

## Vaginal reconstruction with a segment of ectopic ureter – A novel technique

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**Abstract:** Vaginal reconstruction techniques with locally available tissue, flaps and grafts are well known. Even segments of bowel interposed as a substitute for vagina has been described. We describe a unique technique of vaginal reconstruction with ureter in a child who presented primarily with ano-rectal malformation (ARM) and was found to have distal vaginal atresia. Child had nonfunctioning right kidney with ureter opening ectopically in vestibule which was used for vaginal reconstruction. The procedure was done in conjunction with posterior sagittal anorectoplasty.

**Keywords:** Ectopic ureter, vaginal atresia, ureterovaginoplasty, ano-rectal malformation.

### INTRODUCTION

Vaginal atresia occurs due to failure of the Mullerian ducts to reach the urogenital sinus. It can occur in association with ano-rectal malformation (ARM) [1]. In the past many were wrongly diagnosed with rectovaginal fistula which is rare compared to ARM with vaginal atresia [2]. Associated genitourinary malformations are common in these patients. Vaginal reconstruction has been performed using various flaps, segments of bowel [3,5]. However use of ureter for vaginal reconstruction has been very rarely described (only two case reports found cited on PubMed). Ureteric vaginal substitution has its own advantages over local flaps and bowel segments.

### CASE REPORT

The patient was a 7 year old female who presented primarily with the complaints of passing stools from abnormal orifice in the perineum since birth and constipation. The child was not evaluated till then. She did not have associated micturition complaints. On examination the child had an absent anal opening with a vestibular fistula. The child also had an absent vaginal orifice and normal urethral orifice (Fig 1).



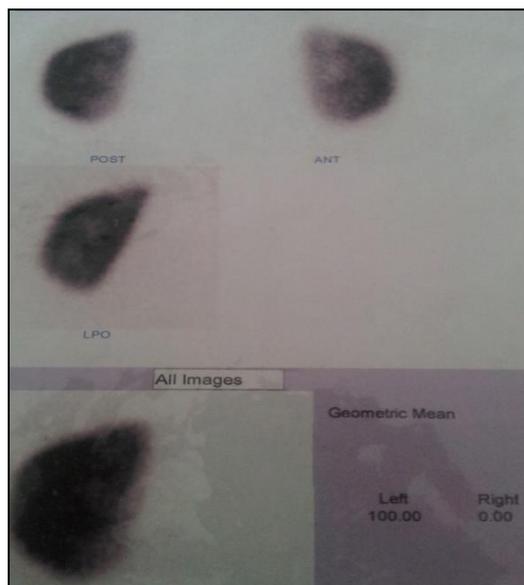
**Fig 1: Preoperative image depicting vestibular fistula (arrow head) with absent vaginal opening (arrow); urethral catheter is insitu**

### Investigations

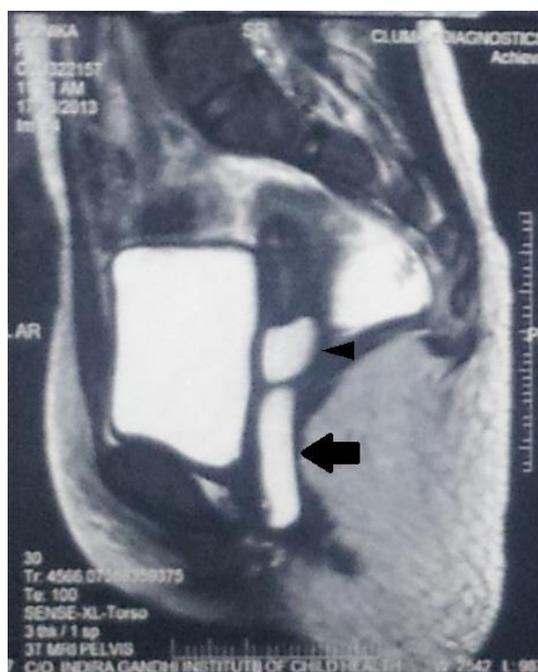
The investigations done and the findings are as follows in Table-1.

**Table-1: Findings of investigation**

<b>Ultrasound scan of abdomen</b>	Non-visualized right kidney and a normal left kidney
<b>Dimer capto succinic acid scan</b>	Nonfunctioning right kidney( <b>Figure 2</b> )
<b>2D – ECHO</b>	Subpulmonic ventricular septal defect with a left to right shunt
<b>Magnetic resonance imaging (MRI) of abdomen and pelvis</b>	Dilated tortuous tubular structure in right paravertebral region with distal end in vestibule suggestive of ectopic right ureter; rudimentary bicornuate uterus with single vagina and agenesis of distal portion of vagina( <b>Figure 3</b> )



