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Summary

Introduction

Persistent cloaca is a rare, congenital anomaly involving the genital, urinary, and rectal organ systems. While prompt bowel diversion is the standard of care, the optimal method of genitourinary decompression is unclear. Bladder outlet obstruction and hydrometrocolpos are common complications that can lead to obstructive uropathy, abdominal distention, infection, perforation, and acidosis. Proposed management strategies include early surgical diversion (vesicostomy, vaginostomy, ureterostomy, nephrostomy) or clean intermittent catheterization (CIC) of the common channel. We hypothesized that CIC is an adequate means of genitourinary decompression and preservation of renal function, regardless of the severity of cloacal anomaly.

Methods

We reviewed all patients with persistent cloaca from a single, tertiary care center from 1995 to 2013. We collected data regarding renal function (serial serum creatinine prior to definitive reconstruction, and baseline estimated glomerular filtration rate [GFR]), presence of hydrocolpos, hydronephrosis, vesicoureteral reflux (VUR) or renal dysplasia, and length of the common channel. A linear mixed model was used to calculate creatinine change over time in relation to method of management and child age. Estimated GFR was calculated using the Schwartz equation for neonates $= 0.45 \times \text{height in cm/serum}$ creatinine in mg/dL. The t test was used for continuous data and Fisher's exact test was used for binomial data. A p value < 0.05 was considered significant.

Results

Twenty-five patients were identified. Nine (36%) patients underwent early surgical diversion versus 16

(64%) managed by CIC prior to formal reconstruction. Seven had short common channels (<3 cm) and 18 had long common channels (≥3 cm). Hydrocolpos was present in 14 (56%) of the patients. When comparing the two management groups, there was no significant difference in hydronephrosis, high-grade hydronephrosis (grades III–IV, p=0.62), any VUR (p=0.33), high-grade VUR (grades III–V, p=0.62), hydrocolpos (p=0.21), or renal dysplasia (p=0.42). No significant differences were found between mean baseline GFR for diversion (22.9 mL/min per 1.73 m²) versus CIC (39.2 mL/min per 1.73 m², p=0.22). There was no difference in creatinine trend between the two groups.

Discussion

Currently, there is no consensus on the initial management of obstructive uropathy and resulting hydrocolpos in newborns with persistent cloaca. In addition to CIC, management strategies include surgical options such as vesicostomy, vaginostomy, or upper tract diversions such as ureterostomy or nephrostomy. Our results suggest that CIC is similar to these other proposed diversion procedures while minimizing morbidity. Creatinine trends over time were similar between the two groups and reached comparable nadirs. Limitations of our study include the retrospective nature of a small sample size. The primary risk is differences between the two groups that we were not able to appreciate. Furthermore, we did not attempt to assess the morbidity of the two different strategies.

Conclusions

CIC is an adequate initial management strategy to decompress the genitourinary tract in patients with persistent cloaca. CIC preserves renal function similar to early surgical decompression.

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Introduction

Cloacal malformations are rare, congenital anomalies involving the rectum and genitourinary organ systems. Clinicians caring for infants with persistent cloaca face significant challenges in the newborn period. There are no standardized treatment guidelines, and failure to recognize and manage bladder outlet obstruction can lead to significant complications, including obstructive uropathy, hydrocolpos, abdominal distention, infection, and metabolic acidosis [1—4].

Definitive correction of a cloacal anomaly involves major surgical reconstruction, often pursued several months after birth. Because of this abnormal anatomy, up to 30% of neonates with this condition have a collection of fluid, urine, and mucous within the vagina, called hydrocolpos [2]. As a result, several concerning complications may arise. The inability of this fluid to drain, combined with the distension of the cavity, puts pressure on the bladder and the insertion of the ureters. Consequently, urine is unable to drain from the kidneys, leading to hydronephrosis and kidney damage [1].

