Cloacal Exstrophy

Gianantonio M. MANZONI and Richard S. HURWITZ

Cloacal exstrophy is the rarest and most extreme form of the exstrophy-epispadias complex occurring only once in 200,000 [33] to 400,000 [36] births. In contrast to classical bladder exstrophy, these patients are born with an extroverted **bladder-and-bowel complex** which occupies a defect of variable size in the lower abdominal wall and perineal region. An omphalocele and severely deranged external genitalia almost always accompany the bizzare bladder/bowel malformation.

In addition, associated congenital anomalies of multiple organ systems are often present, adding to the severity of this devastating condition. Prior to 1960 cloacal exstrophy was thought to be "uniformly fatal in the neonatal period" [32] with no survivors reported despite attempts at surgical reconstruction. These patients were often left to die because of the magnitude of the anomaly and the fatal prognosis.

However, during the past decade, not only have excellent survival rates been achieved, but urinary continence is also possible because of new and innovative reconstructive procedures [2,5,22,26].

A detailed analysis of 34 patients with cloacal exstrophy has provided the basis for a new system of cloacal exstrophy classification into classical and variant groups [9]. This information has also resulted in a coding system which affords a means of efficient communication about the detailed variations encountered in cloacal exstrophy and facilitates the recognition of similarities between cases [21]. The observation that the variations between cases conform to specific recurring patterns has led to the idea of a cloacal exstrophy "pathway" with embryologic implications.

The neonatal investigation of the cloacal exstrophy patient must be performed logically and rapidly, leading to a complete understanding of the complex and unique internal anatomy and to the detection of all associated anomalies. Physical examination, contrast studies, urography and/or ultrasound, radioisotope scans, endoscopic procedures, and karyotype determination will provide the framework of information that will direct management.

The management of cloacal exstrophy is complex and requires a multidisciplinary approach. Correct neonatal surgical treatment is crucial to the final outcome and involves separating the gastrointestinal from the urinary system, closing the abdominal defect, and maximizing conditions for survival. Subsequent reconstructive procedures aim at achieving continence, preserving renal function, correcting orthopedic anomalies, and creating functional genitalia to enhance self-esteem and social acceptability. The surgical principles that form the basis of optimal treatment for these patients are presented.

Anatomical presentation

The main features of classical cloacal exstrophy are an exstrophic central bowel field flanked by two hemibladders, and in 90 % of cases an omphalocele is present. The central bowel field is the ileocecal region and it has three or four orifices. The proximal orifice leads to the terminal ileum which often prolapses producing the "elephant trunk" deformity. The distal orifice leads to a short, blind-ending colon segment that is thought to be the persistent tailgut. One or two appendiceal orifices may be present and occasionally an appendiceal prolapse has been observed. The anus in imperforate. The hemibladders are often asymmetric and each has a ureteral orifice (Fig. 1). Although the hemibladders are usually located on the sides of the central bowel field, not uncommonly they are confluent cranial or caudal to the bowel.

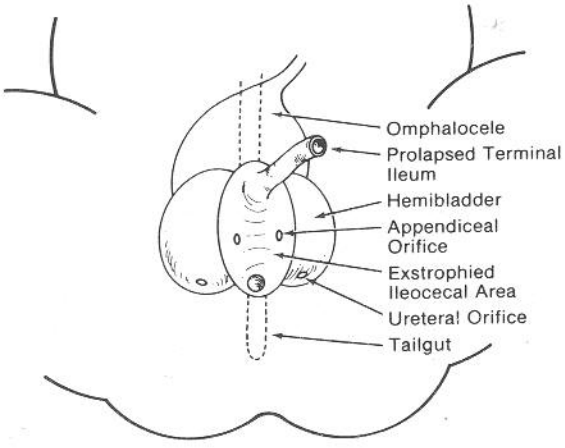


Figure 1. Classical cloacal exstrophy (from Hurwitz RS, Manzoni GM, Ransley PG, Stephens FD 1987 Journal of Urology 138:1060. Copyright Williams and Wilkins, by courtesy of the authors and publisher).

In 30 % of **males** the penis is absent, but more often it is represented by two widely separated, rudimentary unicorporeal structures. About 20 % of males have an epispadias or a fully formed penis, but often these united phallic structures are small and of poor quality. The testes are usually intra-abdominal, but on occasion they are palpable in the groins. Rarely are they fully descended. The scrotum may be absent, widely separated into two hemiscrota, or less commonly bifid.

In **females** the clitoris is usually divided into two widely separated hemiclitori, but the clitoral structures may also be absent or united. The ovaries are normal, while Müllerian fusion anomalies are almost always found. Uterine duplication is present in 95 % of females, vaginal duplication occurs in 65 %, and in 25 % the vagina is absent [33]. Uterine agenesis with fallopian tubes joining the lower ureters has been noted [9].

Associated anomalies

The condition of the cloacal exstrophy patient is often further aggravated by the presence of associated congenital anomalies of multiple organ systems. Associated anomalies were found in 85 % of our patients. They were present in 95 % of the classical cases and 75 % of the variant population, however the types of associated anomalies in both groups were similar [9]. The anomalies found in our review are compared with those of other large series in Table 1 [22,29,30].

Table 1. Associated anomalies (%)

	Soper (1964)	Spencer (1965)	Hurwitz (1987)	Mitchell (1990)
Upper urinary tract	62	56	66	88
Vertebral	72	80	48	ND
Gastrointestinal	ND	83	46	19
CNS (Myelodysplasia)	45	ND	29	75
Lower extremities	18	65	26	31

From Hurwitz RS, Manzoni GM, 1996, reproduced by permission of Butterworths. ND: No Data

Upper urinary tract abnormalities were the most common associated anomalies in both our patients (66 %) and Mitchell's patients (88 %). In both series pelvic kidney was the most common upper urinary tract anomaly (31 %; 44 %) followed by unilateral renal agenesis (21 %; 19 %). Multicystic kidney or tiny dysplastic kidney (10 %), ureteral duplication (7 %), crossed fused ectopia (3 %), and hydroureteronephrosis due to distal ureteral atresia (3 %) were also observed in our patients.

The incidence of **vertebral** anomalies ranges from 48-80 %. These included spina bifida defects, hemivertebrae, sacral agenesis, and kyphosis.

Gastrointestinal anomalies were present in 46 % of our cases, but may be as high as 83 % as reported by Spencer, since most patients have malrotations associated with a universal mesentery. Bowel duplications and anatomic short small bowel were each present in 15 % of our patients. The anatomic short small bowel was noted in five of our patients and did not seem to be related to early gestational age (range 33-40 weeks).

The **central nervous system** is affected in 29-75 of the patients with some form of myelodysplasia. Myelomeningocele (11 %) and meningocele (11 %) were more common than lipomeningocele (3 %) in our series, hovewer, lipomeningocele was far more common (63 %) in Mitchell's patients.

