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POSTERIOR URETHRAL VALVES IN CHILDHOOD: EXPERIENCE IN A CENTER WITH SCARCE FACILITIES

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Objective: Posterior urethral valves (PUV) are the most common congenital causes of lower urinary tract obstruction in male children, but few cases have been reported from Nigeria. In this study we describe our 7-year experience of management of PUV in children in order to increase the awareness of this condition in our environment.

Patients and Methods: This is a report of 41 consecutive children with PUV who were managed at the Jos University Teaching Hospitals (JUTH), Jos, Nigeria, from June 2000 to April 2006. Their age at presentation ranged from 2 days to 15 years (mean: 2.5 years). The relevant clinical, laboratory and radiological data were entered into a database and analyzed.

Results: Twenty-eight patients presented with a condition highly suspicious of PUV, while 7 patients presented with other urological conditions. Six patients presented with non-urological symptoms which caused a delay in diagnosis and institution of treatment. Voiding cystourethrography was diagnostic in all cases. In the majority of patients (n=31), management consisted of transurethral balloon avulsion of the valves yielding a satisfactory outcome in over 80% of the patients. Urethral stricture was the main complication (which responded to serial dilatation) and occurred in 3 patients. The postoperative mortality rate was 2.6%.

Conclusion: Although this study was restricted to one hospital, there appears to be a high incidence of PUV in children in North Central Nigeria. An increased awareness of varied clinical features, a high index of suspicion and simple conservative treatment by balloon avulsion of PUV would improve the outcome.

Key Words: posterior urethral valves, children, treatment, lack of facilities.

INTRODUCTION

Posterior urethral valves (PUV) are the most common congenital cause of lower urinary tract obstruction in male children. Lower urinary tract obstruction secondary to PUV affects the entire urinary tract, but the severity and reversibility of the changes are variable. Much has been written on PUV in developed countries, but in developing countries reports are rare[1,2]. An increased awareness of this condition has considerably improved the prognosis in developed countries[3-8], but the experience in developing countries is still characterized by high morbidity caused by delayed presentation, inadequate facilities for investigation and treatment[2]. We herein report our 7-year experience of management of PUV in Nigerian children in order to increase the awareness of the condition. Aspects of presentation, management and the special problems encountered by urologists in developing countries with limited facilities are highlighted.

PATIENTS AND METHODS

This study reports the management of 41 consecutive children with PUV seen at the Jos

University Teaching Hospitals (JUTH), Jos, Nigeria, between June 2000 and April 2006 with an overall incidence of approximately 6 patients per year. Located in the North Central region of Nigeria, JUTH functions as a primary, referral and teaching hospital with a wide catchment area, serving a population of over 30 million people. Most of the patients were referred from other health centers or district hospitals within the North Central region of Nigeria.

Nineteen patients presented within the first 4 years of the study (approximately 5 patients per year), while 22 were seen during the last 3 years (approximately 7 patients a year), showing an increasing frequency in the later years of this study.

Posterior urethral valves were suspected by the finding of a distended bladder and/or palpable renal masses on careful physical examination. The diagnosis was based on clinical features and the typical finding of a dilated and elongated posterior urethra above the valves on voiding cystourethrography (VCUG).

The relevant clinical, laboratory and radiological data were analyzed.

RESULTS

The age at presentation ranged from 2 days to 15 years (mean: 2.5 years). Seven (17.0%) of the patients were neonates while 21 (51.3%) were infants (Table 1).

Twenty-eight patients presented with a condition highly suspicious of PUV because of urinary features (Table 2), while in 7 patients other urological conditions (chronic renal disease in 2 and urinary tract infection in 5) were the initial diagnosis. In the remaining 6 patients the initial diagnosis was non-urological: septicemia in 2 patients, ascites in 3 and abdominal mass in 1 patient. The 3 neonates with ascites presented within the first week of life and had associated constipation simulating lower intestinal obstruction.

