Case Series

Rare Associations with Posterior Urethral Valves

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Posterior urethral valves are a common cause of congenital bladder outlet obstruction. Known associations include cardiac malformations and gastrointestinal abnormalities. In this case series, we report on two cases of PUV associated with anorectal malformations along with a case of PUV in monochorionic diamniotic twins. We explore the difficulty in achieving a diagnosis and the final management. The association of posterior urethral valves in a patient with anorectal malformation should be suspected in case of associated oligohydramnios or oliguria postnatally. There should be a high index of suspicion in twin pregnancy even if only one of the twins is suspected of bladder outlet obstruction.

1. Introduction

Posterior urethral valves (PUV) are the most common congenital cause of bladder outlet obstruction in children and a leading cause of end-stage renal failure. The estimated incidence of PUV is 1 in 3800 live births in the UK, of which 40% of patients are diagnosed antenatally [1]. Nonurological common anomalies associated with PUV include cardiac malformations, gastrointestinal abnormalities, and aneuploidy in about 40% of cases [2]. We describe 3 cases of PUV with rare associations and their management.

2. Case 1: PUV Associated with Imperforate Anus

A baby boy born at 34 + 6 weeks by emergency C-section weighing 2.735 kg with antenatal scans suggestive of oligohydramnios and dilated bowel loops however with a normal renal tract. At birth, he was found to have an imperforate anus; hence, a sigmoid colostomy was performed.

Postoperatively, he was noticed to have oliguria and worsening renal functions. Interestingly at the same time, the colostomy bag was filling with faeces and watery effluent. Close inspection revealed watery discharge from defunctioning limb of colostomy explaining rising blood urea and creatinine levels and acidosis. We suspected bladder outflow obstruction and regurgitation of urine through the recto urethral fistula. Renal ultrasound scan (USS) showed bilateral hydronephrosis and hydroureters. Perineal USS showed dilated posterior urethra. Attempted urethral catheterisation proved difficult, so a suprapubic catheter was inserted.

Micturating cystourethrogram (MCUG) through suprapubic catheter revealed a dilated posterior urethra and a rectubular fistula (Figure 1). On day 13 of life and after stabilisation of general condition, the patient was taken to theatre for cystoscopic ablation of the valves. The suprapubic catheter was removed, and a urinary catheter was left for one week.

Subsequently, he suffered from persistent nonbilious vomiting along with metabolic alkalosis. Abdominal USS revealed coexisting pyloric stenosis. Laparoscopic pyloromyotomy was performed and made good postoperative recovery. At the age of 4 months, posterior sagittal anorectoplasty (PSARP) with dissection and ligation of the rectubular fistula was done. Under the same anaesthetic, repeat cystoscopy was done to confirm the absence of any residual valves. After regular dilatation and achieving a good size neo-anus, stoma closure was performed at the age of 7 months. On follow-up at 3 years, he is thriving well with improvement of the bilateral hydronephrosis and hydroureter on serial ultrasound scans.
and no history of urinary tract infections (UTI). He has achieved bladder control and has been progressing well with bowel continence.

### 3. Case 2: PUV Associated with Rectoperineal Fistula

This full-term baby boy was born after an uneventful pregnancy and normal antenatal scans. At birth, he was found to have a rectoperineal fistula, accessory nipples, and preauricular skin tags. He underwent primary PSARP on day two of life for the rectoperineal fistula. Echocardiography revealed a patent foramen ovale and tricuspid valve thickening. The rest of his VACTERYL screening—including a renal ultrasound—were normal. Unfortunately, this child later presented with two confirmed episodes of E-coli urinary tract infections in the first 4 months of life. Although a repeat USS did not reveal any anomalies, we proceeded for a MCUG, which showed a dilated posterior urethra suggestive of PUV (Figure 2). Cystoscopic valve ablation was then performed with good postoperative recovery. At the age of 8 years, this child has done well with normal renal function and has achieved both urinary and faecal continence with the aid of regular laxatives.

### 4. Cases 3 and 4: PUV in Monochorionic Twins

We report a case of PUV in monochorionic diamniotic twins born at 36 weeks by Emergency C-section due to breech presentation. Antenatal scans in twin 1 showed bilateral hydroureter and hydronephrosis and oligohydramnios. Postnatal renal USS showed bilateral hydronephrosis with loss of corticomedullary differentiation and a dilated left ureter.

Antenatal scan in twin 2 showed left hydrenephrosis. Postnatal renal USS showed thick wall bladder and bilateral hydronephrosis and hydroureters. MCUG in twin 2 revealed a dilated posterior urethra and grade V left ureteric reflux (Figure 3) While in twin 1 revealed a grossly dilated posterior urethra and a trabeculated urinary bladder (Figure 4). Bladder drainage was achieved using urethral catheters in both twins. Due to the low birth weights of twins and circumstances related to the COVID-19 Pandemic, ablation was performed at 8 weeks of age. Currently, twins are doing well and awaiting check cystoscopy (accepted unit policy in our hospital) and circumcision.
5. Discussion

Posterior urethral valves (PUV) are thought to result from an anomalous insertion of the mesonephric duct into the urogenital sinus, preventing normal migration of these ducts and their anterior fusion forming the abnormal ridges in the membranous urethra [3]. Young’s [4] classification of 3 types of PUV has recently been challenged, and a newer concept was proposed by Dewan and Goh that in bladder outlet obstruction, the uninstrumented urethra looks more like a circumferential obstructing membrane with a small central or eccentric opening named congenital obstructing posterior urethral membrane (COPUM) [5]. Various associations (up to 40%) with PUV have been reported including cardiovascular abnormalities, absent external auditory meatus, bilateral adrenal agenesis, hypospadias, micro/macrophallus, and anterior urethral valves [6–8].

