CASE REPORT

Reconstruction of urethra using appendix in a patient with cloacal malformation

Sandeep Gupta,¹ Barun Kumar,¹ Sandip Basu,² Dilip Kumar Pal¹

¹Department of Urology, Institute of Post Graduate Medical Education and Research Kolkata, Kolkata, India

²Department of Plastic Surgery, Institute of Post Graduate Medical Education and Research, Kolkata, India

Correspondence to

Dr Dilip Kumar Pal, urologyipgmer@gmail.com

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SUMMARY

We report a case of 21-year-old young woman with congenital cloacal malformation. She was operated at the age of 1.5 years separating the rectum from common opening as a two-stage repair. She was incontinent in the earlier part of her life but she became continent to some extent later in early adulthood. She presented with urinary stress incontinence following delivery of dead fetus of 6 months. She underwent multiple investigations revealing common opening of bladder neck and vagina. A multidisciplinary evaluation was done and she underwent closure of common channel and neourethra reconstruction using pedicled appendix. Patient was continent and voiding by herself on discharge.

BACKGROUND

Cloaca is one of the rarest of congenital anomalies. Surgical correction is technically challenging. The goal of separation of genitourinary channels and making the patient continent is met with some major challenges such as complex anatomy and paucity of local tissue.

Our case was unique in the way that patient presented at the age of 21, conceived a fetus of 6 months in spite of the complex anatomy and was continent before the delivery of dead fetus.

Our main goal was to preserve the vaginal continuity, close the fistulous opening, create a neourethra and make the patient continent. The challenge was reconstruction because of the paucity of local tissue.

Most of the reconstruction techniques employed in cloacal repair has used reconstruction of vagina with either local tissue or bowel substitutes. To our knowledge, use of appendix for urethral reconstruction in females with cloacal malformation is first of its kind. Also, the early result is promising and it certainly adds to the armoury of techniques for the complex repair.

CASE PRESENTATION

Our patient is a 21-year-old young woman with a history of single common opening since birth. There was no other congenital anomaly.

She underwent a two-staged repair separating the anal opening. Thereafter, she had a common opening for both bladder and vagina. She never had total incontinence, suggesting a competent bladder neck. However, she had a history of occasional bedwetting and stress incontinence during early childhood. She also had history of cyclical haematuria postpuberty.

At the age of 20 years, she conceived but there was intrauterine fetal death at 6 months of pregnancy for which normal delivery was done. The procedure was uneventful. However, following the procedure, she developed dysuria with stress incontinence for which urological consultation was done.

INVESTIGATIONS

In local examination, the labia majora was ill developed and labia minora was relatively small. There was no urethral opening. On examination under anaesthesia and cystoscopy, bladder neck and vagina were seen to be draining into common channel.

MRI of abdomen and pelvis was done. Length of common channel was more than 3 cm, there was a single left kidney and uterus was bicornuate (figure 1). No other congenital malformation was noted.

TREATMENT

A multidisciplinary team was formed and patient was planned for preservation of vaginal continuity, closure of fistulous opening, creation of a neourethra and make the patient continent. Through transabdominal approach, bladder was bivalved to expose the defect. Bladder was mobilised around the defect and urogenital separation was done.

Figure 1 MRI showing common channel of urethra and vagina.

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Figure 2 Appendix mobilised over its mesentery after appendectomy.

Fistulous opening was closed in two layers after mobilisation. Omentum was mobilised over a pedicled vessel and placed over the defect.

Appendix was identified. It was around 5 cm long and non-inflamed. Appendectomy was done. The appendix was then mobilised with its mesentery, long enough to reach perineum (figure 2). A site for neoexternal urethral meatus was made anterior to vaginal opening and a tunnel was made to reach proximal urethra through pelvic floor. Appendix was taken out over 14 Fr Foley's catheter (figure 3), proximal end anastomosed after spatulation, with bladder neck and distal end merged with labia minora flap to form neourethral meatus. Suprapubic cystostomy was done before closure.

OUTCOME AND FOLLOW-UP

A micturating cystourethrogram done at postoperative day 14 revealed no leak and a patent urethra. The postvoid film showed minimal residual contrast. An ultrasound examination was done, during which the prevoid volume was 180 mL and the postvoid volume was 16 mL.

Patient was taught to do clean self-catheterisation by 14 Fr Foley catheter on a twice-weekly basis. Patient was continent and voiding by herself on discharge. She also had her first proper menstruation 20 days after surgery.

Figure 3 Appendix taken over by 14 fr Foley's catheter.

DISCUSSION

Surgical repair of cloacal malformations has evolved over the years with the advent of more and more reconstructive options.

Early faecal diversion is the rule as single-staged repair is technically difficult. Rectum is generally separated by posterior sagittal anorectoplasty as either a single or two-stage procedure. Although, single-stage repair of both urogenital and anal canal have been reported, urogenital separation is generally performed at later stage. The options for vaginal reconstruction include vaginal pull through, vaginal flap and vaginal replacement with bowel.

Appendix has been used as urinary conduit in long-segment ureteric defects¹ in Mitrofanoff appendicovesicostomy for continent urinary diversion. Submucosa of appendix has been used as ventral onlay in augmented urethroplasty. Appendix has been used as a substitution of urethra in recurrent urethral stricture.²

However, to our knowledge, appendix has not been used as urethral conduit in cloacal repair so far in the published literature. We have used appendix as it provided adequate length as was required in our case. It was non-inflamed and had adequate lumen. Moreover, there was paucity of local tissue in our case to use as local flap.

The early result is promising, however, long-term follow-up and significant number of cases has to be studied before we can come to any conclusion.

Learning points

- ▶ Repair of cloacal malformation is complex and surgically challenging if length of common channel is more than 3 cm.
- ▶ Vermiform appendix can be used as autologous tissue substitute for urethra reconstruction, especially in cases where there is paucity of tissue for local flaps.
- ▶ Routine use of appendix for urethral reconstruction cannot be recommended at this stage. However, it certainly adds to the limited available options.

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Contributors We certify that we have participated adequately in the intellectual content, conception and design of this work as well as the writing of the manuscript. We take public responsibility for the piece of work and have agreed to have our name listed as a contributor. We believe the manuscript represents valid work. Neither this manuscript nor any other manuscript with substantially similar content under our authorship has been published or is being considered for publication elsewhere, except as described in the cover letter. We certify that all the data collected during the study has been presented in this manuscript and that no data from the study has been or will be published separately. We attest that, if requested by the editors, we will provide required information and will cooperate fully in obtaining and providing the information on which the manuscript is based, for examination purposes by the editors or their assignees. We hereby transfer, assign or otherwise convey all copyright ownership, including any and all rights incidental thereto, exclusively to the BMJ case reports, in the event that such work is published by the BMJ case reports. The BMJ case reports shall own the work, including (1) the copyright, (2) the right to grant permission to republish the article in whole or in part, with or without a fee, (3) the right to produce preprints or reprints and translate into languages other than English for sale or free distribution and (4) the right to republish the work in a collection of articles in any other mechanical or electronic format. All mentioned authors were involved in the conception and design, analysis and interpretation of the data. They were involved in drafting the article and revisiting it critically for important intellectual content. DKP, SG and SB were involved in final approval of the version published. All mentioned authors were agreed for accountability of the article and ensure that all questions regarding the accuracy and integrity of the article were investigated and resolved. We give the right to the corresponding author to make necessary changes according to the request of the journal, do the rest of the correspondence on our behalf and act as the guarantor for the manuscript on our behalf. If we do not include an acknowledgement, it means that we have not received substantial

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