Differences in surgical outcomes for patients with craniosynostosis in the US: impact of socioeconomic variables and race

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OBJECTIVE Craniosynostosis is often treated with neurosurgical intervention. The aim of this study was to report and analyze the clinical and socioeconomic characteristics of patients with craniosynostosis and to present current national trends.

METHODS Using the Kids’ Inpatient Database for the years 2000, 2003, 2006, and 2009, the authors identified patients with craniosynostosis using International Classification of Diseases, Ninth Revision, Clinical Modification diagnosis codes and their associated procedure codes. Clinical features, demographics, inpatient procedures, outcomes, and charges were collected and analyzed.

RESULTS Of the 3415 patients identified, 65.8% were White, 21.4% were Hispanic, and 3.2% were Black. More than 96% were treated at urban teaching hospitals and 54.2% in southern or western regions. White patients were younger (mean 6.1 months) as compared with Blacks (mean 10.9 months) and Hispanics (mean 9.1 months; p < 0.0001) at the time of surgery. A higher fraction of Whites had private insurance (70.3%) compared with nonwhites (34.0%–41.6%; p < 0.001). Approximately 12.2% were nonelective admissions, more so among Blacks (16.9%). Mean hospital length of stay (LOS) was 3.5 days with no significant differences among races. Following surgical treatment, 12.1% of patients developed complications, most commonly pulmonary/respiratory (4.8%), wound infection (4.4%), and hydrocephalus (1.4%). The mean overall hospital charges were significantly lower for Whites than nonwhites ($34,527 vs $44,890–$48,543, respectively; p < 0.0001).

CONCLUSIONS The findings of this national study suggest a higher prevalence of craniosynostosis in Hispanics. The higher predisposition among males was less evident in Hispanics and Blacks. There was a significant percentage of nonelective admissions, more commonly among Blacks. Additionally, Hispanics and Blacks were more likely to receive surgery at an older age, past the current recommendation of the optimum age for surgical intervention. These findings are likely associated with a lack of early detection. Although mean LOS and rate of complications did not significantly differ among different races, nonwhites had, on average, higher hospital charges of $10,000–$14,000. This discrepancy may be due to differences in type of insurance, craniosynostosis type, rates of comorbidities, and delay in treatment. Although there are several limitations to this analysis, the study reports on relevant disparities regarding a costly neurosurgical intervention, and ways to diminish these disparities should be further explored.

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KEY WORDS craniosynostosis; outcomes; disparities; complications; national analysis; epidemiology; craniofacial

PREMATURE fusion of skull sutures with resulting cranial distortion is termed craniosynostosis. The deformity most commonly occurs sporadically and in roughly 3–6 people per 10,000 live births.1,2,5,16 The cause of the condition remains elusive, with multiple theories having been suggested, including genetic mechanisms. Craniosynostosis is classified based on the type and number of sutures involved as well as based on the co-occurrence of syndromes. The most common form is sagittal, followed by unilateral coronal, bilateral coronal, metopic, and lambdoid craniosynostosis. There is a male predilection for the condition, particularly with the sagittal type, in...
which males outnumber females 4-fold. Unilateral coronal synostosis occurs slightly more commonly in females (3 to 2). Whether the deformity leads to increased intracranial pressure, decreases in intracranial volume, and hydrocephalus is controversial. However, multiple studies have demonstrated an association of craniosynostosis with increased intracranial pressure. Additionally, the condition has been shown to alter brain structures with a likely associated neurocognitive decline. Although it is questionable whether surgical intervention would have an impact on the neurocognitive delays and alterations in brain morphology, the majority of craniofacial surgeons recommend early surgery (at < 1 year of age) to relieve intracranial pressure increases and improve cosmetic appearance.

In this national database study, we sought to determine the impact of age and socioeconomic factors on outcomes in children receiving surgery for craniosynostosis. Additionally, because this is a costly condition to manage with drastic financial consequences on the medical system, we also analyze these factors with regard to total hospital charges.

