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Management of Posterior Urethral Valves at Kilimanjaro Christian Medical Centre:

A 10 Years Experience.

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Background: Posterior urethral valve (PUV) disorder is an obstructive developmental anomaly in the urethra and genitourinary system of male newborns. A posterior urethral valve is an obstructing membrane in the posterior male urethra as a result of abnormal in utero development. This study was aimed at reviewing the management and outcome of treatment of PUV at KCMC among children presented for treatment during the period from January 1998 to December 2007.

Methods: This was retrospective hospital based review of 56 patients treated for posterior urethral valves. The study was conducted in the Institute of Urology at Kilimanjaro Christian Medical Center, which is a tertiary referral hospital located in northern part of Tanzania. The study population was consisted of all children diagnosed to have posterior urethral valves in the study period. All the children clinically suspected to have posterior urethra valves had the diagnosis confirmed by micturating cystourethrogram and depending on their age, underwent initial vesicostomy and finally posterior urethral valve fulguration as the definitive treatment. Data regarding age at presentation, types of valves seen, clinical presentation, initial treatment given, definitive treatment and complications of definitive treatment of PUV was extracted. Information on outcome of definitive treatment of PUV was also extracted. The information was transferred to a data sheet for analysis.

Results: At presentation 57.1% of patients were aged below two years while 16.1% were above six years of age. Hydronephrosis occurred in 94.6% of patients. A dribbling urinary stream was found in 82.1% of patients. Urinary tract infection was also common, being present in 58.9% of patients. Fifty percent of patients presented with urine retention while vesicoureteral reflux was found in 23.2%. Of the valves seen 96.9% were type I. Initial treatment comprised of vesicostomy (42.9%), initial valve ablation (51.8%) and urethral catheterization 5.3%. Electrofulguration was the mainstay of definitive treatment of PUV and was instituted in all of the definitively treated patients. Urethral stricture as a complication was seen in 8.9% of the patients. Residual valves were seen in 20% of patients. A treatment success rate of 86.7% was observed. A mortality rate of 5.4% was found.

Conclusion: The clinical presentation of PUV and the age at presentation is similar to that seen in Europe and America. Type I valves form the majority of posterior urethral valves. Electrofulguration is the mainstay definitive treatment of PUV at KCMC. The success rate of treatment of PUV is good and compares with that seen in Europe and America.

Introduction

As an anomaly Posterior urethral valves (PUV) is the most common cause of bladder outlet obstruction in male neonates[1]. The reported incidence in United States is between 1 per 8,000 to 1 per 25,000 live births. Reisuke et al[2] in Australia demonstrated that the severity of obstruction caused by this membrane varies depending on its morphology. A posterior urethral valve (PUV) is therefore not limited to male neonates and infants only. It may present later in life, be it in early or later childhood, adolescence period or even in adulthood. The most severely affected patients present early in life or die in utero while the moderately to mildly affected ones present later in life. This anomaly is unique to male children[1,2].

The clinical importance of PUV is that it may cause severe urethral obstruction resulting in widespread damage and dysfunction of the entire urinary tract. The effect of this damage may be reflected on the glomerular filtration, ureteral and bladder smooth muscle function, and urinary continence. Furthermore, PUV is the single most common urologic cause for renal failure and need for renal transplantation in children. Approximately 10-15% of children undergoing renal transplant have PUV as the cause of renal insufficiency, and approximately one third of patients born with PUV progress to end-stage renal disease (ESRD)[1,3].

One retrospective review in Michigan among 47 patients with delayed presentation revealed that 60% presented with urinary dribbling, 40% with urinary tract infection and 13% with voiding pain. Other symptoms were present in less than 10% of the cases and included poor stream, gross haematuria and proteinuria. In the same review, hydronephrosis and vesicoureteral reflux were present in 40% and 33% of the patients respectively at diagnosis. Serum creatinine was elevated in 35%. It was also found that, end stage renal disease developed in 10% of the cases. The severity of presenting signs and symptoms was significantly associated with renal impairment, while patient age at diagnosis was not[4].

