

Case Reports

Unusual Cloacal Exstrophy in a Male Neonate with a Normal Penis and Urethra

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Cloacal exstrophy is the most severe of the ventral abdominal wall defects with an incidence of 1 in 4,00,000 births(1). The anomalies that constitute the *sine qua non* for this condition include exstrophy of the bladder, phallic separation with epispadias, separation of the pubic arch anteriorly, an exstrophic bowel field between the 2 bladder halves, with a rudimentary hindgut and imperforate anus(2). We report a very unusual variation of this condition with fusion condition with fusion of the lower pubic region and a normal single midline penis. Extensive search of the world literature could reveal only one case of a similar kind hitherto(3).

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Case Report

A 14-hour-old male newborn, born by normal vaginal delivery to a primigravida mother at 32 weeks of gestation, was referred to our hospital for the management of multiple congenital anomalies. The baby, weighing 1.7 kg, had a small omphalocele, classical cloacal exstrophy (details given below) with an intact lower pubic area and a normal looking penis and imperforate anus (*Fig. 1*). The length and diameter of the stretched penis were 1.5 cm, and 8 mm, respectively. The scrotum was under-developed and testes could not be palpated in scrotum or groins. The lower extremities assumed the 'frog' position because of the external rotation and abduction at hip joints. Bilateral talipes equinovarus were also noted.

Minor sacral vertebral anomaly was noted on radiograph. Sonographic examination of abdomen revealed solitary normal kidney in the right renal fossa. Blood biochemistry revealed serum creatinine of 1.4 mg dl, serum sodium and potassium levels were 142 and 5.2 milliequivalents/L, respectively. In view of the impending rupture of omphalocele, the child was taken up for surgery after initial resuscitation.

The central bowel field of the cloacal exstrophy was represented by ileocaecal region and 3 orifices were seen. (*Fig. 2*). The proximal orifice led to the terminal ileum which was prolapsing for about 5 cm

producing the typical "elephant trunk deformity". The distal orifice led to a 6 cm long blind-ending persistent tailgut. The blood supply to this tailgut was anomalous coming along the mesentery of cloacal exstrophy. A small 1 cm long vermiform appendix was opening into the ileocaecal plate near the cephalic orifice

The two exstrophied hemi-bladders were confluent with each other caudally. A well formed trigone with normal ureteric orifices and normal bladder neck were seen. A No. 3 Fr ureteric catheter could be easily passed down through the bladder neck and out of penile urethra.

According to the Manzoni *et al* classification of cloacal exstrophy and its variants(4), the present case could be designated as classical cloacal exstrophy Type Ic. The coded form is depicted in Fig. 3.

Other abnormalities noted at surgery included ptosis of right kidney, which otherwise was of normal dimensions, hypoplasia of left kidney and bilateral abdominal testes.

The omphalocele was resected and ileocaecal plate was separated from the exstrophied bladder. The ileocaecal plate was tubularised and closed in two layers. The end of the hindgut was brought out as terminal colostomy. Closure of the bladder was done in 2 layers over a 12F Malecot catheter. Intraoperatively, the neonate had one episode of severe bradycardia lasting for 2-3 minutes, the cause of the same could not be ascertained.

The child was electively ventilated post-operatively. His renal parameters were evaluated daily. Serum creatinine was 1.8 mg dl and serum electrolytes were within normal range. The urine output steadily increased from 44 ml in 24 hrs on first post-operative day to 225 ml.

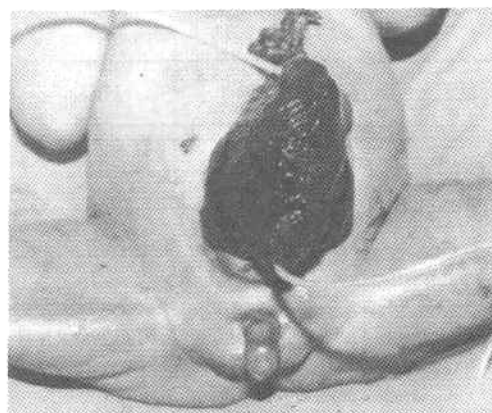


Fig. 1. Classical cloacal exstrophy; the catheters mark the proximal and distal orifices of ileocaecal plate. Note the fusion of the lower pubic region and normal single midline penis.

