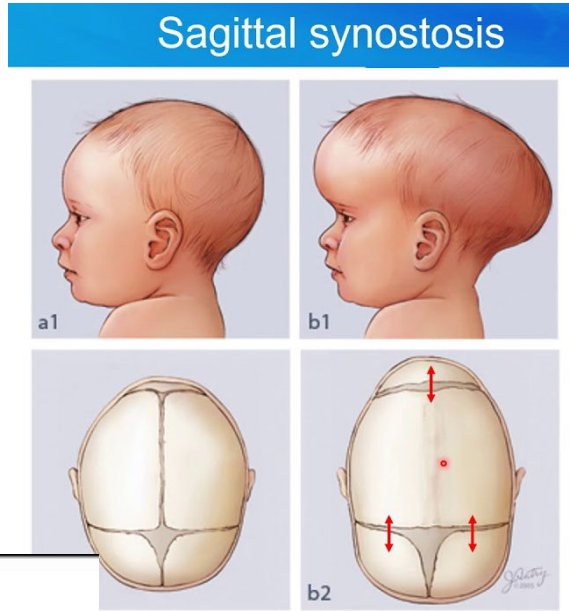
- **Definition**
  - Premature closure of one or more sutures that separate the bones of the cranium
- **Impact**
  - Abnormally shaped head
  - Risk for increased intracranial pressure, which can damage the eyes and brain
- **Epidemiology**
  - 1 in 2000 births

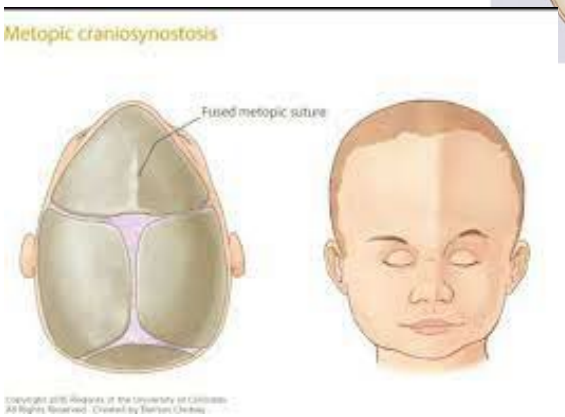
Craniosynostosis (cont'd)

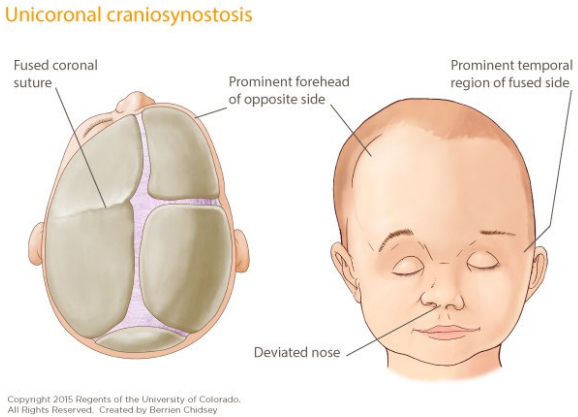
Clinical Background

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

**extend laterally, left and right*







Craniosynostosis
Clinical Background

- **Clinical work-up**
  - General pediatricians screen for it
  - Referral made to craniosynostosis specialists
  - CT or skull XR confirms anatomic diagnosis
  - Treatment and outcomes vary by craniosynostosis type
- **Non-synostosis head deformations**
  - Not all deformations are craniosynostosis
  - Plagiocephaly can be deformational
  - Metopic ridge, sagittal crest can be benign

Craniosynostosis & 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

Q75 - Other congenital malformations of skull and face bones

Q75.0 – Craniosynostosis

Craniosynostosis

Current ICD Diagnosis Code

Q75.0 – Craniosynostosis

Acrocephaly, imperfect fusion of skull, oxycephaly, trigonocephaly

Clinical Classification Challenges:

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

Craniosynostosis Q75.0

Proposed ICD Diagnosis Code Revisions

- **Specify the involved suture**
- **Specify the laterality**
- **Add inclusion terms to suture**

References

Proposed ICD Diagnosis Code Revisions

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

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Thank you!