Risk Factors for Delayed Referral to a Craniofacial Specialist for Treatment of Craniosynostosis

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Objective To assess the impact of age at referral on treatment options in craniosynostosis and to identify risk factors for referral delays in this population.

Study design A retrospective cohort study was performed on patients with an abnormal head shape diagnosis treated at a single academic medical center between January 1, 2004 and January 1, 2014. Newly diagnosed patients with craniosynostosis were identified and referral patterns were examined. A multivariate logistic regression model was used to identify risk factors associated with the range of ages at initial referral.

Results A total of 477 patients were evaluated at our institution, 197 of whom were subsequently diagnosed with craniosynostosis. The median age at initial appointment was 5.6 months (mean 8.2 months). Only 28% of children were referred within 3 months of birth. Patients referred within 3 months of birth were less likely to have had preappointment imaging than those patients referred slightly later (OR 2.53, CI 1.07-5.98, $P = .035$). Several variables were associated with referral after 12 months of age including multiple suture involvement (OR 4.21, CI 1.06-16.68, $P = .041$), minority race (OR 4.96, CI 1.91-12.9, $P \leq .0001$), and referral by a nonpediatrician (OR 6.9, CI 1.73-27.49, $P = .006$).

Conclusions Obtaining imaging before referral to a specialist for abnormal head shape was associated with a delay in evaluation and potentially increases radiation exposure and limits treatment options in patients with craniosynostosis. In addition, children from minority groups, children referred from someone other than a pediatrician's office, and those with multiple suture craniosynostosis are at increased risk of delayed referral. Further studies into the cause of these delays are warranted. *(J Pediatr 2017;186:165-71).*

Craniosynostosis is the most common pediatric cranial malformation requiring operative intervention. In 2010, a multidisciplinary workshop was organized by the Centers for Disease Control and Prevention and the National Foundation for Facial Reconstruction with the goal of developing consensus guidelines for the diagnosis, treatment, and management of craniosynostosis.[1,2] This expert panel recommended that patients with suspected craniosynostosis be evaluated by craniofacial specialists in the prenatal period and no later than 3 months of age. The importance of early referral was stressed, based in part on potential access to newer, time-sensitive techniques. Such techniques, which include endoscopic treatment, rely on the pliability and osteogenic potential of the infant calvaria for remodeling and, thus, cannot be performed reliably once maturation limits these characteristics.

Despite the importance of early evaluation, children with suspected craniosynostosis are frequently referred to see craniofacial surgeons beyond the opportunity window for time-sensitive techniques. Prior literature has reported an average age at initial evaluation between 5 and 12 months of age.[3,4] Although the discrepancy between recommended referral age and actual referral age exists, little research has been done to determine the causality behind referral delays. The goals of this study were to determine the age of referral on treatment options, to identify risk factors for referral delays for the treatment of craniosynostosis, and to consider impact on treatment options.

Methods

A retrospective review was performed of all patients seen by the craniofacial team at the Duke Cleft and Craniofacial Center between January 1, 2004 and January 1, 2014. All patients in the medical record with an *International Classification of Diseases, Ninth Revision, Clinical Modification* diagnostic code for congenital musculoskeletal deformities of skull, face, and jaw (754.0) or anomalies of the skull and face bones (756.0) were selected for detailed chart review and manual confirmation of diagnosis. Patients referred after prior treatment elsewhere and

CT Computed tomography

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patients referred after 10 years of age were excluded from the study. Data were collected for all encounters with these patients from January 1, 2004 to July 1, 2014. The study design and implementation was approved and monitored by the institutional review board.

Chart Review

From the chart review, diagnosis was confirmed by reviewing the clinical notes of the primary plastic surgeon and neurosurgeon involved in the patient's care. A timeline of care was prepared for each patient, with an emphasis on details of the referral process and subsequent course of evaluation and management. By policy within our craniofacial center, an appointment is made for all new referrals related to abnormal head shape within 2 weeks of receipt. Specific data collected in the chart review included date of referral, referring physician (grouped by practice and specialty), date of first appointment with the craniofacial team (plastic surgeon or neurosurgeon), age at initial appointment, date of initial diagnosis, diagnosis and syndromic status, type of craniosynostosis (when applicable), diagnosing physician, whether preoperative imaging was obtained prior to appointment, choice of operative treatment, and amount of time between clinic evaluation and operative treatment. Other variables of interest included sex, race/ethnicity, language spoken at home, insurance status, and average distance from home to hospital.