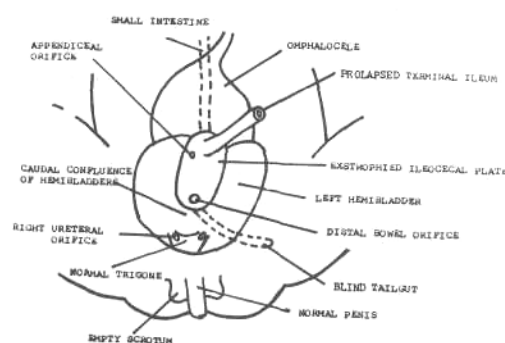


Fig. 2. Line diagram depicting anatomy of the unusual cloacal exstrophy.

On third post operative day, the child had focal convulsions for which meningitis was suspected and crystalline penicillin and amikacin were added to the previous regimen of cefotaxime and metronidazole. The child succumbed to sepsis on 5th postoperative day. The father did not agree for the autopsy.

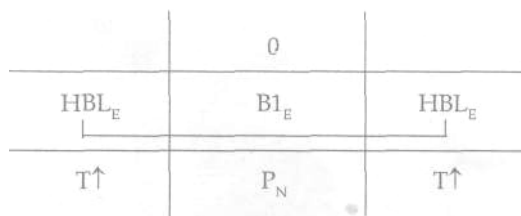


Fig. 3. Coding grid according to Manzoni et al classification (4). Abbreviations used: O - Omphalocele (exomphalos), HBL_E - Hemibladder exstrophy, P_N - Normal penis and T - Undescended testis

Discussion

Although a large proportion of cases of cloacal exstrophy are genotypically males, their phalli are usually duplex and rudimentary. Single midline penis has also been rarely recorded in cases where there is confluence of the hemibladders caudal to the bowel field. But epispadias is the universal rule. Normal single midline penis with normal urethra has been described only once before in 1974(3). Another exception was described by Hall *et al.* Where the neonate had diaphallus, each phallus containing a small central urethra(4). Other rare abnormalities of male external genitalia include 2 cases of hypospadias, both were seen in variant cloacal exstrophy(6,7).

Obviously, the latter cases with normal bladder necks have potential for urinary control and will eventually develop adequate genitalia for sexual gratification. Such cases constitute a very small group of male subjects that do not require gender reassignment in the neonatal age.

The embryological basis of the present case can be explained by Patten and Barry hypothesis of caudal displacement of genital tubercles(8). This permits persistence of more cephalad cloacal membrane with no

mesenchymal migration into this 'unstable' infraumbilical membrane and with a simultaneous incomplete internal urorectal septal division; a disintegration of the cloacal membrane could establish a classical cloacal exstrophy(9). Late and incomplete infiltration of mesoderm would account for variation as seen in our case, where bowel and bladder are exstrophied but confined to the subumbilical region, with normal formation of the urethra and phallus.

Before Rickham's first successful case in 1960(10), there were no survivors despite attempts at surgical reconstruction and cloacal exstrophy was considered to be invariably a fatal anomaly. With technological advances in support devices, such as neonatal intensive care unit monitoring and refinements in hyperalimentation and antibiotic therapy, results have slowly improved, and at present more than 50 survivors of surgical management have been reported from the developed countries(2). In contrast, no elaborate series have been reported from the developing countries such as ours, giving an impression as if we still practice 'no treatment' philosophy. As majority of these patients, if left untreated, would die within a short period due to profuse losses from a short alimentary tract, it would be imperative to attempt reconstruction in every case. No doubt, there would be initial setbacks as the subjects are usually premature and small for dates, but let us not deny these unfortunate children a chance of survival.

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CASE REPORT

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