epispadias: alternative to primary bladder closure.

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The main objective of this study was to evaluate the outcome of management of bladder exstrophy and epispadias with continent urinary diversion. A total of 15 children, 10 females and 5 males underwent continent urinary diversion at Kilimanjaro Christian Medical Centre (KCMC) between 1985 and 1997. Their ages ranged between one month and 13 years with an average of 5.4 years. Eight (53%)of them had exstrophy epispadias complex, 4 (27%) had incontinent epispadias, 2 (13%) presented with neurological conditions and 1 (7%) had traumatic destruction of the bladder neck and urethra. Seven (47%) had Mainz pouch II procedure, 6 (40%) underwent the classical ureterosigmoidostomy while 2 (13%) had appendicovesicostomy. The mean duration of follow up was 3.2 years. Three patients developed mild non-progressive hydronephrosis, which required no surgical intervention. One patient with a solitary kidney developed ureteral stenosis at the implantation site that was undiverted. Metabolic acidosis was well compensated with none of the patients requiring sodium bicarbonate supplements. Only one patient had night soiling that required wearing of diapers. Our experience with continent urinary diversion in children with other benign bladder conditions has been favourable and in our view, it offers a viable treatment method in children with exstrophy epispadia complex.

Introduction

In children with epispadias-exstrophy complex, the aim of management is to achieve urinary continence, preserve upper urinary tracts and provide adequate external genitalia. Despite the fact that various methods of treatment have been reported, the ultimate surgical procedure remains elusive or even debatable. Most urologists in the West prefer bladder closure with or without osteotomy in the first 24-72 hours of life and this is followed by bladder neck reconstruction and bilateral ureteroneocystostomy with reconstruction of the external genitalia at 2 to 5 years. Most of the time additional procedures are required to achieve continence. Continence rate after these procedures has been reported to range from 32 to 80%[4]. However, in most of these series

patients are considered continent even if they are only capable of emptying their bladder by clean intermittent self-catheterisation, which in males can be complicated, by recurrent epididymitis and urethral stricture[5].

Primary bladder closure and subsequent bladder neck reconstruction has also been reported to be complicated by hydronephrosis, reflux and poor preservation of the upper urinary tracts[6].

In developing regions where the health budget is meagre and where patients have to travel long distances to reach referral hospitals manned by consultants, which also makes follow-up of patients difficult, it is not feasible to schedule patients for multiple staged and therefore expensive surgical procedures. It is for those reasons that we decided to seek for simple and appliance free techniques of continent urinary diversion rather than primary bladder closure. The rate of complete continence after ureterosigmoidostomy has been reported to be as high as 92.3%[7]. Since 1992, we have been using the sigma rectum pouch (Mainz pouch II), a modification of ureterosigmoidostomy as our treatment of choice. For the paraplegic paediatric patients, our procedure of choice has been appendicovesicostomy with or without bladder augmentation as these patients have insufficient anal sphincter tone. In this paper we report data obtained from children who underwent urinary diversion over a period of 12 years at Kilimanjaro Christian Medical Centre (KCMC).

Patients and methods

Continent urinary diversion for non-malignant conditions at KCMC during the period under review starting from 1983 were studied. Most of the patients were referrals from district and regional hospitals from all over Tanzania. For the classical ureterosigmoidostomy,a midline laparotomy incision was used, the colon was opened antimesenterically and the ureters were tunnelled submucosaly for antireflux mechanisms as described by Coffey[8].

For the sigma rectum pouch, after opening of the rectum and sigmoid for 10 to 12 cm proximal and distal to the rectosigmoid junction, the median margins were sutured. Fixation of the rectosigmoid junction to the sacral promontory as well as parallel ureteral implantation guaranteed a straight ureteral path and prevented kinking and the subsequent obstruction.

The use of the appendix as a continent vesicostomy (appendicovesicostomy) was first described by Mitrofanoff in 1980[9]. A continent valvular mechanism was achieved with a submucosal tunnel of the mobilised appendix into the bladder with the other end brought out to the skin as a catheterizable stoma.

Postoperatively, the children were followed up by regular upper urinary tract imaging analysis of serum creatinine, blood urea nitrogen electrolytes and rate of bicarbonate usage sphincter control and the need for re-operation. In our study, continence was defined as having regular normal interval use of toilet, voluntary control of the urinary stream and regular normal regular underclothes without diapers.

