Posterior urethral valves in fraternal twins: case report and review of the literature

Ademola O. Talabi[a], Oludayo A. Sowande[a], Olusanya Adejuyigbe[a], Joshua A. Owa[b] and Arua O. Igwe[a]

We report a pair of dizygotic twins with type 1 posterior urethral valves. Concomitant and discordant presentation of posterior urethral valves in twins is rare, with a handful of cases in the literature. The occurrence of identical pathology in dizygotic twins probably suggests the possibility of an inherited trait as it does exist in monozygotic twins. *Ann Pediatr Surg* 14:190–191 © 2018 Annals of Pediatric Surgery.

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Departments of [a]Surgery and [b]Paediatric and Child Health, Obafemi Awolowo University Teaching Hospitals Complex, Ile-Ife, Osun State, Nigeria

Correspondence to Ademola O. Talabi, MBBCh, FWACS, Department of Surgery, Obafemi Awolowo University Teaching Hospitals Complex, Ile-Ife, Osun State 2005, Nigeria
Tel: + 234 803 346 9206; fax: + 234 802 243 1901;
e-mail: ademolatalabi1608@gmail.com

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Introduction

Posterior urethral valves (PUV) are the commonest cause of bladder outlet obstruction in male neonates. The back pressure effect of bladder obstruction may lead to progressive deterioration of upper urinary tract. It has been postulated that ~50% of children with PUV develop end-stage renal disease in 10 years [1,2]. The etiology of PUV is unknown. Some authors suggest that these valves result from abnormal insertion of the mesonephric duct into the urogenital sinus [3,4]. The association between monozygotic twins, nontwin siblings, and PUV has been described in literature. Although there is no established genetic link to this anomaly, it has been postulated that an autosomal recessive pattern of inheritance possibly exists from genetic study of families with affected siblings [1]. There is paucity of reports on the diagnosis of PUV in nonidentical twins. Here, we describe a case of PUV in a set of dizygotic twins who presented with varying degree of bladder outlet obstruction and renal impairment.

Case report

A set of twin boys was born via repeat cesarean delivery to a 28-year-old gravida 2 para 1 woman. They were dichorionic and diamniotic twins delivered at 37 weeks of gestation. The first twin (twin 1) weighed 3.1 kg at birth whereas the second twin (twin 2) weighed 2.8 kg.

Twin 1 initially appeared to be normal and said to have passed urine with a good urinary stream until 7 days after birth when he developed high-grade fever, refused to tolerate feed, and passed bloody urine. There was no bleeding in any other part of the body. Prenatal ultrasound screening did not detect PUV. Baby was not pale, was anicteric, was well hydrated, but febrile (temperature was 38.8°C). There was a palpable suprapubic mass, and both kidneys were ballotable. No Potter's facie was seen. He was placed on continuous bladder drainage with urethral catheter which led to decrease in suprapubic distension. Abdominal ultrasound showed bilateral hydronephrosis, dilated upper urinary system, and significant bladder wall thickening. An intravenous urogram with a micturating phase confirmed PUV with bilateral vesicoureteric reflux worse on the right. Preoperative packed cell volume and serum chemistry findings were normal. Urinalysis showed macroscopic hematuria and reduced specific gravity. Urine culture yielded no growth. At 16th day of life, he had valvotomy using a Mohan's valvotome done at 3, 12, 9, and 6 o'clock positions. Urethral catheter was removed at postoperative day 4, and the patient later passed urine with normal stream. One month after surgery, the patient presented with features of urinary sepsis necessitating admission and use of parenteral antibiotics. Patient was monitored with serial abdominal ultrasound scans, which showed sustained improvement of left upper urinary tract dilatation with, however, worsening dilatation and hydronephrosis on the right. A micturating cystourethrogram showed a right grade V vesicoureteric reflux. He subsequently had a right loop ureterostomy with adequate right renal decompression. Patient has had frequent clinic visits, and serial abdominal ultrasound scans show sustained improvement of both kidneys and good corticomedulary differentiation. Intravenous urogram and serum chemistry showed prompt excretion in both kidneys and good renal function, respectively.

Twin 2 was suspected to have bladder outlet obstruction secondary to PUV following a prenatal ultrasound done at 35 weeks of gestation. This necessitated urethral catheterization at birth. A repeat abdominal ultrasound at delivery showed a distended urinary bladder with thickened (0.3 cm) and trabeculated bladder wall. There were bilateral ureteric (0.11 cm diameter), pelvis, and calyceal dilatations with bilateral hydronephrosis. Preoperative packed cell volume, serum chemistry, and urinalysis findings were normal. Urine culture yielded *Escherichia coli* sensitive to cefuroxime, and he was place on parenteral antibiotic. He subsequently had valvotomy done as described for twin 1. Urethral catheter was removed at postoperative day 4, and patient had good urinary stream. Micturating cystourethrogram done 3 months later showed normal smooth bladder wall, with no evidence of reflux. Patient has since had several clinic visits. Serial serum chemistry and intravenous urogram findings were normal with good somatic growth.

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Discussion

PUV is increasingly being detected by routine prenatal ultrasound in most developed nations. The characteristic findings are bilateral hydronephrosis, a distended bladder, and a dilated posterior urethra (keyhole sign) [1]. Prenatal ultrasound diagnosis and fetal screening in general are not routinely employed in our clime in spite of its availability because of misconception of our patients and lack of thorough fetal scanning on the part of the sinologist [5,6]. In this set of twins, prenatal ultrasound was done to assess gestational age, fetal development and sex. The detection of PUV in one of the twins was an incidental finding, and as such, it was not surprising that the diagnosis was missed in the other twin. Unfortunately, the possibility of identical pathology in the first twin was not considered at birth until he became symptomatic and investigated. The relative delay in the diagnosis of this anomaly in the first twin has underscored the importance of high index of suspicion among twins and siblings of individual with PUV, and routine prenatal screening of all fetuses for possible congenital malformation.

Early diagnosis of PUV is necessary for reducing associated anatomical and functional (compliance) defect of the bladder and upper urinary tracts because renal impairment and other complications begin to develop as early as the antenatal period [1]. In the index case report, this set of twins was detected and treated early in the perinatal period and has been doing well for almost 3 years since birth, though the first twin had severe hydroureter which necessitated loop ureterostomy.

There are few reports in English literature on familial PUV, and in particular on dizygotic twins. Burttet *et al.* [1] in an extensive review of literature found 12 cases of familial nontwin sibling with PUV. Christman *et al.* [7] reported two sets of discordant twin siblings (the first set being dizygotic and the second being monozygotic) in which only one of each pair presented late with PUV. Other researchers [3,4] documented the occurrence of PUV in monozygotic twins. The association of PUV with twins and nontwin siblings has not been elucidated. Some authors opined that the high concordance rate of PUV in monozygotic twins has been taken as evidence for a genetic predisposition to this anomaly [3,4,8]. We also hypothesized that the occurrence of PUV in dizygotic twins may probably suggest the possibility of a genetic trait as does exist with monozygotic and nontwin siblings.

We recommend urologic evaluation of the male sibling and family member of a male child with PUV.

Conflicts of interest

There are no conflicts of interest.

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