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Posterior urethral valves: the scenario in a developing center

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Abstract We have reviewed 233 patients with posterior urethral valves treated in a single center in Calcutta, India, over the last 20 years: 37 were neonates, 75 were between 1 and 12 months, 88 were between 1 and 5 years, and 33 were more than 5 years old when first seen. The clinical presentation and methods employed in diagnosis and assessment are described. Primary endoscopic valve ablation was performed in 140 patients (60%). One or other form of diversion was done in 100 (43%), 93 before and 7 during or after valve ablation. The short- and long-term results have been studied. Eleven patients died during the initial hospitalization, 3 died subsequently, 15 are in end-stage renal disease, 17 are in poor health, and 18 have been totally lost to follow-up. The remaining 169 have been in good health for periods between 1 and 20 years. While our results of primary valve ablation in low-risk patients with responsible parents are as good as anywhere else in the world, we are concerned at our relatively high diversion rate and relatively poor long-term follow up; the methods being adopted to reduce these problems are discussed.

Keywords Posterior urethral valves · Bladder outlet obstruction

Introduction

Although the fundamental principles of diagnosis and management of posterior urethral valves (PUV) are now generally accepted [1–3], there are considerable differences in details from center to center. The topic has been discussed at length in 1990 [4–8] in one journal and again in 1997 [9–14] in another. We propose to highlight how practice in our center has changed over the years and how our current practice differs from that in developed centers.

Materials and methods

We studied the records of 233 patients operated upon in a single center in Calcutta over a period of 20 years, 1979 to 1999. In the first decade the first author alone treated 45 patients; in the second 188 patients were treated by all of us. Patients were referred from the city of Calcutta, all other parts of the state of West Bengal, adjacent Indian States, and adjacent neighboring countries, namely, Bangladesh, Bhutan, and Nepal. We have not included in this study patients who were seen but not admitted or admitted but not treated surgically either because consent for surgical treatment was withheld or because they died before surgical treatment could be offered. The age at first presentation is shown in Table 1.

Reasons for surgical consultation included: (1) frequency of micturition, dribbling, and straining, present in almost all cases; (2) recurrent attacks of urinary tract infection (UTI), present in about two-thirds, most of them after failure of antibacterial therapy; (3) hematuria, present in 11 cases; (4) retention of urine, either unrelied or relieved elsewhere by a catheter in 17; (5) generalized massive abdominal distension due to ascites in 9; (6) palpable kidney or kidneys with a palpable bladder in 13; (7) renal failure, respiratory distress, and septicemia in 37; and (8) renal rickets in 1. The lesion was suspected on prenatal ultrasonography (US) in only 5 patients, and even these were referred after symptoms appeared.

Abdominal US became widely available from 1988, and this was the most important reason for a four-fold increase in referrals in the second decade compared to the first. In the second decade, the vast majority of patients came with an ultrasonogram showing a distended, thick-walled bladder, bilateral ureterohydrouphrosis (UHN), and a dilated posterior urethra. However, occasionally massive unilateral UHN and a normal contralateral system or an empty bladder with massive bilateral UHN was seen.

The diagnosis was generally confirmed by a voiding cystogram (VCG) with contrast instilled through a urethral catheter; the path of the dye was studied while the patient micturated or suprapubic compression was applied. In the vast majority of patients the dilated posterior urethra was delineated and the presence or absence of vesicoureteral reflux established. However, the posterior urethra did not fill in a few patients with massive reflux and the diagnosis could only be confirmed later. Dye was instilled by suprapubic puncture only if we failed to pass a urethral catheter into the bladder. Prior to instillation of contrast, a specimen of urine was

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collected to study the specific gravity, cell profile, and types of bacteria if any together with colony count and antibiotic sensitivity.

Renal function on presentation was assessed by estimation of blood urea and creatinine, measurement of renal cortical thickness on US, renal scintigrams using technetium 99 m Tc DPTA, and in the days before isotopes were readily available, IV urograms (IVU). In the majority of patients, renal function was either adequate at the time of admission or became adequate after relatively simple therapy. However, azotemia persisted in 17 of the 37 patients who were admitted with respiratory distress and septicemia.