Anomalies of the **lower extremities** were present in 25 % of our patients, but these were more prevalent in Spencer's report (65 %). The lower extremity anomalies included congenital hip disclocation, talipes equinovarus, lower limb agenesis, and other severe deformities of the lower legs and feet.

Cardiovascular and **pulmonary** anomalies including cyanotic heart disease and aortic duplication

have been reported but are rare. Duplication of the inferior vena cava has been noted in 4 cases [3].

Classification and coding system

The cloacal exstrophy complex has been classified into two broad categories based only on bowel and bladder patterns (Fig. 2) [21]. Type I includes the classical cloacal exstrophy patterns and type II includes the variants. The **type I** patients may have variations in the surface bowel/bladder relationships which are defined in subgroups IA-hemibladders confluent cranial to the bowel field, IB-hemibladders on the sides of the bowel, and IC-hemibladders confluent caudal to the bowel (Fig. 3). The **type II** patients are also divided into three subgroups: IIA-only the bladder is at variance with the classical pattern, IIB-only the bowel abnormality is different, and IIC-both bladder and bowel differ from the classical pattern. This latter group often contains the most bizarre combination of abnormalities.

The coding system is a method of recording the surface pattern and each of the system variables for a particular case of cloacal exstrophy [21]. It also helps to organize ones thoughts about such complex and unfamiliar anatomy. A grid (Fig. 4) is formed

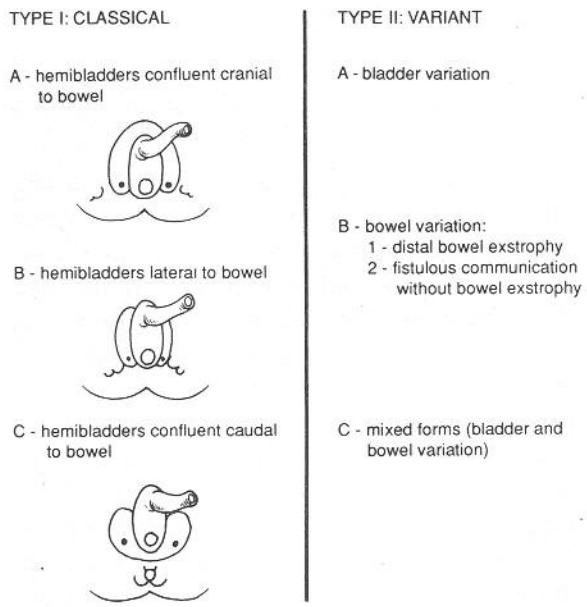


Figure 2. Cloacal exstrophy classification (Modified from Hurwitz RS, Manzoni GM, Ransley PG, Stephens FD 1987 Journal of Urology 138:1061. Coryright Williams and Wilkins, by courtesy of the authors and publisher. Modified version from Hurwitz RS, Manzoni GM, 1996, reproduced by permission of Butterworths).

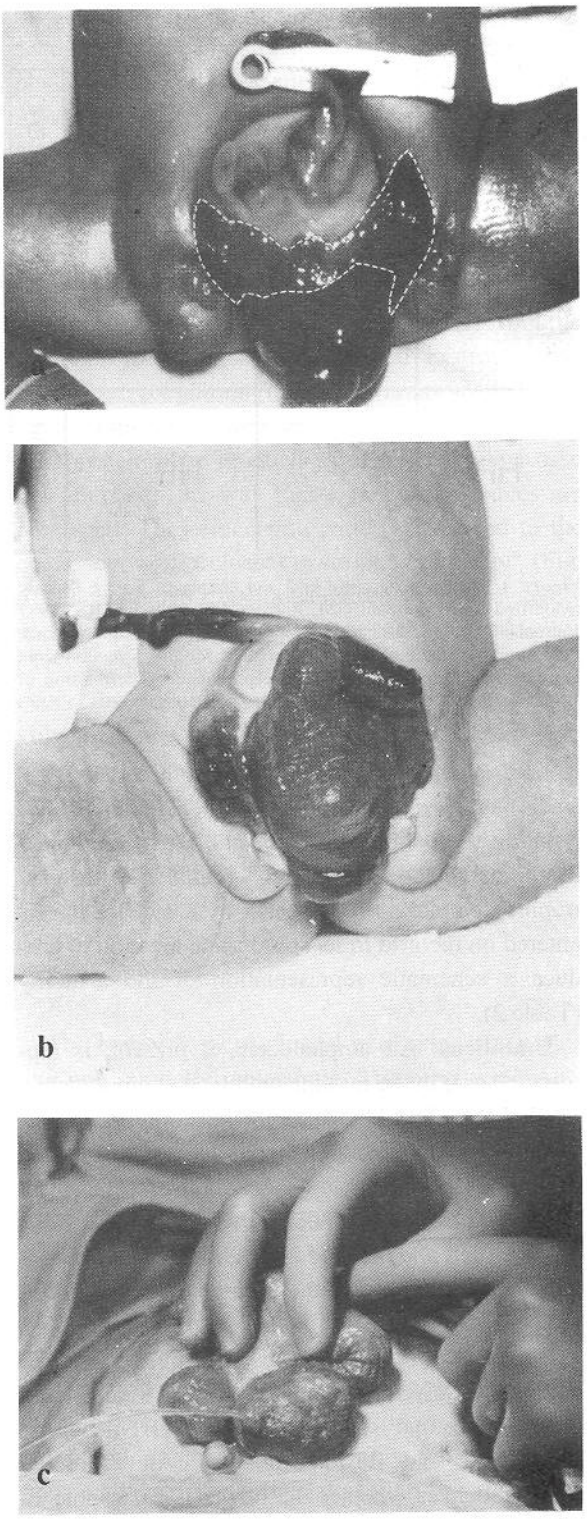


Figure 3. Classical cloacal exstrophy surface patterns. **a)** Bladder (outlined) confluent cranial to bowel, **b)** Bladder on sides of bowel, **c)** Bladder confluent caudal to bowel which is seen more clearly with the bowel retracted superiorly at the time of surgery. Note single midline penis (From Hurwitz RS, Manzoni GM, 1996, reproduced by permission of Butterworths).

