Seminars in Pediatric Surgery 24 (2015) 65–68

Contents lists available at ScienceDirect

Seminars in Pediatric Surgery

journal homepage: www.elsevier.com/locate/sempedsurg

Transition of care in pediatric surgical patients with complex gastrointestinal disease

Eleanor D. Muise, MD, Robert A. Cowles, MD*

Department of Surgery, Yale University School of Medicine, New Haven, Connecticut

ARTICLE INFO

Keywords:
Short bowel syndrome
Intestinal dysmotility
Hirschsprung Disease
Anorectal malformation
Continuity of care
Transitional care

ABSTRACT

Pediatric surgeons provide care for infants and children with a wide variety of conditions throughout the body. Many of these conditions are congenital or occur very early in life, and for this reason, providing continuity of care for these patients into adulthood is an emerging challenge. In the gastrointestinal tract, congenital and acquired conditions are now associated with excellent long-term prognosis; however, little guidance on long-term care exists. The aim of this article is to discuss aspects that are important to transitioning care of pediatric surgical patients with complex gastrointestinal disorders from pediatric to adult practitioners. Transitional care of patients with short bowel syndrome, Hirschsprung Disease, and anorectal malformations will be the focus of this discussion, but the concepts introduced here may translate to other diagnoses as well.

Introduction

The importance of multidisciplinary teams, often led by pediatric surgeons and composed of pediatric gastroenterologists, nutritionists, pharmacists, social workers, and nurses, has been widely accepted in the care of patients with complex gastrointestinal maladies. Effective management aims to provide care driven by subspecialty experts, to coordinate and communicate a treatment plan, and to establish continuity of care to improve patient outcomes.[1] Intestinal rehabilitation programs designed to care for complex patients with short bowel syndrome have resulted in significant reductions in the complications of this disease by implementing protocols in the management of parenteral nutrition and central venous catheter care.[2,3] Patients with Hirschsprung Disease and anorectal malformations are essentially surgically cured, though many continue to face motility and elimination difficulties that persist into adulthood.[4,5] During years of ongoing follow-up with pediatric practitioners, patients and their families develop a close relationship and support system with the care team. As these children mature into adolescence and young adulthood, it is expected that their needs will be better met by transitioning to adult providers for ongoing care.[6] There is a need to establish organized transition from child-centered to adult-oriented care for adolescents and young adults with chronic disease, though little evidence is available to suggest the most optimal transition. This article will highlight specific long-term concerns in patients with short bowel syndrome, Hirschsprung Disease, and anorectal malformation and aims to provide recommendations for transition of care of these complex patients.

Transitional care of adolescent and young adult patients with chronic disease

Children and families of children with chronic illness are enveloped in a rich network of providers and a coordinated support system often from the time of birth. Bonds are formed between families and providers through times of crisis, and often both providers and families are reluctant to transition care to a new group as children grow up. However, adolescents develop very different relationships with parents as well as physicians, and establishing independence and taking on more responsibility for their own care is a vital part of young adulthood. Coordinating a smooth transition from child-centered to adult-oriented care is first the responsibility of the care team. Direct transfer of care from a single pediatric to an adult provider can be successful, provided that medical, psychosocial, and educational needs are addressed.[7] However, transition clinics and transition coordinators may provide the most comprehensive evolution from childhood care to adulthood care by addressing the multiple needs of the patient, family, and practitioners.[8,9]

* Correspondence to: Robert A. Cowles, MD, 333 Cedar St, Farnam Memorial Building Room 131, PO Box 20862, New Haven, CT 06520.
E-mail address: robert.cowles@yale.edu (R.A. Cowles).

http://dx.doi.org/10.1053/j.sempedsurg.2015.01.002
1055-8586/

Timing of transition

Children with chronic disease have often not met developmental milestones on time and may be delayed in psychosocial and psychosexual development, which is especially important for new providers to understand when taking on the care of young adults with chronic conditions.[10] Suggestions for timing of transition include life milestones such as completion of high school, 18 years of age, marriage, pregnancy, or disease-free interval that make transition easier for the patient and the provider.[6,11] Regardless of the proposed age for transition, transition goals should be set forth as early as 14 years of age to begin the process of acquiring health literacy skills and disease-specific knowledge by the patient and the family.[12]

Disease-specific knowledge

Disease-specific knowledge is a key element to assess in an adolescent who is preparing to transition to an adult practitioner, as a lack of understanding of disease progression, concerning symptoms, or complications may delay seeking treatment and may impede care delivery in an adult-oriented setting.[12–14] It is important that providers stimulate and foster that growing autonomy by seeing adolescent patients, at least for a portion of the visit, by themselves, which mimics how care is provided for adult patients and can make everyone more comfortable as a transition approaches.[6] Parental as well as clinician assessment of adolescent readiness to transition to adult care often overestimates adolescent self-efficacy.[12,15] Becoming an independent decision maker with regard to follow-up and continuing care can be a particular challenge in adolescents and young adults with chronic disease, as they may experience psychosocial developmental delay.[10,13]

