

AN UNUSUAL VARIANT OF CLOACAL EXTROPHY

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Cloacal extrophy, which is also called vesico-intestinal fissure, is one of the rare and complex malformations, with a reported incidence between 1 in 200,000 and 1 in 400,000 live births.^{1,2} Classically, cloacal extrophy consists of five components: omphalocele; extrophy of the two hemibladders; lateral cecal fissure which presents between the two hemibladders; imperforate anus; and ambiguous genitalia, but many other variants have also been described.^{3,5} This is a case report of an unusual variant of cloacal extrophy and an outline of aspects of its management.

Case Report

A male newborn, a product of full-term spontaneous vaginal delivery to a 24-year-old hypertensive, diabetic primigravida mother was referred to our hospital because of multiple congenital anomalies. His birth weight was 3.3 kg. Clinically, the neonate was found to have cloacal extrophy and fracture of the right humerus with Klumpke's paralysis secondary to birth trauma. He was also found to have a small ventricular septal defect (VSD) and moderate patent ductus arteriosus (PDA). There was no clinical evidence of meningomyelocele, and the legs were normal. Ultrasound examination of the urinary tract, brain and spinal cord revealed no obvious abnormality, and chromosomal studies were of a normal male pattern.

On the seventh day of life, he underwent examination under anesthesia, and surgery for his cloacal extrophy. The surgery revealed a small omphalocele, two extrophied hemibladders confluent superiorly, extrophied rectosigmoid junction, a complete phallus on the right side and a hemiphallus on the left side. (Figure 1). The right testes was down in the scrotum, the left testes was not palpable, and the he also had bilateral inguinal hernias. The exomphalos membrane was excised, and the urinary bladder was mobilized. As this child had sufficient colon length proximally, it was decided to use the extrophied colon for

bladder augmentation. The colon was divided distally, and a Hegar dilator was inserted through the prolapsed colon, which was divided in the midline anteriorly forming two separate colonic patches, one on each side. The two colonic patches were sutured together and to the two hemibladders, and the augmented urinary bladder was closed anteriorly. The posterior urethra was fashioned by mobilizing two flaps which were closed over a Malecot catheter. The left hemiphallus which was rudimentary was excised, and the complete right phallus was mobilized and swung medially to lie over the newly fashioned posterior urethra. This resulted in a penoscrotal hypospadias (Figure 2). The proximal end of the descending colon was brought out as an end colostomy. The abdominal wall was closed in layers, and an umbilicus was fashioned. Postoperatively, the patient was electively ventilated for seven days, and was discharged home 37 days postoperatively. Three months later, he was readmitted to the hospital, and had bilateral inguinal herniotomies. A micturating cystourethrogram was done at the age of six months. This revealed a good-sized urinary bladder with no vesoureteric reflux. At the age of nine months, the patient had a repair of his penoscrotal hypospadias, and is now passing urine per urethra (Figure 3). He is now waiting for a posterior sagittal anorectoplasty to correct his anorectal malformation.

FIGURE 2 Intraoperative photograph at the end of reconstruction. Note the catheter passing through the penoscrotal hypospadias.

FIGURE 3 Postoperative photograph showing a normal-looking phallus after repair of the penoscrotal hypospadias.

Discussion

Cloacal extrophy is a rare congenital malformation with an incidence of 1:200,000 to 1:400,000, and most reported series are from large referral centers and over a long period of time.^{1,2} Lund and Hendren reported 20 cases of cloacal extrophy which were seen over an 18 year period.⁴ Ricketts et al. treated 12 newborns with cloacal extrophy over a period of 10 years.⁶ Stolar et al treated 14 patients with cloacal extrophy over a 23-year period,⁷ and Cywes treated 24 infants with cloacal extrophy over 30-year period.⁸

The exact embryogenesis of cloacal extrophy is not known, and many theories have been suggested, however, no single theory seems to adequately explain all the abnormalities seen in the condition, but the most accepted theory is that cloacal extrophy results from premature rupture of the cloacal membrane prior to caudal migration

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FIGURE 1. Clinical photograph showing cloacal extrophy. Note the two phalluses, one on each side.

of the urorectal septum, and fusion of the genital tubercles.⁹ Classically, cloacal extrophy is made up of omphalocele, extrophied ileocecal region of bowel, extrophied hemibladders each with its ipsilateral ureter, and anorectal agenesis. The pubic bones are widely separated, and spinal dysraphism is common in these patients.

Commonly in cloacal extrophy, the extrophied bowel is the ileocecal region with little or no large bowel distally, but there are cases like ours where there is colonic extrophy with enough bowel length. In our patient, there was extrophy of the rectosigmoid colon. The presence of enough large bowel is advantageous from the reconstruction point of view, because normally the extrophied ileocecal region and in the presence of short large bowel should be preserved for reconstruction of the anorectal malformation, and not to be used for urinary bladder augmentation. Every effort should be made to preserve all large intestines because not only can they be used for bladder augmentation which is necessary in the majority of these patients to increase the bladder compliance, but they can also be used for vaginal reconstruction in those who require gender reassignment. In addition to this is the valuable absorptive function of the large bowel. In our patient, the extrophied rectosigmoid region was split in the middle to form two colonic patches which were sutured together and used for bladder augmentation. This will obviate the need for a second operation for the purpose of bladder augmentation. Augmentation of the urinary bladder may be performed using the hindgut if enough length is available, ileum, or part of the stomach. In the absence of the large intestine, both small bowel and stomach can be used for bladder augmentation but gastrocytoplasty has been shown to be superior.⁴

Gender assignment is one of the difficult tasks in the management of newborns with cloacal extrophy. Genetic females should normally not raise a problem as they will be raised as females. In genetic males with cloacal extrophy, the phallic structures are usually small and completely bifid, with insufficient phallic tissue to reconstruct an adequate penis. There is now general consensus that genetic males with insufficient phallus be gender reassigned as phenotypic females, and to minimize testosterone imprinting on the nervous system. This should be done in the immediate newborn period with early orchidectomy.⁴⁻⁷

In a large series of 20 patients with cloacal extrophy, 13 (65%) were genetically males, and all but one were given the female gender.⁴ Males with adequate bilateral or unilateral phallic structures should, however, be raised as males. Our patient had an adequate phallus on one side, and a small hemiphallus on the other side. During reconstruction, the small phallus was excised, and the normal-looking phallus was mobilized and swung medially to lie in the midline, overlying the newly fashioned posterior urethra, forming a penoscrotal hypospadias. This is in contrast to the classic repair of cloacal extrophy in

males where an epispadias is created initially after urinary bladder closure. The presence of an adequate well-formed phallus on one side in our patient made it easier for us to convert the urinary repair into a penoscrotal hypospadias. Our patient subsequently had repair of his penoscrotal hypospadias and is now voiding freely per urethra.

The management of newborns with cloacal extrophy has progressed over the years, and now a very reasonable outcome is expected in most cases.^{4,8} This, however, requires a team approach including neonatologists, pediatric surgeons, pediatric urologist, neurosurgeons, geneticists and social workers. Although there are general guidelines in managing newborns with cloacal extrophy, we feel that after thorough evaluation of the anatomical abnormalities, the management should be individualized.

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