

PROTECTIVE FACTORS IN POSTERIOR URETHRAL VALVES IN SAUDI CHILDREN

Khalid Mirza, MD; Vincent Onuora, FRCS; Abdulmohsen Al-Sowailem, FACHARTZ

Posterior urethral valves are the most common cause of bladder obstruction in boys. Clinical presentation may vary, depending upon the degree of obstruction and its sequelae. Pathological changes range from relatively normal upper tracts or a small amount of dilatation to a massive dilatation associated with severe infection or sepsis, azotemia and/or renal dysplasia. In our population they constitute about 17% of chronic renal failure in children.¹ The development of renal insufficiency in patients with posterior urethral valves may be attributed to the high pressure generated by urethral obstruction transmitted to the upper tracts in utero. In this review, four cases of posterior urethral valves are described, in which we identified certain protective factors which provided a "pop-off" mechanism that resulted in preservation of intact renal and bladder function.

Patients and Methods

Case 1

This infant was born with bilateral abdominal masses and mild respiratory distress. Antenatal ultrasound showed bilateral hydronephrosis and oligohydramnios. The initial serum creatinine was 200 $\mu\text{mol/L}$. Ultrasonogram showed bilateral hydronephrosis. A large collection of fluid was seen around the superior pole of the right kidney, measuring 4.4 x 3.1 cm. A similarly large collection was seen around the left kidney. A micturating cystourethrogram (MCUG) showed multiple small bladder diverticulae, a long dilated posterior urethra and a right Grade V vesicoureteric reflux (VUR). A DMSA scan showed 6% function in the right kidney and 94% in the left kidney. The baby underwent cystourethroscopy, vesicostomy and drainage of both perinephric urinomas. The serum creatinine reached 75 $\mu\text{mol/L}$. At the age of two years the vesicostomy was closed and the valve fulgurated. Cystometry showed a normal bladder capacity. Subsequently he underwent a right nephroureterectomy.

At his last follow-up at the age of three years, his serum creatinine was 40 $\mu\text{mol/L}$.

Case 2

A 45-day-old infant was referred to us with failure to gain weight from birth and repeated vomiting of one week's duration. The abdomen was distended, with well-defined masses palpable in both loin regions. The initial serum creatinine was 190 $\mu\text{mol/L}$. A sonogram showed hydronephrotic kidneys with hyperechoic parenchyma. A well-defined urinoma was seen on the anteriomedial aspect of the right kidney (2.6 x 1.1 cm). A cystogram showed multiple diverticulae of varying size, a right Grade IV VUR and a dilated posterior urethra. A DMSA scan showed 7% function in the right kidney and 93% in the left kidney. Cystoscopy showed a Type III posterior urethral valve (PUV). A vesicostomy and drainage of the urinoma by a perinephric drain were performed. On the 10th day after admission, the infant's serum creatinine dropped to 54 $\mu\text{mol/L}$. A repeat ultrasound showed that the urinoma had regressed in size. The vesicostomy was closed and the valve fulgurated. Cystometry showed normal findings for the age. Ultimately, a right nephroureterectomy was performed. At his last follow up at the age of two years, his creatinine was 38 $\mu\text{mol/L}$.

Case 3

A 23-day-old neonate was referred from a peripheral hospital with the complaints of a weak stream of urine, poor feeding, lethargy and abdominal distention. Antenatal ultrasound showed moderate left hydronephrosis. The serum creatinine was 270 $\mu\text{mol/L}$. Ultrasound showed an enlarged left kidney with gross hydropelvis and big urinoma measuring 3.7 cm x 2.5 cm on the posterior superior aspect (Figure 1). A cystogram revealed a left Grade IV VUR, a large urinoma and a prominent posterior urethra. A DMSA scan showed a differential function of 5.6% in the left kidney and of 94.4% in the right kidney. Cystoscopy confirmed PUV. A left retrograde ureteropyelogram showed grossly dilated upper calyces and a collection around the middle calyces (urinoma). A vesicostomy was performed. The serum creatinine reached 52 $\mu\text{mol/L}$. At the age of one year the vesicostomy was closed and the valve fulgurated. Cystometry showed

From the Department of Pediatric Nephrology, Riyadh Medical Complex, Riyadh, Saudi Arabia.

Address reprint requests and correspondence to Dr. Mirza: P.O. Box 9396, Riyadh 11413, Saudi Arabia.

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FIGURE 1. Ultrasound of the left kidney with moderate hydronephrosis and a large perirenal urinoma.