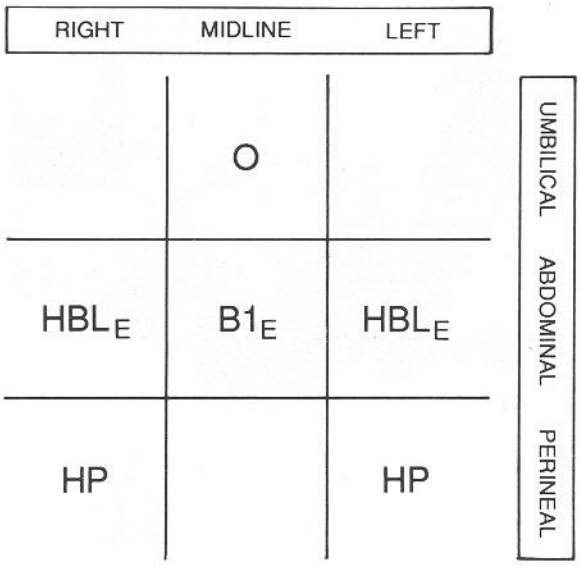


Figure 4. Cloacal exstrophy grid and coding of a type IB case (Modified from Manzoni GM, Ransley PG, Hurwitz RS 1987 Journal of Urology 138:1065. Copyright Williams and Wilkins, by courtesy of the authors and publisher. Modified version from Hurwitz RS, Manzoni GM, 1996, reproduced by permission of Butterworths).

from 3 horizontal (umbilical, abdominal and perineal) and 3 vertical (right, midline, and left) zones defining 9 separate compartments. Each component (umbilical, bladder, bowel, and genitalia) of the exstrophic complex is represented by a symbol that is entered on the grid in its appropriate location to produce a schematic representation of the anatomy (Table 2).

Umbilicus: An omphalocele, if present, is represented as 0 in the midline umbilical compartment.

Bladder: The bladder may be complete or divided into 2 hemibladders and is represented by BL or HBL, respectively. In the exstrophic state a suffix E is added as a lower index letter, while if the bladder is closed and covered a C is used. An additional letter F denotes a fistula to an otherwise closed system.

Bowel: The bowel is represented by the letter B, which is followed by a number 1, 2, or 3 depending on the intestinal segment involved (B1-ileocecal, B2-colonic, and B3-rectosigmoid). An additional suffix indicates whether the bowel is exstrophic E, covered C, or has a fistulous communication F. The bowel fistula may be to the abdominal surface or to a closed or open bladder. An intestinal duplication is represented as D and is followed by a site related number (1, 2 or 3) and by an index letter defining it

Table 2. Cloacal exstrophy coding symbols

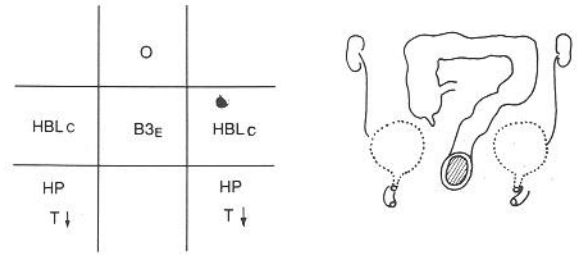
	Coiding	Exstrophied	Covered
Omphalocele			
Present	0		
Absent	-		
Bladder			
Complete	BL	BL$_E$	BLc*
Hemi	HBL	HBL$_E$	HBLc*
Bowel			
Primary			
Ileocecal	B1	B1$_E$	B1F*
Colonic	B2	B2$_E$	B2F*
Rectosigmoid	B3	B3$_E$	B3F*
Duplication			
Ileocecal	D1	D1$_E$	D1c*
Colonic	D2	D2$_E$	D2c*
Rectosigmoid	D3	D3$_E$	D3c*
Anus			
Present	A		
Absent	-		
Female genitalia			
Vagina			
Present	V	V$_E$	
Absent	-		
Clitoris			
Complete	CL		
Hemi	HCL		
Male genitalia			
Phallus			
Complete	P (P$_E$-epispadiac, P$_N$-normal, P$_H$-hypospadiac)		
Hemi	HP		
Testis			
Descended	T ↓		
Undescended	T ↑		

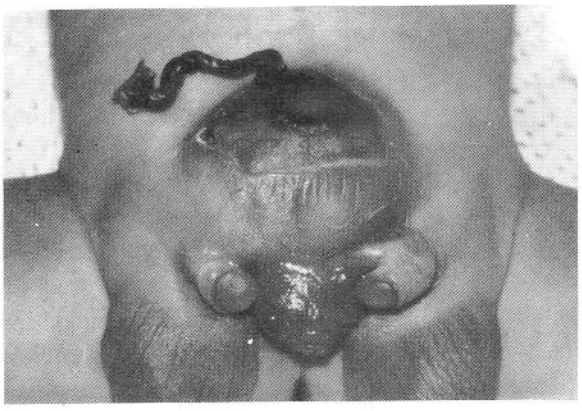
** F-plus fistula.*

+ If fistula is associated add (F), for example BLc(F). (From Manzoni GM, Ransley PG, Hurwitz RS, 1987 J Urol 138:1066. Copyright Williams and Wilkins, by courtesy of the authors and publisher.)

as exstrophied, covered, or associated with a fistula. The anus is usually imperforate. The rare presence of an anus is recorded as an A in the perineal zone. The bowel/bladder relationship is also recorded. When the hemibladders are fused either cranially or caudally to the bowel field, a line will connect the bladder symbols above or below, to indicate the presence of such a fusion.

Genitalia: In the female patient the vagina is indicated by the letter V and it may appear laterally if duplicated or in the midline perineal compartment. The very rare exstrophied vagina is symbolized further by a basal suffix E. The single complete clitoris is noted as CL, while the more common hemiclitoris is HCL. In the male the single complete phallus is represented by P and the urethral pattern is recorded as epispadic Pe, normal Pn, or hypospadic (rare) Ph. A hemiphallus is recorded as HP but no attempt is





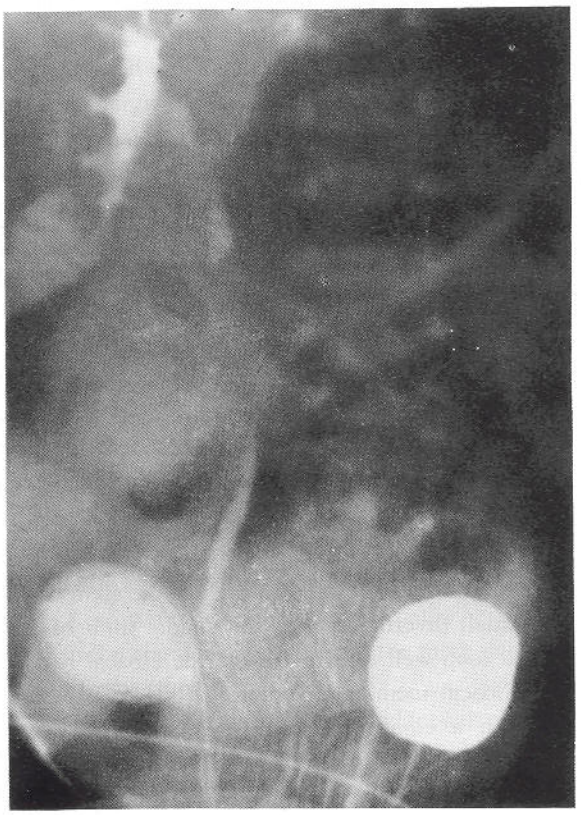


Figure 5 a). Coding and anatomic diagram of a type IIC male with two covered hemibladders and distal bowel exstrophy, **b)** Widely separated hemiphalli each associated with a urethra, and hemiscrota containing descended testes. Distal bowel exstrophy shown, **c)** Cystogram demonstrating two covered bladders and right vesicoureteric reflux (From Hurwitz RS, Manzoni GM, 1996, reproduced by permission of Butterworths).

made to classify the urethra which is occasionally encountered in such structures. When the clitoris or phallus is absent, no symbol is recorded. The usefulness of the coding system in communicating the various types of cloacal exstrophy is demonstrated in a type IB (Fig. 4) and a type IIC case (Fig. 5).

