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ORIGINAL ARTICLE

Transition of care: a growing concern in adult patients born with colorectal anomalies

Shannon Acker[1] · Alberto Peña[1] · Duncan Wilcox[2] · Veronica Alaniz[3] · Andrea Bischoff[1]

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Abstract

Purpose Traditionally, the care of children and adults has been arbitrarily separated into pediatric and adult medicine and surgery. Despite progress in pediatric surgical techniques, patients born with congenital anomalies still suffer from significant functional sequelae, which persist into adulthood. We aim to describe some of the most common problems experienced by adult patients with congenital colorectal malformations.

Methods Following IRB approval, we performed a retrospective database review of all adult patients who were treated by our group from 1983 until 2017.

Results We identified 88 cases. 51 patients had ARM, 18 cloacas, 9 presacral masses, 3 HD, 2 spina bifida and 5 with other diagnoses (3 vaginal anomalies, 1 cloacal exstrophy, 1 obstructed seminal vesical). The specific problems addressed were: complications from previous operations (41), rectal prolapse (25), fecal incontinence (11), gynecologic concerns (12), urologic concerns (6), and recurrent recto urogenital fistula (3). We performed 83 surgical interventions, including 13 rectal prolapse repair, 13 continent appendicostomies, 44 PSARP or redo PSARP, 11 resections of presacral masses, 11 vaginoplasties, 2 examinations under anesthesia, and 2 Mitrofanoff procedures. Five patients were treated medically (bowel management program, obstetric, urologic evaluation).

Conclusion There is a growing need to better prepare adult providers to assume the care of patients born with congenital colorectal disease as these patients transition to adulthood. A collaboration between specialized pediatric referral centers with adult colorectal surgeons, urologists and gynecologists is a potential pathway for the adequate transition of care.

Keywords Transition of care · Anorectal malformation · Hirschsprung

Introduction

Pediatric colorectal surgeons specialize in the care of patients with anorectal malformations (ARM) and Hirschsprung's disease (HD) as well as associated urogenital anomalies. In most cases, these congenital anomalies are diagnosed and successfully repaired early in life, but in many cases their long-term sequela continue beyond childhood. In some cases, these malformations are missed early in life and present during adulthood.

The first posterior sagittal approach to the repair of an ARM was performed in 1980 [1]. As our own patients reach adulthood, the need for proper transition of care is becoming more and more evident.

Patients with ARM often have associated anomalies of the urinary tract or Mullerian structures, which can affect reproductive and obstetric outcomes [2]. In addition to these associated anomalies, known complications of operations aimed at repairing these defects include acquired anorectal atresia, anorectal stricture, anal mislocation, rectal prolapse, posterior urethral diverticulum, vaginal stenosis, and persistent urogenital sinus in patients with cloaca [2–4]. Some of these complications become apparent immediately following

✉ Andrea Bischoff
andrea.bischoff@childrenscolorado.org

[1] Department of Pediatric Surgery, International Center for Colorectal and Urogenital Care, Children's Hospital Colorado, University of Colorado, 13123 East 16th Avenue, Anschutz Medical Campus, Box 323, Aurora, CO 80045, USA

[2] Department of Pediatric Urology, International Center for Colorectal and Urogenital Care, Children's Hospital Colorado, Aurora, USA

[3] Division of Pediatric and Adolescent Gynecology, International Center for Colorectal and Urogenital Care, Children's Hospital Colorado, Aurora, USA

operative repair; however, others, including rectal prolapse or vaginal stenosis, may not arise or become symptomatic until the patient reaches adolescence or adulthood.

Although we often think of ARM and HD as diseases of infancy and childhood, little is known about the needs of these individuals as they transition to adulthood with regard to outcomes of these congenital malformations. The aim of this study is to describe the medical and surgical needs of adult patients born with congenital colorectal disorders. We hypothesize that in the near future, these patients will more often seek the care of adult clinicians including colorectal surgeons, urologists, and gynecologists.