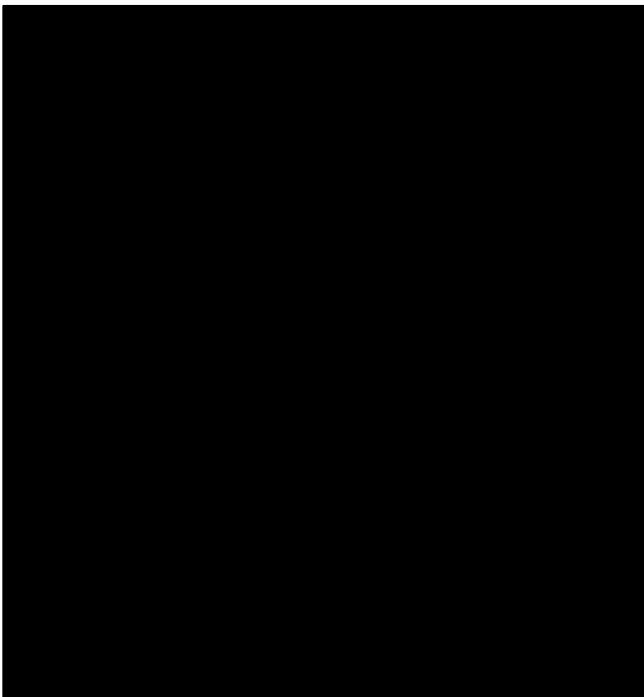


FIGURE 2. MCUG demonstrating a grossly trabeculated bladder and a posterior diverticulum.

normal bladder capacity. At a later stage, a left nephro-ureterectomy was performed. At the age of two years his serum creatinine was 30 $\mu\text{mol/L}$.

Case 4

A nine-month-old boy presented to us with painful dribbling of urine from birth. Abdominal examination

revealed a suprapubic mass. Investigations revealed a serum creatinine of 97 $\mu\text{mol/L}$. A sonogram showed enlarged kidneys (8.2 cm) with hyperechoic parenchyma, dilated pelvicalyceal systems, dilated and tortuous ureters and a thick-walled urinary bladder with a large diverticulum posterior to the bladder. A cystogram showed multiple small diverticulae and a huge diverticulum arising from the base of the bladder (Figure 2). The posterior urethra was grossly dilated and lengthened. No evidence of VUR was seen. A renal DMSA scan showed a differential function of 52% in the right kidney and 48% in the left kidney. The patient underwent valve fulguration and subsequently diverticulectomy. His serum creatinine reached 52 $\mu\text{mol/L}$ and subsequently remained normal.

Discussion

Three anatomical variables have been described in posterior urethral valves which might provide a "pop-off" mechanism: 1) Urinoma/urinary ascites presenting in the newborn period as respiratory distress. 2) The syndrome of posterior urethral valves, unilateral vesico-urethral reflux and renal dysplasia (VURD syndrome), in which the contralateral renal function is maintained in association with reflux into an ipsilateral and usually non-functioning kidney, often exhibiting dysplastic histology. 3) Posterior urethral valves associated with large congenital diverticulae.

Urinary extravasation resulting in either urinary ascites or an isolated urinoma has been described primarily in the neonatal period, and most commonly presents in male infants with posterior urethral valves.² Urinoma formation may be seen in 3%-17% of neonates with PUV.^{3,4} Extravasation presumably results from increased pressure within the obstructed fetal urinary tract, with the rupture of the urinary collecting system, usually at a calyceal fornix. There may be subsequent drainage of the urinoma into the peritoneal cavity, resulting in ascites. It has been speculated that a urinary leak early in gestation with relief of pressure in the early stages of renal development could result in a lesser degree of renal dysplasia and atrophy. In some of these children the respiratory problems due to urinary ascites or urinoma have been wrongly attributed to pulmonary hypoplasia caused by renal obstruction or hypoplasia. Because of the dramatic reversal of respiratory symptoms, immediate aspiration or drainage of urinomas is suggested. In the opinion of Adzick et al.,⁵ however, accumulation into a localized urinoma often provided less adequate decompression and therefore, had a variable effect on renal preservation. On the other hand, Rittenberg et al.⁴ reported well-preserved renal function in three boys with localized urinomas. Our patients had urinomas in addition to VURD, and the beneficial effect of either could not be determined separately. However, it would appear that any factor that provides a release of intra-renal pressure would contribute to preservation of renal function.

In the VURD syndrome characterized by Hoover and Duckett,⁶ and subsequently by Greenfield and associates,⁷ decompression of the obstructed urinary tract occurs via unilateral reflux into a severely dilated and usually non-functioning renal unit. Reflux occurs most commonly on the left side and the associated renal unit is dysplastic. The other non-refluxing kidney typically exhibits good function. Correction of obstruction and excision of the non-functioning reservoir led to normal long-term renal function in all four cases.

Large bladder diverticulae may provide a third type of "pop-off" valve. Bladder diverticulae may be congenital or acquired. Acquired diverticulae are secondary to either functional or anatomical outflow obstruction. They usually are multiple and small, and generally associated with bladder trabeculation. Congenital diverticula typically are few in number, and they are not associated with bladder outlet obstruction. Johnston recognized the "safety valve action" afforded by large congenital diverticulae associated with obstruction, and cautioned that diverticulectomy without simultaneous relief of obstruction may be followed by progressive hydro-ureteronephrosis.⁸

The presence of a pressure "pop-off" is also a favorable sign for ultimate bladder function in boys with posterior urethral valves. This was determined in a recent review.⁹ All our cases showed normal bladder capacity, thereby indicating that these pressure "pop-offs" also helped in the preservation of intact bladder function in our patients.

In conclusion, in all four patients with posterior urethral valves, the renal and bladder function was

apparently protected by coexistent deflationary mechanisms. These were the urinomas, reflux into a functionless kidney and a bladder diverticulum. Thus a thorough evaluation is needed to identify these protective factors in children born with posterior urethral valves.

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