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Case report

## SURGICAL RECONSTRUCTION OF CLOACAL MALFORMATION: A CASE REPORT

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### ABSTRACT

It is a surgical challenge to repair a cloacal malformation. We are reporting a case of cloacal malformation in which we performed a reconstructive surgery. We performed a reconstructive surgery in a 5 yr old patient who had got a colostomy done in the neonatal period. The surgical management included purely a posterior sagittal anorectovaginourethroplasty without an abdominal approach.

**Keywords:** Cloaca, Anorectal malformations, Posterior sagittal anorectovaginourethroplasty.

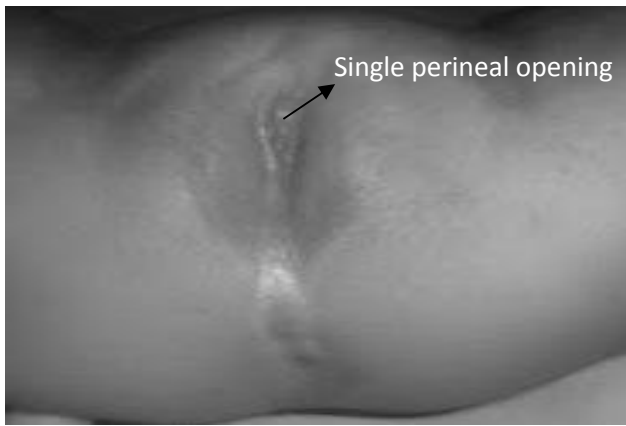
### INTRODUCTION

Persistent cloaca has a wide spectrum of anomalies including those of urogenital and hind gut. It is comes under rare malformations in the Krickenbeck's classification of anorectal malformations<sup>1</sup>. Clinical presentation consists of imperforate anus with a single perineal opening through which urine and meconium are passed. The surgical correction is very tough and challenging and urinary and fecal incontinence are frequent complications following operation. The ultimate goal of treatment includes achieving satisfactory bowel and urinary control as well as normal sexual functions at maturity<sup>1,2,3</sup>.

### CASE REPORT

A five year old female child, with a full term normal hospital delivery, colostomised on 2<sup>nd</sup> day of life for imperforate anus with single perineal opening was referred to us for definitive treatment. Past history and family history was not relevant.

On local examination – A single opening in the perineum was present. The labial folds were fused and the clitoris was well developed. Anal pigmentation was present. Gluteal cleft was not well developed. All sacral pieces could be felt. Per abdomen, a healthy right transverse colostomy was present.



**Fig. 1: Pre-operative picture showing single perineal opening.**

Investigations – CBC, Coagulation profile, Renal function tests, Liver function tests were normal with normal routine urine analysis. **Ultrasound of abdomen** was normal except for small sized left kidney. **Intravenous Urogram** was also normal except for small sized left kidney. **CT scan and MRI scan pelvis** showed atretic vagina. Uterus was normal for age. Both ovaries were normal. **Genitogram** showed that bladder was filled with contrast and was normal. Contrast entered the rectum and entire distal colon was opacified. Internal genitalia could not be visualized. Recto-urinary fistula with common channel opening in the perineum was seen. **Distal cologram** did not reveal a recto-urinary fistula. Rectal ectasia was present.

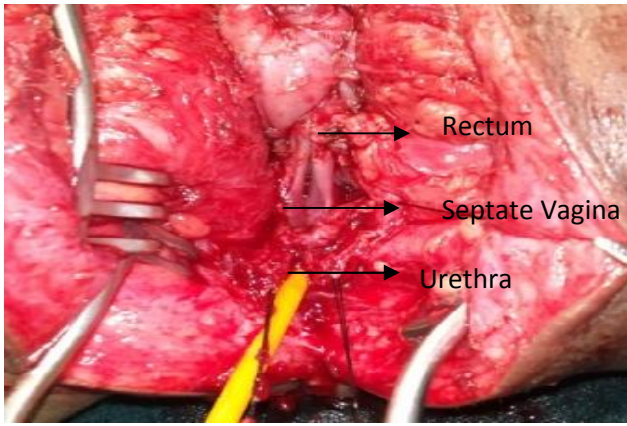


**Fig. 2: Genitogram**

A diagnostic pan endoscopy was performed which showed a short channel cloaca with common channel of about 2.5 cms. Posterior urethra, bladder neck and urinary bladder were

normal. A short channel communicating with cloaca and ending with two visible openings separated by a septum was visualized. Another third tiny opening was also visualized however the scope could not be negotiated into this opening.