Patient Cohorts

Patients were grouped into categories based on age at referral. The age brackets defined for the analyses were (1) less than 3 months of age; (2) 3-6 months; (3) 6-12 months; and (4) greater than 12 months. These age groups were chosen to parallel what is commonly used in clinical decision-making: Current guidelines recommend evaluation by a craniofacial surgeon before 3 months of age, thus, defining the ideal age bracket for referral.[1] Recognizing that certain center-specific variations exist, patients between 3 and 6 months of age may be considered for all forms of cranial vault reconstruction. Patients referred between 6 and 12 months of age may be considered for traditional calvarial vault remodeling but are beyond age limits for less invasive techniques (eg, strip craniectomy, endoscopically assisted strip craniectomy, spring cranioplasty). Finally, those patients referred after 1 year of age are beyond the ideal age for all treatments.

In addition to the age-based categories, patients were additionally classified as being initially referred to a specialist for both evaluation and management or as being referred for management following confirmation of diagnosis (eg, by radiographic evaluation).

Statistical Analyses

Multivariable logistic regression was used to model the relationship between various risk factors and age at referral. The continuous response variable age at first appointment was recoded as a dichotomous outcome to indicate whether or not the appointment occurred at a given age range. A subset of predictors was selected based on the Akaike information criterion and the stepwise selection algorithm from the collected data. The first and second models were defined by patients referred at <3 months of age and compared with patients referred at 3-6 months of age (model 1) and greater than 3 months of age (model 2). An additional model was constructed to assess risk factors for late referral (>12 months of age).

The fit models are of the form: $\log\left(\frac{p_i}{1-p_i}\right) = \mu + \beta_1 x_{1,i} + \cdots + \beta_p x_{p,i}$, where $p_i = \Pr(Y_i = 1 | X_i)$ is the probability of occurrence of first appointment at the specified age range given the predictors (ie, $Y_i = 1$ indicates occurrence of first appointment, and $Y_i = 0$ otherwise). The model expresses the log-odds as a baseline or reference value (μ) plus the predictors' effects (β_j). The multivariable logistic regression models were fit using the maximum likelihood method. Incomplete data sets were excluded from analysis. The significance of the models relative to just fitting an intercept term was assessed through the likelihood ratio test. In addition, the McFadden pseudo R-square was computed. In all cases, the threshold for statistical significance of the *P* values was set to .05. All statistical analyses were performed with R statistical software.[5]

Results

A total of 477 patients were referred between January 2004 and January 2014 with *International Classification of Diseases, Ninth Revision, Clinical Modification* code 754.0 or 756.0 (**Figure 1**). Of these, 197 patients (41%) were confirmed by the consulting surgeon to have craniosynostosis. A referral for evaluation of a general head shape abnormality (n = 165, 84%) was more common than referral with a previously confirmed diagnosis of craniosynostosis (n = 32, 16%). Most referrals (82%) were made by a pediatrician. Of the patients referred for evaluation of a head shape abnormality, 44% were eventually diagnosed with craniosynostosis, 39% with positional plagiocephaly, and 0.002% with a separate craniofacial diagnosis. The remainder of patients evaluated for a potential head shape anomaly (17%) were not diagnosed with a craniofacial abnormality after examination.

The average age at first appointment for all patients referred for "abnormal head shape" was 9.2 months, with a median age of 6.4 months. Of the 197 patients confirmed to have craniosynostosis, the average age at first appointment was 8.2 months, with a median of 5.6 months (**Table I**). Patients with craniosynostosis were, thus, referred later than current guidelines recommend. Assessed by age at referral, 27% of patients were less than 3 months old, 26% were between 3 and 6 months old, 27% were between 6 and 12 months old, and 19% were greater than 12 months of age. These percentages were relatively consistent across years, showing no trend toward improvement (**Figure 2**; available at www.jpeds.com).

Type of craniosynostosis influenced age of initial referral. Unilateral coronal craniosynostosis was referred earlier than any other pattern of craniosynostosis, with an average age of