Results

During the period under review, a total of 15 children underwent continent urinary diversion for non-malignant conditions. There were eight cases of exstrophy epispadia complex, 4 of incontinent epispadias, 2 of neurogenic conditions and one of traumatic destruction of the bladder neck and urethra. There were 10 females and 5 males. Their age at diversion ranged between one month and 13 years with a mean age of 5.4 years and a mean follow up of 3.2 years (range of 1 to 13 years).

Sigma-rectum pouch (Mainz pouch II) was performed in 7 cases, 6 had classical ureterosigmoidostomy while two patients had appendicovesicostomy. Post-operatively, two patients developed superficial wound sepsis. No other early postoperative complications were recorded.

Long-term complication

UPPER URINARY TRACTS:

All patients were followed up with intravenous urography and more recently by renal ultrasound. Mild unilateral, non-progressive hydronephrosis developed in three patients, none requiring surgical intervention. All patients had their serum creatinine and blood urea nitrogen within normal range. No cases of pyelonephritis or urolithiasis were recorded.

One female child with incontinent epispadias who had undergone ureterosigmoidostomy developed stenosis at the ureteral implantation site. This child had a solitary kidney; she was undiverted and simultaneously underwent Marshal Marchet bladder neck reconstruction six months after the primary diversion.

METABOLIC COMPLICATION:

Patients had regular determinations of serum electrolytes (Na+, K+, Cl-) and recently CO2 levels. All patients were metabolically well compensated with none requiring sodium bicarbonate supplements.

CONTINENCE:

All seven children who had sigma rectum pouch were fully continent. Among the six who underwent ureterosigmoidostomy, one had nighttime soiling requiring diapers. This was a female child who had exstrophy epispadias complex and was being considered for conversion to sigma rectum pouch. The two paraplegic patients who were done appendicovesicostomy were fully continent, doing intermittent catheterisation through their stomas. No stoma complications had developed.

Discussion

In most developed countries, the standard treatment for exstrophy epispadia complex is primary bladder closure during the neonatal period with subsequent bladder neck reconstruction at the age of 2 to 5 years. Often additional procedures are required to achieve continence. Despite these multiple procedures however, continence rate vary, ranging between 32 and 80%. Primary bladder closure and subsequent bladder neck reconstruction is not always successful. The reason for the failure of primary bladder closure is the fact that creation of a functional sphincter mechanism is ultimately impossible. To achieve complete continence, further procedures beyond Young - Dees-Leadbetter bladder neck reconstruction are often necessary. These additional procedures include intestinal bladder augmentation, a pubovaginal sling or artificial sphincter implantation[10]. In those cases where outlet resistance is high, clean intermittent self-catheterisation is required. This is an additional risk procedure especially for boys in whom it has been associated with increased rate of epididymitis, urethral stricture and even perforation of the augmented bladder[5]. Another hypothesis for failure of primary bladder closure may be insufficiency of the detrusor muscle itself. Hollowell et al[11] demonstrated urine leakage due to involuntary detrusor contractions in 10 out of 18 patients with poor continence.

Normal bladder function during filling and contractile voiding was demonstrated in only 8% of their patients. They therefore concluded that "evolution to completely normal lower urinary

tract function after bladder neck reconstruction is unrealistic expectation for the majority of children with exstrophy and severe epispadias."

In our series, metabolic complications were minimal and so were changes in the upper urinary tracts. Continence rate after sigma rectum pouch was 100% while after ureterosigmoidostomy, it was 83%; findings which were comparable with the continence rates of 80 - 100% reported in literature from elsewhere. Since 1992, we have been performing sigma rectum pouch in preference o the classical ureterosigmoidostomy. For the paraplegics with insufficient anal sphincter unction, our procedure of choice has been appendecovesicostomy. The overall continence rate among our 15 children was 93%.

In conclusion, we have presented our 12 years' experience with primary urinary diversion in children with exstrophy epispadias complex. A continence rate of more than 90% was achieved with good preservation of upper tracts as well as adequate metabolic compensation. Our experience has therefore been most favourable and we conclude that continent urinary diversion is a viable alternative treatment method for children with bladder exstrophy epispadia complex.

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