It is a normal practice that in any case, management usually begins with placement of a small transurethral catheter to provide unobstructed vesical drainage. In the unusual situation in which the newborn urethra seems too small to accommodate the available endoscopes, an elective vesicostomy is appropriate and safe. Indeed, Walker preferred a vesicostomy to endoscopic ablation in the neonate, believing that this technique is safer and reduces the incidence of urethral strictures. It is well documented that the definitive management of posterior urethral valves is endoscopic destruction of the urethral valves. However, the initial management depends on the degree of renal insufficiency as well as the age of the child. It is accepted that older children who present with voiding dysfunction or urinary tract infection but have satisfactory renal function are easily and effectively treated initially by endoscopic destruction of the urethral valve alone. Decisions concerning selective surgery for persisting VUR, lingering UVJ obstruction (uncommon), or detrusor dysfunction are made on an individual basis[1,3,5].

The surgical complications of posterior urethral valves include urethral stricture, urethral diverticula and urinary incontinence. Careful endoscopic destruction of valves, even in the newborn, has not been associated with a significant incidence of urethral stricture. However, the development of dense urethral strictures following transurethral ablation of posterior urethral valves is a well known complication, especially in the neonate. In one series evaluating 85 boys with posterior urethral valves no stricture were found during follow-up. In the same series urinary incontinence associated with transurethral incision occurred in 3 patients and a small urethral diverticulum was found in 1. In this series the complication rate associated with the transurethral incision itself was 5%. The fact remains that many children with posterior urethral valves have significant degrees of parenchymal dysplasia and demonstrate gradual but progressive loss of renal function during their lifetime. It is important that parents be aware that the diagnosis of posterior urethral valves means a long-term commitment to surveillance and care and that even today the eventual outcome is unclear in many instances[6].

The aim of this study was to bring to light facts that had not been documented before pertaining to the challenges encountered in managing PUV which is a difficult and a life threatening condition at Kilimanjaro Christian Medical Center.

Patients and Methods

This was a retrospective, descriptive hospital based study covering a ten years period from January 1998 to December 2007. Files of the patients who were treated for posterior urethral valves in the urology department of the KCMC hospital between January 1998 and December 2007 and met the inclusion criteria were retrieved from the KCMC medical records and relevant data was extracted. Age at presentation was taken as that age at which the patient first reported to KCMC for treatment

with the initial urinary symptoms that ultimately led to the diagnosis of PUV and subsequent treatment. Information on presence of hydronephrosis was derived from the abdominal ultrasound results. In case of misplaced ultrasound result sheet, the description made in the file by an attending specialist quoting the ultrasound results or made by a registrar during a major ward round was taken as valid information. This also applied to vesicoureteral reflux which was derived from MCUG results. Urinary bladder distension was taken as a clinical diagnosis.

Urinary ascites was strictly an ultrasound diagnosis considering the difficulty of assessing a child with respect to presence of ascites as all children with suspected PUV undergoes an ultrasound examination. Respiratory distress is a clinical diagnosis; the description of an attending physician was taken as valid information. Urinary tract infection was taken as prior history of recurrent fever associated with any one of the urinary symptoms like crying on micturition, painful micturition, straining on micturition, dribbling urinary stream or incontinence of urine. Laboratory evidence of urinary tract infection was considered valid even if the history lacked any indication of urinary tract infection. Dehydration is a clinical diagnosis and an attending physician's description was considered valid.

Electrolyte abnormalities are biochemical diagnoses and biochemical test results were referred to. Dribbling urinary stream was taken from the attending physician's description of the presenting complaint or amplification of the same. Failure to thrive was taken from a doctor's description of the patient's presenting complaints or the amplification of the presenting complaints. Urinary incontinence was taken from the attending physician's description of the patient's presenting complaints or amplification of the presenting complaints.