**Fig 2: DMSA scan showing nonfunctioning right kidney**

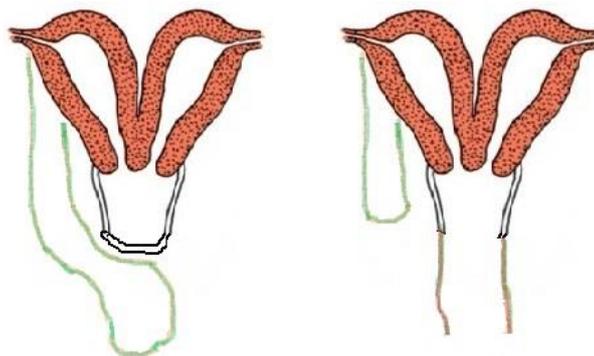


**Fig 3: MRI abdomen showing dilated tubular structure with distal end in vestibule suggestive of ureter (arrow), proximal patent vagina (arrow head)**

The child subsequently underwent cystoscopy which showed left hemi-trigone and a normal left ureteric orifice and an absent right ureteric orifice. A right transverse colostomy was done as the first stage of surgery. Later the child was planned for posterior sagittal ano-rectoplasty (PSARP) and vaginal reconstruction at the same time. At surgery, after mobilization of the rectum, the bulbous right ureter ending in the vestibule was identified. It was opened

and a portion of the wall was dissected. The dissected wall was sutured to the vaginal end to reconstruct the distal portion of the vagina (Figure 4). The rest of the procedure including right nephrectomy with excision of the residual ureter was completed and the child had an uneventful recovery in the post-operative period.

The child is on regular follow-up and evaluation has shown a good caliber vagina (Figure 5).



**Fig 4: Line diagram depicting transfer of distal portion of ureter to form the neo-vagina**



**Fig 5: Postoperative follow-up image at one year**

## DISCUSSION

Failure of the Mullerian ducts to reach the urogenital sinus contributes to congenital vaginal atresia. This can present as complete atresia, proximal atresia, or distal atresia, and each form has a different clinical picture. Distal atresia results when there is a failure of the sinovaginal bulbs, arising from the urogenital sinus to proliferate [1]. Occasionally ARM may be associated with Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome. Many of these patients in the past were wrongly diagnosed with rectovaginal fistula which is rare compared to ARM with vaginal atresia [2]. In females the ectopic ureter may enter anywhere from the bladder neck to the perineum or into the vagina, uterus, or even the rectum. Single system with an ectopic ureter may present with severe hydronephrosis reflecting distal obstruction. This may be associated with a non-functioning renal unit [3, 4].

The type of vaginal reconstruction required depends on the level of atresia and may range from non-operative dilatation to simple cutback vaginoplasty to total vaginal replacement. The appropriate timing of vaginal reconstruction has remained controversial with both proponents and opponents of early and late reconstruction. Early vaginal reconstruction may carry

an increased risk of vaginal stenosis as noted by some surgeons whereas delayed reconstruction is associated with psychological concerns as advocated by others. Options for construction of neo-vagina include local skin grafts from buttock, labial flaps, muscle flaps and intestinal substitutes. Skin grafts and local flaps are associated with dryness and dyspareunia. Intestinal substitutes commonly employed are sigmoid colon and ileum [5, 6]. In children with ARM and vaginal atresia some surgeons have advocated utilization of rectum to construct the vagina and proximal bowel pull through for the neo anus as a single stage procedure. Bowel vaginal reconstruction is not associated with the problem of dryness and dyspareunia due to mucous secretion which acts as a lubricant. However excess mucous secretion remains a problem, atleast for initial few months.

Probably the first description of vaginal reconstruction with ureter was described by Saalfeld *J et al.*; [7] in 1973 in a 10yr old girl with gynecological, urological, arterial and skeletal anomalies. Their patient had a dysplastic left kidney with partial duplication of dilated ureters. Lower portion of the dilated left ureter was incorporated into ureterovaginoplasty. Another case was reported by Gosalbez *et al.*; [8] in 1998. The patient was a 17yr old girl with a nonfunctioning right

kidney with grade IV reflux. They had detubularized almost the entire length of the right ureter after transecting proximally at the ureteropelvic junction and distally at the ureterovesical junction, preserving its blood supply. Our patient had a vestibular fistula with distal vaginal atresia with an ectopically opening right ureter into vestibule. We utilized only the distal most part of the ureter to reconstruct the vagina.

### CONCLUSION

Anatomy of the genitourinary and pelvic organs to rule out associated anomalies in these patients is best delineated by MRI. Use of ureter for vaginal reconstruction has the advantages of providing a mucosa lined substitute with no mucous production. There is no loss of bowel segment and local tissue is used as in our case with no adverse effects. Major limitation of the technique is finding a suitable patient with vaginal atresia and dilated ureter with non-functioning kidney.

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