

POSTERIOR URETHRAL VALVES: A REPORT BASED ON 10 PATIENTS

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Bladder outflow obstruction is a well recognised problem in the newborn, infants and children. In Pakistan a large number of these patients suffer from stone in the urinary bladder which are treated by vesicolithotomy. However a group develops outflow obstruction due to a number of other causes and posterior urethral valves in the male is one such cause.

Posterior urethral valves are congenital mucosal folds originating at the distal end of verumontanum extending obliquely on either side of the prostatic urethra. Developmental anatomy has been a controversial subject (Young, 1972).

Functionally these are one way valves producing varying degrees of outflow obstruction with understandable devastating clinical implications.

These valves were first described by Young et al (1919). Their original observations were based on 5 postmortem cases and the types described by them are not valid today. Since their description posterior urethral valves were considered as rare entities. Marion's disease was thought to be the main cause of bladder outflow obstruction. With the introduction of wide angle fiber optics Marion's disease is rarely diagnosed and valves are considered to be the commonest form of outflow obstruction in children (Williams, 1968).

In Pakistan our attention was drawn to the urinary problems of pediatric age group when we acquired the pediatric fiber optic resectoscope. In this paper we shall highlight the problems of management of patients with posterior urethral valves in Pakistani setting. A plea for early recognition and treatment has been made.

Material and Methods

This study includes patients referred to the Department of Urology, Mayo Hospital, Lahore, over a period of one year (January 1977-78). During this period 10 cases of posterior urethral valves were diagnosed.

Following routine documentation, clinical examination and laboratory investigations, 8 of the patients underwent micturating cystography. Dilatation of the prostatic urethra was considered as diagnostic for the presence of the valves.

All patients were subjected to cystourethro-scopy and the presence of the valves was confirmed.

Results

A total of 10 male patients were documented. Their age ranged from 22 days to 12 years. Five patients were less than one year of age on admission.

All mothers reported urinary flow problems in children immediately after birth. In older children episodes of retention and difficulty of micturition were significant. Two children subsequently developed retention with overflow. Bouts of fever though reported were not a common feature.

Palpable urinary bladder was an important finding in 8 patients and in 4 bilateral renal enlargement was also significant.

Five patients had blood urea over 45 mg% of whom 2 had levels of over 100 mg%.

Micturating cystography done in 8 patients was diagnostic. The dilated onion shaped prostatic urethra

was found in all cases. Urinary bladder was enlarged showing marked hypertrophy and diverticula. In 4 patients marked bilateral third degree reflux with gross dilatation of the ureters was a prominent feature. IVP done in one patient showed similar findings.

On cystoscopy 7 patients had type I and 3 type III valves. All ten valves were resected with the pediatric resectoscope. In 3 patients complete flow of urine was not achieved and postoperative micturating cystograms were done. The results of these in one was normal, one showed stricture urethra at the perineal urethrotomy and one had residual valves which were resected.

Followup over a period of 2 years showed 4 deaths, of whom 2 patients died in the immediate postoperative period, one after 6 months and one 2 years later. Six patients had good results and are symptom free to date.

Discussion

Posterior urethral valves are developmental anomalies of the prostaticomembranous urethra in males resulting in outflow obstruction. The degree of obstruction varies in individual cases and determines the final outcome.

At birth failure to pass urine for more than 10 hours should arouse suspicion. Hydronephrosis, reported in one of our patients, is usually found in severe cases. The child is usually still born with associated congenital malformations and dysplastic kidneys which cannot sustain life (William, 1968).

In infancy diagnosis can be extremely difficult. Loss of weight, unexplained fever, failure to thrive and bouts of diarrhoea are significant findings. Palpable urinary bladder and kidneys in these cases give a clue. Perirenal urinary leakage and urinary ascitis are recognised complications distorting the clinical picture (Mooney et al., 1975; Scott, 1976).