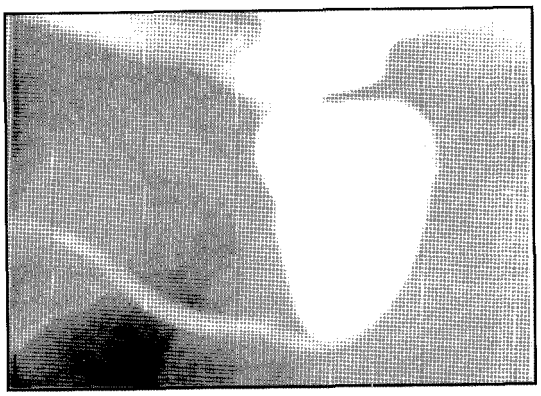

Fig. 1: A voiding cystourethrogram in a child with posterior urethral valves.

The main clinical features were distended bladder, poor stream, dribbling and palpable loin masses (kidneys). The presenting symptoms and signs are summarized in Table 2. Non-urological features like infection, uremia and failure to thrive were common in the neonates and infants. Abdominal distension was due to urinary ascites in 3 neonates and a distended bladder and enlarged kidneys in 11 older patients. Hypertension was noted in 5 patients in whom frankly purulent urine drained at the time of vesicostomy. Nine showed bilateral vesico-ureteric reflux (VUR). One 4-year-old boy presented with severe anemia, pyocystis and renal failure. He died in spite of suprapubic cystostomy at which thick purulent urine was drained.

Voiding cystourethrography (as described by Waterhouse[9]) was diagnostic in all cases (Fig. 1). Preoperative abdominal ultrasonography showed varying degrees of hydronephrosis, a thickened bladder wall with trabeculation and a few cases of sacculation. The initial chemistry showed high levels of serum urea (Table 3). The serum creatinine level was examined in 15 patients in whom the urea level was above 15 mmol/l; four of them had renal failure with creatinine levels ranging from 320 µmol/l to 720 µmol/l (average: 240 µmol/l) and required peritoneal dialysis. Preoperative urine culture results were available in 36 patients. The most common organism grown was Escherichia coli which was found in 36.1% of our patients. (Table 4).

Table 1: Age Distribution in 41 Children with Posterior Urethral Valves.

Age	No. of patients	%
In months		
0-1	7	17.0%
2-6	12	29.3%
7-12	9	22.0%
In years		
1-5	7	17.0%
6-10	4	9.8%
11-15	2	4.9%
Total	**41**	**100.0%**

Table 2: Clinical Features in 41 Children with Posterior Urethral Valves.

Clinical Features	No. of patients	%
Urinary Features		
Distended bladder	40	97.6 %
Poor stream	38	92.7 %
Dribbling	35	85.4 %
Palpable renal mass	14	34.1 %
Hematuria	8	19.5 %
Pyuria	5	12.2 %
Acute urinary retention	2	4.9 %
Non-urinary features		
Abdominal distension	19	46.3 %
Anemia	16	39.0 %
Dehydration	12	29.3 %
Vomiting	9	22.0 %
Failure to thrive	7	17.0 %
Fever	6	14.6 %
Edema	6	14.6 %
Constipation	5	12.2 %
Ascites	3	7.3 %

Table 3: Serum Urea Results in 41 Children with Posterior Urethral Valves.

Serum urea (mmol/l)	No. of Patients	%
4.5 - 6.6	11	26.8 %
6.7-15.0	15	36.6 %
15.1-25.0	9	22.0 %
25.1-35.0	4	9.7 %
35.1-45.0	2	4.9 %

Eleven patients with negative preoperative urine cultures had significant infection during the post-operative period.

