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Colorectal anomalies in patients with classic bladder exstrophy

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Abstract

Aim: This study aims to determine the proportion and type of colorectal anomalies that occur in children born with classic bladder exstrophy (CBE).

Methods: All patients in a database of 1044 patients with the bladder exstrophy-epispadias-cloacal exstrophy complex were reviewed. Those with CBE had their complete medical records reviewed. Children noted to have gastrointestinal malformations were identified, and all aspects of their history were extracted.

Results: A total of 676 patients were identified with CBE, of whom 12 patients were identified who had a concomitant colorectal anomaly. In this population, the proportion of colorectal anomalies is 1.8%. The most common gastrointestinal anomaly was imperforate anus in 8 patients. Two patients had severe rectal stenosis requiring serial dilations, and 2 patients were born with congenital rectal prolapse.

Conclusions: Excluding all variants and cloacal patients with exstrophy, isolated colorectal anomalies occur at a rate of 1.8% in children born with CBE, a 72-fold increase compared with the general population. Although uncommon in this rare birth defect, the morbidity is significant and warrants prompt evaluation and treatment of both the genitourinary and colorectal anomalies.

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Bladder exstrophy presents as a spectrum of disorders from mild exstrophy variants, such as a superior vesical fissure; to complete epispadias; then classic exstrophy; and finally, cloacal exstrophy. Classic bladder exstrophy (CBE) is the most common form of the exstrophy spectrum. Although generally considered to be a primary urologic malformation, the defect is 1 failure of the abdominal wall and pelvis to close and involves severe defects in the bony pelvis, pelvic organs, and pelvic floor musculature [1]. The widely splayed pelvic bones cause an underlying lack of support of the pelvic genitourinary (GU) organs while also affecting the rectum and the distal colon because the puborectalis muscle is outwardly rotated and more posteriorly located than normal [2]. In addition, recent longitudinal studies of patients with exstrophy in adulthood now recognize that there is significant pelvic floor dysfunction causing issues with defecation and fecal continence that must be addressed [3].

There exists a subset of patients with exstrophy who have an anatomical anomaly that is not necessarily part of the exstrophy spectrum in addition to the CBE. Cadeddu et al [4] in 1997 described a 6.7% proportion of congenital spinal

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anomalies seen in children born with CBE resulting from defective closure of the neural tube early in fetal life. Renal anomalies such as horseshoe kidney, pelvic kidney, and solitary kidney exist, although extremely rarely [5]. From a gynecologic standpoint, the vaginal canal has a more vertical than horizontal lie; and the cervix inserts more distally on the superior vaginal wall close to the introitus [6]. However, in CBE, reported gastrointestinal (GI) anomalies are exceedingly rare because patients with GI malformations such as imperforate anus and omphalocele are more almost universally associated with cloacal exstrophy than CBE.

With a patient with exstrophy database containing more than 1000 children, we have noted an association between CBE and colorectal malformations. Therefore, we reviewed our experience with the bladder exstrophy population to determine the proportion of colorectal anomalies that present concomitantly in newborns with CBE.

1. Materials and methods

We reviewed 1044 patients in an institutionally approved database containing all patients with a diagnosis of exstrophy evaluated at our institution between 1974 and 2010. The inclusion criteria for this study were a diagnosis of CBE and complete and available medical records with a comprehensive, documented physical examination. Patients were excluded if they had an exstrophy variant, complete epispadias, cloacal exstrophy, or classic exstrophy with any associated spinal or neurologic anomaly. A total of 676 patients with CBE were identified for the study.

Each patient had their complete medical record evaluated for any evidence of evaluation for or treatment for any GI issue. All colorectal anomalies that were noted were confirmed to be congenital in nature. All reports were reviewed by the senior author to ensure diagnostic accuracy. Each colorectal malformation was classified by the location and the intervention required. Proportion of colorectal malformations was calculated for the entire cohort of patients evaluated at our institution for CBE.

2. Results

A total of 1044 patients with the exstrophy-epispadias-cloacal exstrophy condition have been evaluated or treated at our institution for a 36-year period. Of these, 676 have CBE, with the remaining 368 patients having cloacal exstrophy, epispadias, an exstrophy variant, or incomplete records for evaluation.

Of this cohort of 676 patients, boys were represented in a 2.5:1 ratio to girls, 482 patients to 193, respectively. Of those patients whose data on ethnicity/race were available for review, the CBE population was 93.7% white, 3.3% African American, 1.6% Hispanic, 0.7% Native American, and 0.7% Asian/Indian.

Table 1 Associated colorectal anomalies

Colorectal malformation	No. of patients	% affected	% CBE cohort
Rectal stenosis [a]	2	16.7	0.3
Rectal prolapse [b]	2	16.7	0.3
Imperforate anus	8	66.6	1.2

Identified colorectal anomalies associated with patients presenting with CBE.

[a] Requiring operative dilation of the rectum.

[b] Congenital before primary closure.

Of the 676 patients, 12 patients or 1.8% were identified as having a colorectal anomaly in addition to CBE (Table 1). The most commonly identified GI anomaly was an imperforate anus in 8 patients (1.2%), followed by severe rectal stenosis in 2 patients (0.3%) and congenital rectal prolapse in 2 patients (0.3%). All 8 patients with imperforate anus underwent initial colostomy to manage their imperforate anus. At the time of this review, 6 patients had undergone reconstruction with some type of pull-through repair; and 2 patients are still awaiting reconstruction. In these 6 patients, anorectal reconstruction was performed at a mean age of 11 months with a range of 3 to 24 months; where operative reports were available for review, posterior sagittal anorectoplasty was the surgical approach used. Of the 2 patients awaiting reconstruction, 1 child is 6 months old, waiting reconstruction within the next 6 months; and the other patient is 4 years from a Third World country where access to medical care has delayed his reconstruction. Both patients with severe rectal stenosis required serial dilations to facilitate defecation; however, neither required formal surgical correction. The rectal innervation in these patients was confirmed to be normal via evaluation with a nerve stimulator. In the 2 patients born with congenital rectal prolapse, the prolapse was present before any closure procedures or surgical reconstruction of the bladder exstrophy defect. Both patients with rectal prolapse required repair after their bladder was successfully closed.

3. Discussion

The existence of GI malformations in the exstrophy complex is well documented; however, most of these malformations present in patients with more severe forms of the disease, such as cloacal exstrophy [7]. Traditionally, the most common form of bladder exstrophy, CBE, has been classified primarily as a GU malformation. However, for the past 3 decades, patients presenting with CBE have also been found to have significantly higher proportions of associated congenital anomalies than the general population. Most notably to date is the 6.7% proportion of isolated spinal abnormalities in patients with CBE [4]. The presence of exstrophy variants reemphasizes the spectrum of severity

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