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VIEWPOINT
Craniofacial/Pediatric

New ICD-10 Diagnosis Codes to Improve Craniosynostosis Classification

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REVIEW OF THE CLINICAL CONDITION: CRANIOSYNOSTOSIS

Craniosynostosis (CS) is the premature closure of one or more major cranial sutures that occurs in approximately one in 2000 infants born every year. In addition to the various skull and orbital deformities, neurocognitive delay and elevated intracranial pressure (ICP) are associated. Management (both surgical and nonsurgical) is a function of factors including but not limited to the affected suture(s), age at presentation, neurocognitive status, degree of skeletal deformity, and evidence of elevated ICP.

Pediatric clinicians are routinely the first to screen infants and children for abnormal head shapes. If CS is suspected, time-sensitive referral to either a plastic/craniofacial surgeon or a pediatric neurosurgeon is warranted to confirm the diagnosis with physical examination and, possibly, a head computed tomography.

NEED FOR IMPROVED CLASSIFICATION AND FORMATION OF A COLLABORATIVE WORKING GROUP

Despite the heterogeneity of CS, only one International Classification of Disease, 10th Revision (ICD-10) diagnosis code, Q75.0, exists for all types.

Members of the American Society of Pediatric Neurosurgeons, the American Society of Craniofacial Surgeons, and the American Academy of Pediatrics (AAP) requested a revision of the current CS ICD-10-CM diagnosis code (Q75.0) to provide more clinical granularity.[1,2] To that end, a multi-institutional working group of pediatric neurosurgeons, plastic surgeons, and pediatricians was assembled to revise Q75.0.

The working group solicited mentorship from international authorities on the ICD system, including members of the AAP Committee on Coding and Nomenclature and the Centers for Disease and Control (CDC) ICD-10 Coordination and Maintenance Committee (CDC-CMC). Together, the working group revised the Q75.0 code to include the necessary clinical granularity, including the type of CS (ie, sagittal, coronal, metopic, lambdoid, other, and not specified), laterality (ie, unilateral, bilateral, not specified), and number of sutures (ie, single, multiple) (Table 1).

PRESENTATION TO THE CDC AND PREVENTION

The working group presented the initial version of the proposed revisions at the Spring 2021 ICD-10 CMC meeting. The CDC-CMC committee oversees and affirms all proposed revisions to ICD diagnosis codes. The committee provided immediate feedback and stewarded an open period for public comment on the proposed revisions. The working group responded to the feedback, revised the proposal accordingly, and re-presented the updated revisions at the Spring 2022 CDC-CMC meeting.

The CDC-CMC supported the revised proposal, stewarded a second open period for public comment (during which no new revisions were suggested), and officially endorsed the proposal.

NEXT STEPS

Public release of the new CS ICD system for widespread use is set for October 2023. The complete table of newly revised codes can be found on the CDC website by clicking on the link to ICD-10-CM FY2024 Addenda PDF, downloading the zip file ICD10CM-addedum-2024.zip, and going to pages 45–46.[3] A summary of the revised codes is reproduced in Table 1.

Improved classification of the type of CS is essential for several reasons, including (1) accurate measurement and assessment of worldwide trends in the epidemiology of CS types, (2) improved accuracy in

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Table 1. Summary of ICD Diagnosis Code Revision for CS (Q75.0)

Q75.00 CS unspecified
Q75.001 CS unspecified, unilateral Q75.002 CS unspecified, bilateral Q75.009 CS unspecified
Q75.01 sagittal CS
Q75.02 coronal CS
Q75.021 coronal CS, unilateral Q75.022 coronal CS, bilateral Q75.029 coronal CS, unspecified
Q75.03 metopic CS
Q75.04 lambdoid CS
Q75.041 lambdoid CS, unilateral Q75.042 lambdoid CS, bilateral Q75.049 lambdoid CS, unspecified
Q75.05 multisuture CS
Q75.051 cloverleaf skull (includes Kleeblattschaedel skull) Q75.052 pansynostosis Q75.058 other multisuture CS Excludes: coronal CS, bilateral (Q75.022) Excludes: lambdoid CS, bilateral (Q75.042)
Q75.08 other single-suture CS

hospital billing and coding, and (3) more precise and accurate evaluation of treatment and clinical outcomes of CS.[4]

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DISCLOSURE

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