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Assessment of Epidemiological Trends in Craniosynostosis: Limitations of the Current Classification System

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Background: Craniosynostosis affects 1 in 2,000 live births, which makes it one of the most common craniofacial abnormalities in the United States. Despite this fact, few national epidemiologic reports exist, although US and European studies have reported an increased incidence of metopic craniosynostosis. The aim of our study is to analyze the National Inpatient Sample (NIS) to support those conclusions.

Methods: We identified hospitalizations from 1998 to 2012 by using the ICD-9-CM diagnosis code for congenital anomalies of skull and face bones (756.0) and procedure codes related to craniosynostosis repair (2.01, 2.03, 2.04, 2.06).

Results: We analyzed data from 37,815 hospitalizations and 49,505 reconstructive procedures. There was a 61.6% increase in the number of hospitalizations related to craniosynostosis repairs. There was a 180% increase in bone graft to skull procedures, 109% increase in other cranial osteoplasty, 54% increase in formation of cranial bone flap, and a 6% decrease in opening of cranial suture.

Conclusions: We observed a steady rise in the number of craniosynostosis repairs performed, but whether this is a result of a true increase in incidence, better diagnosis, or change in treatment patterns needs further research. The current classification system does not provide information about the specific suture affected (metopic, sagittal, etc.), the type of repair performed (endoscopic, fronto-orbito advancement, etc.), and whether the repair is a primary procedure or a revision. More descriptive diagnosis and procedural codes are imperative to improve the epidemiologic and outcomes data of craniosynostosis in the United States. *(Plast Reconstr Surg Glob Open 2020;8:e2597; doi: 10.1097/GOX.0000000000002597; Published online 23 March 2020.)*

INTRODUCTION

Craniosynostosis is defined as the premature fusion of one or more cranial sutures resulting in abnormal calvarial development and morphology.[1] This group of defects is further classified according to the suture involved and the associated malformations.[1] Surgical procedures increase the cranial vault space by creating a more normal head shape, thus correcting the craniosynostosis condition.

The incidence of craniosynostosis is estimated to 1 in 2,000 live births.[2] The frequency of the subtype of suture involved varies significantly among single-suture craniosynostoses, with the sagittal suture being the most commonly affected.[3] Moreover, the surgical repair varies according to the type of suture involved and can range from endoscopic assisted suturectomy, to fronto-orbito advancement (FOA) to total cranial vault remodeling. Epidemiological studies in Europe and some in the United States have noted a shift in the distribution of the different subtypes of craniosynostosis with a dramatic increase in the incidence of metopic synostosis.[1,2,4–7] Although reports from national datasets have been able to address global clinical outcomes 30 days after surgery, including length of stay and blood loss for example,[8,9] clinical correlation has proved difficult to compare "apples to apples" because current ICD codes do not distinguish between different types of craniosynostosis. The aim of our study is to evaluate the evidence available, at a national level, to support conclusions made by previous urban studies regarding the shifting trends of metopic craniosynostosis in the United

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States, and to highlight opportunities for improvement in coding which would allow more accurate data collection and outcome analysis.

METHODS

Database

This retrospective, cross-sectional study used data from the National Inpatient Sample (NIS) database between 1998 and 2012. The NIS database is the largest publicly available, all-payer inpatient healthcare database in the United States.[10] It is part of the Healthcare Cost and Utilization Project group of databases sponsored by the Agency for Healthcare Research and Quality (AHRQ), a division of the US Department of Health and Human Services.[10] Each hospital included in the NIS database provides information about all hospitalizations, allowing for nationally representative estimates. Information such as primary reason of hospitalization, type and number of procedures, outcomes, and demographic characteristics are included in the database.