Methods

Data Source and Patient Selection

Patients between 0 and 20 years of age who received a craniosynostosis diagnosis and underwent a surgical intervention between 2000 and 2009 were included in this study. Data were obtained from the Healthcare Cost and Utilization Project of the Agency for Healthcare Research and Quality. The Kids’ Inpatient Database (KID) is a nationally representative database that samples 80% of pediatric discharges and 10% of uncomplicated births. This large sample size allows an ideal statistical power to detect and evaluate rare conditions among hospitalized children. Discharges are weighted based on the sampling scheme to permit inferences for a nationally representative population. In 2009, the most recent year for which the KID is available at this time, the KID contained documented information for 7.4 million weighted discharges. From 4121 hospitals in 44 states. Using International Classification of Diseases, Ninth Revision, Clinical Modification (ICD-9-CM) codes, we identified those patients with a primary diagnosis of craniosynostosis (ICD-9-CM code 756.0) accompanied by an ICD-9-CM procedure code for surgical repair (2.01, 2.03, 2.04, and 2.06). The following complications were also considered in this analysis: cardiac, infection, peripheral vascular, neurological, stroke, pulmonary, thromboembolic, and hydrocephalus.

Patient and Hospital Characteristics

A patient’s age, sex, race, type of admission, insurance payer, median income, and year of diagnosis were documented. Only patients younger than 3 years of age were identified. Patients’ pre-existing comorbidities were evaluated using Elixhauser methodology, a well-established system for identifying comorbidities from administrative databases through the use of a comprehensive set of 30 comorbidity measures with accompanying ICD-9-CM codes. Hospital characteristics such as bed size, teaching status, region, and location were also considered. Patients were categorized into 3 separate groups based on race. Patient age and race and the differences in complications, nonroutine discharge (nonhome), and charges were the main outcomes of interest. Secondary outcomes included a patient’s average length of in-hospital stay and total hospital charges. Most common complications and additional conditions associated with craniosynostosis were described in detail.

Statistical Analysis

Univariate analysis was used to evaluate differences between patients in the different groups. Patient-level characteristics, hospital-level characteristics, and outcomes were compared between these cohorts using chi-square and Student t-tests. A p value ≤ 0.05 was considered statistically significant. United States nationwide estimates were performed using SAS PROC SURVEY methodology. All statistical analyses were conducted in SAS (version 9.2, SAS Institute).

Results

Demographics

According to the KID, a total of 3415 patients with craniosynostosis were discharged in 2000, 2003, 2006, and 2009 in hospitals across the nation. The average patient age was 6.9 months (Table 1). More males (66.0%) were represented in the cohort. Whites (65.8%) followed by Hispanic (24.1%) and Black (3.2%) patients were the leading race categorizations. Approximately 29.3% of patients did not have a race recorded. Most cases were admitted as elective (65.9%), some were emergency or urgent admissions (12.2%), and type of admission was not listed in others (21.8%).

Univariate Analysis

White patients were statistically younger (mean 6.1 months) as compared with Black (10.9 months) and Hispanic (9.1 months) patients (p < 0.0001; Table 1). More Whites (70.3%) had private insurance as compared with Blacks (41.6%) and Hispanics (34%; p < 0.0001, Table 1). Fewer Whites (13.9%) lived in zip codes with lowest quartile median incomes as compared with Blacks (32%) and Hispanics (22.9%; p = 0.006). Hospital-level characteristics for all patients as well as cohort-specific patients are documented in Table 2. Most patients (48.3%) were discharged from large bed-size hospitals and teaching hospitals (96.5%), and more patients were discharged from Midwest (32.5%) and Western (38.2%) hospitals. The majority of patients were treated at urban hospitals (98.1%). A similar proportion of patients was discharged in each of the 4 years of which the data were collected; 23.9% of cases corresponded to 2000 and 25.4% to 2009.

Outcomes

Patient outcomes are summarized in Table 3. A total of 2 patients (0.05%) died prior to discharge in this cohort and 412 patients (12.1%) experienced a complication during the hospital stay. Figure 1 presents the specific com-
complications; wound infections (4.4%) and pulmonary (2.0%) and respiratory (2.8%) complications were most common. The average length of hospital stay was 3.5 days. The average total charge for the hospital stay was $35,765. Blacks and Hispanics tended to have longer hospital LOSs, although the differences were not statistically significant. However, these groups had statistically significantly higher mean hospital charges as compared with White patients ($44,890 and $48,543 vs $34,527, respectively; p < 0.0001).

