Journal of Pediatric Surgery (2013) **48**, 2043–2046

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Journal of Pediatric Surgery

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Fate of the uterus in 46XX cloacal exstrophy patients

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Received 10 November 2012; revised 2 February 2013; accepted 5 February 2013

Key words:
Cloacal exstrophy;
Didelphic uterus;
Hysterectomy;
Hemihysterectomy;
Pregnancy;
Hematometrocolpos

Abstract

Purpose: Genetically female cloacal exstrophy (46XX CE) patients develop complications later in life due to their abnormal uterine anatomy, resulting in various invasive gynecologic procedures. Furthermore, they have difficulty becoming pregnant, and if they do conceive, they are unlikely to carry the pregnancy to term. We performed this review to determine the rate of gynecological complications, the fate of the uterus, and the rate of pregnancy in 46XX cloacal exstrophy patients.

Methods: All charts for 46XX CE patients treated by us were reviewed following IRB approval. Patient age at last follow-up, surgical management of the uterus, uterine complications, and pregnancies were recorded.

Results: The charts of all 16 of our 46XX CE patients who survived past the neonatal period were reviewed. Two patients underwent hemi-hysterectomy (HH): 1 for an atretic hemi-uterus at birth, another for abnormal uterine insertion at 3 years. A third patient initially had HH for hydrometrocolpos leading to ureteral and colonic obstruction at 14 years but she required a completion hysterectomy a year later. Four patients underwent total hysterectomy (TH) at the outset: 2 neonates for a diminutive uterus with extreme disparity in the halves, another for uterine prolapse at 1 month of age, and a fourth for hematometrocolpos at 16 years of age. Six patients reached adulthood without requiring gynecologic intervention; one of these six is now being managed at another institution. Two patients are prepubertal and one was lost to follow-up. The only patient in the series who became pregnant miscarried at 11 weeks gestation.

Conclusion: Out of 13 post-pubertal patients 6 have retained the entire uterus and another 2 had a hemi-hysterectomy. One patient who became pregnant miscarried at 11 weeks gestation. We believe it is appropriate to avoid ablative genital surgery as far as possible and for these patients to become pregnant after detailed discussion with physicians experienced in the care of 46XX CE patients.

Cloacal exstrophy (CE) is a devastating condition that appears to be the result of a very early developmental defect of the caudal eminence. These patients typically present with

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0022-3468/$ – see front matter © 2013 Elsevier Inc. All rights reserved.
http://dx.doi.org/10.1016/j.jpedsurg.2013.02.040

an omphalocele, imperforate anus with a shortened hind gut, and an open cecal plate flanked by open hemibladders on either side. In addition many patients also have deformities of the spinal cord, kidneys and ureters. The most reliable data regarding incidence of CE were collected by Feldcamp et al. [1] who collated information from 18 surveillance programs in 24 countries. The incidence varied from 2·25/100,000 births in Wales to 0·37/100,000 births in South America. The overall incidence (live and still births) was 0·76/100,000 while for live births it was 0·54/100,000. The sex of 47 of 186 patients in the cohort was not known. Of the 75% in whom it was known the male:female distribution was 1:1·14.

Cloacal exstrophy patients invariably require numerous operations and lengthy hospitalizations and they may suffer severe morbidity and poor quality of life [2]. Genetically female CE patients (46XX CE), have normal ovaries but their Müllerian ducts do not fuse, resulting in uterine and vaginal duplication [3–5]. Surgeons have performed various operations ranging from ablation of the septum to removal of 1 of the 2 halves of the uterus (hemi-hysterectomy or HH) to total hysterectomy (TH). If the uterus is left *in situ*, complications such as uterine prolapse and hydro- or hematometrocolpos have been reported. Studies on long-term outcomes in CE patients typically evaluate quality of life pertaining to urinary and fecal continence and gender assignment and reconstruction. There is a paucity of literature regarding management and outcomes of the internal genitalia in genetic females.

We have tried to determine the fate of the uteri, incidence and type of complications, and rate and outcome of any pregnancies in 46XX CE patients.

1. Methods

After approval from the Internal Review Board (IRB# 2010-14334) a retrospective chart review was performed on all 46XX CE patients treated by the authors (JR, EC, EY, MR) from July 1978 to December 2009. Patient data including anatomy of the internal genitalia, surgical management of the uteri, and follow-up status were collected.

2. Results

The charts for 17 46XX CE patients were identified. One neonate with anencephaly died in the neonatal period and was excluded from the study. The remaining 16 patients had a median follow-up age of 19 years with follow-up ranging from 3 to 34 years. We were unable to determine if CE was identified in any patients antenatally. All these babies were assigned a female sex prior to referral to us. Each of these patients had didelphic uteri as determined by imaging studies or at operation (Table 1).

Fig. 1 depicts the timing and indications for operative interventions in all seventeen 46XX CE patients. One neonate underwent TH at birth for diminutive hemiuteri with great disparity in size between the halves, while in another only the atretic half was removed (HH). Three patients required surgical intervention in the prepubertal period. Two had TH, in one patient with a high meningo-myelocele for uterine procidentia and in another for abnormal insertion of the uteri. In the third patient, 1 side of the didelphic uterus inserted into the bladder and it was removed (HH). Two of our patients are prepubertal and they

Table 1 Uterine operations and follow-up for patients with cloacal exstrophy.

N	Uterine operations	Age at last follow-up (years)	Follow-up information
1	None	0	Anencephalic, anatomy not defined
2	TH for uterine prolapse at 3 mo	1	High myelomeningocele no function below umbilicus
3	None	3	Prepubertal
4	None	3	Prepubertal
5	None	7	In another state, now in her 20s
6	TH for small disparate uterus at birth	14	
7	None	17	
8	None	18	
9	None	19	Miscarriage after 11 week pregnancy
10	HH for abnormal uterine insertion at 3 yo	20	
11	TH for hematometrocolpos at 16 yo	20	
12	None	21	
13	None	23	At another center
14	HH for hydrocolpos with ureteral and colonic obstruction at 14 yo CH for TOA and salpingitis at 15 yo	23	
15	HH for atretic hemi-uterus at birth	24	Sexually active
16	None	29	Married in 2011, adopted baby in 2012
17	TH for abnormal uterine insertion at 3 yo [6]	34	

HH = hemi-hysterectomy; TH = total hysterectomy; CH = completion hysterectomy.