Methods

Following approval of the Colorado Multiple Institution Review Board, we performed a retrospective review of all adult patients seen at the International Center for Colorectal and Urogenital Care at the Children's Hospital Colorado (CHCO), since its creation on February 2016, as well as all adult patients who underwent an operation by Doctors Alberto Peña and Andrea Bischoff between 1983 and 2015. Data were retrospectively obtained from the electronic medical records (2016–2017) as well as our own database (1983–2015), and included: demographics, original diagnosis and operative interventions, reason for consultation and medical or surgical intervention performed. Patients were divided into two groups, those born prior to 1990 and those born after 1990 to determine if the problems of these patients have changed over time as operative techniques for repair of these malformations have become standardized. Comparisons were made between groups using two way ANOVA, Fisher's exact test or Mann–Whitney *U* test where appropriate, with significance set at $p < 0.05$. Statistical analysis was conducted in Prism 7.0 (GraphPad Software, Inc. La Jolla, CA, USA).

Results

We identified 120 adult patients who were either evaluated and managed medically or managed surgically between 1983 and 2017. Thirty-two adult patients were excluded from our analysis either because they were operated on for an acquired condition (cancer, trauma, inflammatory bowel disease), as opposed to a congenital anomaly ($n = 24$) or because there were insufficient data in our records ($n = 8$). We included 88 patients in our final analysis, 33 males (37.5%) and 55 females (62.5%). Age ranged from 18 to 60 years (median 26 years) at the time of consultation. Seventy-three patients were born before 1990 and the remaining 15 were born in 1990 or later. Demographic characteristics of the study population are presented in Table 1. The majority of patients had a previous diagnosis of anorectal malformation ($n = 51$; 57.9%) or cloaca ($n = 18$, 20.4%). Other diagnoses included presacral mass ($n = 9$; 10.2%), spina bifida ($n = 2$, 2.3%), Hirschsprung's disease ($n = 3$, 3.4%), or other diagnoses ($n = 6$, 6.8%). The majority of patients ($n = 74$, 84.1%) underwent a previous operative repair of their defect as infants and children. Neither of the patients with spina bifida had undergone a previous perineal procedure. Twelve additional patients (13.6%) had not undergone a previous perineal operation. The majority of these patients were diagnosed with a congenital anomaly as an adult including 8 with a presacral mass (Fig. 1), two women diagnosed with rectovestibular fistula (Fig. 2), one man with a cystic seminal vesicle causing obstructive symptoms, and one woman with a

Table 1 Demographic characteristics of adult patients who sought care from our pediatric colorectal surgery team

	Born before 1990 ($n = 73$)	Born after 1990 ($N = 15$)	*p value*
Age at consultation, median (standard deviation)	29 (11.4)	23 (2.7)	0.003
Female, *n* (%)	45 (62%)	10 (67%)	0.78
Diagnosis, *n* (%)			0.31
Anorectal malformation	45 (62%)	6 (40%)	
Hirschsprung's disease	2 (2.3%)	1 (6.7%)	
Spina bifida	0 (0%)	2 (13.3%)	
Cloaca	13 (17.8%)	5 (33.3%)	
Presacral mass	8 (11.0%)	1 (6.7%)	
Other diagnoses	5 (6.8%)	0 (0%)	

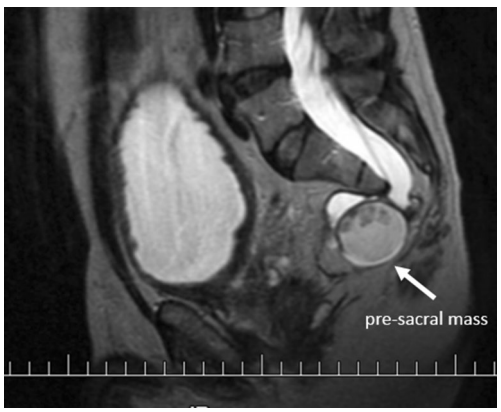

Fig. 1 Missed presacral mass in a 51-year-old male patient previously operated on for an anorectal malformation. A simple sacral radiograph showing a sacral defect should make the clinician suspicious about the presence of a presacral mass, which can be ruled out with a pelvic MRI. The presacral mass can be dealt with at the same time as the anorectal malformation