Once the diagnosis was confirmed, immediate decompression of the urinary tract was established by transurethral catheterization with an infant feeding tube of size 8F, if the patient was a neonate or infant. In older children a size 10-12 Foley's catheter was used for bladder drainage. In critically ill children, gentamycin was given in an appropriate dose according to age and severity of renal impairment, pending the results of urine culture and bacterial sensitivity tests; the antibiotic was subsequently changed when necessary. Patients whose renal function and infection improved on this regimen then had ablation of the valves. Patients whose renal condition did not improve required further continuous urinary tract drainage. Their valves were ablated later only when their renal function had improved or when the patients had been fully treated for any existing septicemia.

In patients with elevated serum urea and urinary infection, preliminary drainage via a urethral catheter for a period of 2 - 5 weeks or vesicostomy was done until satisfactory drainage was achieved. The urethral catheters were changed every two weeks until the upper tracts had significantly reduced in size. None of the patients had upper tract drainage by ureterostomy or nephrostomy.

Management consisted of transurethral balloon avulsion of the valves[10,11] in 31 patients using a Foley's catheter, and transvesical (retropubic) excision of the valves in 9. One patient died after suprapubic cystostomy and did not have definite valve ablation. Transvesical resection of the valves was practised within the first year of this study, but was abandoned subsequently in favor of balloon avulsion of the valves which is a cheaper and easier method with shorter duration of the procedure.

The balloon avulsion technique is described as follows: with the patient fully anesthetized, under sterile conditions in theater, an appropriately-sized self-retaining Foley's catheter (usually size 5-8F in neonates and 12-14F in older children) is passed into the bladder until urine drains freely. The balloon is distended with saline to the maximum volume indicated on the catheter. The surgeon applies steady but gentle traction to the catheter, while an assistant steadily but slowly withdraws the saline from the balloon using a syringe. This process is continued until the surgeon feels a 'give', indicating that the balloon has slipped from the bladder through the bladder neck into the posterior urethra. At this point about 2 - 3 ml of saline is pushed back into the balloon and the traction is continued gently until the catheter comes out through the urethra. The balloon should come out intact, but if the balloon ruptures the procedure is repeated until the balloon comes out intact. Each time the balloon is inspected for the presence of the avulsed valves which appear as pinkish, flimsy strands of tissue, resembling blood capillaries, on the balloon. The entire process is repeated between 2 to 4 times until there is evidence that the valves have been avulsed. After successful valve ablation, pressure on the suprapubic area produces voiding of urine very easily and

Table 4: Urine Culture Results in 36 Children with Posterior Urethral Valves.

Organism	Number of Patients	%
E. coli	13	36.1%
Klebsiella	5	13.9%
Proteus	3	8.3%
Staph. Aureus	2	5.6%
Pseudomonas	2	5.6%
No growth	11	30.5%

freely. The catheter is left in situ to drain the bladder for another 48 hours after which it is removed, unless otherwise indicated.

Voiding of urine could be re-evaluated by VCUG within the first week of valve ablation in 33 (80.5%) of the 41 patients (26 with balloon avulsion and 7 with transvesical valve resection), and showed satisfactory results defined as restoration of normal urine flow with a good stream and normal blood urea. Seven patients were lost to follow-up after the intervention and one patient died of renal failure. Post valve ablation VCUG showed complete resolution of the reflux in the 9 children who had reflux before the ablation.

At last follow-up ranging from 2-7 years, 28 of the 33 children had a satisfactory outcome. Three patients developed minor residual strictures which responded to serial dilatation, while 2 developed occasional dribbling which improved and eventually stopped in one. None of the patients had complete incontinence after ablation/avulsion of the valves.

DISCUSSION

The incidence of PUV ranges from 1:5000 to 1:8000 in male infants[12,13]. Posterior urethral valves are probably not as rare in Africa as the paucity of reports in the English literature may suggest[1,2]. Although the incidence in Nigeria is unknown, recent reviews of childhood obstructive uropathy in Nigeria suggest that the condition is certainly not uncommon[14]. In this study the rate of between 3 and 8 children with PUV diagnosed per year compares with rates of 10 per year in London[3] and 15 per year in the United States of America[4]. PUV constituted 15% of all congenital abnormalities of the genitourinary system with a hospital incidence of 1.7 per 1000 in a study by Abdurrahman et al. in Zaria, in North Western Nigeria[14].

