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Cloacal exstrophy: A single center experience

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Abstract *Objective:* Cloacal exstrophy is an exceedingly rare and complex anomaly. The records of 23 patients treated in a tertiary care center with limited infrastructure were analyzed for anatomic types, associated anomalies, surgical procedures adopted, and the outcome.
Materials and methods: There were 14 males. Seventeen babies were preterm with an average weight of 1.92 kg. The time of presentation, gestational age, birth weight, position of the hemibladders and associated malformations were noted. Reconstruction procedures involved dismantling of the hemibladders and primary turn in, tubularization of the bowel with an end colostomy, and reconstruction of the abdominal wall. Results of the primary surgical repair, bowel function, and outcome of secondary procedures were analyzed.
Results: The position of hemibladders was lateral in 11, upper confluent in 4 and lower confluent in 8. Associated anomalies were noted in 19 patients. Four patients presented late (>5 days). Five died preoperatively, all had major associated anomalies. Four of them were preterm with average weight of 1.4 kg. Two patients refused surgery. Single-stage surgical reconstruction was done in 15 patients. Five patients died postoperatively because of associated anomalies, prematurity, and sepsis. One patient is waiting for surgery. Six patients had follow-up at 3—42 months and are awaiting further reconstruction. Four patients were lost to follow-up.
Conclusions: Prematurity, late presentation, and sepsis are the major causes of high mortality noted in this series. In our experience, single-stage reconstruction without osteotomy gives satisfactory results.

Introduction

Cloacal exstrophy, which is also called vesico-intestinal fissure, is a rare and complex congenital malformation, with a reported incidence between 1 in 200,000 and 1 in 400,000 live births [1,2]. Classically, cloacal exstrophy consists of an omphalocele, exstrophy of the two hemibladders, lateral cecal fissure which presents between the

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two hemibladders, and imperforate anus; many other variants have also been described [3—5]. This article describes management of cloacal exstrophy patients in a remote tertiary care center in a developing country, where patients often present late because of poor socio-economic conditions and geographical isolation. Patient demographies, associated malformations, treatment outcome, and relevant literature are discussed. Risk factors and difficulties encountered during management of this patient population are also discussed.

Material and methods

Twenty-three patients with cloacal exstrophy presented to us between January 2001 and April 2012, out of which 14 were males (Table 1). The time of presentation, gestational age, birth weight, position of the hemibladders, and associated malformations were noted. Reconstruction procedures involved dismantling and closure of the two hemibladders, tubularization of the bowel patch, fashioning of an end colostomy, excision of the omphalocele, and reconstruction of the anterior abdominal wall. The recti muscles were detached with the periosteum from the pubic bones and brought to the midline. Results of the primary surgical repair, bowel function, and outcome of secondary procedures are analyzed.

Results

Four patients presented more than 5 days after birth. Seventeen were preterm, and the average weight was 1.92 kg. The hemibladders were laterally placed in 11, upper confluent in four, and lower confluent in eight patients (Figs. 1 and 2). All had an omphalocele. Meningomyelocele (MMC) was noted in 6 patients, lipo-MMC in 4 (Fig. 3), spina bifida occulta (SBO) in 3, congenital heart disease (CHD) in 5, bilateral talipes deformity (CTEV) in 6, hydronephrosis and Meckel's diverticulum in 2 and high arched palate and solitary kidney in 1 patient each. Associated anomalies were present in 19 patients (Table 2). Five patients died preoperatively. Four of them were preterm with an average weight of 1.4 kg. All had major associated anomalies (Table 3).

Fifteen patients underwent single-stage reconstruction (Figs. 4 and 5). Of the nine male patients, gender reassignment was done in two because of small phallic size. One patient had a large omphalocele, so the sac was not excised at the time of primary repair to avoid a tight abdominal wall closure (Fig. 6). Patients who had a tight closure were electively ventilated for the initial 3—5 days. The lower limbs were kept in the adducted position with a plaster cast or adhesive tap for 2 weeks. Five patients died postoperatively: three of them were preterm with average weight of 1.5 kg; two had lipo-MMC; and three had major renal anomalies. One patient had dehiscence of the bladder repair. Of the survivors, MMC repair was done in one patient and detethering of the cord was done in two patients. Corrective surgery for CHD was done in two patients at another center. Epispadias closure was done in one patient. Four patients were lost to follow-up. Six patients are doing well, with follow of 3—42 months: three of them are forming solid stools and are awaiting further reconstructive procedures. Five patients had a minimum of 1-year follow up.

Discussion

Cloacal exstrophy is a rare congenital malformation and most reported series are from large referral centers and over a long period of time [1,2]. Lund and Hendren [4] reported 20 cases of cloacal exstrophy seen over an 18-year period. Ricketts et al. [6] treated 12 newborns with cloacal exstrophy over a period of 10 years. Stolar et al. [7] treated 14 patients with cloacal exstrophy over a 23-year period, and Cywes [8] treated 24 infants with cloacal exstrophy over 30-year period. In the present series, 13 out of 23 patients were from a single community, in whom other rare congenital malformations have also been reported [9,10]. During this period, 26 patients with classic bladder exstrophy were treated at our center.

The exact embryogenesis of cloacal exstrophy is not known, and many theories have been suggested. The most accepted theory is that cloacal exstrophy results from premature rupture of the cloacal membrane prior to caudal migration of the urorectal septum, and fusion of the genital tubercles [11]. The pubic bones are widely separated, and spinal dysraphism is common in these patients [3—5]. Associated anomalies occur in 95—100% of cases in some series [12]. Vertebral anomalies accompanying cloacal exstrophy range from 48% to 75% of patients in various reports [12,13]. Mathews et al. [14] reported spinal abnormalities in 34 of their 37 cases of cloacal exstrophy; lipo-MMC was noted in 17 cases. Twenty-two cases had vertebral and 18 had renal abnormalities [14]. Greene et al. [15] found spina bifida in all the 13 cases of cloacal exstrophy in their report. In our series, only 12 patients (52.2%) had spinal abnormality. This figure would be much higher, for we could not evaluate all the patients for associated anomalies. One reason for higher incidence of associated malformations in recent times may be due to improved survival of these babies, enabling thorough evaluation in later infancy. Other associated malformations, such as congenital hip dislocations, talipes equinovarus, and other foot and leg anomalies have been reported in 9—30% of patients in various reports [12—17].

Evaluation of the genitalia and gender assignment should be made by the consultant pediatric urologist who will be involved in reconstruction. In male infants with cloacal exstrophy, the phallus is usually divided. The hemicorpora may be unequal in size. A midline phallus is rarely observed. Recent understanding of the psychology of children who have had gender reassignment has fueled interest in male gender assignment if adequate unilateral or bilateral corporeal tissue is present [14]. Earlier studies advocated female gender assignment for male neonates with minimal phallic structures, but recent studies have shown that testosterone imprinting occurs much earlier in utero and any postnatal gender reassignment would have an adverse impact on the child's developing psychology [1,6,18]. A trained pediatric psychiatrist/psychologist familiar with treating patients with gender ambiguity should be an integral part of the management team to help the parents and later the developing child deal with