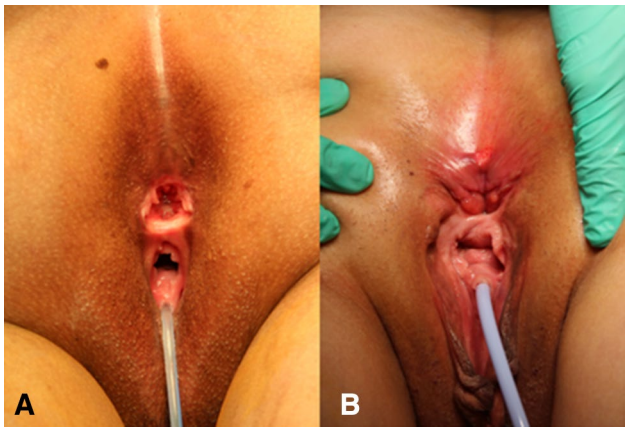

Fig. 2 Undiagnosed recto-vestibular fistula in: **a** 25 years of age and **b** 54 years of age. Both patients suffered from severe constipation and fecal incontinence throughout life

Table 2 Reason for consultation among adult patients seeking care with a pediatric colorectal surgeon

	Born before 1990 ($n=73$)	Born after 1990 ($N=15$)	*p value*
Reason for consultation			0.057
Complication related to a previous operation, *n* (%)	40 (54.8%)	1 (13.3%)	
Rectal prolapse, *n* (%)	22 (30.1%)	3 (20%)	
Request for assistance with bowel management, *n* (%)	6 (8.2%)	5 (33.3%)	
Gynecologic concern, *n* (%)	8 (11.0%)	4 (26.7%)	
Urologic concern, *n* (%)	4 (5.4%)	0 (0%)	
Recurrent fistula, *n* (%)	3 (4.1%)	0 (0%)	
Other problem, *n* (%)	15 (20.5%)	0 (0%)	

114 separate reasons for consultation in 88 patients

diagnosis of vaginal agenesis without a previous attempted repair.

The reason why these patients sought consultation usually fell into one of the following categories: rectal prolapse, complication of a previous operation, request for assistance due to fecal incontinence and constipation, a gynecologic or urologic concern, or recurrent recto-urogenital fistula. Many patients presented with more than one complaint. Complication of a previous operation was the most common reason for consultation in the group born before 1990 (54.8%) followed by rectal prolapse (30.1%). Among the younger age group, the most common reasons for consultation included assistance for the management of fecal incontinence (33.3%) and gynecologic concerns (26.7%). Data regarding reason for consultation are presented in Table 2.

Eighty-three of the 88 patients underwent an operation, 2 of which were performed with the assistance of 1 of our adult colorectal surgery colleagues. Operative procedures performed included rectal prolapse repair, Malone procedure, neo-Malone (Fig. 3) or Malone revision, posterior sagittal anorectoplasty (PSARP) (either an initial operation or a repeated operation), excision of a presacral mass, examination under anesthesia, vaginoplasty, Mitrofanoff, or other procedure. Forty-four of the patients underwent a PSARP or a redo PSARP, all but 1 of whom was born before 1990. Among those born after 1990, the Malone procedure, rectal prolapse repair, and vaginoplasty were the most common operations performed. The number of patients requiring each of these interventions are presented in Table 3.