Cohort

Hospitalizations during the study period were identified using the ICD-9-CM procedure codes for craniosynostosis surgical repair accompanied by an ICD-9-CM diagnosis code for congenital anomalies of skull and face bones, as previously described by Nguyen et al.[11]

The procedure codes defined by the ICD-9-CM included: opening cranial suture (02.01), formation of cranial bone flap (02.03), bone graft to the skull (02.04), and other cranial osteoplasty (02.06) (Table 1). Only hospitalizations involving these surgical repair procedure codes and a diagnosis of anomalies of the skull and face bones (diagnosis code 756.0) were selected. The procedure codes for craniotomy (01.24) and craniectomy (01.25) were not included due to their significant overlap with the repair of neurological diagnosis that may decrease the overall accuracy of the data estimates. The age of patients in our study ranged from birth to 6 years.

To ensure that the data in our study remained consistent in the way data were obtained, and to analyze long-term epidemiological trends in craniosynostosis hospitalizations, we limited our study to NIS data between 1998 and 2012 originating from ICD-9 codes in a *sample of hospitalizations* from participating hospitals.[10] Starting in 2012, the NIS restructured its source of data, and it became *a sample of discharges*, rather than a sample of hospitalizations. Due to the changes in the way data were reported in the NIS, only hospitalizations up to 2012 were included in our study. Data from the ICD-10 classification system were not included in this study because the NIS first began using it in 2015, which was outside of our study period for the aforementioned reasons.[10]

Table 1. Demographic Variables of Children with Hospitalizations Related to Craniosynostosis Repair, 1998–2012, National Inpatient Sample

Variables	(n = 37,815)	
	Number	%
Age		
0–4 mo	4,945	13.0
5–12 mo	14,533	38.4
1–3 y	8,069	21.3
4–6 y	2,572	6.8
Unknown	7,695	20.3
Race		
White	18,731	49.5
Black	1,459	3.9
Hispanic	5,581	14.8
Other	2,631	7.0
Unknown	9,414	24.9
Sex		
Male	24,405	64.5
Female	13,036	34.5
Unknown	374	1.0
Insurance status		
Private	21,169	56.0
Government	14,041	37.1
Self-pay	457	1.2
Other	2,076	5.5
Unknown	73	0.2
Year of hospitalization		
1998	1,894	5.0
1999	2,556	6.8
2000	1,857	4.9
2001	2,233	5.9
2002	2,025	5.4
2003	1,858	4.9
2004	2,498	6.6
2005	4,131	10.9
2006	2,571	6.8
2007	2,017	5.3
2008	2,433	6.4
2009	2,590	6.8
2010	3,321	8.8
2011	2,770	7.3
2012	3,060	8.1
Hospital type		
Nonteaching	2,723	7.2
Teaching	31,828	84.2
Unknown	3,263	8.6
Hospital region		
Northeast	6,020	15.9
Midwest	8,257	21.8
South	12,383	32.7
West	8,095	21.4
Unknown	3,060	8.1

Patient Characteristics

We analyzed patient factors including outcomes (length of stay and charges), age, race, sex, insurance status, year of hospitalization, hospital type, and region for each hospitalization involving craniosynostosis repair. We also examined the type of procedure performed for each age group and the cumulative number of procedures during hospitalization for each age group and year of hospitalization. Hospital charges were adjusted based on the 2012 inflation rate. The independent and dependent variables were summarized using descriptive statistics due to the single sample and categorical nature of the data.

RESULTS

A total of 37,815 hospitalizations and 49,505 procedures for craniosynostosis repair occurred from 1998 through

2012 (Table 1). There was a 61.6% increase in the number of hospitalizations related to craniosynostosis repairs in the United States with an average increase, over the 15-year period, of 4.1% per year. The year with the highest craniosynostosis repairs during this period was 2005, with 4,131 hospitalizations and 5,583 procedures. There was a shift in the frequency of each procedure during the study period with a 180% increase in the rate of bone graft to skull, 109% increase in other cranial osteoplasty, 54% increase in the formation of cranial bone flap, and a 6% decrease in the rate of the opening of cranial suture.