Differences in child-centered and adult-oriented care

Recognizing the major differences between child-centered care and adult-oriented care is essential in orchestrating a smooth transition. Adult practitioners see patients by themselves and rely on subspecialty referral and coordination by primary care providers, whereas pediatric practitioners maintain a more multidisciplinary, family-focused approach.[13] Appointments for adult patients may be shorter than pediatric patients, and providers are likely to rely on adult patients to provide a summative history and to be knowledgeable about recent test results.[13] Adult providers may have different goals for visits compared with their pediatric counterparts, for example, valuing self-efficacy and the ability to undergo procedures without sedation remains 2 of the top concerns for providers, while patients and families express anxiety surrounding the transition, desiring a more coordinated and comprehensive transition than is likely feasible.[7,11] Becoming aware of the differences in the way that care is provided empowers patients, families, and providers to address expectations and concerns early.

Transitional care model

Transition clinics suggest that focusing on increasing adolescent knowledge, promoting self-efficacy, and even arranging a single combined visit with pediatric and adult practitioners in the adult care office is beneficial to decrease family anxiety about transition and increase adherence to the plan of care.[7,14–16] A collaborative approach in which the pediatric surgeon remains available and involved with the adult gastroenterologist in addition to the adult surgeon suggests the highest likelihood of patient and family satisfaction and the best outcome for the patient.[11] Involving a transition coordinator or a coordinated care nurse may address the needs of providers, patients, and families by helping to streamline appointments and coordinate adult and pediatric teams.[8] A transition coordinator who meets with the patient and family throughout the transition process can assess patient knowledge, facilitate joint appointments between pediatric and adult practitioners, and assist patients with follow-up.[9] Of note, several combined medicine–pediatrics residency programs in the United States facilitate subspecialty multidisciplinary clinics, and these may be a model for the future of caring for young adults with chronic disease.[17]

Patients, families, and providers will feel more comfortable transitioning to an adult provider who is familiar with complex gastrointestinal disease and who has been provided relevant medical records.[6] Furthermore, it is important to help patients and families identify reliable sources of information beyond the physician, including patient associations or disease-specific support groups, as follow-up visits may be less frequent in adult care centers.[18]

Considerations in the transitional care for specific pediatric gastrointestinal surgical conditions

Short bowel syndrome

Short bowel syndrome (SBS), or short gut syndrome, is most often a result of an initial insult that requires massive bowel resection. A central tenet in the standard treatment of infants and children with SBS is the provision of safe parenteral nutrition (PN) via a tunneled, long-term central line. PN is lifesaving and provides caloric, fluid, and electrolyte needs for adequate homeostasis and growth.[19] Despite this, long-term PN carries the known risks of catheter-associated blood stream infections, PN-associated liver disease (PNALD), and venous thrombosis, especially when used over a long period of time.[2,3,20] When possible, enteral nutrition is optimal, as direct mucosal stimulation by food promotes intestinal adaptation that can partially or fully compensate for functional loss of the resected bowel.[21] Dysmotility and intestinal dilatation can predispose to luminal bacterial overgrowth and sometimes sepsis.[22–24] While many children with SBS are able to eventually achieve enteral autonomy, some remain PN-dependent over the long-term and some, albeit off PN, have persistent requirements for fluids, electrolytes, and other supplements.[21,25–30]

The challenges involved in the long-term care of patients with SBS are only briefly reviewed above. Transition of care for patients with SBS as they reach adulthood must occur carefully. Young adults with SBS have multiple needs that often vary from patient to patient, and ongoing follow-up is important with surgery, gastroenterology, nutrition, and possibly endocrinology, hepatology, and transplantation teams.[2,31–34] As has been successful in pediatric SBS programs, a formal intestinal failure team caring for other adult patients with SBS would likely provide the optimal ongoing care and expertise for young adult patients with SBS.

Hirschsprung Disease

Hirschsprung Disease (HD) is one of the most common defects in colorectal function, affecting roughly 1 in 5000 live births. Characterized by aganglionosis of the rectum extending proximally for a variable distance into the colon, HD is most often limited to the rectosigmoid, but total colonic aganglionosis and even more rarely panintestinal aganglionosis have been reported.[35] Treatment involves a pull-through procedure that is most commonly performed in neonates or infants. Historically favorable functional outcomes have been reported in the pediatric literature; however,