



# Posterior urethral valves in a newborn with imperforate anus: clinical presentation and management

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**Abstract** Anorectal malformations are frequently associated with urinary tract abnormalities, which generally consist of vesicoureteral reflux, renal agenesis, and bladder dysfunction. Posterior urethral valves associated with anorectal malformations are exceedingly rare. We report the third case described in literature and the unique management.

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## 1. Case report

A 4011-g male infant was delivered at 37 weeks by cesarean birth to a 24-year-old woman because of the sudden development of oligohydramnios. The neonate was in respiratory distress upon delivery and required intubation for mechanical ventilation. Examination revealed a male neonate with a soft, mobile abdominal mass measuring approximately 9 cm in diameter and an imperforate anus. An abdominal ultrasound revealed a normal urological tract but noted a large mass extending from the pelvis into the right upper quadrant (Fig. 1). The neonate was taken to the operating room, and after a Foley catheter was inserted, he underwent abdominal exploration. Findings included a massively dilated rectosigmoid colon containing yellow watery meconium. He underwent an end sigmoid colostomy and rectal mucus

fistula. A malrotation was noted and a Ladd procedure was performed, but the appendix was left in place for a possible future appendicostomy.

A cystogram was performed postoperatively because the patient had oliguria with a palpable bladder (Fig. 2). This revealed that the Foley catheter had passed through a large rectourethral fistula into the rectum. Because of the inability to drain the bladder, a suprapubic tube was placed at the bedside under fluoroscopic guidance, with excellent return of urine. Posterior urethral valves were suspected. Cystoscopy and fulguration of the posterior urethral valves were performed at 20 days of life. The rectoprostatic fistula was found to be located above the valves (Figs. 3,4). A Foley catheter was placed at that time. The suprapubic tube was removed on day 25. The patient was then discharged home with the Foley catheter in place for 1 week on postoperative day 30.

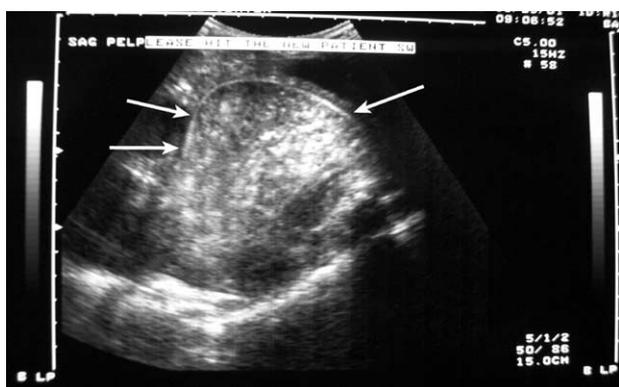
A posterior sagittal anorectoplasty with rectal tapering was performed at 4 1/2 months of age. The colostomy was taken down at 7 months of age, where an 8-cm segment of very dilated distal sigmoid was resected.

A follow-up ultrasound and voiding cystourethrogram at 1 year of age demonstrated normal urological anatomy and no significant postvoid residual (Fig. 5). He is

Posterior urethral valves were encountered in a newborn boy with imperforate anus. This extremely rare combination is presented along with the management and outcome.

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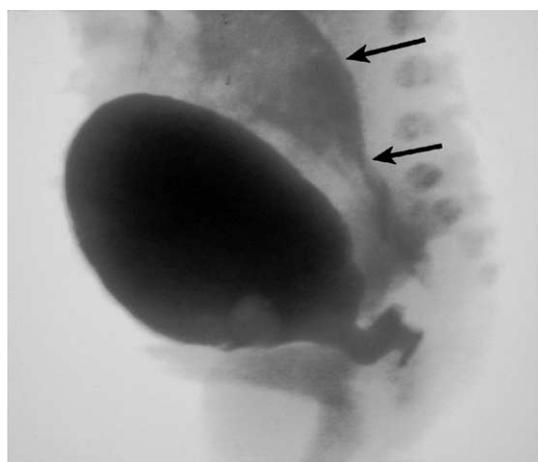
**Fig. 1** Abdominal ultrasound revealing markedly dilated intestinal loop containing heterogeneous material.

undergoing a bowel management program for asymptomatic constipation.