**Operative findings:** The patient was in jackknife position. A posterior sagittal approach was used. The incision was taken from midsacrum upto the single perineal opening. All the structures were divided in midline. The coccyx was split. The endopelvic fascia was incised. The rectal pouch was visualized and the rectum was opened posteriorly; however no fistulous communication could be seen. Rectum was separated off vagina so that it could reach the perineal skin. The common channel was incised. Per-urethral catheter had entered into the vagina. The urethra was visualized and bladder was catheterized. Entire anomaly was laid open and all openings into the common channel were seen. Vagina was septate. The vaginal septum was excised and the septate vagina was disconnected from the common channel. Common channel was repaired over a 10F Foley's catheter to create a new urethra. Vagina was quite high and was sharing a paper thin wall with the urethra. Hence decision was made to create a vaginal substitute. Various options were available like using bowel segments, pedicle graft, split thickness skin graft etc. But the surgery was performed by perineal route alone and abdomen was not opened. Surgery was completed by doing an ano-rectoplasty which could accommodate a 12 no Hegar's dilator. Rectum was placed in the middle of the sphincter muscle complex which was identified by muscle stimulator. The perineal body was reconstructed anteriorly. The child was turned supine and a Y-V plasty was performed using labial skin flaps which were sutured to the native vagina to create a new vagina over a 10F red rubber catheter. Post operatively, feeds were started after 6 hours. Post operative period was uneventful.



**Fig. 3: Intra-operative findings**



**Fig. 4: Post-operative reconstruction**

## DISCUSSION

A persistent cloaca is a complex anorectal and genitourinary malformation, in which the rectum, vagina, and urinary tract meet and fuse, creating a cloaca, a single common channel<sup>4</sup>. Cloaca's probably occur in 1 in 20,000 live births<sup>5,6</sup>. This defect has been considered one of the most formidable challenges in pediatric surgery. The goals of treatment include achieving bowel and urinary control, as well as normal sexual function, intercourse, menstruation and obstetric issues. The repair of persistent cloaca represents a serious technical challenge and should be performed in specialized centers.

History of the procedure. Hendren has published the most comprehensive and authoritative reports on the subject of cloacal malformation repair. Hendren has described the posterior sagittal anorectovaginourethroplasty (PSARVUP). For more complex anomalies, an abdominal

approach is added to mobilize a very high vagina or rectum.

The etiology of persistent cloaca is unknown. More than 80% of all patients with a cloaca experience an associated urogenital anomaly. Rich et al report the following associated urogenital anomalies<sup>9</sup> - Cloaca, Rectovesical fistula, Rectoprostatic fistula, Rectovestibular fistula, Rectobulbar fistula, Rectoperineal fistula and all malformations.

In most centers, a single stage reconstruction is the definitive treatment though some surgeons prefer a two staged approach where they opt for repairing the anorectal anomaly initially and perform urogenital sinus repair at a later date. The planning of surgical management includes carefully identifying the anatomy specially measuring length of common channel, level of insertions of urinary channel, vagina and rectum, and associated urogenital anomalies. Depending upon the length of the common channel, cloacal malformations are categorized into two groups. Patients having common channel lesser than 3 cm (more than 60% of entire group) can be repaired by posterior sagittal approach. In patients having a length of the common channel more than 3 cm, it would be difficult to perform total urogenital mobilization from perineum to repair the malformation, so the common channel is left intact to be used as urethra, and colovaginoplasty along with anorectoplasty is performed. The major complication is fecal and urinary incontinence. The incidence is usually higher in cases of higher confluence where an abdomino-perineal approach is used<sup>1,2,3,7,8</sup>. Prognostic factors include quality of the sacrum, quality of the muscles, and length of the common channel. Total urogenital mobilization is a technique devised by Alberto Pena; it allows mobilization of the urethra and vagina as one structure. This is possible in patients with the more benign types of cloacae. If total urogenital mobilization does adequately lengthen the vagina, the vagina and urethra must be separated,

which is a technically challenging maneuver. Vaginourethral fistulae are more likely after this plane is dissected. Approximately 50% of patients have various degrees of vaginal or uterine septation. These can be totally or partially repaired during the main operation or deferred for definitive repair until puberty. Approximately one third of the patients have obstructed Müllerian structures, which can lead to severe problems due to retrograde menstruation, amenorrhea in patients with atretic uteri or hydrometrocolpos.

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