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Cloacal Exstrophy: A Rare Case Report

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Abstract

Cloacal exstrophy is an extremely rare congenital malformation resulting in an exstrophy of the urinary, intestinal, and genital organs and associated with anomalies of other organ systems. Cloacal exstrophy occurs in approximately 1 out of 200,000 pregnancies and 1 in 400,000 live births. The underlying cause is not known. The physical characteristics are the result of a developmental abnormality during embryonic growth in which the cloacal membrane is not replaced by tissue that will form the abdominal muscles. Our patient a term male neonate, delivered by lscs, in bokaro general hospital, bokaro, jharkhand, presented at birth with cloacal exstrophy and respiratory distress. The defect was covered with sterile silicon gauzes and waterproof dressing and patient was shifted to the scbu (special care new born unit) setup. Multidisciplinary team of surgeons (pediatric general surgeons, pediatric urologists, pediatric orthopedists) will create a treatment plan based on the type and the extent of child's condition.

Keywords: cloacal exstrophy, epispadius, bladder exstrophy, rare congenital anomaly.

Introduction

Cloacal exstrophy is a severe birth defect wherein much of the abdominal organs (the bladder and intestines) are exposed. It often causes the splitting of the bladder, genitalia, and the anus. It is sometimes called OEIS complex. Omphalocelecloacal exstrophy-imperforate anus-spinal defect syndrome. Diagnostic tests can include ultrasound, voiding cystourethrogram (VCUG), intravenous **Pyelogram** (IVP), nuclear renogram, computerized axial tomography (CAT Scan), and magnetic resonance imaging (MRI). Cloacal exstrophy is a rare birth defect, present in 1/200,000 pregnancies and 1/400,000 live births. It is caused by a defect of the ventral body wall mesodermal migration is inhibited and folding fails.





Conclusion

Improvements in perinatal management have increased survival in cloacal exstrophy. The focus of reconstruction has shifted to reduction in the numbers of incontinent stomas, assistance with ambulation and improved cosmesis. All of these goals are achievable using a multidisciplinary approach to the management of this complex anomaly.