Prenatal diagnosis was registered when the patient had ultrasonographic evidence of posterior urethral valves like prenatal hydronephrosis, distended urinary bladder and oligohydramnious in the prenatal period. Types of valves were recorded according to the cystoscopic findings at the time of valve ablation or at diagnostic cystoscopy. The surgeon either clearly stated the type as type I or type III or gave a description that the researcher matched with one of the known types e.g. a description that states the finding of two fleshy valves originating distal to the verumontanum towards the membranous urethra implies type I valves.

The type of initial management, type of definitive treatment and complications of surgical treatment were derived from the documentation in the files. Regarding the outcome of surgery, serum creatinine before treatment was defined as the serum creatinine level at presentation. Serum creatinine level after treatment was defined as the serum creatinine level in the last visit. The stream of urine after treatment conformed to the patient's, parents or care taker's definition and the clinician's as documented in the file.

Results

A total of 56 files of patients who underwent treatment of posterior urethral valves in the urology department at KCMC between January 1998 and December 2007 were retrieved from the medical records. These patients had a mean duration of follow up of 88 weeks with a standard deviation of 104. The minimum duration of follow up was 0 weeks and the maximum was 416 weeks. Forty five patients (80.4%) among these received definitive treatment of posterior urethral valves that is valve ablation either as an initial treatment or after an initial vesicostomy. The diagnosis was made by MCUG and confirmed by urethrocystoscopy at the time of definitive treatment. On average 6 patients were seen each year.

It was found that thirty two patients (57.1%) presented at the age below 2 years. Fifteen patients (26.8%) presented between age 2 and 5 years. The remaining 9 patients (16.1%) presented at the age 6 years and above (Table 1). The most common clinical presentation was hydronephrosis which

occurred in 53 patients (94.6%). Of these, 50 patients (94.3%) had bilateral hydronephrosis and the remaining 3 patients (5.7%) had unilateral hydronephrosis which was exclusively on the left side (Table 2). Dribbling urinary stream was the next commonest presentation occurring in 46 patients (82.1%) while urinary ascites was the least occurring in 1 patient (1.8%). Vesicoureteral reflux was found to be present in 13 patients (23.2%). In 11 of these patients the information regarding the side of reflux was available and showed that 8 patients (72.7%) had left vesicoureteral reflux while only 3 patients (37.3%) had right vesicoureteral reflux. Table 2 summarizes the findings.

In only 32 among 46 patients who underwent urethrocystoscopic evaluation at the time of intended definitive treatment could information on the type of valves be obtained. Diagnostic urethrocystoscopies were not done routinely. However one patient underwent urethrocystoscopy for intentions of proceeding with valve ablation but the procedure was abandoned due to tight urethra especially at the meatus. The patient though initially presented with acute urinary retention did not come back for further treatment. It was found that 31 patients (96.9%) among the 32 patients had type I posterior urethral valves. One patient (3.1%) had type III posterior urethral valves.

It was found that among the 56 patients studied, 24 patients (42.9%) had vesicostomy as their initial treatment while 29 patients (51.8%) had valve ablation as their initial treatment. Among the 24 patients who had vesicostomy, 20 of them had definitive treatment later during their follow up. However, 4 patients did not receive definitive treatment because either they had not reached the optimum age for adequate urethral caliber or they got lost to follow up or died.

Table 1. Age Distribution at presentation

Age in Years	Number of Patients	Percentage
0 - 1	32	57.1
2- 5	15	26.8
6 - 10	7	12.5
11-15	2	3.6
Total	56	100.0

Table 2. Distribution According to Clinical Presentation and Investigation Findings

Presentation	Number of Patients	Percentage
Hydronephrosis	53	94.6
Dribbling urinary stream	46	82.1
Urinary infections	33	58.9
Urinary bladder distension	28	50.0
Vesicoureteral reflux	13	23.2
Failure to thrive	12	21.4
Electrolyte abnormalities	7	12.5
Urinary incontinence	4	7.1
Respiratory distress	3	5.4
Dehydration	2	3.6
Urinary ascites	1	1.8

Table 3. Distribution According to Type of Initial Treatment Given

Initial Management	Number of Patients	Percentage
Vesicostomy	24	42.9
Initial Valve Ablation	29	51.8
Urethral catheter	3	5.3
Total	56	100.0

