Cloacal Exstrophy with Intravesical Phallus: An Intra-operative Revelation in a case of OEIS Complex

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Abstract. Cloacal exstrophy is a complex congenital malformation and presents more often than not as a spectrum of anomalies. Gender assignment is difficult in these babies because of the ambiguity of the genitalia and the major surgical undertaking which may make the salvage of certain delicate structures an arduous task. Awareness of the variety of ways in which the phallus may be positioned with respect to the exstrophy patch aids surgeons in further reconstruction. We describe a neonate with an extremely rare variant of an intravesical phallus (eight published cases of this entity were found in literature) and discuss the implications of the same.

Keywords: Cloacal exstrophy, Intravesical phallus, Staged management

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Introduction

The many synonyms of cloacal exstrophy including "vesicointestinal fissure" and the descriptive term, "OIES complex", introduced by Carey, Greenbaum and Hall in 1978[1-2] denote the same anatomic defect. The multitude of variations around the central theme of Omphalocele, Exstrophy, Imperforate anus and Spinal defects makes reconstruction a daunting task. Gender assignment is challenging because of the innate ambiguity of the genitalia which accompanies this most severe form of the exstrophy-epispadias defect. The entity of the intravesical phallus (revealed more often than not during surgical exploration) aids surgeons in taking the correct decision for the child entrusted in their care. We describe a neonate with this extremely rare variant and discuss the implications of the same.

Case Report

A 1.4 kg neonate presented to us on the first day of life with cloacal exstrophy, omphalocele, sacral agenesis and bilateral congenital talipes equinovarus. After initial stabilization, a thorough physical examination revealed an omphalocele with a large exstrophy patch which contained a prolapsed ileal segment (the "elephant trunk" deformity) (Fig. 1) and a caecal patch bounded on both sides by the hemibladders. A single ureteric opening was visible within the edematous mucosa. The genitalia were ambiguous and a possibility of widely separated bifid clitoris and labio/scrotal folds was kept in the absence of palpable testes. Aphallia was evident.

An ultrasound of the abdomen revealed orthotopic location of radiologically normal kidneys. An infantogram showed diastasis pubis and sacral agenesis which correlated with a 5

![Fig. 1. Pre-operative photograph of the cloacal exstrophy with prolapsed ileal segment (the "elephant trunk" deformity) (asterix) and widely separated labio/scrotal folds (arrow head)](image-url)
x 4 cm, soft, globular lump located in the midline just above the shallow natal cleft, possibly a lipoma. The baby was taken up for surgery after routine haematological and blood chemistry panels. The dilated ileum was found to open into the right side of the caecal patch. As in the classical description, two appendiceal openings and approximately 10 cm of distal atretic colon were found towards the left. The hemibladders flanked the intestinal structures and there was no evidence of gross cystitis cystica. Both ureteric orifices could be comfortably cannulated with 5 Fr infant feeding tubes. A 2 cm phallus-like structure was noted to be ensconced within the vesical area just deep to the symphysis pubis and adherent to the caecum by a fibrotic band of tissue (Fig. 2). The left testis was seen in the inguinal canal, no attempt was made to dissect out the right testis.

Our team was unprepared for this surgical surprise of an intravesical penis. As a lack of accurate anatomic information would have made single stage surgical reconstruction potentially dangerous, a decision was taken to separate the urogenital and intestinal tracts and tackle this unusual entity after further growth of the child. The dilated terminal ileocaecal region and the proximal 2 cm of the distal colon were resected and an end to end ileocolic anastomosis was achieved with 5-0 PDS suture. The blind ending colon was pulled through the pelvic floor into the site of the future neoanus and the bowel hung for 2 cm (Fig. 3) after resecting the distal 1 cm. This hanging bowel will be trimmed at a later date after auto-anastomosis of the bowel to the pelvic floor has taken place. Thus a colostomy was avoided and the twin aims of adequate bowel decompression and diverting the faecal stream from the perineal wound were achieved. The bladder patch was reconstructed and sutured to the abdominal wall thus creating a classical bladder exstrophy (Fig. 3).

The child continues to do well in the post-operative period and is thriving.

**Discussion**

This uncommon variant of phallic morphology in cloacal exstrophy is the ninth case of its kind. Apart from this, the reported continuum of phallic abnormalities include, widely separated hemicorpora, epispadias, diphallia, bifid penis and aphallia. We are aware of eight cases in world literature and each description is unique which only adds to the fact that this is a spectrum of malformations. Arunachalam et al described a 46 XY child with an entrapped and buried penis. Although the child developed multiple post-operative complications and died, a biopsy from the tip of the phallic structure was confirmatory.

A review of the prevailing literature was presented in the light of gender assignment by Tomaszewski et al. All the cases described had anatomic similarities in terms of fused bladder plates. This differs from our experience as the bladder plate was cleaved completely by the bowel patch. The penile morphology in the case studies described previously consisted of partially or fully fused corpora or a solitary corporal body covered completely by urothelium. Our baby had fused corpora and the crura could be seen disappearing under the widely separated pubic bones. Though the penis

![Fig. 2. Per-operative findings after prolapsed ileal segment was reduced inside the abdomen showed intravesical phallus (arrow) and bilateral ureteric openings catheterised by no 5 infant feeding tube](image1)

![Fig. 3. Operative photograph after separation of the bowel plate from bladder plate showing intravesical phallus (arrow), reconstructed bladder patch to classical bladder exstrophy (arrow head) and pulled through hanging bowel from neoanus (asterix)](image2)
was hidden within mucosal folds, it was not covered by urothelium and had adequate skin cover.

Though antenatal diagnosis was possible in two cases,[7,8] this entity is usually discovered at the time of bladder closure. The embryology of cloacal exstrophy is the subject of much controversy and even the origin of the intra-exstrophic phallus is disputed. Lakshmanan et al[7] theorized that the abnormal anterior abdominal wall mechanically approximates the exstrophic bladder tissue and thus traps the phallus within it. In the present case, the band connecting the base of the phallus to the caecal patch may have had a role in halting the natural evolution of the penile tissue and the fusion of the bladder plates. The paucity of reported cases could be due to a case of mistaken identity. The urothelial covering coupled with the mucosal edema in a clinically diagnosed aphallic patient can lead the surgeon to excise it as a bladder hamartoma. Pathological examination in such cases has shown a hamartoma of squamous mucosa surrounding erectile tissue.[8]

The true incidence of cloacal exstrophy is unknown but it does account for 10% of the exstrophic defects.[9] An improvement in overall survival and continence rates in the modern era makes it imperative that we strive to give these babies a wholesome and productive life. A greater degree of knowledge about such variants could influence reconstruction and the choice of gender in children presumed to be aphallic. Genetic males need not be assigned a female sex which may go against testosterone imprinting and consign them to a life beset by psychosexual issues along with the other long-term complications of this anomaly.

References