Challenges in the Management and Outcome of Posterior Urethral Valve in Aba, Nigeria

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Abstract: Posterior urethral valves are the commonest cause of bladder outlet obstruction in boys. The incidence varies from 1 in 5000 to 25,000 live birth worldwide. The incidence in lower and middle-income countries is unknown due to paucity of data. The aim of this study was to review the presentation, management and outcome of posterior urethral valve at the Abia State University Teaching Hospital Aba, Nigeria. Patients and Methods: Fifteen boys with posterior urethral valve seen at the Abia State University Teaching Hospital Aba from January 2016 to June 2018 were included in the study. Data collected includes demographic data, clinical features, and duration of symptoms, serum biochemistry, radiological diagnosis, surgical management and outcome. Results: The patient age ranged from 2 weeks to 2 years, Median age of 6 months. 7(46.6%) of the patients were neonates. The commonest symptoms were voiding dysfunction, distended bladder and anaemia. Duration of symptoms was 2weeks in 6 (40%) patients, 4 weeks in 4(27) patients and 8weeks in 5(33%) patients. 2(13%) patients had grade 2 vesicoureteric reflux on expressive cystourethrogram. All the patients had foley catheter avulsion of posterior urethral valve. There was no case of frank haemorrhage but haematuria was noticed which resolved spontaneously. 14(93.3%) out of 15 patients reported consistent good urinary stream without evidence of renal deterioration. A patient (6.6%) had persistent straining at micturition requiring a repeat Foley catheter balloon avulsion. All mothers were satisfied with the outcome of treatment due to good post-operative urinary stream. There was complete resolution of the vesicoureteric reflex at 6 months post valve ablation expressive cystourethrogram. The post-operative urinary stream, serum electrolyte, and urea and creatinine status remained normal during follow up period of 6 months. Conclusion: Posterior urethral valve is a common cause of bladder outlet obstruction in boys, high index of suspicion will lead to early diagnosis, late presentation still persist due to poor knowledge of health care workers in our environment.

Keywords: Posterior Urethral Valve, Challenges, Management, Outcome

1. Introduction

Posterior urethral valves are the commonest cause of bladder outlet obstruction in boys. The incidence varies from 1 in 5000 to 25,000 live birth worldwide [1]. The incidence in lower and middle-income countries is unknown due to paucity of data. The study of the embryologic origin and classification of PUV has led to a myriad of theories, with varying types and amounts of experimental and anatomic evidence. The male urethra is known to complete development at week 14 of gestation. The valves form prior to this time, producing some level of obstruction once urine production has begun for the remainder of fetal development. Valves were first described by Morgagni in 1717 and were described again by Lagenbac in 1802 from the examination of autopsy specimens [2]. The first scientific description of valves, their embryology, and their role in urinary tract pathology was presented by Tolmatschew in 1870. Perhaps the most famous description of valves came from Young in 1919, at which point the well-known PUV classification method was described [2] According to the Young classification, there are three types of PUV, with Type I being by far the most commonly encountered. Type I (95%) represents sail-like folds from the verumontanum distally
along the urethra. Type II valves likely have only historical significance and are not considered to be a clinical entity, but rather hypertrophied urethral folds. Type III (5%) valves represent a cannulated septum thought by some to represent an incomplete dissolution of the urogenital membrane. Since that original classification, the description of PUV embryology has teetered between claims of abnormal integration of the Wolffian ducts into the urethra and exaggerations of normal folds seen in normal urethral development [2]. Interesting recent work on the topic claims that Type I and II valves represent the same entity, and are actually a COPUM (congenitally obstructing posterior urethral membrane) whose appearance may vary depending on the instrumentation or imaging studies performed. The term “Cobb’s collar” has been applied to a distinct congenital urethral stricture occurring distal to an external urethral sphincter. COPUM appears to be a result of a persistent urogenital membrane, although the embryology of PUV is far from clear and an area fertile for research [2].