Table 4. Complications of Definitive Treatment of PUV

Surgical Complication	Number of Patients	Percentage (n = 45)
Urethral Stricture	4	8.9
Residual Valves	9	20
Total	13	28.9

Table 5. Serum creatinine level before and after treatment

Definitive Treatment status	Serum creatinine before treatment		Ratio of normal to raised serum creatinine	Serum creatinine after treatment		Ratio of normal to raised serum creatinine
	Normal	Raised		Normal	Raised	
Yes	26	17	1.5	22	4	5.5
No	4	4	1	NA	NA	NA

Three patients received neither of the above treatment except for urethral catheterization. One of them underwent a cystoscopy with confirmation of the valves but fulguration of the valves was deferred till the urethra attains adequate size to accommodate available resectoscopes. The patient did not come back. The other two patients died before such treatment could be instituted as they presented in moribund condition, one with respiratory distress from pneumonia and urosepsis and the other from respiratory distress only. Table 3 summarizes the findings.

It was found that 45 patients (80.4%) underwent a definitive treatment of the posterior urethral valves which is valve ablation. In 11 patients (19.6%) the treatment had not been given for various reasons. Three of them died before the institution of the definitive treatment, one of these dying shortly after vesicostomy.

Another three of the untreated 11 patients underwent vesicostomy as an initial treatment and were waiting to reach an optimum age for adequate urethral caliber before definitive treatment. Four patients among the untreated 11 got lost to follow up, three of them after an initial vesicostomy and the other one after a cystoscopic evaluation confirming PUV type I but with the urethra too small to admit available resectoscopes. The patient did not turn up for the next scheduled visit. In the remaining one patient the information was not complete. He underwent vesicostomy after confirmation of PUV by MCUG. Information on subsequent treatment was missing but he had his vesicostomy closed as per new file notes and was voiding with a good stream per urethra and

hydronephrosis was resolving. All the 45 patients (100%) who received definitive treatment had electrofulguration as the mode of valve ablation as a primary procedure. One patient had a cold knife incision of the valves at a repeat valve ablation.

Urethral stricture as a complication occurred in 4 patients (8.9%) among the 45 patients who underwent electro fulguration of the vales. No patients developed urinary incontinence as a complication of surgical treatment of the valves. Among the 45 patients who had definitive treatment of PUV, 9 patients (20%) had residual valves which required repeat valve ablation. Seven of these patients had good stream of urine after repeat valve ablation, two of whom were lost to follow up immediately post operatively. Disregarding the residual valves as a complication of definitive treatment, the overall complication rate was 8.9% comprising urethral stricture alone. Table 4 shows the findings. It was found that the ratio of the number of patients with normal serum creatinine to those with raised serum creatinine increased approximately four times after treatment from 1.5 to 5.5 signifying the efficacy of the treatment (Table 5).

Among the 45 patients who underwent definitive treatment of PUV, 39 patients (86.7%) voided with a good stream. Two patients (4.4%) had a weak stream and 1 patient (2.2%) could only dribble. One patient (2.2%) remained diverted by suprapubic cystostomy. In two patients the information regarding the stream of urine after treatment was not available as the patient was lost to follow up immediately after discharge (Table 6).

Three patients died during the study period giving a mortality rate of 5.4%. All the deaths occurred among children aged 5 years and below. Note that the ages referred to here are the ages of patients at their last follow up visit or admission (for those who did not attend for follow up) and do not necessarily correspond with the age at presentation unless the patient died shortly at presentation. It was found that respiratory distress, urosepsis and renal failure overlapped as the causes of death each contributing two thirds of the causes.

