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Congenital cloaca: Long-term follow-up results with emphasis on outcomes beyond childhood

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ABSTRACT

Persistent cloaca remains a challenge for pediatric surgeons and urologists. Reconstructive surgery of cloacal malformations aims to repair the anorectum, urinary tract, and genital organs, and achieve fecal and urinary continence as well as functional genital tract capable for sexual activity and pregnancy. Unfortunately, even in most experienced hands these goals are not always accomplished. The endpoint of the functional development of bowel, urinary, and genital functions is the completion of patient's growth and sexual maturity. It is unlikely that there will be any significant functional improvement beyond these time points. About half of the patients with cloaca attain fecal and urinary continence after their growth period. The remaining half stay clean or dry by adjunctive measures such as bowel management by enemas or ACE channel, and continent urinary diversion or intermittent catheterization. Problems related to genital organs such as obstructed menstruations, amenorrhea, and introitus stenosis are common and often require secondary surgery. Encouragingly, most adolescent and adult patients are capable of sexual life despite often complex vaginal primary and secondary reconstructions. Also, cloacal malformation does not preclude pregnancies, although they still are quite rare. Pregnant patients with cloaca require special care and follow-up to guarantee uncomplicated pregnancy and preservation of anorectal and urinary functions. Cesarean section is recommended for cloaca patients. The self-reported quality of life of cloaca patients appears to be comparable to that of female patients with less complex anorectal malformations.

Repair of a congenital cloaca is a major challenge for pediatric surgeons. The condition is rare; reported incidences range around 1:20,000–50,000, therefore, most pediatric surgeons encounter only few cases of cloaca during their entire career. The management of cloacal malformations was revolutionized by the posterior sagittal approach (PSARP) and total urogenital mobilization procedures of Alberto Peña.[1,2] This approach enabled full reconstruction of the urinary tract, genital tract, and rectum and anus during one operative session. Before the PSARP era, many patients with cloaca initially underwent only anorectal reconstruction and the remaining urogenital sinus was left untouched or was repaired later. The outcomes of these operations were usually dismal.[3]

In the literature, there is scarcity of reports on long-term follow-up of cloaca. Furthermore, most reports present only small number of cases or the outcomes are displayed as a part of general population of anorectal malformations. No controlled or prospective studies on long-term functional outcomes are available, all reports in the literature are retrospective institutional reviews. Only a few reports include more than 100 patients.[4,5] Especially, there is very little information about functional status and quality of life of cloaca patients after the development of puberty and beyond their growth period.

The aim of this review is to summarize the reported late outcomes in patients with a congenital cloaca, with the focus on those who are at their puberty or beyond that. Moreover, the reported outcomes from the literature are compared with the author's own patients of a similar age range. Of a total of author's 89 patients with cloaca, 27 are adolescent or adults.

The importance of analyzing the outcomes at this age lies in the fact that at puberty or beyond that the development of anorectal and urinary tract functions are likely to approach its endpoint. The development of gynecological organs is also completed and the outcomes and prospect of sexual life and fertility can be assessed.

Anorectal function

Most of the reports describing anorectal function in patients with cloaca present the outcomes by pooling all age groups. The

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Table 1
Bowel function in adolescents and adults with cloaca.

	Continent (%)	Bowel management/ACE (%)	Medication (%)	Stoma (%)	Soiling (%)
Davies (2010)[12] ($n = 15$)	27	33	13	27	n.r
Couchman et al.[13] ($n = 19$)	58	5	n.r	37	n.r
Rintala (2015) ($n = 27$)	52	22	0	11	15

n.r, not reported.

usual lower cut-off age is 3–4 years, as these are considered the time points when children usually are toilet-trained and evaluable for bowel function.

It is very clear that the bowel function in a preschool-aged child is different from that of a patient who has gone through puberty. A certain degree of improvement in the bowel function in patients with anorectal malformations has been reported by numerous authors.[6–8] It is most likely that this holds true also in patients with cloaca. The small number of patients in almost every cloaca series have precluded the evaluation of the role of aging in relation to bowel functional outcomes.

Versteegh et al.[9] have recently published a systematic review on the functional outcomes in cloaca patients. This collective review has pooled the data on anorectal function for a total of 263 patients. The length of follow-up in this pooled patient series ranged from 5 months to 31 years. The largest series[10] included in the report had 156 patients with pertinent data on bowel function. In this series, the patients were older than 3 years. This systematic review reported bowel function by using Krickenbeck criteria.[11] None of the studies included in this systematic review were prospective, the data collection was a retrospective case note review in all.

