



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)

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Dear Editor:

The purpose of our letter is to bring attention to the national epidemiological studies who reported an increase in the incidence of metopic craniosynostosis and to raise awareness about the importance of developing improved International Classification of Disease (ICD) codes for craniosynostosis [1–3]. Reports of epidemiological shifts in single-suture synostosis are not limited to the USA. In the last decade, epidemiological studies across Europe have also reported a similar increase in the incidence of metopic craniosynostosis. A study done in France by DiRocco et al. revealed almost a 4-fold increase in metopic craniosynostosis, compared with a 1.7-fold increase in sagittal craniosynostosis [4]. A nine-year study using data from centers throughout Europe found a statistically significant increase in the incidence of metopic cases in both absolute numbers and in comparison with other craniosynostosis subtypes [5]. As a means to provide an insight into the views of physicians involved in the surgical management of patients with craniosynostosis, we will also share the results of a US national survey we designed to assess the perspectives of pediatric neurosurgeons (PNS) and craniofacial plastic surgeons (CPS) regarding the epidemiology of

craniosynostosis and the current craniosynostosis repair coding practices.

We performed a focused PubMed search of studies investigating the epidemiology of craniosynostosis in the USA in the past 10 years (2008–2018). We identified three studies done in large cities such as Atlanta [1], Philadelphia [2], and Michigan [3]. The studies ($n = 1628$) indicated a statistically significant increase in the incidence of metopic synostosis ranging from 6 to 12% increase per year [1–3]. The subjects included in these studies were limited to only three cities, and as a result, it is not plausible to extrapolate their data to adequately represent the US population.

To further understand the clinical impact of the reported increases in the incidence in metopic craniosynostosis, we sent out surveys to members of the American Society of Craniofacial Plastic Surgeons (ASCFS) and the American Society of Pediatric Neurosurgeons (ASPN). A total of 34 out of 136 (25%) craniofacial plastic surgeons and 23 out of 130 (18%) pediatric neurosurgeons responded to the survey. When subjectively recalling which type of single-suture craniosynostosis (SSC) is the most common five years ago vs. now, the percentage of CPS who identified sagittal to be the most common type previously vs. now decreased from 91 to 64%. The percentage of PNS who identified sagittal to be the most common type previously vs. now decreased from 100 to 87%. All respondents who reported that sagittal craniosynostosis is no longer the most prevalent SSC stated that metopic craniosynostosis is currently the most common SSC affected. The majority of craniofacial plastic surgeons (76%) and pediatric neurosurgeons (86%) agreed that it would be valuable to design International Classification of Disease (ICD) codes that reflected the specific suture involved in the craniosynostosis defect (e.g., metopic) rather than head shape (e.g., trigonocephaly). Over 95% of those surveyed agreed that there needs to be a more adequate classification system, rather

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than collectively referring to the cases as “craniosynostosis,” so that the national incidence of craniosynostosis subtypes would not be based on their subjective recall alone, but instead on a highly descriptive national database.

The responses obtained from members of the ASCFS and ASPN support the previously reported increases in the incidence of metopic craniosynostosis. Furthermore, the responses also indicate a consensus amongst both CPS and PNS regarding the need for a more granular classification system in upcoming ICD coding updates which distinguishes between the various sutural fusions. ICD codes are managed and offered to member states for ratification by the World Health Organization (WHO). The ICD-11 coding system will be presented at the World Health Assembly in May of 2019 and will come into effect on January 1, 2022. The ICD-10 code for craniosynostosis (Q 75.0) will be changed to (LB70.0Y) in the ICD-11 update, but unfortunately, only one diagnosis code remains for craniosynostosis, regardless of the suture affected.

Currently, most centers must *reinvent the wheel* to produce a unique registry to track craniosynostosis subtypes. Databases such as the National Inpatient Sample (NIS), the National Surgery Quality Improvement Project (NSQIP), and the Kid’s Inpatient Database (KID) track patients at the national level, but since their data is based on ICD-9 and ICD-10 codes, all cases of craniosynostosis are lumped together into one category. Precise classification is the cornerstone of outcomes research, but with our current system of classification, we are asked to compare “apples to apples.” The 18th Congress of the International Society of Craniofacial Surgery (ISCFS) in Paris is approaching, and this letter is intended to foster the continued dialog between craniofacial surgeons and pediatric neurosurgeons to advocate to the World Health Organization, with one voice, for a more specific classification of craniosynostosis.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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