Drainage of the urinary tract remains the prime concern to the pediatric urologist in the newborn period in order to prevent further renal impairment [5]. Surgical drainage or diversion of the urinary tract is commonly undertaken in patients with cloacal anomalies in the form of suprapubic tube, vesicostomy, vaginostomy, and nephrostomy tube in order to bridge these patients to definitive reconstruction later in life [6-8]. Use of vesicostomy was first proposed by Duckett [9] in 1973, and the procedure is guite effective and simple to perform. However, the patient must undergo an operation under anesthesia, and it carries the potential complications of stenosis of the tract or prolapse of the bladder [10]. More recently, clean intermittent catheterization (CIC) of the common channel has been proposed as a less-invasive method to drain the urinary tract, relieve upstream obstruction, and protect or maintain renal function [11].

The purpose of this study is to compare our institutional experience with two different management strategies for obstructive uropathy in infants with persistent cloaca: early surgical diversion (vesicostomy, vaginostomy, nephrostomy) versus CIC of the common channel. We hypothesized that CIC of the common channel provides for adequate urinary tract decompression and preservation of renal function compared with surgical diversion. Our aim was to compare renal function in newborn cloaca patients who were treated by these two different management strategies.

Materials and methods

After institutional review board approval, we retrospectively reviewed the medical records of all patients diagnosed with persistent cloaca and managed at Children's Hospital Colorado from 1995 through 2013. Baseline data included common channel length, drainage strategy, initial serum creatinine values, and imaging characteristics. Initial serum creatinine values were recorded between 24 and 72 h of birth to avoid maternal influence. Birth height was recorded to calculate baseline glomerular filtration rate

(GFR) bγ the Schwartz equation for neonates = $0.45 \times \text{height in cm/serum creatinine in mg/dL}$ [12]. This equation was chosen as it relies on two variables that were commonly available (as opposed to newer variations that rely on a blood urea nitrogen test or cystatin C, among other variables). Diversion procedures included nephrostomy tube, ureterostomy, vesicostomy, or vagi-Imaging characteristics included nostomy. ureteronephrosis grade using the Society for Fetal Urology (SFU) classification system [4], vesicoureteral reflux (VUR) grade, renal dysplasia, and the presence of hydrocolpos.

Follow-up creatinine values were recorded to assess the trend until definitive cloacal reconstruction. A linear mixed model was used to model the creatinine change over time in relation to method of management (diversion versus CIC) and child age. The two-sided Student t test was used to compare continuous data and the Fisher exact test was used for binomial data. A p value <0.05 was considered significant.

Results

Twenty-five infants with persistent cloaca were identified at our institution. Nine infants who underwent surgical diversion were compared with 16 managed by CIC alone. A description of the clinical characteristics of each group is shown in Table 1. Of note, all patients who underwent diversion received this treatment within a week of birth. Seven patients had short common channels (<3 cm) and 18 had long common channels (≥3 cm) and this was not significantly different between groups. There was no significant difference between groups with regards to the presence or absence of hydronephrosis or high-grade hydronephrosis (SFU grades III and IV), the presence or absence of VUR or high-grade VUR (grades III—V),

Table 1 Characteristics of the 25 patients with cloacal anomaly in this study, including breakdown of associated genitourinary findings, including hydronephrosis, vesicoureteral reflux, and the presence of hydrocolpos and renal dysplasia.

	CIC	Diversion	р
N	16	9	
Common channel length			
Short (<3 cm)	5/16	2/9	1.00
Long ($<$ 3 cm)	11/16	7/9	1.00
Hydronephrosis			
Any	12/16	8/9	0.62
High grade (SFU III—IV)	7/16	6/9	0.38
VUR			
Any	5/16	6/9	0.33
High grade (III—V)	4/16	3/9	0.62
Renal dysplasia	7/16	2/9	0.40
Hydrocolpos	7/16	7/9	0.21
Mean estimated initial GFR \pm SD39.2 \pm 15.322.9 \pm 23.10.22 (mL/min per 1.73m²)			

 $\mbox{CIC} = \mbox{clean intermittent catheterization; GFR} = \mbox{glomerular filtration rate; SFU} = \mbox{Society for Fetal Urology; } \mbox{VUR} = \mbox{vesicoureteral reflux.}$

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