**Discussion**

Using the KID for 4 discrete years of data spanning 2000 to 2009, we conducted an analysis of the demographics and outcomes of children surgically treated for craniosynostosis in the US. Differences in prevalence were found across races, sexes, and geographic regions. Among the sampled cohort, we found a higher prevalence of males, Hispanics, and patients from the Western or Midwestern regions. Our data showing a male predilection is consistent with prior studies showing that males outnumber females for sagittal synostosis (the most common craniosynostosis phenotype) and metopic synostosis, but not other subtypes of craniosynostosis. A likely higher prevalence in Hispanics (21.4%) is discerned when compared with the percentage of Hispanics in the general population of approximately 12%–16% during the study period. However, there may have been other variables or confounding factors that may have contributed to the higher number in the present study. Explaining the regional and ethnic disproportionalities is complex and likely involves the interplay between both genetic and environmental influences. Although fibroblast growth factor pathway genes have been implicated in the pathogenesis of craniosynostosis and family history is a strong risk factor, no clear genetic relationship exists. At the same time, environmental factors such as maternal smoking, nitrosatable drug use, vitamin intake, and hormonal factors such as hyperthyroidism have also been shown to play roles in premature cranial suture closure, as well as advanced maternal age.
and factors influencing fetal head constraint such as macrosomia, breech presentations, and multiple births. 1,8,11,21 Thus, demographic variabilities of craniosynostosis may be due to higher proportions of predisposed populations or higher rates of environmental exposures and maternal-fetal comorbidities. Further studies are needed to differentiate between the predisposing causes that lead to the racial and regional disproportions of craniosynostosis.

Although females made up a minority of patients (33.0%) and the overall trend was for males to be afflicted, the sex predisposition was less apparent among Blacks and Hispanics. While fewer than a third of Whites were females, more than 40% of Hispanics and almost half of Blacks were female. This interesting finding could also be explained by genetic and environmental elements and type of craniosynostosis, although it highlights a point for future investigation. Further examination of the data indicate that Blacks and Hispanics tended to be older at the time of surgery. Hispanics were on average 3 months older at presentation and Blacks were almost 5 months older than Whites. There are number of possibilities that may explain this finding. First, Hispanic and Black patients may have presented later for intervention, either because of lack of access to health care or poor medical care. For instance, these patients may have been less likely to be referred for specialty consultation or were referred at a later stage of the disease. This could also be one of the reasons for the much lower prevalence of the condition in Blacks.

### Table 2. Hospital characteristics of 3415 patients with craniosynostosis who underwent surgical repair at US hospitals during 2000, 2003, 2006, and 2009

<table>
<thead>
<tr>
<th>Outcomes*</th>
<th>All</th>
<th>White (65.8)</th>
<th>Black</th>
<th>Hispanic (21.4)</th>
<th>Other†</th>
<th>p Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>No. of cases (%)</td>
<td>3415</td>
<td>1588 (65.8)</td>
<td>77 (3.2)</td>
<td>517 (21.4)</td>
<td>232 (9.6)</td>
<td>0.003</td>
</tr>
<tr>
<td>Hospital bed size (%)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Small</td>
<td>446 (13.9)</td>
<td>208 (13.6)</td>
<td>†</td>
<td>74 (14.6)</td>
<td>30 (13.4)</td>
<td></td>
</tr>
<tr>
<td>Medium</td>
<td>1218 (37.8)</td>
<td>536 (34.9)</td>
<td>15 (20.3)</td>
<td>299 (58.9)</td>
<td>98 (43.7)</td>
<td></td>
</tr>
<tr>
<td>Large</td>
<td>1556 (48.3)</td>
<td>790 (51.5)</td>
<td>53 (71.6)</td>
<td>135 (26.6)</td>
<td>96 (42.9)</td>
<td></td>
</tr>
<tr>
<td>Teaching hospital (%)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>0.58</td>
</tr>
<tr>
<td>Yes</td>
<td>3106 (96.5)</td>
<td>1484 (96.7)</td>
<td>72 (98.6)</td>
<td>498 (98.0)</td>
<td>213 (95.5)</td>
<td></td>
</tr>
<tr>
<td>No</td>
<td>114 (3.5)</td>
<td>50 (3.3)</td>
<td>†</td>
<td>†</td>
<td>†</td>
<td></td>
</tr>
<tr>
<td>Hospital region (%)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td>Northeast</td>
<td>457 (13.4)</td>
<td>309 (19.5)</td>
<td>†</td>
<td>50 (9.7)</td>
<td>55 (23.5)</td>
<td></td>
</tr>
<tr>
<td>Midwest</td>
<td>1109 (32.5)</td>
<td>490 (30.9)</td>
<td>22 (28.6)</td>
<td>21 (4.1)</td>
<td>43 (18.5)</td>
<td></td>
</tr>
<tr>
<td>South</td>
<td>545 (16.0)</td>
<td>297 (18.7)</td>
<td>32 (41.6)</td>
<td>†</td>
<td>22 (9.5)</td>
<td></td>
</tr>
<tr>
<td>West</td>
<td>1304 (38.2)</td>
<td>492 (31.0)</td>
<td>14 (18.2)</td>
<td>442 (85.5)</td>
<td>112 (48.3)</td>
<td></td>
</tr>
<tr>
<td>Hospital location (%)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td>Rural</td>
<td>61 (1.9)</td>
<td>31 (2.0)</td>
<td>†</td>
<td>†</td>
<td>†</td>
<td></td>
</tr>
<tr>
<td>Urban</td>
<td>3159 (98.1)</td>
<td>1503 (98.0)</td>
<td>74 (100.0)</td>
<td>508 (100.0)</td>
<td>219 (98.2)</td>
<td></td>
</tr>
</tbody>
</table>