The cloacal exstrophy pathway

One of the most fascinating observations from the study of multiple cases of cloacal exstrophy has been the recognition that the variations between cases conform to well recognized patterns within each system. The uniqueness of an individual case lies simply in the way these standard variables are combined. These recurring patterns have led to the construction of a cloacal exstrophy "pathway" (Fig. 6). Each framed diagram represents a case actually seen in our series, while unframed examples are cases we did not observe but which we expect do occur. Progression from proximal to distal bowel involvement and from the exstrophied to the covered forms are expressions of time related embryological events. It would appear that there is a spectrum in the development of these anomalies with classical bladder exstrophy, classical cloaca, and bladder duplication possibly being peripherally related [11].

Embryology

The embryology of cloacal exstrophy is complex and only partially understood. The most widely accepted theory was proposed by TB Johnston in 1913. He believed that cloacal exstrophy was due to premature rupture of the cloacal membrane at any time between the first appearance of the membrane and the completion of the subdivision of the cloaca. Furthermore, he felt that the time at which the cloacal membrane ruptured determined the variety of the anomaly.

At 2 1/2 to 3 weeks of gestation the cloacal membrane covers not only the cloacal area, but also extends along the allantoic diverticulum as far as the body stalk [35]. The urorectal septum has not yet developed and the primitive hindgut which contains the primordia of the terminal ileum, cecum, and colon occupies essentially the same position as the posterior wall of the cloaca, while the bladder primordial area is located over the anterolateral aspect

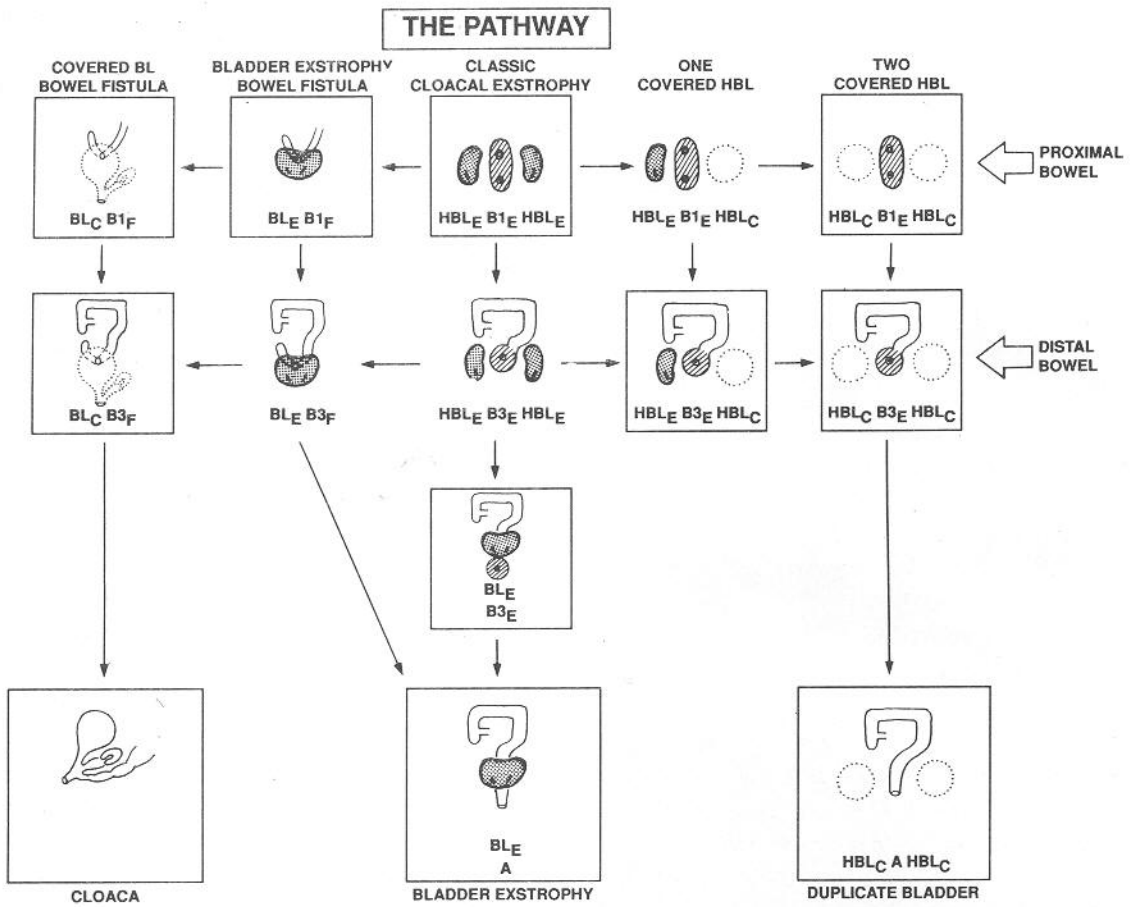


Figure 6. Cloacal exstrophy pathway. The pathway demonstrates how variations between cases conform to recognizable patterns. Starting with the classical pattern, progression to the covered forms, distal bowel forms, or combinations involving both covered and distal bowel variations may occur. Framed diagrams represent cases actually seen in our series (From Hurwitz RS, Manzoni GM, 1996 reproduced by permission of Butterworths).

of the cloaca. Rupture of the cloacal membrane at this very early time exposes the undivided cloaca and allows it to evert, resulting in the classical pattern of a central bowel field composed of hindgut primordia and two lateral hemibladders [15].

In the patient with classical cloacal exstrophy, all or the majority of the hindgut derivatives are trapped within the undivided cloacal complex. The ultimate ceco-appendiceal location may be proximal, at, or distal to the site of bowel exstrophy depending on the position of these hindgut primordia at the time of extroversion. This variaton in ceco-appendiceal position is well illustrated in the cases described by Magnus [19]. However, her explanation for these findings (strangulated loops of bowel) has been criticized for being at variance with accepted theories [14,23].