The most frequently performed surgical procedure during the study period was other cranial osteoplasty (44.6%), followed by the opening of cranial suture (28.5%), bone graft to the skull (16.1%), and formation of a cranial bone flap (10.9%). The most frequent procedure in the combined 0- to 4-month age group was opening of cranial suture (50.3%). Other cranial osteoplasty was the most frequent procedure for the 5- to 12-month age group (46.5%), 1- to 3-year age group (47.4%), 4- to 6-year age group (54.7%), and the unknown age group (39.1%) (Table 2). Most hospitalizations required only one type of procedure for the repair (73.1%) (Table 3). Most hospitalizations were comprised of patients aged 1 year or younger (n = 19,478). Infants 4 months and younger constituted 13.1% of the sample, whereas the most prevalent age group was 5–12 months at 38.4%. Caucasians (49.5%) and men (65%) were predominant in the sample.

Table 2. Surgical Procedures to Repair Craniosynostosis, 1998–2012, National Inpatient Sample

Variables	Opening of Cranial Suture (n =14,088)		Formation of Cranial Bone Flap (n = 5,387)		Bone Graft to Skull (n = 7,973)		Other Cranial Osteoplasty (n = 22,057)	
Age Categories	Number	%	Number	%	Number	%	Number	%
0–4 mo	3,036	21.6	353	6.6	441	5.5	2,205	10.0
5–12 mo	5,309	37.7	2,212	41.1	2,799	35.1	8,959	40.6
1–3 y	1,998	14.2	1,456	27.0	2,405	30.2	5,273	23.9
4–6 y	260	1.8	287	5.3	960	12.0	1,818	8.2
Missing	3,486	24.7	1,080	20.0	1,368	17.2	3,802	17.2
Year								
1998	1,165	8.3	192	3.6	309	3.9	888	4.0
1999	1,095	7.8	408	7.6	398	5.0	1,431	6.5
2000	614	4.4	192	3.6	388	4.9	1,140	5.2
2001	871	6.2	333	6.2	480	6.0	1,200	5.4
2002	780	5.5	408	7.6	392	4.9	979	4.4
2003	710	5.0	311	5.8	321	4.0	1,037	4.7
2004	998	7.1	336	6.2	482	6.0	1,390	6.3
2005	1,327	9.4	487	9.0	917	11.5	2,852	12.9
2006	796	5.7	328	6.1	609	7.6	1,536	7.0
2007	739	5.2	254	4.7	345	4.3	1,246	5.6
2008	853	6.1	371	6.9	559	7.0	1,436	6.5
2009	859	6.1	246	4.6	639	8.0	1,807	8.2
2010	1,130	8.0	643	11.9	499	6.3	1,947	8.8
2011	1,058	7.5	584	10.8	770	9.7	1,309	5.9
2012	1,095	7.8	295	5.5	865	10.8	1,860	8.4

Table 3. Cumulative Surgical Procedures to Repair Craniosynostosis, 1998–2012, National Inpatient Sample

	No. Procedures							
	1 Procedure (n = 27,634)		2 Procedures (n = 8,798)		3 Procedures (n = 1,257)		4 Procedures (n = 126)	
Variables	Number	%	Number	%	Number	%	Number	%
Age Categories								
0–4 mo	3,965	14.4	874	10.0	103	8.2	4	3.2
5–12 mo	10,375	37.5	3,622	41.2	484	38.5	52	41.3
1–3 y	5,480	19.8	2,159	24.5	389	30.9	42	33.3
4–6 y	1,914	6.9	569	6.5	85	6.8	5	4.0
Missing	5,900	21.4	1,576	17.9	196	15.6	24	19.0
Year								
1998	1,341	4.9	457	5.2	86	6.9	10	7.9
1999	1,881	6.8	583	6.6	83	6.6	9	7.3
2000	1,428	5.2	380	4.3	49	3.9	0	0
2001	1,684	6.1	452	5.1	92	7.4	5	3.9
2002	1,557	5.6	406	4.6	57	4.5	5	3.7
2003	1,387	5.0	427	4.8	40	3.2	4	3.5
2004	1,866	6.8	557	6.3	75	6.0	0	0
2005	2,895	10.5	1,048	11.9	160	12.8	28	21.9
2006	1,985	7.2	488	5.5	84	6.7	14	11.2
2007	1,499	5.4	467	5.3	51	4.0	0	0
2008	1,751	6.3	582	6.6	95	7.5	5	3.8
2009	1,743	6.3	740	8.4	102	8.1	5	3.8
2010	2,580	9.3	607	6.9	112	8.9	22	17.6
2011	1,905	6.9	784	8.9	76	6.1	5	3.7
2012	2,130	7.7	820	9.3	95	7.6	15	11.9

