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ORIGINAL ARTICLE

Clinical experience with persistent cloaca

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Purpose: Persistent cloaca is one of the most severe types of anorectal malformation. Appropriate initial drainage is difficult due to their various malformations and hydrocolpos or dilated urinary bladder. Corrective surgery also differs among individual patients. We describe our experiences with the surgical management of children with persistent cloaca. **Methods:** We retrospectively reviewed 16 children diagnosed with persistent cloaca at Asan Medical Center. **Results:** Sixteen patients were managed in their neonatal period. Twelve patients had enlarged bladder or vagina at birth. Three patients, who did not undergo cystostomy or vaginostomy at first operation, had earlier complications after surgery or required drainage tube insertion. One patient who did not undergo hydrocolpos drainage died of sepsis and complications. Nine patients underwent corrective surgery; posterior sagittal anorectovaginourethroplasty using the Pena method. Three patients required additional operations due to complications after surgery. **Conclusion:** Patients found to have anatomical malformations before colostomy, as well as hydrocolpos and bladder enlargement, require a vaginostomy with or without a cystostomy to reduce complications. Follow-up is required in patients with hydrocolpos and bladder enlargement to determine whether vaginal drainage improves dilated bladder. Continuous long-term follow-up examination is required to determine the long-term results of corrective surgery.

Key Words: Persistent cloaca, Anorectal malformation, Hydrocolpos, Posterior sagittal anorectovaginourethroplasty

INTRODUCTION

Persistent cloaca is one of the most severe types of anorectal malformations occurring in newborn infants. Appropriate drainage is difficult because the degree of malformation differs among individuals and difficulty to determine the exact anatomical structure in each patient due to hydrocolpos or dilated urinary bladder and colon. Later reconstruction is also dependent on individual characteristics and type of the surgical procedure performed. We describe here our experiences with surgical management of the children with persistent cloaca.

METHODS

Children diagnosed with persistent cloaca at the Asan Medical Center from 1998 to 2010 were retrospectively identified, and their clinical histories and radiological and surgical findings were analyzed.

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RESULTS

Clinical features

Sixteen patients were managed in their neonatal period (Table 1). Eleven infants prenatally diagnosed as following abnormalities, 3 were persistent cloaca, 8 were other disease (Tables 2, 3). Associate anomalies were combined in 3 patients, 1 patient combined the vertebral, anorectal, cardiac, esophageal atresia with tracheoesophageal fistula, renal and radial and limb anomalies (VACTER) syndrome. Chromosomal spreads were performed in 9 patients, with 1 having a 47.XX + marker chromosome. The father of this patient had the same chromosomal abnormality, as well as a surgical history of imperforate anus.

Table 1. Initial findings of hydrocolpos and dilated bladder at their neonatal period after birth in all sixteen patients

	Hydrocolpos	Dilated bladder	No. of patients
	+	+	4
	+	−	3
	−	+	5
	−	−	4
Total	7	9	16

Management in neonatal period

In 4 patients, hydrocolpos and dilated bladder were detected at the time of birth. In 3 of these patients, cystostomy and vaginostomy were performed immediately after birth, they had no further problems other than urinary tract infections. One patient (no.2) with a foley catheter during 2 months after birth underwent cystostomy and vaginostomy due to persistent extension of the vagina and bladder.

Three patients had hydrocolpos only without any bladder enlargement. One patient (No.5) who did not undergo vaginostomy initially underwent a cystostomy and vaginostomy at 30th day after birth, but died of ventriculitis and hydrocephalus 6 months later due to sepsis caused by urinary tract infection. One patient (No.6) who underwent a vaginostomy had no further problems for 4 months. Another patient (No.7) was underwent a colostomy at another hospital did not require further treatment.