In children apart from signs and symptoms of chronic renal failure, recurrent urinary tract infection and micturition problems become prominent. Hypertension which settles after valves resection has been commonly noted (Evins and Lorenzo, 1979). Diagnosis at this stage is relatively easy. However the delay in diagnosis may lead to extensive renal damage.

The ultimate outcome of these patients depends upon the following factors:-

- (a). Degree of obstruction
- (b). Duration of obstruction
- (c). Urinary tract infection
- (d). Ureteric reflux
- (e). Other congenital malformations

Mild cases have progressed to adult life with minimal symptoms (Martin et al., 1977) but these are rare. The diagnosis is based on micturating cystogram findings. These are dramatic in severe cases. The signs of back pressure in the prostatic urethra, bladder, ureters and the kidneys are prominent (figures 1, 2 and 3).



Fig. 1. Micturating cystourethrogram showing marked dilatation of the prostatic urethra.



Fig. 2. Micturating cystogram showing posterior urethral valve obstruction with severe back pressure. Urinary bladder has diverticulae and severe reflux of the right ureter is prominent.



Fig. 3. Third degree of reflux and bladder distention in a case of posterior urethral valve.

In mild cases the findings may be equivocal and urethroscopy is mandatory to confirm the diagnosis. Valve resection has been the main objective of the management schedule. Valves resection with pediatric resectoscope is generally the accepted mode of treatment today. This requires a degree of expertise since normal mucosal folds can be confused with the valves. The extent of resection is also

important since the urethra at this age is very thin and rupture is a possibility. In infants the resectoscope has to be introduced through the perineum and subsequent stricture formation can occur (one patient in the present series developed stricture urethra). Incontinence and higher rates of stricture formation has been reported by other workers.

Valve resection may be incomplete, as in one case in our series, depending upon the experience of the surgeon. Kimbrough and Wyker (1977) have shown that compression cystourethrogram immediately after resection will be able to indicate the completeness of resection. This unfortunately was not possible in our series.

In Pakistan, with lack of pediatric Uro-logical facilities, the management of these cases needs to be reevaluated. Valve resection through the bladder is not possible. V-Y plasty and transpubic approach have been tried but failed to give adequate results. Hook resection (Will-Iiams et al., 1973) and rupture of the valves with distended Foley's catheter (Kalicinsky et al., 1978) seem to be worth a trial but fluoroscopic control may not be available in most Pakistani hospitals.

Before we acquired the resectoscope set we operated on two valve patients (not included in this series) through the perineum. The bulbous urethra was opened and a nasal speculum was introduced. The prostatomembranous urethra was then maximally dilated rupturing the urethral valves. Perhaps this may be the ideal approach in Pakistan for the treatment of this problem and needs further work.

In the present series 50 percent of the patients were admitted with renal failure. Bilateral 3rd degree reflux and recurrent urinary tract infection were prominent sequelae of obstruction. It was earlier felt that urinary diversion in the form of bilateral nephrostomies, cutaneous ureterostomy and rarely cystostomy were necessary to improve renal function before undertaking valve resection. Cystostomy, as we have experienced, is a poor form of drainage and open to severe urinary tract infection is thus contraindicated. Pinto and associates (1978) favour cutaneous urethroostomy but it appears that this was in response to poor results of primary valve resection in their series of 6 children. We feel that primary resection is simple and gives adequate results. Kalicinsky et al (1978) and Evins and Lorenzo (1979) hold the same view.

The 3rd degree bilateral reflux seen in 4 of our patients carried poor prognosis and proved to be the most difficult management problem. Control of urinary tract infection was almost impossible in 2 patients. Antibiotic therapy with deteriorating renal function and weekly followup proved difficult. Reflux following relief of obstruction usually cures itself but in patients with persistent reflux and dilated ureters reflux curing operations do-not improve the results (Johnston, 1979).

Four deaths reported in this series are really not very encouraging. We feel that the unnecessarily delayed diagnosis leading to severe renal damage was the main reason for poor results.

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