It is assumed that for the derivatives of the hindgut to develop they must be separated from the cloaca by the partitioning process of the urorectal (perhaps better termed "urohindgut") septum. The extent to which the urorectal septum has separated the posterior hindgut from the anterior urogenital sinus at the time of cloacal membrane rupture determines the amount of hindgut differentiation that will be present. In the normal human embryo, the cecal primordium is first seen early in the sixth week as a small diverticulum on the caudal limb of the midgut loop well separated from the cloacal region [34]. Cloacal membrane rupture at this time would result in a cloacal exstrophy variant with distal bowel involvement. The short blind-ending bowel segment distal to the exstrophic site is thought to be derived from the persistent post anal or tailgut. It may form from a process of distal subdivision of the cloaca by lateral ingrowths of mesoderm [15] or, because of its fixed distal location, result merely as a consequence of primitive hindgut eversion at a

slightly more proximal position.

The formation of the three surface patterns of classical cloacal exstrophy can be explained on the basis of the position and extent of the cloacal membrane relative to the underlying bladder primordium. The size and position of the bladder primordium may also be important factors. A detailed discussion of the embryogenesis of the classical and variant surface patterns of cloacal exstrophy has been presented elsewhere [11].

Neonatal assessment

The cloacal exstrophy team

The complex clinical evaluation and management of these newborns inevitably requires a multidisciplinary approach. Fluid and electrolyte as well as nutritional and metabolic problems demand the close attention of the neonatologist. The surgical team should be composed of a pediatric surgeon and a pediatric urologist who are thoroughly familiar with the principles and options of treatment. Often orthopedic and neurosurgical evaluation and management are required. Social and psychiatric support services can be invaluable in helping the emotionally traumatized parents cope with the overwhelming situation.

Investigation

The immediate neonatal evaluation plays a crucial role in the strategy for successful surgical reconstruction. A superficial or incomplete assessment can lead to potentially harmful and dangerous surgical decisions, making further treatment even more difficult. Neonatal evaluation should be directed towards the genitourinary and gastrointestinal systems and also towards the detection of associated anomalies. A meticulous physical examination can distinguish the classical (type I) from the more unusual variant forms (type II).

The pattern of the exstrophied bladder can usually be recognized, but a cystogram may be required for those rare cases with a covered hemibladder(s) to determine the anatomy and exclude the presence of vesicoureteric reflux (Fig. 5C). Contrast studies and/or endoscopy are often required to define the precise anatomical bowel pattern (i.e. segment involved). Early renal assessment is mandatory with an ultrasound while subsequent evaluation is achieved either with an IVP or nuclear medicine studies. Abdominal and sacral x-rays are useful for identification of vertebral defects while MRI may be subsequently required for better definition and diagnosis of spinal cord dysraphisms. Since gender identification is often impossible because of the severe derangement of the external genitalia, a karyotype will enable the surgeon to anticipate the findings of the internal genitalia and properly counsel the parents regarding possible gender reassignment.

Management

History

Cloacal exstrophy was considered a uniformly fatal malformation only 30 years ago. No survivors were reported before 1960 despite attempts at surgical reconstruction. These dismal results led to an attitude that these infants were hopeless and surgical treatment was often withheld. In 1960, Rickham generated renewed interest in surgical reconstruction when he reported the first surgically treated survivor [27].

However, the continued high mortality rates and disappointing results during the 1960's led many surgeons to readopt the no treatment philosophy. Reconstructive efforts emerged again in the 1980's with excellent survival rates in the 80-100 % range [2,9,22,26,36], probably as a result of technological advances in suppot systems such as neonatal intensive care and refinements in hyperalimentation and antibiotic therapy.

Although it is now generally accepted that almost all cloacal exstrophy patients should undergo reconstruction and be given the chance to survive, the unanswerable moral and ethical questions about the consequences of treatment versus nontreatment inevitably arise when there are multiple severe associated anomalies. When one considers that the life saving reconstructive efforts are often the beginning of an enormous life-long physical and emotional burden for the patient and the family, one cannot help but wonder whether heroic efforts in these severest cases are the kindest course of action.

Surgical reconstruction

The surgical management of cloacal exstrophy can be divided into two general phases: 1) the neonatal closure of the defect which may be done in one or two stages and 2) the subsequent reconstructive procedures to achieve continence, preserve renal function, and create functional genitalia. Because of the extreme variability of these cases, management must be highly individualized. However, there are certain general guidelines that apply to all cloacal exstrophy patients. (A film demonstrating the principles of surgical management is also available [8].)

Phase 1: Neonatal closure

The important considerations at the time of initial closure include determining whether a 1 or 2-stage closure is most appropriate, and deciding upon the optimal management of the omphalocele, bowel, hemibladders, and genitalia, and whether gender reassigment is required [7]. The management options for each component are shown in the algorithms (Fig. 7,8,10). In selected cases the longer, more involved single-stage closure may be appropriate if the patient is in excellent condition and has favorable anatomy with minimal associated anomalies. However, many of these patients are premature, small for dates, and have such severe associated anomalies that it may be difficult for them to withstand an extensive single-stage reconstruction in the newborn period.

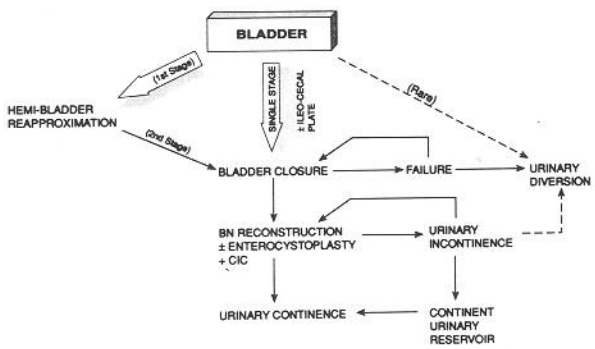


Figure 7. Options in the management of the bladder

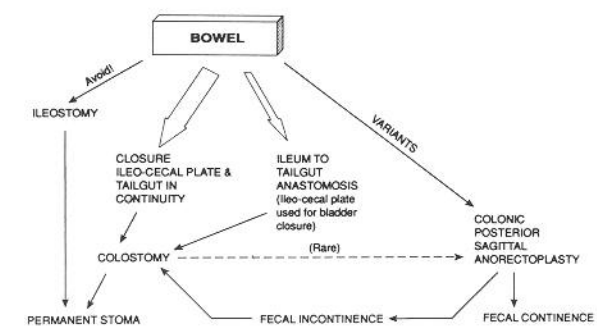


Figure 8. Options in the management of the bowel.