The preponderance of infants (51.3%) in this study is in agreement with the report of Willams, et al.[3], while in the report of Hendren only 14% of the patients were infants[4]. Neonates comprised 17.1% of the patients in this study, whereas neonates constituted one third of the patient population in other reports[3,14], which may be explained by the late presentation of our patients due to poverty and ignorance.

In most cases the correct diagnosis was suspected after a detailed history and on careful clinical examination by demonstrating a distended bladder and/or palpable kidney masses. In 13 patients there was a considerable delay in the initial diagnosis caused either by ignorance on the part of the referring clinician, or by non-specific presenting symptoms. For instance, in 6 (14.6%) of our patients, various non-urological initial diagnoses caused a considerable delay (average 8 days) in the institution of satisfactory treatment. Williams, et al.[3] also emphasized the possibility of misleading presenting symptoms in neonates and infants, such as neonatal ascites. Therefore, any obscure case of abdominal swelling in children should

be investigated by intravenous urography and VCUG in order to detect any underlying PUV. An increased awareness of the condition and its varied clinical presentation will help early recognition with an improvement in prognosis. A distended bladder and/or palpable kidney masses in any male child presenting with either typical or obscure urinary symptoms should arouse a high index of suspicion of PUV in the clinician.

A simple method of VCUG as described by Waterhouse[9] was used in this series and was diagnostic in all cases. It is occasionally assumed that sophisticated rapid-recording equipment is required for satisfactory visualization of PUV[4,5] which may inhibit attempts to carry out this procedure in some centers with limited radiological facilities in developing countries. The absence of such apparatus should not present any handicap to performing this essentially simple study and making a positive diagnosis as early as possible.

Most studies from Europe and North America report that VUR occurs in 40-50% of children with PUV. In contrast, in a study of 17 African patients by Van Den Bulcke and Hennebert[1] there was no demonstrable reflux. The authors concluded that the uretero-vesical junction in Africans may be anatomically different, and that the African bladder responds differently to lower urinary tract obstruction. In 22% of the patients of this study (mainly older children) voiding cystourethrography demonstrated VUR. This compares favorably with the 29% earlier reported by Garg et al.[15] from Northwestern Nigeria. Considering that the majority of VUR occurred in the older children, two possibilities can be taken into consideration: either the VUR resulted from longstanding increased intra-vesical pressure, or there may be an element of selection in that infants with early gross reflux become severely ill earlier, never reach hospital, and are therefore never diagnosed. Future studies are needed to elucidate in more detail the problem of VUR in Africans.

The majority of our patients were uremic at the time of admission, mainly because of delay in presentation and/or a delayed diagnosis of PUV due to non-specific presenting features. In some instances there was some further delay in offering definitive treatment even after the diagnosis was confirmed because of the patient's poor general condition requiring urethral catheterization for a period of 2 – 5 weeks or vesicostomy before definitive treatment could be undertaken.

The most difficult problem in the management of PUV is the infant or neonate presenting with severe sepsis and uremia. A good treatment outcome therefore depends on initial effective drainage of the urinary tract (especially in the presence of VUR) and appropriate antimicrobial treatment for a variable length of time depending on the response. Sepsis in the presence of uremia is usually fatal unless appropriate treatment is started immediately[16]. In critically ill children gentamycin, in appropriate doses, according to age and severity of renal impairment, should be started pending the results of urine culture and bacterial sensitivity tests. The antibiotic may subsequently be changed if necessary. Patients who improve on this regimen should have ablation of the valves. Patients who do not improve may require upper tract drainage[3,6,7]. However, this form of drainage is not always successful, especially in patients with grossly infected upper tracts and frankly purulent urine, who may tolerate general anesthesia very poorly. We agree with Garg et al.[15] that once the diagnosis is confirmed, immediate decompression of the urinary tract should be established by urethral catheterization. The patients (including those with VUR) fared well on varying lengths of continuous bladder drainage either by indwelling urethral catheter, suprapubic cystostomy or a vesicostomy, and therefore did not require ureterostomy or nephrostomy.