Prenatal ultrasound screening of posterior urethral valve has significantly increased early diagnosis and management of the pathology in high income countries [3]. The consequential pathology due to back pressure from untreated obstruction from posterior urethral valve leads to thickened bladder wall, trabeculation, sacculation, diverticulum and hydroureter. The anomaly is associated with high mortality and morbidity including urosepsis, overflows urinary incontinence, chronic kidney diseases (CKD), hypertension, chronic anaemia, failure to thrive, poor quality of life and even death [3]. The Pop-off mechanisms such as vesicoureteral reflux syndrome, large bladder diverticulum, patent urachus and perineal urinoma may help in reducing the back pressure, abating renal failure and subsequent death [1, 2, 3]. The early detection and intervention in high income countries helps to ameliorate the complications from posterior urethral valve but late presentation and intervention in middle and low-income country accounts for high morbidity and mortality. Endoscopic ablation of the posterior urethral valve under direct vision using resectoscope remains the gold standard. These facilities are not readily available in some centres in low income countries such as Nigeria [1, 2, and 3]. Long-term management of PUV constitutes a challenge in the practice of paediatric surgeons/urology in our environment due to lack of specialized centres as well as the paucity of trained paediatric urologists and nephrologists to evaluate and monitor these children [6]. In the few highly specialized centres where valve ablation is done, most patients fail to turn up for follow-up visits due to an erroneous belief that it has been cured following the initial relief of the obstruction. The aim of this study was to review the presentation, management and outcome of posterior urethral valve at the Abia State University Teaching Hospital Aba, Nigeria.

2. Patients and Methods

Fifteen boys with posterior urethral valve seen at the Abia State University Teaching Hospital Aba from January 2016 to June 2018 were included in the study. Data collected includes demographic data, clinical features, and duration of symptoms, serum biochemistry, radiological diagnosis, surgical management and outcome. Data collected were analysed using statistical package for social scientist software version 17 percentages and proportions.

3. Results

The patient age ranged from 2 weeks to 2 years, Mean age of 6 months. 7(46.6%) of the patients were neonates as shown in Table 1.

<table>
<thead>
<tr>
<th>Age in months</th>
<th>Number of patients</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 day--------1 month</td>
<td>7</td>
<td>46.6</td>
</tr>
<tr>
<td>2 months------12 months</td>
<td>5</td>
<td>33.3</td>
</tr>
<tr>
<td>13 months------24 months</td>
<td>3</td>
<td>20%</td>
</tr>
</tbody>
</table>

The commonest clinical features were voiding dysfunction characterised by [straining in 10 (66%), poor stream 8(53%) and dribbling 6(40%)] in 15(100%), Distended bladder in 12 (80%) patients and anaemia in 10 (66.7%) as shown in Table 2.

<table>
<thead>
<tr>
<th>Clinical features</th>
<th>Number of patients</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Voiding anomalies</td>
<td>15</td>
<td>100%</td>
</tr>
<tr>
<td>Distended bladder</td>
<td>12</td>
<td>805</td>
</tr>
<tr>
<td>Anaemia</td>
<td>10</td>
<td>66.7%</td>
</tr>
<tr>
<td>Fever</td>
<td>8</td>
<td>53.3%</td>
</tr>
<tr>
<td>Ballotable kidneys</td>
<td>6</td>
<td>40%</td>
</tr>
<tr>
<td>Failure to thrive</td>
<td>6</td>
<td>40%</td>
</tr>
<tr>
<td>Patent urachus</td>
<td>2</td>
<td>13.3%</td>
</tr>
</tbody>
</table>

Duration of symptoms was 2 weeks in 6 (40%) patients, 4 weeks in 4(27%) patients and 8 weeks in 5(33%) patients. The urine of the patients was evaluated for microscopy, culture and sensitivity. Common isolates on urine microscopy and culture were Escherichia coli (640%), klebsiella in 4(27%) and no isolates in 5(33%). All the patients had abdomino-pelvic ultrasound with emphasis on the kidney, ureter and bladder. 4 patients had hydronephrosis, 8 patients had hydroreuter and 3 patients thickened bladder wall. 2 (13%) of the prenatal ultrasounds were suggestive of PUV by showing bilateral hydronephrosis. Micturating cystourethrogram was done in all the patients which confirm the diagnosis posterior urethral valve. Two patients had bladder diverticular, 6 patients had left hydronephrosis, 3 patients had right hydronephrosis, and 2 (13%) patients had grade 2 vesicoureteric reflux.