For various reasons, five patients did not undergo an operation following consultation in our clinic. One pregnant woman sought consultation to discuss our recommendations regarding mode of delivery given her previous repair of an ARM. One man, who had a stoma all his life, sought consultation for possible pull-through procedure; however, on further work up he was found to have an ileostomy with no residual colon, which is a contra-indication for a pull-through in a patient without the anus. One patient was offered a Malone procedure but did not follow up, another was offered repair of his rectal prolapse but also did not follow up, and a final patient has been seen in our bowel management clinic and is currently satisfied with the regimen of constipating diet and enemas to control her fecal incontinence related to her history of cloaca.

Discussion

Our results indicate that patients who received an operation after 1990 were less likely to seek care due to a complication of a previous operation. This is likely the result of improved surgical techniques, as pediatric surgeons have become more familiar with the "new surgical approach" to repair ARM. Patients born after 1990 were more likely to seek consultation for the assistance with bowel management, urologic or gynecologic concerns. Few patients born after 1990 underwent a reoperation for the same congenital defect, a finding that supports our belief that the procedures performed after 1990 have been technically superior. We also believe that the group of patients born after 1990 are a reflection of the ongoing improvement of the management of ARM patients.

The most common reason for consultation among patients born after 1990 was fecal incontinence. Our protocol of management for this problem consists in finding an individualized enema that is given through the rectum once a day, keeps the patient artificially clean of stool in the underwear [5–7]. If successful, which occurs 95% of the time, the patient is offered a continent appendicostomy (Malone procedure) [8] to allow the administration of an antegrade enema. It is important to remember

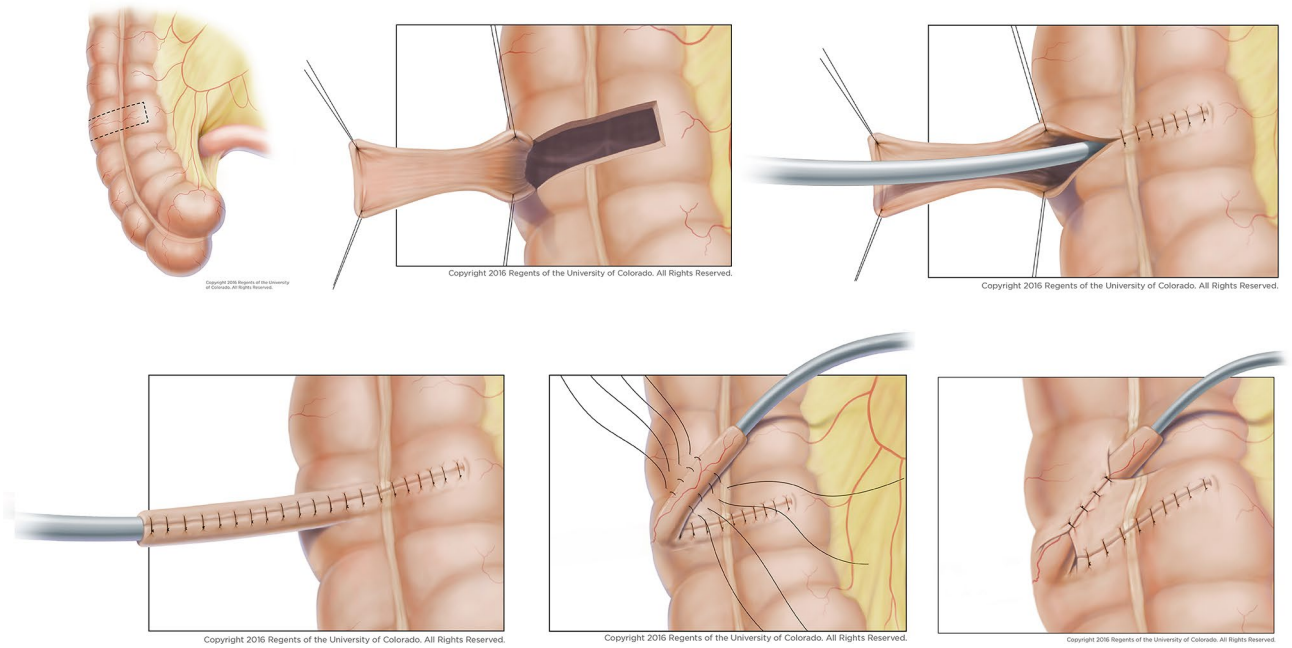

Fig. 3 Neo-appendicostomy technique for patients without an appendix

Table 3 Operative interventions performed on adult patients by pediatric colorectal surgeons