Different methods of valve ablation have been described, including ablation of the valves under imaging control[3] and bouginage. Some authors use perineal urethrotomy to

fulgurate the valves with a blunt metal hook, through a plastic aural speculum[16]. Indwelling catheters have been used by others to relieve the obstruction by causing pressure necrosis[17,18]. With the development of pediatric endoscopic equipment; most workers have adopted endoscopic resection[3,4,6]. However, the unavailability of this equipment in our center presents a problem. In the present study the majority of the valves were avulsed/ disrupted using catheter balloons, although a few patients had retropubic excision of their valves. This technique produced satisfactory results and can therefore be recommended for adoption in hospitals where specialized, sophisticated, expensive radiological or endoscopic equipment is unavailable.

The mortality rate was low - 2 out of 41 (4.9%) - as compared with 24% reported by Garg et al.[15], and 16.2% reported by Williams and co-workers[3]. Sepsis in the presence of uremia was the cause of death in the 2 patients in this series. This emphasizes the need to adequately and effectively control sepsis and uremia as urgently as possible to achieve a good prognosis in such children.

In conclusion, although this study was restricted to one hospital, there appears to be a high incidence of PUV in children in North Central Nigeria. Increased awareness of varied clinical features, a high index of suspicion and simple conservative treatment by balloon avulsion of PUV would improve the outcome.

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RESUME

VALVES DE L'URETRE POSTERIEUR CHEZ L'ENFANT : EXPERIENCE DANS UN CENTRE AVEC DES EQUIPEMENTS RARES

Objectif: Les valves de l'urètre postérieur (PUV) sont les causes congénitales les plus communes d'obstruction du bas appareil urinaire chez les enfants masculins, mais peu de cas ont été rapportés au Nigeria. Dans cette étude nous décrivons notre expérience de sept ans de prise en charge de PUV chez les enfants afin d'attirer l'attention vers cette pathologie dans notre environnement.
Patients et méthodes: C'est une revue de 41 enfants consécutifs présentant des PUV qui ont été contrôlés aux hôpitaux de l'enseignement universitaire de Jos (JUTH), Jos, Nigéria, de juin 2000 à avril 2006. Leur âge à la présentation s'est étendu de 2 jours à 15 ans (moyenne : 2,5 ans). Les résultats des examens cliniques, de laboratoire et les données radiologiques ont été saisis dans une base de données et analysés.
Résultats: Vingt-huit patients se sont présentés dans une condition hautement suspecte de PUV, alors que 7 patients se présentaient dans d'autres tableaux urologiques. Six patients se sont présentés avec des symptômes non-urologiques qui ont causé un retard dans le diagnostic et le traitement. La cystourétrographie mictionnelle a été réalisée dans tous les cas. Dans la majorité des patients (n=31), une avulsion transurétrale par ballon des valves a donné des résultats satisfaisants chez plus de 80% des patients. Le rétrécissement urétral était la complication principale (qui a répondu à la dilatation périodique); elle a été vu chez 3 patients. Le taux de mortalité postopératoire est de 2.6%.
Conclusion: Bien que cette étude ait été limitée à un hôpital, il semble y avoir une incidence élevée de PUV chez les enfants au Nigéria central et du nord. Une meilleure connaissance des divers tableaux cliniques et un traitement conservateur simple par avulsion par ballon des PUV amélioreraient les résultats.

Mots Clés : valves urétrales postérieures, enfants, traitement, manque d'équipements.

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