All the patients had Foley catheter avulsion of posterior urethral valve. There was no case of frank haemorrhage but haematuria was noticed which resolved spontaneously. 14 (93.3%) out of 15 patients reported consistent good urinary stream without evidence of renal deterioration. A patient (6.6%) had persistent straining at micturation requiring a repeat Foley catheter balloon avulsion. All mothers were
satisfied with the outcome of treatment due to good post-operative urinary stream. There was complete resolution of the vesicoureteric reflux at 6 months post valve ablation expression cystourethrogram. The post-operative urinary stream, serum electrolyte, and urea and creatinine status remained normal during follow-up period of 6 months.

4. Discussion

The incidence of posterior urethral valve has remained stable over some period. The indexed study described the experience in the management of posterior urethral valves in a tertiary care centre in Abu, Nigeria. The prevalence of PUV and admission rate in the surgical unit of this institution was estimated at 7 cases per annum. Jaja et al. [3] in Portharcourt South-South Nigeria observed that it accounted for 1 in 2447 children seen in their hospital. Uba et al. [4] in Jos North-Central Nigeria, Angwafo et al. [5], Tambo et al. [1] and Chiabi et al. [6] in Cameroon reported 3-4 cases per annum while Talabi et al. [7] in Ile-South —West Nigeria reported 54 cases over a 15-year period and Orumuah et al. [8] in Benin Nigeria reported 44 cases over a 10-year period. However, widespread use of prenatal ultrasound evaluation has significantly increased its early diagnosis and management in developed countries. [7-9]. Two (13%) of the prenatal ultrasounds, there was suspicion of PUV. This is consistent with reports by Okafor et al. [11], Talabi et al. [7] and Tambo et al. [1] of 6.5%, 8.1% and 11.2% respectively. Ohagwu et al. [12] showed that previous ultrasound, long distances to service providers, considerably heavy financial cost, long waiting periods, and unsatisfactory previous scan experience are major barriers to prenatal ultrasound in Nigeria, and these barriers have indirectly made prenatal diagnosis of PUV difficult in our setting. Low level of detection of PUV by prenatal ultrasound scan can also be explained by the lack of experts in prenatal imaging hence missed diagnosis.

Most patients in developed countries are now diagnosed by the postnatal evaluation after a diagnosis of prenatal hydronephrosis but despite the availability of ultrasound in low and middle income countries, prenatal diagnosis of posterior urethral valve is still very low. Post natal diagnosis based on clinical presentation and radiological investigations persist in developing countries. [7-9]. The mean age of presentation 6 months suggests that patients with this condition present late in our environment. More than half of the patients managed within the period presented after the age of one month. Similar findings were also noted in previous studies in southwestern [7] and eastern Nigeria [9] in which 57.1% and 52% of patients, respectively, presented after the age of 1 year. Factors which may have contributed to delayed presentation noted in the region include sub optimal prenatal ultrasound scan utilization poverty, ignorance on the part of parents and paucity of specialist care.

All the patients in this study presented were voiding dysfunction characterised by [straining in10 (66%) , poor stream8(53%) and dribbling6(40%)]. This similar findings were noted by Tambo et al and Chiabi et al.in cameroun where dribbling 11 (61.1%), dysuria 8 (44.4%) and pollakiuria 6 (33.3%) and dribbling (61.7%) and dysuria (54%) respectively. a Bhauvik et al. [17], in India noted that dribbling occurred in79% in there series. Again, the similarity was noted in studies conducted by Odetunde [9] et al. in Enugu South East Nigeria and Orumuah [8] et al. where 100 and 92% respectively presented with voiding anomaly. It is therefore important to have a high index of suspicion of PUV in neonates and infants presenting with urinary symptoms. The bladder/urine retention, which is comparable to a study conducted in a Nigerian tertiary hospital in 2015 by Orumuah [8] et al. where about a third of participants had a palpable bladder.

In this setting, participants were not routinely subjected to screening tests to detect other congenital anomalies due to financial constraints and the limited availability of screening facilities. However, associated congenital anomalies noted included four umbilical hernias (26%), 4 inguinal hernia (26%), 2 hydrocele (13%), and one cryptorchidism (6%). Similar congenital malformations were described in studies by Tambo et al. [1], Chiabi et al.6 and Orumuah et al.8. Recurrent fever occurred in 53% of patients in this index study which is due to urinary tract infection secondary to urinary stasis caused by the posterior urethral valve. Odetunde et al noted that the presence of ballotable kidneys suggests back pressure effect of the lower urinary obstruction with attendant hydronephrosis. This predisposes to stasis of urine and colonization by bacteria with attendant urinary tract infection and fever which was present in over 90% of their patients.