* Missing data rates: bed size (0.06%), teaching (0.06%), and rural (0.06%).
† Other race = Asian/Pacific Islander, Native American, and other unspecified groups.
‡ Exact value omitted to protect patient privacy.


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<td>517 (21.4)</td>
<td>232 (9.6)</td>
<td>NE</td>
</tr>
<tr>
<td>Deaths (%)</td>
<td>†</td>
<td>†</td>
<td>†</td>
<td>†</td>
<td>†</td>
<td></td>
</tr>
<tr>
<td>Complications (%)</td>
<td>412 (12.1)</td>
<td>199 (12.5)</td>
<td>†</td>
<td>64 (12.4)</td>
<td>29 (12.5)</td>
<td>0.99</td>
</tr>
<tr>
<td>Comorbidity (%)</td>
<td>77 (2.3)</td>
<td>37 (2.3)</td>
<td>†</td>
<td>18 (3.5)</td>
<td>†</td>
<td>0.48</td>
</tr>
<tr>
<td>LOS (Mean [SEM])</td>
<td>3.5 (0.1)</td>
<td>3.4 (0.2)</td>
<td>4.1 (0.3)</td>
<td>3.7 (0.2)</td>
<td>3.7 (0.2)</td>
<td></td>
</tr>
<tr>
<td>Median (IQR)</td>
<td>3 (2–4)</td>
<td>3 (3–4)</td>
<td>3 (2–4)</td>
<td>3 (2–4)</td>
<td>3 (2–4)</td>
<td></td>
</tr>
<tr>
<td>Total charges ($)</td>
<td>26,857</td>
<td>26,184</td>
<td>25,618</td>
<td>43,342</td>
<td>37,819</td>
<td></td>
</tr>
</tbody>
</table>

NE = not estimable due to limited sample size for this outcome.
* Other race = Asian/Pacific Islander, Native American, and other unspecified groups.
† Exact value omitted to protect patient privacy.
in this patient population. Second, parents’ perceptions, knowledge, and awareness of the condition may influence the timing for presentation for medical attention, a factor that may be further affected by cultural and educational backgrounds. Third, the difference in ages could be explained by a discrepancy in the type and complexity of suture synostosis among the different races. Unlike sagittal synostosis, coronal, metopic, or heterogenous synostoses are more likely to undergo operations at an older patient age. Thus, Black and Hispanic patients could more likely harbor these other types or more complicated synostoses that require delayed treatment, a possibility that future studies should explore.

The implications of this study provide insight into the effects of craniosynostosis on health care costs and the interplaying socioeconomic factors. In our sample population, 12.2% of admitted cases were nonelective (urgent or emergency basis), with a higher risk in Black patients as compared with other races. This may be, in part, due to the disparities in health care access and in insurance types between the different populations, as only about 30%–40% of Hispanics and Blacks had private insurance compared with more than 70% of Whites. These values are comparable to the national percentages of health insurance types among the different races.7 The finding is also consistent with the delayed presentation for surgery in the Hispanic and Black populations as compared with Whites. Barriers to health care in the underserved populations may lead to both delayed diagnoses and surgical treatment, which in turn may also increase the risk of complications. However, the Black population carries a disproportionately lower risk of craniosynostosis (only 3.2% of cases) and thus may represent more atypical cases with complicated pre- and postoperative courses, with hindrances in health care access and intervention likely a contributor.