The mean length of hospital stay was 4.2 days, with an average cost of $67,962 per hospitalization. The overall payer mix was predominantly private insurance (56%), followed by government (37.1%) and other (5.5%). The surgeries were performed at teaching hospitals 84.2% of the time. Hospitalizations varied by geographical region: Northeast (15.9%), Midwest (21.8%), South (32.7%), and West (21.4%). Variables reported as unknown in the NIS database relevant to this study included age (20.3%), race (24.9%), sex (1%), insurance status (0.2%), hospital type (8.6%), and region (8.1%) for all hospitalizations.

DISCUSSION

Previous studies have addressed the lack of specificity in the subtype of craniosynostosis captured by Current Procedure Terminology (CPT) codes alone, which seem to provide inadequate information about the diagnoses and procedures patients undergo.[9,12,13] Ultimately, the shortage of detail obtained from the current national databases may lack in both utility and clinical relevance for craniofacial surgeons involved in the treatment of patients with craniosynostosis. These databases are also deficient in the information they provide in regards to the affected suture and the approach utilized to perform the operations (eg, open versus endoscopic and FOA). Our study brings attention to the fact that ICD coding, just like previously seen with CPT codes, is also inadequate in providing sufficient information about craniosynostosis subtypes as well as the surgical method used in each craniosynostosis repair.

The aim of our study was to evaluate the evidence available, at a national level, to support conclusions made by previous urban studies regarding the shifting trends of metopic craniosynostosis in the United States. We found notable limitations in the data we were able to extract from the NIS, which uses ICD coding as its raw data source. Under the ICD-9-CM coding system, all types of craniosynostosis were grouped with other unspecified craniofacial abnormalities using the code 756.0, representing a diagnosis of "congenital anomalies of the skull and face bones," which do not give us any information about the specific craniosynostosis subtype (eg, metopic, coronal, etc.). The lack of specificity in the ICD-9 code 756.0 representing congenital anomalies of the face and skull bones means that our dataset includes anomalies such as absence of skull bones, acrocephaly, congenital anomalies of the forehead, Crouzon's disease, hypertelorism, imperfect fusion of the skull, oxycephaly, platybasia, cleidocranial dysplasia, craniofacial microsomia, and amniotic band syndrome among others that fall under both code 756.0, and procedure codes 2.01, 2.03, 2.04, and 2.06. Because the ICD-9 diagnosis codes do not distinguish between the different suture types, we were not able to accurately account for the number of hospitalizations related to each subtype of craniosynostosis, and thus, we were not able to accomplish our initial objective of evaluating the conclusions made by previous urban studies regarding the shifting trends of metopic craniosynostosis in the United States.