	Born before 1990 ($n=73$)	Born after 1990 ($N=15$)	*p value*
Operation performed, *n* (%)			0.1
Rectal prolapse repair	10 (13.7%)	3 (20%)	
Malone procedure or revision of prior malone	7 (9.6%)	6 (40%)	
PSARP or redo PSARP	43 (58.9%)	1 (6.7%)	
Resection of presacral mass	10 (13.7%)	1 (6.7%)	
Examination under anesthesia	2 (2.7%)	0 (0%)	
Vaginoplasty	9 (12.3%)	2 (13.3%)	
Mitrofanoff	2 (2.7%)	0 (0%)	
Other procedure	2 (2.7%)	0 (0%)	

98 total procedures as 15 patients underwent 2 different procedures

that prophylactic appendectomy is contra-indicated in patients with colorectal conditions since the appendix is useful to perform a Malone and/or Mitrofanoff procedure. If operating on a patient who previously underwent an appendectomy, the surgeon must be prepared to do a neo-appendicostomy (Fig. 3). We found that our fecally incontinent adult patients were unaware of these procedures, even when they were previously seen by adult colorectal surgeons, which highlights the need for collaboration with our adult counterparts. We predict that the procedure will be adopted by more adult colorectal surgeons, as they become familiarized with it, as well as with the necessary enema adjustments to assure success.

Another common reason for consultation was gynecologic concerns. Many of these patients complained of vaginal stenosis and required a vaginoplasty or introitoplasty. Other reasons for consultation were obstetric concerns, including feasibility and safety of vaginal delivery following a perineal operation during childhood; risk of cesarean section in patients with a complex anatomy of the abdomen due to previous procedures including Malone, Mitrofanoff, and bladder augmentation. At the same time that our surgical techniques have improved, the field of reproductive endocrinology and infertility has also progressed. We anticipate that discussions regarding fertility and pregnancy management will increase in frequency among adult patients with congenital colorectal anomalies. For this reason, we have included a gynecologist who sees both children and adults as an integral member of our team. We will continue to work with our adult gynecologic colleagues to facilitate care and counseling of these women.

Our data is limited by a variety of factors. Adult patients previously seen for consultation that did not require operation between 1982 and 2015, were not recorded in our database. Because this is a single-center, retrospective review, we have not captured the rate at which our patients seek consultation with other providers; such as male patients who sought consultation with a urologist for concerns of erectile dysfunction or lack of ejaculation, even when we have evidence that it happens in some of our patients [9]. We are an

international colorectal referral center and as such, expect that the population of patients seen in our series is likely not reflective of what other pediatric surgeons may see in their clinics in terms of complexity. However, we do think that the general trend of adult patients seeking assistance for bowel management, genito-urinary concerns, as well as to discuss fertility and pregnancy management will increase. With these data, we now have a better understanding and can better anticipate the future needs of this group of patients and coordinate the transition of their care with adult providers.

Conclusions

There is a clear trend for adult patients born with congenital malformations to seek help for the management of their anatomic or functional sequela. The most common needs of adult patients born with congenital colorectal anomalies who seek care from adult clinicians include: complications from prior operations, undiagnosed anorectal malformation, patients with "permanent stoma" requesting a stoma pull-through, rectal prolapse repair, bowel management for fecal incontinence and constipation, and sexual and reproductive concerns. Efforts should be concentrated in these areas. A collaboration between specialized pediatric referral centers with adult colorectal surgeons, urologists and gynecologists is a potential pathway for the adequate transition of care.

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