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Transitions in care from pediatric to adult general surgery: Evaluating an unmet need for patients with anorectal malformation and Hirschsprung disease☆,☆☆,★,★★

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ABSTRACT

Background: The provision of timely and comprehensive transition of care from pediatric to adult surgical providers for patients who have undergone childhood operations remains a challenge. Understanding the barriers to transition from a patient and family perspective may improve this process.

Methods: A cross-sectional survey was conducted of patients with a history of anorectal malformation (ARM) or Hirschsprung Disease (HD) and their families. The web-based survey was administered through two support groups dedicated to the needs of individuals born with these congenital abnormalities. Categorical variables were compared using Chi-squared and Fisher's exact test with Student's t test and ANOVA for continuous variables.

Results: A total of 118 surveys were completed (approximately 26.2% response). The average age of patients at time of survey was 12.3 years (SD 11.6) with 64.5% less than 15 years old. The primary diagnosis was reported for 78.8% patients and included HD (29.0%), ARM (61.3%), and cloaca (9.7%). The average distance traveled for ongoing care was 186.6 miles (SD 278.3) with 40.9% of patients traveling ≥30 miles; the distance was statistically significantly greater for patients with ARM ($p < 0.001$). With regards to ongoing symptoms, 44.1% experience constipation, 40.9% experience diarrhea, and approximately 40.9% require chronic medication for management of bowel symptoms; only 3 respondents (3.2%) reported fecal incontinence. The majority of patients, 52.7% reported being seen by a provider at least twice per year and the majority continued to be followed by a pediatric provider, consistent with the majority of the cohort being less than 18 years of age. Conversations with providers regarding transitioning to an adult physician had occurred in fewer than 13% of patients. The most commonly cited barrier to transition was the perception that adult providers would be ill-equipped to manage the persistent bowel symptoms.

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Conclusion: Patients undergoing childhood procedures for ARM or HD have a high prevalence of ongoing symptoms related to bowel function but very few have had conversations regarding transitions in care. Early implementation of transitional care plans and engagement of adult providers are imperative to transitions and may confer long-term health benefits in this patient population.
Level of evidence: Level IV, case series with no comparison group.

Since the early 1900s, pediatric surgeons have operated on congenital and acquired anomalies previously assumed to be nonsurvivable with steadily improving outcomes [1]. The likelihood of a child born with a congenital birth defect today reaching adulthood has increased considerably owing to improvements in surgical techniques, medical therapies, neonatal intensive care, and provision of safe anesthesia. As a result, the improved prognoses for diseases such as imperforate anus, congenital diaphragmatic hernia, and esophageal atresia, however, have produced a population of adult survivors with unique challenges. While childhood outcomes are reported for some surgically treated congenital anomalies, there are limited data on adult outcomes, with provision of disease-specific care into adulthood limited by poorly defined medical support teams [2,3].

Teenagers and young adults pose a unique challenge as they transition physically and mentally into adulthood, seeking independence from parental supervision and guidance in healthcare and other aspects of their lives. This population is particularly vulnerable to lapses in healthcare and for patients with Hirschsprung Disease (HD) and anorectal malformations (ARM), they are often affected by ongoing surgical and physiological challenges [4,5]. Multiple studies have documented the costs of lapses in transitional care and potential for increased health care resource utilization associated with adverse outcomes [6]. Recommendations dating back to 2002 in a joint statement by the American Academy of Pediatrics, the American Academy of Family Physicians, and the American College of Physicians, described a framework to ensure a smooth transition to adult healthcare [7]. Unfortunately, adoption of transitional programs and application of these principles, especially in pediatric general surgery, have been limited.

The aim of this study was to examine both patient and parent experiences with transitional care for those who have undergone a childhood operation for HD or ARM.

1. Methods

This study was conducted as a survey of adolescent patients and parents of pediatric patients with a history of HD or ARM. The study protocol and electronic survey were reviewed by the Institutional Review Board of the State University of New York, University at Buffalo and informed consent was obtained through survey completion (UB IRB:00000834). The survey was created using a Delphi approach and collaboration by members of the American Academy of Pediatrics Section on Surgery's Delivery of Surgical Care Committee. The survey was reviewed by key representatives from the organizations through which subjects were identified prior to distribution.

1.1. Patient selection

Subjects were identified for inclusion in the study based on involvement with one of two nonprofit organizations dedicated to patients with congenital anorectal or colon disease. The Pull-thru Network (PTN) was founded in 1988 and is one of the largest organizations in the world dedicated to the needs of those born with an anorectal malformation or colon disease (http://pullthrunetwork.org). The organization tracks nearly 300 families, including young adults, who have undergone surgical correction of the aforementioned conditions. REACH (Research, Education, and Awareness for Children with Hirschsprung's Disease) is a nonprofit organization committed to improving the lives of children and families living with Hirschsprung Disease (http://www.reachhd.org). REACH was established in 2011 and supports affected families throughout adolescence and into young adulthood. Regardless of their relationship with the healthcare system, PTN and REACH correspond with patients for many years after the surgical correction of their pediatric disease.

Patients were included who were affiliated with one of the two organizations and who had undergone anorectal/colorectal pull-through procedures for HD, cloaca, and ARM. Organization affiliation varied from passive receipt of quarterly newsletters to active participation in online support groups and interactive networks. There were no age restrictions on participation with consent and assent tailored to participants older than and younger than 18 years of age, respectively, and parents. Surveys were administered to both patients and parents as the organization databases did not distinguish between the two in list serve and newsletter distribution.

1.2. Data source

Once contacted by the participating network coordinators or liaisons, an electronic survey was administered using the SurveyMonkey® interface. The survey was anonymous but participants were given the option of providing contact information for answer clarification and outreach as needed. The survey was designed to address the congenital/pediatric surgical condition, initial care received, and current medical condition including complications associated with the principal disease process and/or procedure. Additionally, the survey asked about experience with transitioning from pediatric to adult providers and any barriers, real or perceived, which had been encountered or anticipated (Appendix 1).

1.3. Statistical analysis

This is a descriptive study and no comparative statistical procedures were performed. No power calculation was performed based on a convenience sample of voluntary survey respondents. All descriptive analyses were completed using Microsoft Excel 2010 software. Categorical variables were compared using the Pearson's Chi-squared and Fisher's exact test, with Student's t test for continuous variables. For all tests, 2-sided p values <0.05 were considered statistically significant.

2. Results

2.1. Participant characteristics

Patient and parent characteristics are shown in Table 1. A total of 118 surveys were completed by participants of PTN and REACH for an estimated response rate of 26.2%. Greater than 70% of patients were less than 18 years old and surveys were completed by parents with patient assent. The average age of patients at time of survey completion was 12.3 years (standard deviation 11.6); there was no significant difference in the age of patients based on primary diagnosis ($p = 0.31$). For patient confidentiality, participants were not required to select the organization with which they are affiliated. Of the respondents who reported a

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