Dilated bladder without hydrocolpos was detected in 5 patients. In one patient (No.12) of these, a cystostomy was performed 18 days after birth, because urine was drained via a colostomy. Intermittent urinary drainage was performed through a colostomy in 1 patient (No.8) and internal urethrotomy was performed in another 1 patient

Table 2. Clinical characteristics, anatomical variations and combined abnormalities of all sixteen patients

Case	Prenatal diagnosis	Hydro-colpos	Dilated bladder	Uterus	Vagina	Combined anomaly
1	Hydronephrosis[a)]	+	+	Hemiuterus	Normal	
2	Cloaca	+	+	Hemiuterus	Hemivagina	
3	Cloaca	+	+	Normal	Normal	TAPVR, 47XX
4	Cloaca[a)]	+	+	Normal	Normal	EA with TEF
5	Cyst, hydronephrosis	+		Hemiuterus	Hemivagina	
6	Meconium peritonitis	+		Hemiuterus	Hemivagina	
7		+		Hemiuterus	Hemivagina	
8			+	Hemiuterus	Normal	
9	Mass		+	Hemiuterus	Hemivagina	
10	Horseshoe kidney		+	Hemiuterus	Hemivagina	
11			+	Hemiuterus	Normal	
12	Mass		+	Hemiuterus	Hemivagina	
13				Normal	Normal	
14	Mass			Normal	Blind pouch	
15				Hemiuterus	Vaginal septum	
16	Duodenal atresia			Mullerian agenesis		VACTER[b)]

TAPVR, total anomalous pulmonary venous return; EA with TEF, esophageal atresia with tracheoesophageal fistula.
[a)]Case performed shunt procedure. [b)]Combined with esophageal atresia, tracheoesophageal fistula, duodenal atresia, tetralogy of Fallot, pulmonary artery atresia, malrotation, club hand.

Table 3. Performed surgical procedures including initial and later operations, their clinical course and follow-up survey

Case	Initial procedure	Later procedure and clinical course	Follow-up
1	Colostomy, cystostomy vaginostomy		Loss after 2 mo
2	Colostomy foley catheter (+)	Cystostomy, vaginostomy (right) at 60 day vaginostomy (left) at 75 day	32 mo[a]
3	Colostomy, cystostomy vaginostomy		Expire due to TAPVR at 16 mo
4[b]	Colostomy, cystostomy vaginostomy	Waiting for corrective surgery	4 mo
5	Colostomy[c] vagina-only aspiration	Urological sepsis ventriculitis hydrocephalus cystostomy, vaginostomy (both) at 40 day	Expire due to sepsis at 6 mo
6	Colostomy vaginostomy		Loss after 4 mo
7	Colostomy		8 yr[a]
8	Colostomy	Intermittent urinary aspiration	6 yr[a]
9	Colostomy foley catheter (+)	Foley catheter removal at 3 day	Loss after 1 mo
10	Colostomy	Internal urethrotomy at 20 day	3 yr 10 mo[a]
11	Colostomy		21 mo[a]
12	Colostomy	Cystostomy at 18 day	Loss after 4 mo
13	Colostomy		11 yr[a]
14	Colostomy		11 yr[a]
15	Colostomy		10 yr[a]
16[d]	Colostomy	Internal urethrotomy at 8 mo	3 yr[a]

TAPVR, total anomalous pulmonary venous return.
[a]Cases performed corrective surgery and their follow-up periods. [b]Case performed tracheoesophageal fistula ligation and esophagoesophagostomy. [c]Misdiagnosed case as "high type imperforate anus" at initial colostomy, thereafter diagnosed as cloaca at postoperative 16 days. [d] Duodenoduodenostomy, tracheoesophageal fistula ligation, gastrostomy.

(No.10) with stenosis of the bladder neck. The remaining 2 patients did not require any other procedures.

The remaining 4 patients did not show enlargement of the vagina and bladder. Three patients of these did not require any urinary or vaginal catheter insertion until corrective surgery. The fourth patient (No.16) underwent an internal urethrotomy at 8th month after birth because of hydronephrosis caused by repeated urinary tract infection and bladder neck stenosis.

