

## PERSISTENT CLOACA: UROLOGICAL THERAPY

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### Abstract

*Purpose:* To evaluate urinary tract function and continence in patients (pts) with persistent cloaca.

*Materials and Methods:* surgery was performed in 32 patients with cloaca, 3 months to 16 years old.

*Group A.* In 12 pts urethral and vaginal reconstructions and pull-through of rectum were done together during the primary operation. 5 pts had VUR, 3 neurogenic bladder (NB), 2 had solitary kidneys. Bladder augmentation n=1 and VUR-correction n=2 were done as secondary operations.

*Group B.* In 20 pts primary surgery for the cloaca was done at other institutions. They presented with persistent urogenital sinus (UGS) n=14, vesico-vaginal fistula n=2, vesico-urethral fistula n=2, reconstructed urethra n=2. Other urinary pathologies: VUR n=13 pts, NB n=7, solitary kidneys n=6, megaureter n=4. Only 2 pts were continent. Urethral reconstruction n=15, bladder augmentation/substitution n=5, vesico-cutaneostomie n=5, VUR-correction n=5 were performed.

*Results:* *Group A.* There are no pts with renal insufficiency. All patients are dry spontaneously n=7, or by catheterisation via urethra n=5. *Group B.* Reflux-nephropathy leading to renal insufficiency was seen in 3 pts. One patient had undergone renal transplantation. Another was transplanted after the re-do operation for cloaca and died 6 month after transplantation. The 3rd was lost to follow-up. 7 patients are continent, 9 are socially continent, and 2 are incontinent for urine.

*Conclusions:* The outcome of urinary tract function is better when urethral and vaginal repairs and the pull-through of the rectum are done together (group A). To prevent the damage of the urinary tract long-term follow-up is essential. Some of these patients need secondary operations to achieve urinary control or to correct reflux.

**Key words:** persistent cloaca, posterior sagittal approach, urinary tract function, incontinence

### Introduction

Persistent cloaca is a complex malformation with a wide anatomical spectrum.

The surgical procedures enable the correction of cloacal anomalies with good prognosis regarding urinary and bowel control and genital function. However, persistent cloaca is a rare malformation and for many pediatric surgeons it is difficult to get substantial experience with this anomaly.

Consequently the anatomy of malformation may be misinterpreted with inadequate timing of operation and inadequate procedures. The obtained results are frustrating both for patient and surgeon leading to frequent reoperations.

### Material and methods

During a 12 year period (1993-2004), 32 girls with persistent cloaca have been operated. There were 12 primary operated children (Group A) and 20 reoperated patients (Group B). All children from group B presented after having had surgery for persistent cloaca elsewhere ranging from 1 to 14 previous surgical procedures.

A review of clinical and radiological records was undertaken. The parents and the patients were interviewed. All patients were seen in the last 5 years.

**Group A:** The age at the operation was between 2 months and 1 year with a mean of 7 month. Eleven patients had colostomies and 1 patient vaginostomy before definitive correction. There were 5 children with double vagina and double uterus (2 with hydrocolpos), one with vaginal agenesis and one with vaginal and uterine agenesis. One girl had a perineal lipoma, and one absence of both minor labia. In 5 children the common channel was longer than 3 cm. In 7 children sacral dysplasia was present. Five patients had VUR (8 renal units), 3 neurogenic bladder (NB) all with sacral dysplasia, 2 solitary kidneys, and one ureteral triplication with one megaureter. In all these patients urethral and vaginal reconstructions and the pull-through of the rectum were done together at primary correction. The reimplantation of the megaureter in the child with ureteral triplication was performed during primary correction of the cloaca.

**Group B:** All children underwent at least one correction for persistent cloaca before presentation to our institution: pull-through of the rectum in 12 patients, surgery for the UGS in 2, pull-through of the vagina instead of the rectum in 1, pull-through of the common channel in 1, multiple operations (up to 14) in 4 patients. Fourteen presented with UGS (one of them with bladder agenesis), vesico-vaginal fistula in 2 (one of them with urethral atresia), urethro-vaginal fistula in 2, reconstructed urethra n=2, both of them with low bladder volume.

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Thirteen patients had VUR (21 renal units), 7 NB (6 in combination with sacral dysplasia), 6 solitary kidneys, 4 megaureter, and one had bladder diverticula. There was no information about the common channel (CC) in 14 patients, in 6 the CC was more than 3 cm long. 12 patients had sacral dysplasia.

There were 11 girls with double vagina and double uterus, 4 with vaginal atresia, 3 with atresia of the cervix. One girl had stenosis of the vaginal substitute (ileum segment), 2 absence of both minor labia and one had no clitoris. Renal failure occurred in 3 girls as consequence of undetected hydrometrocolpos leading to obstructive uropathies. One of them already had renal transplantation at presentation, another girl had kidney-transplantation 6 month after the reoperation for cloaca.

The age at reoperation in Group B was between 7 months and 16 years with a mean of 7 years. In 15 patients urethral reconstruction was performed. Other undertaken procedures on urinary tract: bladder augmentation/neobladder in 5, continent vesicostomy according to Mitrofanoff procedure (1) in 5, VUR-correction in 5 (8 units).

The urethra was reconstructed together with the vagina and rectum in 14 girls, and together with the vagina in 1. All reconstructions were performed under protective colostomies or ileostomies. Bladder augmentation or bladder substitution were performed in 5 patients. The same number of patients received continent cystostomies and 3 were provided with MACE (2). Two patients had vaginoplasty only (Table 1).

Table I.