Our study did reveal an increased number of hospitalizations and surgical procedures for craniosynostosis repair over the 15-year period between 1998 and 2012. The cause for this increase is unknown and further research is needed to determine if this is a true increase in incidence over time, or if it is due to other factors, such as better diagnosis practices or changes in treatment patterns. Most procedures were performed on children younger than 1 year (51.1%) with bone graft to skull (180%) and other cranial osteoplasty (109%) procedures demonstrating the highest increases for the study period. Among age categories, those aged 4–6 years had the highest percentage (54.7%) of other cranial osteoplasty procedures. This age group may have the most clinically interpretable findings because "other cranial osteoplasty" (code 02.06) likely represents a filling the skull voids that remained from the primary repair and would be correspond to CPT codes 62141-7.[14,15] Interestingly, there was a higher percentage of hospitalizations related to craniosynostosis repairs in the South compared in comparison to other regions in the United States. Academic centers were the predominant hospitals performing these procedures, likely due to the complexity of the surgical repairs. The regionalization of particular surgical procedures into select centers has been shown to be beneficial. A study by Salazar et al[16] found that in multiple geographic regions, surgeries on infants had better outcomes when performed in specialized centers with high patient volume.

There are various limitations in the dataset due to the lack of specificity in the ICD classification system available to use in this study. Although *other cranial osteoplasty* was shown to be the most frequently performed procedure overall (44.6%), a major limitation of the ICD-9 coding system is that we do not have a way of assessing the true meaning of code 02.06, which further illustrates the need for more descriptive procedure codes in our current system of classification. Because opening of cranial sutures accounted for 28.5% of the total procedures performed during the study period, then one may hypothesize that only 28.5% of patients in our cohort actually had a diagnosis of craniosynostosis, as opening of the cranial sutures tends to be the first step in its surgical correction. The NIS dataset is also incomplete by the fact that basic information such as age is missing, which is highlighted by the fact that other cranial osteoplasty was done in an "unknown age" group in 39.1% of cases. The lack of detail in the NIS may arise due to difficulties associated with differences in the way hospitals record and report their data to the NIS. The CPT codes also failed to account for major surgeries used for the correction of craniosynostosis, including cranial vault remodeling, FOA, and posterior cranial vault distraction followed by FOA, which also show that the NIS dataset from 1998 to 2012 has inadequate information to accurately assess the shifting trends in the epidemiology of craniosynostosis. Taking into account the discrepancies pointed out by our study, the ICD-9 coding system and its procedure codes do not allow for a comprehensive selection of the patient records that would precisely reflect the cases of craniosynostosis in the United States. As a result, our initial patient selection methodology falls short of being an accurate reflection of the true number of cases of craniosynostosis at the national level.

Although the ICD-9 codes had inherent limitations to account for the subtypes of craniosynostosis at a national level, the lack of specificity in the classification system was not fully addressed by the ICD-10-CM coding update. The ICD-10-CM coding system expanded the ICD-9-CM code 756.0 into 3 classifications: craniosynostosis (Q75.0), hypertelorism (Q75.2), and congenital malformations of skull and face bones (Q75.9). Although we are now able to track cases of craniosynostosis as a single disorder with ICD-10-CM, there is still a lack information about the specific craniosynostosis subtype involved (eg, metopic, coronal, etc.). Although our study is limited by its restricted dataset only including data until 2012 and its basis on an ICD-9 coding system that does not possess sufficient information, the fact that the ICD-10 system also lacks crucial information to create an accurate dataset, may actually mean that the craniofacial surgery community at large does not yet have the necessary raw data to adequately assess the shifting national trends in the epidemiology of craniosynostosis.

From the data in this study, it is difficult to extrapolate which suture was being addressed and which ICD-9-CM code corresponded to specific types of reconstructive procedures such as "FOA," which is often used in metopic and coronal craniosynostosis remodeling. It may be beneficial for the upcoming ICD-11 code updates to reflect the suture-specific types of craniosynostosis. It may be beneficial for the upcoming ICD-11 code updates to reflect the suture-specific types of craniosynostosis. The addition of an identifier for nonprimary repairs (ie, revisions and skull void defects) may also be useful in estimating the incidence of primary surgical craniosynostosis repair. A more granular classification system implemented in the CPT, or the upcoming ICD-11 would enable further investigation into the epidemiological trends of craniosynostosis subtypes. Innovative changes to our national registries may create a deeper understanding of this relatively common abnormality, as well as the contributing factors behind the shifting epidemiological trends reported in previous studies.

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