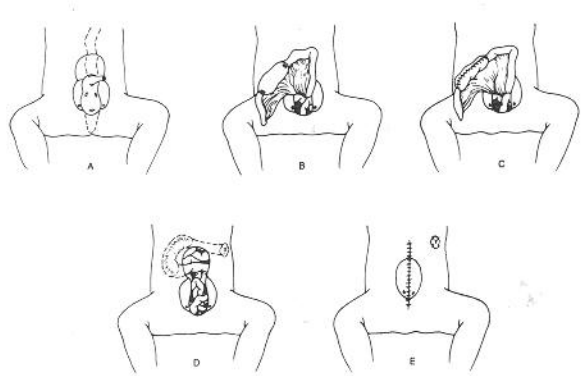


Figure 9. Cloacal exstrophy closure. First stage of a 2-stage procedure. **a)** Presenting cloacal exstrophy defect, **b)** Excision of omphalocele and separation of bowel from hemibladders, **c)** Exstrophic ileocecal region closed in continuity, **d)** Colostomy created from end of distal colon segment, **e)** Hemibladders reapproximated and omphalocele defect closed. (From Hurwitz RS, Manzoni GM, 1996, reproduced by permission of Butterworths).

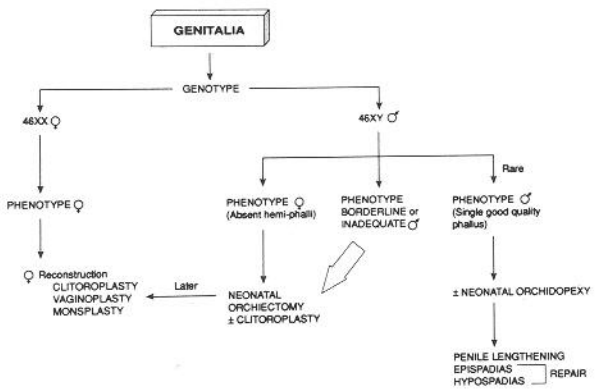


Figure 10. Options in the management of the genitalia.

During the first stage of a two-stage procedure the omphalocele is excised, the bowel is separated from the hemibladders, the exstrophic ileocecal region is closed in continuity with the tailgut and a colostomy (tailgut-ostomy) is created at its distal end. The hemibladders are then reapproximated in the midline to create a single exstrophic bladder (Fig. 9). This helps to close the lower abdominal defect, especially if the omphalocele is large. The need for prosthetic materials to achieve closure has been reported occasionally [7]. When the patient is stable and thriving, bilateral osteotomies (posterior iliac or anterior

innominate) and bladder closure are performed similar to a standard exstrophy closure.

The other option is single stage neonatal bladder closure. If the hemibladders are very small, consideration can be given to leaving the exstrophied ileocecal plate between them as an autoaugmentation. The terminal ileum and tailgut would then be detached from the exstrophied ileocecal region, rejoined, and a tailgut-ostomy formed. Since the urethra is usually absent, construction of a neourethra can be challenging. At the time of bladder closure a urethra may be made by tubularizing locally available tissues such as perineal and paraexstrophy skin [5,9]. During later reconstructive procedures a variety of anatomic parts including vagina [5], ureter, tapered ileum, and stomach [22], an ileal nipple [6] and tubularized bladder [18] have been used to create a continent urethra.

Since at least 25-50 % of these patients have a life-threatening short bowel syndrome, it is extremely important to maximize the total bowel length by preserving and using all available bowel in forming the distal gastrointestinal tract. Many patients managed initially with a terminal ileostomy have been plagued by massive fluid and electrolyte losses and malnutrition. Patients with ileostomies have required longer hospital stays for gastrointestinal complications and prolonged use of parenteral hyperalimentation has been necessary [12,26].

This problem is often markedly improved by adding on the tailgut [2]. This small segment of bowel which averages 10 cm in length in classical cases has actually been observed to greatly enlarge with recognizable taeniae when it is used initially as a fecal colostomy [1]. It is important to emphasize that the tailgut segment is valuable and should not be excised. Although we believe it should be used primarily for functional bowel length, in selected cases it may be used for vaginal or urethral reconstruction, bladder augmentation, or as a colon conduit.

Another consideration in the management of the bowel is the location of the colostomy. During the neonatal closure the colostomy should be positioned on the abdomen instead of being brought to the perineum by a pull-through technique. Since the chance of fecal continence is small, the stoma should be located where it can be managed easily with an appliance. The exception might be an infant with an extra long tailgut and no neurological deficit, but even in this instance, an initial end colostomy followed later by a posterior saggital anorectoplasty [24], for optimal placement of the colon within whatever functional musculature there is, would seem preferable to an initial "blind" pull-through.

Since the phallic structures in males with cloacal exstrophy are usually rudimentary and widely separated, attempts at penile reconstruction have generally been very unsatisfactory. Reports of long term follow-up of cloacal exstrophy patients raised as males have documented the disastrous results that occur when male gender identity is accompanied by an inadequate phallus [2,7,13]. There is therefore general agreement that almost all male cloacal exstrophy patients should be converted to females. When the phallus is a single midline structure, pressure from parents to retain male gender may be considerable and the temptation to reconstruct rather than to convert may be great. However, the presence of a single phallus is not in itself sufficient to assure adequate phallic development. Only in the rare situation where a good size phallus is present with corporeal tissue that responds to androgen stimulation would maintenance of male gender seem appropriate [10]. When gender reassignment is necessary, neonatal orchiectomy should be performed. This can be done as part of an early abdominal operation if the testes are intraabdominal or as part of an early genital reconstruction if present in the groin. Early orchiectomy is believed to minimize testosterone imprinting on the developing nervous system [31].

The creation of a vagina in gender reassigned males or in females with vaginal agenesis should be deferred until the post pubertal period. Although there might be some psychological advantages with an early procedure, these early attempts are probably more ornamental than functional.

Phase 2: Later reconstructive procedures

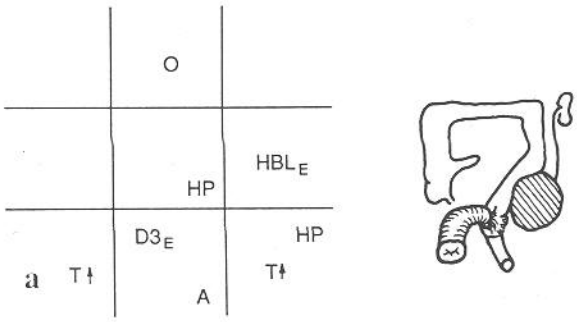
Reconstructive procedures following neonatal closure focus on developing a socially acceptable quality of life. The goals are to achieve urinary and fecal continence and provide functional genitalia.