The gold standard for the diagnosis of PUV is VCUG 13-19. This was done in all cases and demonstrated dilatation and elongation of the posterior urethral in 15(100%). This is similar to Tambo et al. [1] 88.9%, and by Talabi et al. [6]. where it showed dilated and elongated posterior urethral valves in 83.8% of the cases. Van Den Bulcke and Hennebert [20]: In a study of 17 African children found no demonstrable reflux and concluded that the ureterovesical junction in Africans may be anatomically different and that the African bladder responds differently to lower urinary tract obstruction. In contradistinction to their findings, VCUG demonstrated vesicoureteric reflux in 13% of this series. This compares favorably with 29.7% by Talabi et al in Ile South West Nigeria the 22% reported by Uba et al. Jos in North Central Nigeria. A much higher figure was reported in India [18] and Iran. [19] Future prospective studies are necessary to elucidate the presence of VUR in African boys with PUV.

There have been various operative techniques described for PUV resection. In this study, all the patients benefited from Foley catheter valve avulsion. Valve avulsion resulted in the relief of obstruction in all but one (6%) patients that had residual valves and thus needed repeat valve ablation to attain satisfactory passage of urine. However, in a study conducted by Mirshermirani et al. [18] and Sudarsanan et al [19] where 15.3 and 13% respectively had residual valves. Complications such as urinary retention, urinary extravasation, urethral bleeding and urethral stricture have
been reported to be associated with the procedure. However, these were not recorded in this series within the period of follow-up. However, Tambo et al noted urethral stricture in two (11.2%) patients which was comparable to 7.1%, 5% and 8.2% obtained by Mirshermirani et al [18], Shittu et al. [15] and Sudersanan et al. [19] respectively. Other means of valve ablation include the use of an endoscopic loop resectoscope, endoscopic valve ablation, hook diathermy, valvotome, and cold knife urethrotome [21]. Close follow-up is needed after valve ablation to ensure proper bladder function in the children. For example, persistent severe reflux that prevents efficient bladder emptying, poorly functioning bladders, or UVJ (ureterovesical junction) obstruction due to detrusor hypertrophy may require an alternate procedure (such as urinary diversions) to maximize emptying and protect renal function. In addition, antibiotic prophylaxis is crucial to avoid urinary tract infections, in order to prevent any further damage to already tenuous kidneys.

The long term bladder function of these indexed study cannot be ascertain at this moment due to short period of follow up. However, there have been many classifications of bladder function in patients with PUV, as well as the development of the concept of the “valve bladder” [22]. The description of the valve bladder syndrome by Peters is useful in following these patients long term [23]. Of course, all children with PUV are different and do not necessarily follow any schedule in terms of deteriorating bladder function, but it is clear that different patterns of bladder pathology are seen specifically in children with PUV. Peters divided these into three groups: hyper-reflexic, small noncompliant and myogenic failure [23]. These categories tend to proceed in order, with children beginning with hyper-reflexic bladders (that may respond to anticholinergic therapy) and then often descending into small noncompliant bladders that may need medical therapy and catheterization, or even augmentation. Interestingly, overnight catheter drainage has proven to be protective in some patients, by reducing the strain of high urine output on the urinary tract, with resultant normal bladder function in these children following transplant.

There was no mortality in this series. However, Talabi et al in Ife South West Nigeria noted a mortality of 13.5% where uremia with superimposed infection was the cause of death in three patients while overwhelming sepsis alone was the cause of death in the remaining two patients in their series. This underscores the need to adequately and effectively control infection and uremia as urgently as possible to achieve a good outcome in such children which was consistent with 12.5% reported by Okafor et al in Enugu South East Nigeria but in contrast to 4.9% by Uba et al in Jos North Central Nigeria

5. Conclusion

Posterior urethral valve is a common cause of bladder outlet obstruction in boys, high index of suspicion will lead to early diagnosis, late presentation still persist in this environment due to poor knowledge of the disease by health care workers in this environment. Capacity buildings inform of further training of pediatric surgeon/urologist and procurement of the endoscopic equipment will enhance outcome. Public health education and health insurance will ameliorate the cost of treatment and reduce morbidty resulting from late presentation of the patients.

References