Re-do Procedures in “group B”	Nr. of patients
Posterior sagittal anorectovaginourethroplasty (PSARVUP)	4
PSARVUP, bladder augmentation (Mainz-Pouch I) and Mitrofanoff	1
Posterior sagittal anorectoplasty (PSARP) and TUM	5
PSARP, TUM, bladder augmentation and Mitrofanoff	1
PSARP, vaginal substitution with ileum, urethra substitution with ileum, and bladder augmentation with ileum	1
PSARP, vaginal substitution with sigma and urethroplasty	2
PSARP, vaginal substitution with ileum, Mitrofanoff and MACE	1
PSARP, vaginoplasty from UGS, neobladder from ileum, Mitrofanoff, and MACE	1
TUM	1
Vaginal introitoplasty, bladder augmentation with ileum, Mitrofanoff and MACE	1
Vaginal substitution with ileum	1
Vaginal introitoplasty of vaginal substitute (ileum segment)	1

## Results

Only the urological status will be analysed in spite of the fact that all patients from group A and most patients from group B had simultaneously correction of urethra, vagina and rectum.

Postoperative evaluation was undertaken clinically in all and radiologically in most patients.

**Group A:** All patients have good renal function. In 1 girl the urethra retracted after TUM. She also has neurogenic bladder with VUR III in a solitary kidney. Although 7 patients from group A had sacral dysplasia, there are only 3 children with neurogenic bladder dysfunction.

Seven patients void with good urinary control and are dry. Five empty their bladder by intermittent catheterisation via the urethra and are also dry.

**Group B:** Obstructive uropathies leading to renal insufficiency were seen in 3 patients. All 3 girls had undetected hydrometrocolpos as newborns with obstructive uropathies. One patient presented with a transplanted kidney, another one was transplanted after re-do operation for cloaca, but died 6 months following transplantation. The 3rd patient waiting for transplantation, was seen 5 years ago before she was lost to follow-up. Other 5 girls had reflux nephropathies leading to nephrectomy in one.

There were 11 girls with sacral dysplasia, 6 of them with neurogenic bladder, 1 with ileum-neobladder and 1 with bladder agenesis, and 3 with normal bladder function. But there was also one girl with neurogenic bladder, without sacral dysplasia.

At last follow-up, there were 7 continent patients. Nine patients achieved continence by intermittent catheterisation via the urethra or a catheterisable channel, and 2 were still urinary incontinent.

## Discussion

Persistent cloaca is an anomaly with an extremely complex anatomy. It consists of a spectrum of anomalies of the genitourinary tract and anus and rectum. All three organ systems have a common confluence, termed the cloacal channel (3).

The malformation can be corrected satisfactory in most patients, providing them with urinary and bowel control and good sexual function (4). But most patients need very sophisticated surgical procedures (4,5,6).

The surgeon must have vast experience in the treatment of anorectal and genitourinary malformations. Treatment of a newborn with cloacal malformation should start with a colostomy. Moreover, satisfactory evacuation of urine must

be achieved, as some children may present with infravesical obstruction due to hydrometrocolpos or obstruction of the common channel (7). Vesicostomy or vaginostomy should be considered in such cases. In our series, there were 3 girls (all in group B) in which unrecognised hydrometrocolpos led to urinary obstruction and subsequently resulted in renal failure.

The anatomy must be clarified by clinical examination, sonography, x-ray studies, MRI, and endoscopy (8, 9). The timing of surgery is important: the patients should be in good condition, having an accurate diagnosis before any procedure. A correct diagnosis will allow the experienced surgeon to repair the entire malformation and avoid a persistent urogenital sinus (5). In most patients from “group B” the cloacal malformation was misinterpreted as anal atresia with recto-vaginal fistula. Consequently 14 of them presented with persistent UGS. The other 4 presented with vesico-vaginal, or urethro-vaginal fistula. Only 2 girls had a functional urethra, but they also needed correction because of vaginal pathology.

As for primary operation, the posterior sagittal incision provides good exposure of the anatomy, protecting the nerves and vessels which enter the pelvis posteriorly and laterally. Yet scar tissue and the absence of organs, e.g. urethra, vagina, uterus, require other approaches and types of reconstructions (see Table I).

The re-do operations were performed by posterior sagittal approach in all patients from group A and in 16 patients from group B. To achieve urinary continence in one patient from group A and in 5 patients from group B,

bladder augmentation or bladder substitution were carried out. There were 5 patients needing a Mitrofanoff procedure to become dry by intermittent catheterization. Additionally a bladder neck plasty in one and a colpo-wrap (10) procedure in another girl were done.

Primary reconstruction of the urethra, the vagina and the rectum simultaneously during the first operation gave better results regarding urinary continence. All patients from group A are dry: 7/12 void spontaneously and 5/12 by intermittent catheterization.

In contrast, the results are less good in patients with multiple procedures regarding both, continence and renal function. In group B only 7/18 patients void spontaneously, 10/18 are on intermittent catheterization, 2/18 are incontinent, one child died after renal transplantation.

The likelihood of needing continuing intermittent catheterization is greater in those with severe sacral anomalies (3). That was true also in our patients.

Accomplishment of definitive repair involves not only anatomical reconstruction, but also postoperative urinary control, including the initiation of intermittent catheterization under repeated urodynamic evaluations (11).

The well-being of the patients is markedly affected by incontinence and also by renal failure. The lack of compliance to regular catheterizations is an absolute contraindication for continent urinary diversion (12).

Therefore, before providing the patient with continent urinary diversion the surgeon has to be sure that the patients or their caregivers will be able to empty the bladder by intermittent catheterization.

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