Corrective surgery and follow-up

Among total 16 patients, 9 patients underwent corrective surgery and were followed-up for a median 72 months (range, 21 months to 11 years) (Tables 4, 5). Two patientswho combined with cardiac anomaly and hydrocephalus, died without correction and 1 patient is currently awaiting corrective surgery. Remaining 4 patients were lost to follow-up.

Among 9 patients performed corrective surgery, 7 patients underwent Pena procedure as posterior sagittal anorectovaginourethroplasty (PSARVUR), and 3 patients of these required an additional abdominal approach for hysterectomy and colostomy dissection. Remaining 2 patients underwent PSARUR due to Mullerian agenesis.

The median length of the common channels was 3.2 cm (range, 2 to 4.5 cm). The vaginal switch method was performed in 1 patient with a hemiuterus and hemivagina. The uterus and vagina of 1 patient had to be removed because of high located vagina and damage during dissection. Four patients underwent 3 procedures respectively, consisting of 1) a colostomy with a cystostomy or vaginostomy, 2) corrective surgery, and 3) colostomy restoration. Four patients required additional procedures after corrective surgery due to complications such as vesicovaginal fistula, urethral stone or urethral stricture. One patient combined with several associate abnormalities died of seizure in two years after corrective surgery.

Five patients with decreased renal function were assessed by vesicoureteral reflux and 99 mTc-dimercaptosuccinic acid tests. All live patients were able to do self-voiding. Two patients are trained by timed voiding, and since the rest two were below 3-year-old, it was un-

Table 4. Results including complications, their urinary and fecal functions after corrective surgery

Case	CC (cm)	Age (mo)				Decline of renal function	VUR	Voiding pattern	Support enema
		Total correction		Complication					
2	4.5	PSARVUP (sacrifice left vagina,uterus)	11			Not done		Self[b]	2 yr 8 mo
7	4.0	PSARVUP + laparotomy (vaginal switch)	13	Bladder stone remove urethrotomy[a] (×2 times)	22 25 72	Mild (Lt)	G3 (Lt)	Good	8 yr
8	4.0	PSARUP + hysterectomy	15	Urethral stone remove + urethroplasty	28	Moderate (Rt)		Timed[c]	6 yr
10	2.0	PSARUP	11			Mild		Good	3 yr 10 mo
11	2.5	PSARVUP	10			Not done		Self[b]	1 yr 9 mo
13	2.5	PSARVUP	13			Not done		Good	Good (11 yr)
14	3.5	PSARVUP	15	UVF repair	43	Mild (Lt)	G4 (Lt) G1 (Rt)	Timed[c]	11 yr
15	2.5	PSARVUP + laparotomy	14	VVF repair	63	Not done		Good	10 yr
16	3.5	PSARUP	13	Expire due to seizure	36	Severe (Lt)	G4 (Lt)		

CC, length of common channel; VUR, vesicoureteral reflux; PSARVUP, posterior sagittal anorectovaginourethroplasty; PSARUP, posterior sagittal anorectourethroplasty; UVF, urethrovaginal fistula; VVF, vesicovaginal fistula; Rt, right; Lt, left; G, grade.
[a]Due to urethral stricture. [b]Self voiding but could not evaluated because they were less than 3 years of age. [c]Timed voiding required some interval to empty their bladder.

Table 5. Comparison between patients according to length of common channels

	Length of common channel	
	<3 cm (n = 4)	≥3 cm (n = 5)
Over than 3 times of operative procedures	2	5
Requiring cases an additional abdominal approach	1	2
Vesicoureteral reflux	0	3
Decreased renal function on DMSA	1	4
Constipation to need supportive intermittent enema	3	5
Complications after total correction[a]	1	3

DMSA, 99mTc-DMSA dimercaptosuccinic acid.
[a]Needed operative procedure due to vesicovaginal fistula, urethral stricture, urethral stone.

available to evaluate their voiding control. Only 1 patient was able to defecate without any supportive enema, and 8 patients needed enemas.