Although rates of complications did not differ among different races, mean overall hospital charges were significantly higher for Hispanics and Blacks than for Whites by $10,000–$14,000. This may be due to differences in insurance type and means of support outside the hospital, differences in rates of comorbidities between the races (higher in Hispanics and Blacks), and the requirement for more complex surgical procedures due to delayed treatment as discussed. The results do indicate, however, that craniosynostosis surgery is relatively safe with low mortality (0.05%) and low morbidity. The most common complications were wound infection and pulmonary/respiratory. The rates of other measured complications (neurological, cardiac, hydrocephalus, etc.) all fell below 2.0%. Moreover, the median LOS was 3 days. Beyond measures such as prophylactic antibiotics to reduce local infection incidence and the increasing use of less invasive procedures, the low rates of mortality and acute complications from this surgery mainly stem from the coordinated care provided by multidisciplinary teams of pediatric specialists.13,15

Although surgical intervention for craniosynostosis is costly, lack of early surgical treatment can cause progression of both functional and aesthetic deformities in the child, with an increased risk for emergency complications. If craniosynostosis goes uncorrected, it increases the risk of increased intracranial pressure and its related problems such as adverse effects on mental function.6,12,18,23 Delays in surgery beyond 9 months to 1 year result in abnormal growth and asymmetry of the facial bones, which can lead to complications such as strabismus in certain types of craniosynostosis.15 In addition, as the patient ages, especially beyond the age of 9 months to 1 year, the calvarial bone becomes less malleable to surgical shaping and less readily reossifies to fill in surgical defects, and thus surgical intervention becomes more complicated with the need for more comprehensive surgical approaches and for metal fixation and fillers.15,16 Studies have shown that early intervention before 1 year of age has resulted in bet-
Younger age during craniosynostosis surgery has correlated with less morbidity, lower cost, and shorter hospital stay. Although there are clear benefits to earlier surgical correction, surgery before 3 months of age is not recommended due to greater risk of blood loss. Furthermore, infants who undergo surgical correction before 6 months of age may have higher rates of reoperation. Thus, surgical intervention is generally recommended between 6 and 9 months. It is noteworthy that although the majority of our sampled White patients received surgical treatment within this time frame, the mean age of surgery was beyond 9 months for both Hispanics and Blacks. These delays in surgical management may need to be addressed to allow for better outcomes and reduced hospital costs.

Limitations of the Study
Results of the current study were obtained in a retrospective manner from a national database. Only certain patient information is accessible through the system. Elements such as type of synostosis, syndromic versus non-syndromic, preoperative symptoms, and radiological signs cannot be determined. Different types of procedures or operative approaches such as endoscopic versus open repairs cannot be differentiated. Additionally, it is not possible to verify long-term outcomes such as cosmetic improvement and clinical relief. Also, coding errors can and do occur, resulting in over- and underestimations. There may also be missing data, as was the case in the current study with missing data in areas such as race and type of admission. It is also worth mentioning that for analysis of comorbidities, we used the Elixhauser metric and there are no previous studies that have examined this methodology in the pediatric population. Given the small rates of comorbidities based on this metric in the studied population, it is very likely that we are underestimating the actual comorbidity rates in these patients. This study was directed at evaluating whether racial disparities exist in the population of young children undergoing craniosynostosis surgery. Future studies could widen the inquiry to include all races and could further subdivide patients into groups such as ethnicities and religious affiliations to determine if cultural and/or religious beliefs are a contributor to the disparities.

Conclusions
In this national study of young children treated for craniosynostosis in the US, we found disparities in age at surgery, type of admission, and hospital costs among different races. Although the condition is well-known to more likely affect males than females, the predilection was less apparent among Hispanics and Blacks. The study is suggestive of a higher prevalence of craniosynostosis among Hispanics. A higher percentage of urgent or emergency admissions was present among Blacks. Moreover, Blacks and Hispanics were older at the time of surgery, beyond the recommendation of 6–9 months for surgical intervention. On average, Hispanics and Blacks incurred 30%–40% higher hospital costs compared with Whites. These differences could be explained by poor access to medical care, insurance status, variations in the type of synostosis and pathophysiology, cultural and socioeconomic beliefs, and the confounding factors and limitations of the current analysis. In the current state of health care changes and reform, future research should further investigate these possible disparities.

References
Disclosure
The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions
Conception and design: Shweikeh. Acquisition of data: Foulad, Nuño. Analysis and interpretation of data: Shweikeh, Nuño. Drafting the article: Shweikeh, Foulad. Critically revising the article: Shweikeh, Adamo. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Drazin. Statistical analysis: Nuño. Study supervision: Drazin, Adamo.

Supplemental Information
Online-Only Content
Supplemental material is available with the online version of the article.
Supplementary Table 1. http://thejns.org/doi/suppl/10.3171/2015.4.PEDS14342.

Previous Presentation
Results from this study were presented as an abstract at the 42nd Annual AANS/CNS Section on Pediatric Neurological Surgery in Toronto, Canada, on December 3, 2013.

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