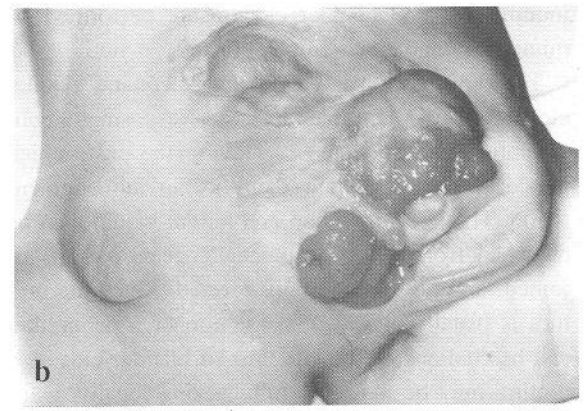
Urinary continence is accomplished by creating a large capacity, low pressure reservoir with adequate outlet resistance. Bladder augmentation, bladder

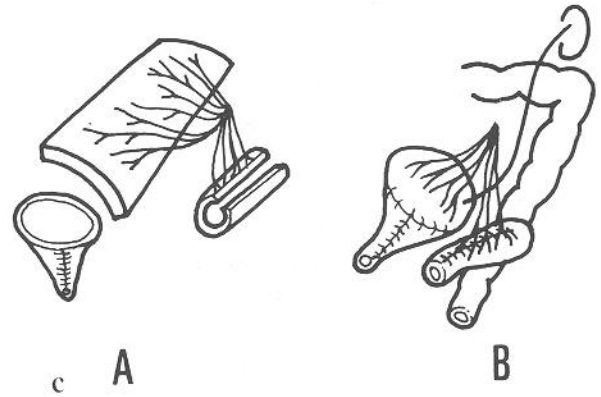
neck reconstruction, creation of a continent urinary reservoir, and the use of the Mitrofanoff principle [4] are the basic tools in achieving these goals. Therefore all tubular structures that might otherwise be discarded such as appendices, fallopian tubes, ureters, and bowel duplications, should be preserved for possible Mitrofanoff continence mechanisms.

An easily catheterizable channel is essential since almost all of these children will rely on clean intermittent self catheterization (CIC). The timing for this surgery must take into account a number of factors including age and motivation of the child, acceptance of CIC, parental support, and social environment.

A limiting factor is the amount of bowel that can be used safely for reconstructive procedures. In variants with distal bowel involvement or patients who have intestinal duplications (types II B,C) both ileal and colonic segments have been used without problems [25] (Fig. 11). In the classic ileocecal presentation, the short bowel syndrome usually resolves and nutritional status stabilizes by age 3 [12]. At this point some bowel can be "borrowed" for reconstructive purposes, although the amount that can be used is unknown. In the presence of limited intestinal availability, stomach is the segment of choice for lower urinary tract reconstruction. Stomach has been used successfully both for augmentation and for creating a continent urinary reservoir in cloacal exstrophy patients [22]. Other ad-







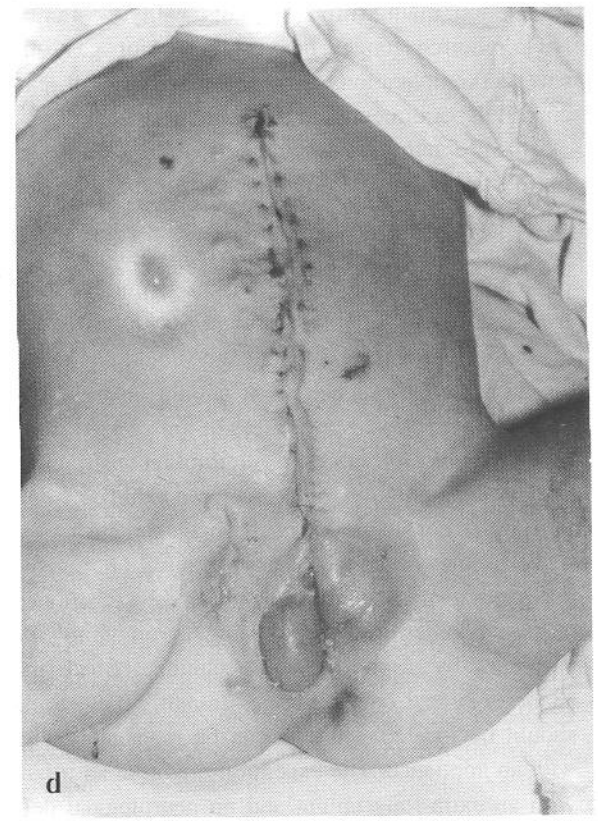


Figure 11. This type IIC male demonstrates utilization of excess bowel from an exstrophied colonic duplication for both augmentation and vaginoplasty. **a)** Coding grid and presenting anatomy, **b)** Single left sided exstrophic hemibladder and exstrophied distal colonic duplication are visible. Left sided ectopic anus is also present. Bifid hemiphallic structures and a left hemiscrotum are visible between the exstrophied hemibladder and bowel, and an ectopic hemiscrotum is noted on the right. The right kidney was absent, **c)** Reconstruction using part of the duplicate bowel for augmentation enterocystoplasty (A) and part for vaginoplasty (B), **d)** Immediate post-operative result (From Hurwitz RS, Manzoni GM, 1996, reproduced by permission of Butterworths).

vantages of stomach include less acidosis in patients with renal failure, less mucus production, decreased urinary tract infections, and the ability to relatively easily create a submucosal antireflux or continence mechanism.

Genital reconstruction to create a vagina in the teenage years must also be anticipated. Even if vaginal construction was done during one of the earlier operations, formal revision after puberty will usually be required [10]. Although techniques for vaginal construction using intestine [16] and skin grafts [28] have been described, experience with post pubertal vaginal reconstruction in cloacal exstrophy patients is limited.

Results and outcome

Important criteria in the assessment of outcomes for cloacal exstrophy patients include degree of preservation of vital organ function to sustain life (gastrointestinal, renal) and quality of life factors including urinary and fecal continence, adequacy of genital appearance and function, adequacy of abdominal wall appearance, and degree of psychosocial adjustment. Outcome information is now available in the literature regarding survival, and short term outcomes regarding urinary continence are beginning to emerge. However, information about the other important quality of life factors is scarce.

Diamond and Jeffs were the first to report satisfactory urinary continence after functional bladder closure in 3 of 7 evaluable patients who had dry intervals of 3 to 4 hours [2]. Mitchell and associates have also reported excellent success in achieving continence in 10 of 12 patients (83 %) using stomach and a variety of mechanisms for outlet resistance [23]. Ricketts and associates reported 3 of 11 patients continent of urine [26].

Fecal continence has been reported in only two patients [12] and one of these was a cloacal exstrophy variant [17]. Although voluntary fecal control should not be expected in these patients because of their associated sacral and bowel anomalies, "social" control with an enema program can be achieved in some of these patients (variants) with modern abdominoperineal pull-through procedures [26].

A new procedure to facilitate bowel evacuation has been recently reported. Based on the Mitrofanoff principle, a cutaneous non-refluxing appendicocecostomy is created through which an antegrade enema is performed intermittently to achieve fecal control [20].