We performed transurethral endoscopic valve ablation without prior diversion in 140 patients; 21 were neonates, 44 were between 1 and 12 months, 57 were between 1 and 5 years, and 18 were over 5 years of age. Selection was based on urethral size and state of health assessed after a short period of bladder decompression through a urethral catheter, IV fluid therapy, and IV antibiotics. Primary ablation was performed if a suitable endoscope was available and if there was clinical improvement with a fall in blood urea and creatinine levels, disappearance of white cells and bacteria in the urine, good corticomedullary differentiation on US, and reasonable renal function on either scintigraphy or IVU. With improved sonographic equipment, we have dispensed with these last two tests preoperatively in neonates.

The smallest endoscope available in the early years of the study was a no. 10 Storz, and hence, neonates and small infants whose urethras did not permit the passage of this instrument were initially diverted; we did not perform a perineal urethrostomy. In 1984 we acquired a no. 8 Storz endoscope and most neonates became eligible for primary fulguration. Fulguration was done by the method described by Hendren [15]. In 1992 no. 10 Storz pediatric resectoscopes and in the following year a no. 7 Wolff endoscope and a no. 9 Wolff resectoscope arrived. At this point, some of us switched over to transurethral resection (TUR) even in neonates.

After valve ablation, we left an indwelling catheter for a few days and the patients went home after they passed urine normally. Antibiotics were continued for a variable period of time, depending on the results of urine examinations. US was repeated usually within 3 months, earlier if necessary, and renal scintigraphy or IVU was done at a convenient time. A VCG was repeated in patients who had reflux or persisting dysuria or UTI. The importance of clinical assessment and appropriate investigations at regular intervals was explained to every family before their patient left the clinic.

Results

Two patients died; both were neonates treated in 1984 just after we had acquired a no. 8 endoscope. In retrospect, we had misjudged their fitness for primary ablation. Diversion was performed concurrently in 3 patients and subsequent to ablation in 4, for reasons that will be discussed presently; all survived. Initial ablation was found to be incomplete in some patients and was completed at the second or third attempt. Revisions were more often necessary in neonates and older patients whose valves had been fulgurated, not resected.

Nephroureterectomy had to be performed prior to valve ablation in 2 patients. Both had grade 5 reflux, 1 had pyonephrosis with septicemia, and the other was found to have a totally dysplastic kidney during attempted diversion. Both did well subsequently. Twelve ureteroneocystostomies (UNC) were done after valve ablation in 9 patients; there was persistent grade 5 reflux with UTI in 7 and persistent unilateral ureterovesical obstruction in 2.

Of the 23 patients alive after primary valve ablation in the first decade, 4 were lost to follow-up soon after. Fourteen were well when last seen at periods varying from 1 to 10 years after ablation, but only 5 responded to a request to report for this study. Of the 115 patients treated in the second decade, 15 have been lost to follow-up.

Five patients, including 1 with prune-belly syndrome and 1 who had bilateral UNC, are known to have gone into end-stage renal disease; 3 were advised surgery on their ureters but the families did not give consent. Most of the others, including 8 who had UNC, lead normal lives; however, a few have dysuria, frequency, hesitancy, mild incontinence, and recurrent episodes of UTI. Most families agreed to give antibiotics for infection, but only one-half would consent to periodic US, VCG, isotope scans, or urodynamic studies and even fewer for check cystoscopies or surgery for reflux. The patient who presented with renal rickets is doing well with long-term alpha-calciolod therapy. In recent months, some urodynamic studies have been carried out; these have helped us to rationalize our pharmacologic manipulation and surgical management of persistent frequency, dribbling, and reflux.

Diversion

One hundred patients (43%) had urinary diversion; 88 (38%) were considered unfit for valve ablation either because of urethral size or because they had persisting and fulminating infection, uncontrolled azotemia, or massive reflux and were primarily diverted by us. As mentioned earlier, 7 were diverted during or after valve ablation. The remaining 5 were referred after diversion elsewhere. Percutaneous nephrostomy, vesicostomy, and ureterostomy were the three forms of diversion performed.

Nephrostomy

Percutaneous nephrostomy was done in 2 infants and 1 8-year-old for large pyonephrotic masses in the right loin. The 1st improved rapidly and had valve ablation soon after; he is now 11, and is doing well. The 2nd had ascites and azotemia, and a vesicostomy was done in addition; subsequently valve ablation was done and the vesicostomy was closed. He is now 9 and is doing well. The 3rd had bilateral reflux and azotemia, and a ureterostomy was done on the left side. He improved rapidly and has recently had valve ablation.