The association of PUV with anorectal malformations is exceedingly rare and can pose a diagnostic and therapeutic challenge. Few reports [6, 9, 10] exist of this association; however, only one report had a presentation similar to our challenge. The data used to support the findings of this study are included within the article.

6. Conclusion

Although exceedingly rare, the association of posterior urethral valves in a patient with anorectal malformation (ARM) should be suspected in case of associated oligohydramnios, postnatal oliguria, or watery effluents in colostomy bag with rising renal function and acidosis. There should also be a high index of suspicion in twin pregnancy even if only one of the twins is suspected of bladder outlet obstruction.

Data Availability

The authors declare that they have no conflicts of interest.

References

[1] E. Brownlee, R. Wragg, A. Robb, H. Chandran, M. Knight, and L. McCarthy, “Current epidemiology and antenatal presentation of posterior urethral valves: outcome of BAPS CASS National Audit,” Journal of Pediatric Surgery, vol. 54, no. 2, pp. 318–321, 2019.
[2] T. T. Chao and J. S. Dashe, Posterior Urethral Valves, Elsevier Inc., Second edition, 2017.
[3] A. Krishnan, A. De Souza, R. Konijeti, and L. S. Baskin, “The anatomy and embryology of posterior urethral valves,” The Journal of Urology, vol. 175, no. 4, pp. 1214–1220, 2006.
[4] H. H. Young, W. A. Frontz, and J. C. Baldwin, “Congenital obstruction of the posterior urethra,” The Journal of urology, vol. 3, no. 5, pp. 289–366, 1919.
[5] P. A. Dewan and D. G. Goh, “Variable expression of the congenital obstructive posterior urethral membrane,” Urology, vol. 45, no. 3, pp. 507–509, 1995.
[6] S. A. Hayden, P. D. Russ, D. H. Pretorius, M. L. Manco-Johnson, and W. H. Clewell, “Posterior urethral obstruction. Prenatal sonographic findings and clinical outcome in fourteen cases,” Journal of Ultrasound in Medicine, vol. 7, no. 7, pp. 371–375, 1988.
[7] J. Carvell and R. Mulik, “A case of hypospadias, anterior and posterior urethral valves,” Journal of surgical case reports, vol. 2013, no. 2, 2013.
[8] K. L. Rao, B. Eradi, and P. Menon, “Anterior and posterior urethral valves: a rare association,” Journal of Pediatric Surgery, vol. 38, no. 7, pp. 1–2, 2003.
[9] C. E. Carlton, F. J. Harberg, and F. M. Fry, “Urologic complications of imperforate anus,” The Journal of Urology, vol. 109, no. 4, pp. 737–739, 1973.
[10] M. P. Singh, A. Haddadin, R. B. Zachary, and D. W. Pilling, “Renal tract disease in imperforate anus,” Journal of Pediatric Surgery, vol. 9, no. 2, pp. 197–202, 1974.
[11] L. M. Burttet, “Posterior urethral valves disorder in non-twin siblings: case report and literature review,” Medical and Surgical Urology, vol. 3, no. 2, 2014.
[12] V. J. Baldwin and V. J. Baldwin, Anomalous development of twins, Pathology of Multiple Pregnancy, 1994.
[13] J. V. Thomalla, M. E. Mitchell, and R. A. Garett, "Posterior urethral valves in siblings," *Urology*, vol. 33, no. 4, pp. 291–294, 1989.

[14] P. M. Livne, J. Delaune, and E. T. Gonzales, "Genetic etiology of posterior urethral valves," *The Journal of Urology*, vol. 130, no. 4, pp. 781–784, 1983.

[15] G. M. Maruotti, A. Agangi, P. Martinelli, and D. Paladini, "Early prenatal diagnosis of concordant posterior urethral valves in male monochorionic twins," *Prenatal Diagnosis*, vol. 26, no. 1, pp. 67–70, 2006.

[16] F. Morini, M. Ilari, A. Casati, A. Piserà, L. Oriolo, and D. A. Cozzi, "Posterior urethral valves and mirror image anomalies in monozygotic twins," *American Journal of Medical Genetics*, vol. 111, no. 2, pp. 210–212, 2002.

[17] A. O. Talabi, O. A. Sowande, O. Adejuyigbe, J. A. Owa, and A. O. Igwe, "Posterior urethral valves in fraternal twins," *Annals of Pediatric Surgery*, vol. 14, no. 3, pp. 190–191, 2018.