## 2. Discussion

The occurrence of abdominal distension in a child with an anorectal malformation is common. This is usually because of meconium in the distal bowel and is seen in both high and low lesions. However, the patient presented with posterior urethral valves, causing significant retrograde urinary flow through the high rectoprostatic fistula and into the rectum. The dilated rectosigmoid loop was palpable as an abdominopelvic mass and contained urine and meconium as observed during operative exploration. This fortunately responded to treatment because persistent reflux of urine into the colon can cause life-threatening electrolyte imbalances [1].

The sudden decrease in amniotic fluid was a first sign to the existence of a fetal urological abnormality and likely represented a sudden, complete obstruction of the urethra caused by the posterior urethral valves. The



**Fig. 2** Cystogram showing flow of contrast into rectosigmoid colon.

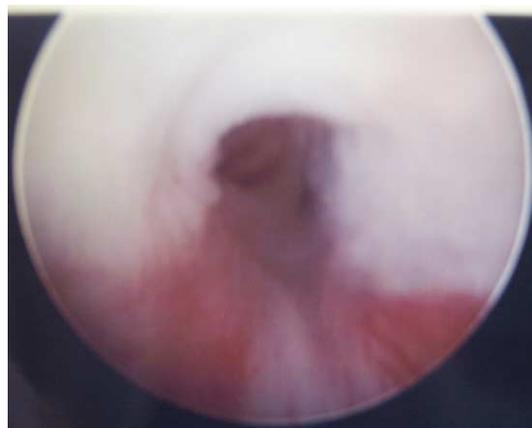


**Fig. 3** Cystoscopy demonstrating occlusion of prostatic urethra with urethral valves.

existence of a path of retrograde urinary flow through the rectoprostatic fistula likely helped prevent hydronephrosis and renal damage.

Urological malformations are seen in approximately 50% of patients with anorectal malformations, depending on the location of the fistula [2]. The common defects include neurogenic bladder dysfunction, vesicoureteral reflux, and renal agenesis. There have been only 2 previous reports of posterior urethral valves occurring with imperforate anus; however, neither describe the clinical picture or management [3,4]. Its presence in this patient likely caused the oligohydramnios, which necessitated the early emergent cesarean delivery.

This neonate's good functional outcome, as measured by his manageable degree of constipation, suggests that his rectal segment maintained reasonable function despite its marked dilation initially. It has been reported that those patients who are found to have large, dilated bowel either before or after primary correction of imperforate anus are



**Fig. 4** Appearance of urethra after fulguration of valves.



**Fig. 5** One-year follow-up cystourethrogram demonstrating normal urological anatomy.

more likely to experience bowel function difficulties [5-8]. This is thought to be because of a functional defect in the dilated segment, and tapering or resection of this area has been recommended, as was done in this case.

The concurrent presentation of imperforate anus and posterior urethral valves is extraordinarily rare. In this case, the posterior urethral valves were diagnosed postoperatively as a result of oliguria leading to bedside cystography. A preoperative voiding cystourethrogram was not performed because of the normal genitourinary ultrasound and the child's requiring mechanical ventilation for respiratory distress. However, if the cystoscopy had been performed

intraoperatively after finding the megarectosigmoid with liquid meconium, the valves could have been diagnosed and treated earlier, thus avoiding any delay and potential electrolyte imbalances.

In a patient who is noted prenatally to have a large cystic abdominal mass, the differential diagnosis should include imperforate anus with rectoprostatic fistula and posterior urethral valves. Oligohydramnios, oliguria, and pulmonary hypoplasia in the setting of imperforate anus may indicate the presence of posterior urethral valves. Finally, if liquid yellow meconium is encountered at the time of colostomy, a genitourinary evaluation including voiding cystourethrography or cystoscopy is indicated for definitive therapy.

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