Case Report

Cloacal Exstrophy: a rare and complex anomaly.

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Abstract:

Cloacal exstrophy is a very rare and complicated birth defect. It occurs once in every 200,000 births. It is a complex anomaly of the urogenital tract and intestinal tract resulting in exstrophy of both bowel and bladder; the most severe form of anomaly is in the exstrophy–epispadias complex. Its features include omphalocele, imperforate anus and exstrophy of two hemibladders, between which lies the everted cecum. A small colon ends blindly in the pelvis, and the terminal ileum often prolapses out of the exposed cecum.

We are reporting a case of cloacal exstrophy with lumbosacral meningomyelocele and deformity of left foot which was managed successfully.

Key Words: Cloacal exstrophy, meningomyelocele, tailgutostomy.

Introduction:

Cloacal exstrophy represents one of the most severe congenital anomalies compatible with intrauterine viability. It is exceedingly rare, occurring 1 in 200,000 to 400,000 live births (Hurwitz et al, 1987). Most recent reports indicate a male-female sex ratio of 2:1 (Gearhart & Jeffs, 1998). Inheritance of cloacal exstrophy is unknown, because offspring have never been produced by patients with this disorder.

Cloacal exstrophy is often referred to by different labels including: vesico intestinal fissure, exstrophy of the cloaca, exstrophy of the spalancia, and OEIS Complex, (O-Omphalocele, E-Exstrophy of the cloaca, I-Imperforate Anus and S-Spinal Defects).

Pathogenesis:

It occurs due to failure of two concomitant mesodermal migrations. First, the urorectal septum fails to develop and divide the urogenital sinus from the rectum; second, the mesodermal proliferation forming the infraumbilical abdominal wall and genital tubercle fails to develop. The failure of these two events to occur results in exstrophy of both bladder and intestine.

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Associated anomalies:

Cloacal exstrophy is commonly associated with other anomalies including cardiovascular, central nervous system, omphalocele (70-90%), vertebral anomalies (46%), upper urinary tract (42%), malrotation (30%), lower extremity anomalies (30%), double appendix (30%), absent appendix (21%), short small bowel (19%), small bowel atresia (5%) and abdominal musculature deficiency (1%; Spencer, 1965). Upper urinary tract anomalies include pelvic kidney, horseshoe kidney, hypoplastic kidney and solitary kidney (Jeffs, 1987). Vertebral malformations include sacralization of L5, congenital scoliosis, sacral agenesis, and interpedicular widening.

Case Report:

A 2.2 kg male baby was born by normal vaginal delivery after full term gestation to a 23 year unbooked primigravida. There was no significant antenatal history of any infection, drug intake or radiation exposure to mother. The marriage was non consanguineous. The baby cried immediately after birth. There was no cyanosis, tachypnoea or added cardiac sounds. Baby was hypothermic and examination showed presence of omphalos below umbilicus, protruding and exposed bowel and bladder plates in lower part of abdomen. Prolapsed terminal ileum through the open cecal plate in midline appeared like trunk of elephant. There was a prominent buldge...
visible on the lower abdomen on crying. The hind gut opening was seen on the lower part of the open bowel while the anal opening was absent. There was diastasis of the pubic bones with each half of phallus attached to the corresponding pubic bones and open bladder mucosa was seen in between. Both testis were undescended with wide open internal inguinal rings. There was a big lumbosacral meningomyelocoele with paraparesis and deformed left foot.

The baby was resuscitated and investigated for other associated anomalies. After counseling sessions with parents new born was taken for surgery. The hemi bladder plates were mobilized and sutured together to form the bladder after putting catheters in both the ureters. The central cecal plate was tubularised in continuity with the ileum and the shortened hind gut and a “tailgutostomy” was done. The pubic bone on both sides were mobilized and after tubularization of the urethra they were approximated together by heavy prolene suture and rectus sheath closure was done simultaneously to close the exomphalos (Fig. II.).

Postoperatively the baby was kept on elective ventilation for 48 hrs and later weaned off. Recovery was uneventful (Fig. III).

In the second stage, the big meningocele was operated and sheath and muscle cover provided. Baby is on regular follow up with us.

Discussion:

Prenatal diagnosis of cloacal exstrophy:

In 1998, Austin et al, reviewed 20 patients with this abnormality and proposed major and minor criteria for the prenatal diagnosis of cloacal exstrophy, based on the frequency of occurrence rather than the severity. A criterion was considered major if it was present in more than 50% of cases. The gestational age for diagnosis of cloacal exstrophy ranged between 15 and 32 weeks. Major diagnostic criteria included nonvisualization of the bladder in 91%, a large midline infraumbilical anterior wall defect or a cystic anterior wall structure in 82%, an omphalocele in 77%, and a myelomeningocele in 68%. Minor criteria included lower-extremity defects in 23%, renal anomalies in 23%, ascites in 41%, widened pubic arches in 18%, narrow thorax in 9%, hydrocephalus in 9%, and a single umbilical artery in 9%.

Embryology and Anatomy:

Typically, after 4 weeks of life, the urorectal septum divides the cloaca into an anterior urogenital sinus and a posteroanorectal canal. Simultaneously,
the cloacal membrane is invaded by lateral mesodermal folds at approximately 4 weeks of gestation. It is postulated that if a mesodermal invasion does not occur, the infraumbilical cloacal membrane persists, with subsequently poor abdominal wall development. Because of its inherent instability, the cloacal membrane eventually ruptures. If it does so before the urorectal septum descends at 6 to 8 weeks of gestation, then cloacal exstrophy results. However, in the developing embryo, a stage similar in appearance to cloacal exstrophy does not exist. Therefore, the anomaly must not represent an arrest in development, but more likely some form of embryogenetic defect. According to Muecke (1964), an abnormally extensive cloacal membrane produces a wedge effect, serving as a mechanical barrier to mesodermal migration, which results in impaired development of the abdominal wall, failure of fusion of the paired genital tubercles, and diastasis of pubis. Exstrophy of the cloaca results when the wedge effect occurs before the formation of a urorectal septum at 6 weeks.

Management:

Immediate management is directed to the medical stabilization of the infant. Evaluation and appropriate management of associated malformations should be undertaken. For infants who have few other associated malformations and are medically stable, staged closure can be considered. The bowel should be moistened with saline and covered with protective plastic dressing. Evaluation of the genitalia and gender assignment should be made by a gender assignment team, including a pediatric urologist, pediatric surgeon, pediatrician, and pediatric endocrinologist. Consultation of social worker, pediatric orthopedic surgeon and other disciplines should be obtained. In a large medical center with experience in dealing with these patients, these multiple consultations should be done in a short period of time.

The initial operation consisted of separating the bowel from the bladder to create an intestinal stoma; closing the omphalocele; and reapproximating, closing, or leaving the extrophied bladder undisturbed. The importance of creating a “tailgutostomy” instead of an ileostomy to prevent problems with diarrhea, dehydration, and acidosis is emphasized.