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Achieving urinary continence in cloacal exstrophy

Ranjiv Mathews, MD

From Johns Hopkins Hospital, Baltimore, Maryland.

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Cloacal exstrophy is the most significant urological anomaly compatible with life. The development of urinary continence is compromised significantly by the anatomic and neurological deficits that are a part of this complex anomaly. Most children with cloacal exstrophy can eventually be made continent by the use of current reconstructive techniques. This article summarizes some of the challenges and reconstructive methods to permit eventual continence in children with cloacal exstrophy.

Continence in cloacal exstrophy continues to remain a challenge. Complete continence in cloacal exstrophy can only considered if the patient is continent of urine and stool. The multisystem nature of cloacal exstrophy makes the potential for voided urinary continence and stool continence with defecation relatively rare. Children who are able to achieve voided continence and defecatory stool continence typically have variants of cloacal exstrophy. Although general principles can be suggested for achieving continence in children with cloacal exstrophy, the wide variation in anatomic defects and innervations make an individualized approach imperative for success.

As recently as the 1970s long-term survival of children with cloacal exstrophy was considered exceptional.[1] There was a rapid improvement in overall survival from 20% in the 1960s and 1970s to 90% in the 1980s.[2] Initial attempts at reconstruction were directed at preservation of life and the use of incontinent diversions was considered the norm. With the marked improvement in survival, efforts turned to improving quality of life. The first reports of achieving urinary continence were reported in the 1980s.[3] In the initial report by Diamond and Jeffs, functional continence with a dry interval for urine was achieved in 43% of patients.[3] Because of the relatively rarity of the condition, reports on continence remained single-case experiences.[4,5]

Address reprint requests and correspondence: Ranjiv Mathews, MD, Johns Hopkins Hospital, 600 N Wolfe Street, Baltimore, MD 21287.
E-mail address: Rmathews@jhmi.edu.

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In an effort to develop a logical management strategy, Manzoni et al[6] developed a grid for identifying the anatomic abnormalities present in classical cloacal exstrophy. Ability to achieve continence is determined by the extent of the anomalies present. Despite efforts to classify cloacal exstrophy patients into treatment groups, the best surgical outcomes appeared to be achieved by the use of an individualized approach.[7]

Evaluation for potential continence

Reconstruction for cloacal exstrophy is performed with a staged approach. Initial bowel diversion (terminal ileostomy/colostomy) is performed along with approximation of bladder halves or closure of the bladder. In girls, reconstruction of the external genitalia is typically performed at the time of bladder closure; however, many girls will require further surgical management to improve the cosmetic outcomes of the external genitalia. Boys typically have epispadias reconstruction performed at 6-12 months of age. Bladder growth is evaluated yearly by the use of gravity cystograms under anesthesia. Patients who have significant spinal defects should have urinary tract evaluation similar to other forms of spinal dysraphism. Encouragement of early and complete bladder emptying along with the use of anticholinergics will permit improvement in bladder capacity over time. Most series indicate that despite close follow-up many children

will require additional reconstructive procedures for eventual continence.[8]

Spontaneous voiding per urethra is noted in a minority of patients with cloacal exstrophy. Some patients are able to be dry with intermittent catheterization per urethra; however, most require eventual incontinent or continent urinary diversion. Because of lack of bladder growth many children with require augmentation cystoplasty in conjunction with continent urinary diversion or bladder neck reconstruction to be dry. When considering timing and type of surgical reconstruction many factors should be considered.

Social considerations

The maturity and desire of the child to be continent are crucial to the success of any operative reconstruction. Children who lack the capacity, because of physical or mental limitation, to participate in a catheterization regimen, may be best managed with incontinent diversions. Parental support can help with insuring that the child is compliant with the required care of the continent or incontinent diversion. Nursing assistance in the immediate postoperative period can also be helpful in getting patients and family comfortable with care of the urinary diversion.

Anatomic considerations

Bladder factors

The capacity of the bladder and the presence or lack of contractility are important in the consideration of the appropriate surgical reconstructive procedure. Evaluation of the bladder by the use of preoperative urodynamics can discern the capabilities of the bladder. A large-capacity, noncontractile bladder may be managed with a bladder outlet procedure and intermittent catheterization through the urethra or a continent channel. A small-capacity, high-pressure bladder, by contrast, will most like require augmentation along with increased outlet resistance, to provide continence.

Urethral factors

The male urethra following epispadias reconstruction and the female urethra following bladder neck reconstruction can be difficult to negotiate, making intermittent catheterization per urethra difficult to accomplish. Patients who have normal perineal sensation may also have difficulty complying with a urethral catheterization regimen. Children who are wheelchair bound and have upper-extremity deficits may also be unable to manage a catheterization regimen per urethra.

Skeletal/neurological factors

Significant contractures of the spine and the resulting abdominal changes can make surgical reconstruction challenging. The reduction in available abdominal wall and the need for separation of the bowel stoma from the urinary diversion (continent or incontinent) require individualization of surgical reconstruction. Careful preoperative planning is therefore crucial for post operative success.

The single most important factor in the achievement of voided urinary continence is the level and degree of the spinal anomaly present. Many children with cloacal exstrophy have significant spinal anomalies that affect bladder contractility making the development of voided continence unlikely.[9] Preoperative urodynamic evaluation to determine bladder contractility may be beneficial to determine if a patient can develop voided continence.

Bowel factors

Many children with cloacal exstrophy may have a preexisting significant reduction in available bowel length and have varying degrees of malabsorption. When considering the use of a bowel segment to increase bladder capacity, the possible impact on bowel function should be carefully considered. If any hindgut segments remain, they may be considered for use in urinary tract augmentation.

Techniques for achieving urinary continence

Incontinent diversions

Children who have limited ability to catheterize because of physical or mental limitation are candidates for incontinent diversion in the form of an ileal or sigmoid conduit. This form of diversion is associated with significant potential for long-term upper tract deterioration because the ureters are reimplanted with no effort to prevent reflux. Persistence of bacteriuria and the potential for stone formation are reasons why this management has been relegated to those patients who are not candidates for continent urinary systems. In addition, the need to have a second appliance with children that frequently already have a bowel appliance makes this the least desirable approach for most children who can have a continent urinary reconstruction.

"Single-stage" reconstruction

"Complete" closure of the bladder in conjunction with tapering of the bladder neck has been suggested as a single procedure to achieve continence. Patients have initial reconstruction with approximation of the bladder halves to convert the patient into a classic bladder exstrophy. The bladder is then reconstructed as with classic exstrophy. In the initial report by Lee et al,[10] 6 patients underwent reconstruction using this approach. All patients had osteotomy for reconstruction. Two patients developed hydronephrosis and were noted to have an associated bladder septum. A third patient required ureteral reimplantation because of the development of recurrent urinary tract infection in the context of vesicoureteral reflux. Only one of 6 children eventually had spontaneous voided continence. All