Table 6. Urine Stream of Patients after Surgical Treatment

Stream of urine	Number of patients	Percentage
Good	39	86.7
Poor	2	4.4
Dribbling	1	2.2
No stream	1	2.2
Lost to follow up	2	4.4
Total	45	100.0

Discussion

In this study 56 files of patients were reviewed. Not all of them had the complete required data. Some of the patients got lost to follow up soon after discharge from initial treatment. Definitive treatment was instituted in 80.4% of the patients, which are 45 patients out of the 56 patients studied. An average of 6 new cases per year was noted. This is in keeping with a similar review done in Melbourne, Australia, between 1950 and 1979 where an average of 5 patients was seen per year[7]. In children's hospital of Philadelphia an average of 8 patients with PUV were seen per year in a span of ten years[8]. However a higher prevalence was observed in the Netherlands where an average of 25 patients was seen per year in a span of 5 years[9].

The age distribution of the patients with PUV is in keeping with the trend observed in other published series. Most patients present in the neonatal and infancy periods, followed by toddlers and lastly school age boys[1,3]. In the present study 57.1% of patients presented in the first year of life representing the neonates and infants, 26.8% of patients presented between the age of 2 and 5 years which represents the toddlers and the remaining 16.1% presented at age between 6 and 15 years representing the school age boys. In Melbourne, Australia, among 135 patients, 51.1% of them presented at age 0-1 years, 20 % presented between 1-5 years and 28.9% presented at age above 5 years[7].

The clinical presentation reveals a picture similar to that seen in some series in Europe and America. In this study 53 patients (94.6%) had hydronephrosis. In Atlanta Georgia, of the 22 patients studied, all (100%) had hydronephrosis. The prevalence of hydronephrosis in this study, however, is more than twice comparing with the Michigan study where it was 40%[4]. In this study 50 (94.3%) of the 53 patients who had hydronephrosis presented with bilateral hydronephrosis and only 3 patients presented with unilateral hydronephrosis all involving the left kidney. This compares with findings in Georgia where of the 22 patients, 18 (81.8%) had bilateral hydronephrosis and the remaining 4 (18.2%) had unilateral hydronephrosis[10]. The preponderance of left hydronephrosis and vesicoureteral reflux was observed in this study. Similar findings have been documented in other series. The syndrome of unilateral vesicoureteral reflux, hydronephrosis and a nonfunctioning kidney has been recognized and shows propensity to the left side [8]. Patients with no hydronephrosis are said to have a milder form of the disease according to Hendren's clinical classification system[9].

Vesicoureteral reflux was present in 13 (23.2%) of the 56 patients. In standard textbooks it is said to be present in 30 to 40% of the patients[3,5,]. In the Netherlands a rate of vesicoureteral reflux of 21.8% was found among 124 patients studied [9]. As stated earlier the propensity of vesicoureteral reflux to the left side has been recognized[17, 34]. In this study, of 13 patients with VUR, 11 patients could be stratified into left VUR or right VUR. Of these11 patients, 8 had left VUR and 3 had right VUR demonstrating propensity to the left.

In this study dribbling urinary stream was seen in 42 patients (82.1%) and urinary tract infection was seen in 33 patients (58.9%). This compares with a review of 47 patients in Michigan where 60% presented with urinary dribbling and 40% presented with urinary tract infection[4]. In the Netherlands urinary tract infection was found in 30% of 124 patients reviewed[9].

Type I posterior urethral valves was found to be the commoner of the two types accounting for 96.9% of the patients who underwent urethrocystoscopy at the time of valve ablation. This correlates with the generally accepted rates documented in standard textbooks of urology where type I posterior urethral valves is said to account for more than 95% of the cases and type III accounts for the remainder[3]. However in Ohio Crooks found the prevalence of type I posterior urethral valves among 36 patients studied to be 80.6%[11]. Type I posterior urethral valves is said to be associated with good prognosis[3].