DISCUSSION

Persistent cloaca is a malformation in which the urinary, genital and digestive organs remain open toward the telomeric site of the perineum, mainly the rear of clitoris, through a common channel. This condition is due to abnormal development of the urogenital septum and is very rare, with a frequency of one per 50,000-125,000 newborns [1,2]. Since persistent cloaca has various clinical manifestations, it is not easy to diagnose by prenatal ultrasonography. Therefore, many of our patients were diagnosed as intraperitoneal cyst or hydronephrosis.

Since the patients with persistent cloaca can affect their mortality in neonatal period and the outcomes of corrective surgery and renal function, the appropriate diagnosis and treatment of these patients are very important [3-6]. A study of 361 patients with persistent cloaca identified 3 pitfalls in neonatal period management: 1) failure to recognize and manage hydrocolpos, 2) colostomy or vesicostomy problems and 3) misdiagnosis as imperforate anus with rectovaginal fistula and intersex [7]. The most important initial treatment is drainage of hydrocolpos, which can cause enlargement of the bladder. Subsequent cystostomy is recommended only when the bladder continues to be enlarged [7,8]. Other studies have found that 30% of children with persistent cloaca also have hydrocolpos [4,7], that suppression of trigone of bladder causes

hydroureter and hydronephrosis and that infection of undrained hydrocolpos itself can result in perforation, sometimes accompanied by pyocolpos [9]. Since our study involved only a few patients, it is difficult to determine the efficacy of cystostomy and vaginostomy. Nevertheless, children who did not undergo drainage of hydrocolpos died of sepsis and complications, suggest that such drainage is essential. If self-voiding is possible, the need for cystostomy should be determined after assessing the degree of bladder enlargement at follow-up. Since we have not conduct vaginostomy only if both the vagina and bladder were enlarged, it is necessary to determine whether vaginostomy alone can cause bladder enlargement to disappear.

It is also most important to accurately determine the anatomical structure of each patient and to establish a treatment plan before cystostomy or vaginostomy. Urological defect is present in 68% children with persistent cloaca [6] with 33 to 50% having hemiuterus and hemivagina [6,10,11]. Various malformations were present in our patients, such as hemivagina, vaginal septum, blind pouch vagina, and Mullerian agenesis. Since hydrocolpos might not be drained appropriately, radiographic examination may be required before colostomy. A cystoscope can be used to measure the structure of the genital system and the length of the common channel, and a laparoscope can be used to diagnose hemiuterus and degree of enlargement.

Since 1982, corrective surgery has been performed using PSARVUR [6,12]. In addition, total urogenital mobilization can be performed by mobilizing the urogenital sinus from the pelvic cavity [13,14]. Using this method, it is easy to relocate the vagina down to the perineum when the length of the common channel is less than 3 cm, but, in patients with a common channel longer than 3 cm, there may be technical difficulties because the length may not be sufficient even after avulsion of both the vagina and urethra [15]. Other methods may be better in these patients, including vaginal switch and bowel replacement [9,11,16,17]. Although all patients underwent revision surgery using PSARVUR, several also required a laparotomy, increasing the number of patients with common channel $\geq$3 cm who are eligible for corrective surgery. Several patients also required additional operations due to complications after surgery. These children should be repeatedly examined for complications of the urinary system and renal function and require continuous bowel training. Since all patients had not reached puberty, we could not fully analyze the functions and results of the genital systems, suggesting the need for longer term follow-up.

In conclusion, three of twelve patients who had enlarged bladder or vagina at birth did not undergo cystostomy or vaginostomy at first operation, and they had earlier complications after surgery or required drainage tube insertion. Patients with anatomical malformations before colostomy, as well as hydrocolpos and bladder enlargement, require a vaginostomy with or without cystostomy to reduce complications. Follow-up is required in patients with hydrocolpos and bladder enlargement to determine whether vaginal drainage improves dilated bladder. Continuous long-term follow-up survey is required to determine the long-term results of corrective surgery.

CONFLICTS OF INTEREST

No potential conflict of interest relevant to this article was reported.

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