In the last 30 years, cloacal exstrophy has evolved from a uniformly fatal malformation to a severe deformity with a 90-100 % survival rate. The focus is now on quality of life issues and on striving to provide these patients with continence and functional genitalia. It has been shown that with aggressive and innovative surgical procedures urinary continence is possible in these patients. As more surgeons become familiar with these newer techniques, it is likely that urinary continence in cloacal exstrophy will become the rule rather than the exception. Fecal continence continues to be elusive. Posterior saggital anorectoplasty may prove to be successful in selected patients, but fecal continence in the majority must await new developments. The lessons of raising male cloacal exstrophy patients with inadequate genitalia have been clearly learned. Gender reassignment in all but the most unusual male will avoid the disastrous problem of the sexually inadequate male, but the future social and sexual potential of these gender converted males and their female counterparts with vaginal agenesis remains uncertain.

The creation of a functional vagina after puberty is a major challenge and an area where limited information and guidence exists. As more patients with cloacal exstrophy reach puberty, experience with vaginal construction techniques will increase, and our goal of achieving functional genitalia for cloacal exstrophy females will hopefully be realized.

Acknowledgment

We wish to express our gratitude to the Pediatric Urology and Pediatric Surgery consultants from The Great Ormond Street Hospital for Children, London for providing many of the cases we have been privileged to review.

References

1. Colodny AH: Editorial comment. J Urol 133:782, 1985
2. Diamond DA, Jeffs RD: Cloacal exstrophy: a 22-year experience. J Urol 133:779, 1985
3. Diamond DA: Cloacal exstrophy: associated anomalies. Dialogues in Pediatric Urology 3:6, 1990
4. Duckett JW, Synder HM: Continent urinary diversion:

variations on the Mitrofanoff principle. J Urol 136:58, 1986
5. Gearhart JP, Jeffs RD: Techniques to create urinary continence in the cloacal exstrophy patient. J Urol 146:616, 1991
6. Hendren WH: Ileal nipple for continence in cloacal exstrophy: experience with 5 cases. J Urol (Suppl). 145:246A, 1991
7. Howell C, Caldamone A, Synder H, Ziegler M, Duckett J: Optimal managemet of cloacal exstrophy. J Pediatr Surg 18:365, 1983
8. Hurwitz RS, Muenchow SK: Surgical reconstruction of cloacal exstrophy (film). Wexler Film Productions, Inc, Los Angeles. (Available from American College of Surgeons/Davis+Geck Surgical Film-Video Library, Danbury, Connecticut, AUA/Norwich Eaton Video Library, Wilkes-Barre, Pennsylvania, National Library of Medicine, Bethesda, Maryland and British Medical Association Film Library, London), 1985
9. Hurwitz RS, Manzoni GM, Ransley PG, Stephens FD: Cloacal exstrophy: a report of 34 cases. J Urol 138:1060, 1987
10. Hurwitz RS: Cloacal exstrophy: Management of the genitalia. Dialogues in Pediatric Urology 3:5, 1990
11. Hurwitz RS, Manzoni GM: Cloacal exstrophy. In: O'Donnell B, Koff SA (Eds). Paediatric Urology, 3rd edn. Butterworths, London, 1996
12. Husmann DA, McLorie GA, Churchill BM, Ein SH: Management of the hindgut in cloacal exstrophy: terminal ileostomy versus colostomy. J Pediatr Surg 23:1107, 1988
13. Husmann DA, McLorie GA, Churchill BM: Phallic reconstruction in cloacal exstrophy. J Urol 142:563, 1989
14. Johnston JH: Letter to the editor. J Pediatr Surg 5:276, 1970
15. Johnston TB: Extroversion of the bladder, complicated by the presence of intestinal openings on the surface of the extroverted area. J Anat 48:89, 1913
16. Laub DR, Laub DR, Biber S: Vaginoplasty for gender confirmation. Clinics in Plastic Surg 3:463, 1988
17. Lobe TE: Fecal continence following an anterior-sagittal ano-enteroplasty in a patient with cloacal exstrophy. J Pediatr Surg 19:843, 1984
18. Lobe TE, Smey P, Anderson GF: A neo-urethral enteroplication for urinary continence in a case of cloacal exstrophy. J Pediatr Surg 20:616, 1985
19. Magnus RV: Ectopia cloacae -a misnomer. J Pediatr Surg 4:511, 1969
20. Malone PS, Ransley PG, Kiely EM: Preliminary report: the antegrade continence enema. Lancet 336:1217, 1990
21. Manzoni GM, Ransley PG, Hurwitz RS: Cloacal exstrophy and cloacal exstrophy variants: a proposed system of classification. J Urol 138:1065, 1987
22. Mitchell ME, Brito CG, Rink RC: Cloacal exstrophy reconstruction for urinary incontinence. J Urol 144:554, 1990
23. Muecke E, Marshall VF: Letter to the editor. J Pediatr Surg 5:276, 1970
24. Peña A, de Vries PA: Posterior sagittal anorectoplasty: important technical considerations and new applications. J Pediatr Surg 17:796, 1982
25. Ransley PG, Manzoni GM, Hurwitz RS: The management of the bowel in cloacal exstrophy. Dialogues in Pediatric Urology 3:2, 1990
26. Ricketts RR, Woodard JR, Zwiren GT, Andrews HG, Broecker BH: Modern treatment of cloacal exstrophy. J Pediatr Surg 26:444, 1991
27. Rickham PP: Vesico-intestinal fissure. Arch Dis Child 35:97, 1960
28. Sadove RC, Horton CE: Utilizing full-thickness skin grafts for vaginal reconstruction. Clinics Plastic Surg 3:443, 1988
29. Soper RT, Kilger K: Vesico-intestinal fissure. J Urol 92:490, 1964
30. Spencer R: Exstrophia splanchnica (exstrophy of the cloaca). Surgery 57:751, 1965
31. Stolar CJH, Randolph JG, Flanigan LP: Cloacal exstrophy: individualized management through a staged surgical approach. J Pediatr Surg 25:505,
32. Swan H, Christensen SP: Exstrophy of the cloaca. Pediatrics 12:645, 1953
33. Tank ES, Lindenauer SM: Principles of management of exstrophy of the cloaca. Am J Surg 119:95, 1970
34. Warwick R, Williams PL: Gray's anatomy. 35th British Edition. WB Saunders, Philadelphia, 1973. p.173-174
35. Wyburn GM: The development of the infra-umbilical portion of the abdominal wall, with remarks on the aetiology of the ectopia vesicae. J Anat 71:201, 1937
36. Ziegler MM, Duckett JW, Howell CG: Cloacal exstrophy. In: Welch KJ, Randolp JG, Ravitch MM, O'Neil JA, Rowe MI (Eds). Pediatric Surgery. Year Book Medical Publishers, Chicago, 1986. p.764-771

Gianantonio M. Manzoni, MD
Section of Pediatric Urology
Ospedale di Circolo
Varese