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Research Article

CASE REPORT: A RARE CLOACAL EXSTROPHY VARIANT – DESCRIPTION OF ANATOMY AND ITS MANAGEMENT

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ABSTRACT

Cloacal exstrophy is an extreme and rare form of the exstrophy epispadias complex and may have varied presentations. The treatment of various entities of cloacal exstrophy may vary markedly. Cloacal exstrophy canbe diagnosed antenatally. Management of cloacal exstrophy requires multi disciplinary team approach at tertiary centers.These children require long term supportive care nd parental counselling.

INTRODUCTION

Cloacal exstrophy is the most extreme and rare form of the exstrophy epispadias complex with an incidence of one in every 300,000 live births [1]. Its variant is a seldom seen entity and is 10 times more infrequent (2). Although cases of classic epispadias and bladder or cloacal exstrophy are usually easily recognized, variant presentations of the complex can be confusing and difficult to categorize due to the scarcity of the malformation. No definite genetic defect or environmental factor has been identified to suggest occurrence of such malformation (3,4). Most often, affected pregnancies are terminated, while those that continue there is risk of infant mortality shortly after delivery (5,6).

Over the many years, the focus has drastically shifted from patient survival to improving patient outcomes and better quality of life despite its complexity with particular focus on gender assignment, higher level of physical and social independence and mobility. The priority is given to urinary, gastrointestinal, and genital reconstruction. (7)

Case

2.9 kg full term male child presented to us with defect in lower abdominal wall since birth and dribbling of urine. On examination, there was exposed complete bladder plate, ureteric orifice was identified and cannulated, bilateral hemi-scrotum with bilateral testis present. A fistulous opening was seen at inferior aspect of bladder plate. On cannulation meconium staining was present. Phallus was seen on right side with med penile hypospadiac urethra. There was glanular tissue present on left side.

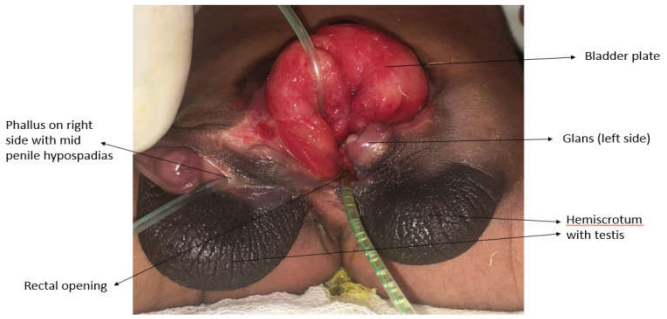

Figure clinical photograph

X-ray pelvis showed wide pubic diastasis. Per rectal dye study was suggestive of normal bowel pattern.

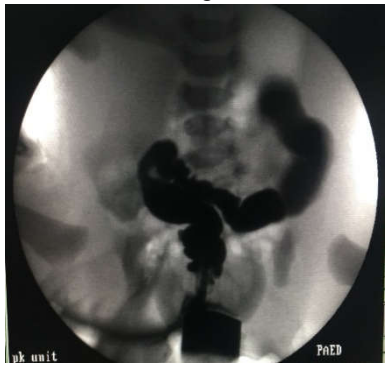

Figure per rectal dye study

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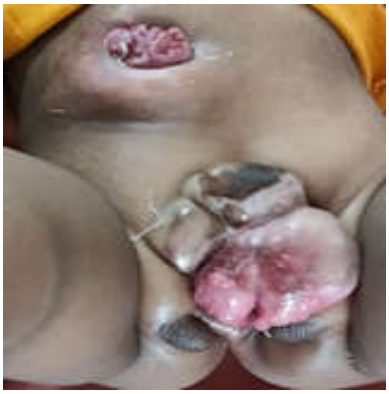
Patient underwent loop transverse colostomy on 2nd day of life.



MRI abdomen and pelvis demonstrated cloacal exstrophy with omphalocele containing bladder plate. Infra-umbilical abdominal wall defect of size 2.7x5.3 cm. Pubic diastasis of approximately 8 cm. A linear muscular structure seen extending from sigmoid colon reaching caudal aspect of infraumbilical defect - ?distal colonic/rectal opening. Levator ani muscle was thinner on right side whereas external anal complex was thinner on left side with partial sacral agenesis.

At 2 years of age bladder exstrophy repair with standard bowel pull through procedure was done. Osteotomy was not done as there was wide gap of approximately 8cm between symphysis pubis.

DISCUSSION

The argument regarding classification of variant presentations is the fact that treatment of the various entities varies dramatically. Therefore, any erroneous initial diagnosis can have deleterious impact on the long-term outcome. (8)

In 1987 Manzoni et al - devised a classification system for cloacal exstrophy (2 types). Type I - classic cloacal exstrophy which was further subclassifed according to positioning of the hemibladders relative to the everted bowel. Type II - cloacal exstrophy variant, and further subclassified on basis of bladder variations, bowel variations and mixed bladder-bowel variants. (9)

Cloacal exstrophy can often be diagnosed antenatally on ultrasound. The ultrasound like feature identified are non-visualization of the bladder, a large infraumbilical midline anterior wall defect or a cystic anterior wall structure and lumbosacral anomalies. (10).

The classic cloacal exstrohy is described as two exstrophied bladder halves and a strip of the exstrophied cecum in between and is generally accompanied by prolapsed ileal segment. Male genitalia is usually found to be an epispadiac penis on widely separated pubic bones. Whereas in female, the Mullerian duct orifices may be exstrophied below the bladder mucosa and duplicate vagina, bifid clitoris. Zderic and colleagues proposed management plan for cloacal exstrophy neonates involves surgery after stabilizing infant preferably within first 48 to 72 hours of life. In early correction the risk of bacterial colonization is decreased in exposed viscera and it might lessen need for pelvic osteotomy. Multistage procedure is commonly acknowledged as producing the best long-term outcomes. The goal of surgery to secure abdominal wall along with bladder closure, reconstruction of genitalia both functionally and cosmetically acceptable, preservation of renal function along with urinary and fecal continence (11).

Initial years are focused on growth and development of these children. The urinary tract should drain freely to prevent recurrent urinary tract infections. Epispadias and bladder neck repair is done between 3 to 5 years depending upon bladder capacity. (12)

CONCLUSION

Cloacal exstrophy variant is rare and challenging diagnosis affecting multiple systems and thus requiring multi disciplinary team approach that can be managed in tertiary centers. The different aspects of optimal treatment of cloacal exstrophy must address timing and type of repair to genital reconstruction and quality-of-life issues.

With advancement in medical and surgical management, hyperalimentation, survival and continence rates have drastically improved however, these children require lifelong supportive care.

Parents counselling and support is an important task as patient require multiple admissions, surgeries and prolonged follow up.

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