In this study the initial treatment given was of mainly two types namely vesicostomy and initial valve ablation. These are acceptable modalities of initial treatment of PUV worldwide. Vesicostomy is said to be an effective and safe temporizing measure to manage babies with posterior urethral valves[12]. Vesicostomies are closed after valve ablation usually when the child has grown and the urethra can accommodate the size of available resectoscope. This may occur at the age one year and above. In this review 45 patients (80.0%) underwent definitive treatment of PUV either as an initial treatment or after an initial vesicostomy. All of these patients underwent electrofulguration of the valves as a primary treatment. Of the 9 patients who underwent repeat valve ablation for residual valves one underwent a cold knife incision of the residual valves. The accepted standard of treatment worldwide is ablating the valves under direct vision[13]. Electrofulguration was done by applying an electrocoagulating current to the valve through a resectoscope loop or Bugbee electrode. It is

recommended that the current delivered should be just enough to fulgurate the valves leaving the surrounding urethra and the sphincter intact thus preventing subsequent stricture formation and stress incontinence respectively[11]. Nijman and Scholtmeyer [9] recommended cold knife incision as the mode of valve ablation especially in neonates to prevent urethral stricture.

Regarding the complications of surgical treatment of PUV, urethral stricture was found to be the sole surgical complication of PUV in this series at a rate of 8.9%. Urethral stricture is a recognized complication of ablation of PUV. Similar findings were obtained in Columbus, Ohio, where a urethral stricture rate of 8.3% was observed post fulguration[11]. In Florida in a review of 28 patients treated by electrofulguration a higher rate was observed whereby 7 patients (25%) developed urethral stricture and all of these involved patients who were treated at age <1 year. No stricture developed in any of the patients older than 1 year at the time of treatment[12]. On the other hand Nijman, using a cold knife, reported no incidence of stricture in his series of 85 boys treated for PUV. The neonates and low birth weight babies are recognized to be at a higher risk for urethral stricture complication and so are the infants[14].

The outcome of surgical treatment of posterior urethral valves shows improvement of patients in terms of the level of serum creatinine. The study demonstrated a rise in the ratio of patients with normal serum creatinine to patients with raised serum creatinine eleven fold after treatment from 0.5 before treatment to 5.5 after treatment. The four patients with persistently elevated serum creatinine after treatment may represent a subset of patients with concomitant supravesical obstruction either at the ureterovesical junction or the pelviureteric junction. These are patients that may benefit from initial supravesical diversion to stabilize them before the definitive valve ablation or will require corrective surgery even after initial definitive valve ablation that is neoureterocystostomy or pyeloplasty[3].

The outcome of treatment as regards stream of urine showed a success rate of 86.7% with good urine stream. This is an acceptable success rate. The patients who had unsatisfactory stream of urine are those who had urethral stricture complication and included 2 patients with poor stream and one patient with dribbling stream. The other patient remained diverted by suprapubic cystostomy after an unsuccessful anastomotic urethroplasty. This demonstrates the morbidity associated with PUV[3].

An overall mortality rate of 5.4% was observed in the studied patients. Mortality was observed among the under fives only. No mortality was observed in age groups above 6 years. In a series of 135 patients in Melbourne, 22 patients died making an overall mortality rate of 16.3%[7]. In Ohio, of the 36 patients studied one patient died at home following valve ablation making a mortality rate of 2.8% in the series. The cause of death was thought to be sudden infant death syndrome[11].

The causes of death overlapped. These were respiratory distress, urosepsis and renal failure each contributing 66.7% of the causes. This contrasts with literature from the current standard text books which states that currently with diagnosis made early prenatally respiratory distress accounts for most deaths. In our study none of the patients studied was diagnosed to have PUV prenatally. This is probably the reason why urosepsis and renal failure featured among the causes[3].

Conclusion

The presentation of PUV at KCMC is comparable with the presentation elsewhere in the world. Vesicostomy and initial valve ablation are the common modalities of initial treatment of PUV at KCMC. Electrofulguration of the valves is generally the sole and acceptable modality of definitive treatment of posterior urethral valves adopted at KCMC. Worldwide, electrofulguration is the standard treatment against which any other treatment should be compared. Urethral stricture was the sole complication of surgical treatment of PUV and it occurred at a rate of 8.9%. This rate compares with studies done in Europe and America and is within acceptable limits. The success rate of 86.7% of treatment of PUV compares favorably with published results worldwide.

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