Voluntary bowel movements were reported in 108 of the 188 patients (57%) in whom this factor of bowel function was analyzed. The presence of voluntary bowel movements ranged between 41% and 60%. Only one study in this systematic review reported patients with total continence. Peña et al. found 26 out of 156 (17%) patients to be totally continent at last follow-up.

More or less total fecal incontinence was found in 33% of the patients (range in the studies 14–41%) in this systematic review. The actual severity of incontinence was not graded in the articles included in the systematic review. The reports also did not describe any criteria for continence or incontinence. It is still obvious that many, if not most, of these patients required some sort of bowel management to stay clean. The reported bowel management modalities included regular enemas and formation of an ACE stoma. In 17% of the patients (range in the studies 5–22%) the quality of bowel control was so poor that the patients chose a permanent colostomy. Some degree of fecal soiling was reported in 71% of the patients (range in the studies 14–83%). Again, the severity of soiling was not graded. The consequences of soiling was not analyzed in the present reports.

Constipation was a very common problem in patients with cloaca. Overall, more than half of the patients suffered from constipation requiring medical management or enema program. The incidence of constipation ranged between 30% and 88%. Constipation was managed medically by oral laxatives (29% of the patients) or by an enema bowel management program.

Some kind of bowel management, either by retrograde bowel washouts or by antegrade washouts through an ACE appendicostomy were used on average by 38% of the patients. The need for bowel management ranged between 14% and 65% in the reports of the systematic review. The indications for bowel management were fecal incontinence in most cases or intractable constipation.

The reported bowel function outcomes in adolescents and adults are somewhat different than in the series that have pooled functional data from all age groups (Table 1). However, the data on bowel function in patients beyond their childhood is extremely scarce. Davies et al.[12] had 15 cloaca patients in their series of patients with anorectal malformations and a mean follow-up of 26 years. A total of 4 (27%) of their 15 patients were spontaneously continent, five (33%) had an ACE or used retrograde washouts, two (13%) used medication to augment continence, and four (27%) had a permanent stoma. Couchman et al.[13] identified 19 cloaca patients with a mean follow-up of 22 years. Fecal continence was reported by 11 (58%) of these patients. The remaining eight patients had an ileostomy or colostomy (7, 37%), or ACE conduit (1, 5%) for bowel evacuation. In the author's personal series, 27 patients are adolescent or adults. The mean age of the patients is 23 years (range: 13–40 years). Of these 27 patients, 14 (52%) have developed spontaneous continence and do not require any medications or protective aids. Six patients (22%) have an ACE formation for bowel emptying. Four patients (15%) suffer from soiling that occasionally requires use of protective pads. Three patients,[11] two with a complete sacral agenesis and another with a poorly functioning ACE conduit, have opted for a permanent stoma formation.

In the few studies concerning adolescent or adult patients with a cloaca, none have been reported to suffer from constipation. In the author's series this was also the case, none of the 27 patients reported constipation nor used any laxatives to augment bowel emptying.

It appears that the fecal continence rate in adolescents and adults is more favorable than in children. Several factors are likely to contribute to this finding. Adolescents and adults are maximally adapted to their initially deficient anorectal function and have usually developed strategies to stay clean. These include dietary modifications, optimized stooling habits, and sometimes, use of antipropulsive medication. The absence of constipation also promotes better continence because the risk of overflow incontinence is eliminated. In childhood, overflow incontinence is probably the most common cause of fecal soiling in these patients.

Urinary tract function

It is not unexpected that patients with cloaca commonly have urinary tract dysfunction. Urological as well as spinal and spinal cord anomalies are very common in cloaca patients. Urodynamic studies have revealed that a significant proportion of patients with anorectal malformations have neurovesical dysfunction prior to surgery.[14,15] This is especially true in patients with a cloaca.[16] Moreover, the extensive surgery required to repair a cloacal malformation can deteriorate the bladder function even more than in other patients with anorectal malformations.[16]

In the systematic review of Versteegh et al.[9] concerning cloaca patients of all ages, urinary tract function was reported in 332 patients. This review pooled patients from nine studies. Spontaneous voiding was achieved on an average by 46% (138/299) of the assessed patients. In the studies included in the systematic review, the percentage of patients with spontaneous voiding ranged from 22% to 54%. Intermittent catheterization to empty the bladder was reported by 42% (141/332) of the patients (range in the studies