Comparative study between vesicostomy and fulgeration of posterior urethral valve in uraemic infants

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Abstract

Introduction: Posterior urethral valves is the most common structural cause of urinary outflow obstruction in pediatric practice leading impaired renal function in a third of patients after the resolution of obstruction.The issue of urinary diversion is somewhat controversial as endoscopic valve fulgeration is considered the treatment of choice.If the infant is too small, cutaneous vesicostomy can be performed as a temporary measure. The vesicostomy has proved to be a safe and efficient treatment, with long-term results in preserving renal function equal to those of primary valve ablation.

The aim of our study: is to evaluate and compare the outcome of posterior urethral valve fulgeration versus vesicostomy in patients during first six months of life with serum creatinine above 0.5mg/dl in terms of renal functions and upper urinary tracts dilatation.

Patients and methods: This study included 30 patients.The 1st group is classified as group A were treated by fulgeration and the 2nd group is classified as group B were treated by vesicostomy.All the patients were evaluated after management as regards improvement in their creatinine levels, attacks of urinary tract infections, improvement in hydronephrosis and reflux grades.

Results: After management laboratory and radiologic improving occurred in most cases with varying degrees.Vesicostomy was the better surgical option with better results.There were statistical significant differences towards vesicostomy group in postoperative serum creatinine level and degree of reflux, otherwise both groups showed almost same results as there was no significant statistical difference between both groups regarding post operative attacks of urinary tract infections and changes in hydronephrosis in U/S.

Conclusions: Management of PUV can either be done by vesicostomy or 1ry valve fulgeration even in young infants during first six months of life with creatinine level above 0.5mg/dl.In young infants with high serum creatinine level and severely affected upper urinary tracts with either hydronephrosis or reflux with or without urinary tract infections; vesicostomy should be considered as the treatment of choice.

Key words: Posterior urethral valve-fulgeration-vesicostomy- uraemic

LIST OF FIGURES

List Of Tables

LIST OF ABBREVIATIONS

COPUM: Congenital obstructing posterior urethral membrane

CRF: Chronic renal failure

DMSA: Dimercaptosuccinic acid

DTPA: Diethylenetriaminepentaacetic acid

ESRD: End-stage renal disease

MAG3: Mercaptoacetyl triglycine

PUV: Posterior urethral valve

RNC: Radionuclide cystography

UTIs: Urinary tract infections

VCUG: Voiding cystourethrography

VUR: Vesicoureteral reflux

Contents

INTRODUCTION

Historical background

Posterior urethral valve (PUV) was 1st described by Morgagni in 1717. [1]

Earliest description is credited to Langenbeck in 1802, who descriped valve-like folds in autopsy specimens without interfering any clinical significance. [2]

After thirty years the subject was discussed by Velpeau (1832), describing anatomical specimens with valve-like folds that might be of clinical importance.70 years later comprehensive discussion of valves in 1870 by Tolmatschew who recognized this as a pathological entity and propose theory about its embryology. [3]

Young et al described it as a clinical and pathological condition in 12 patients in 1919. [4].

This classic anatomical description of the valves was the subject of a recent review. [5]

Posterior urethral valve is the commonest cause of congenital structural obstruction of urethra in pediatric practice, with incidence of 1 in 3,000 to 1 in 8,000 male births.Elevated renal function is the major long term complication, it occurs in 1/3 of patients after relief of obstruction.[6]

In spite of decrease in mortality rate there is still considerable long-term morbidity due to secondary pathology of the valves.

Up to 43% of patients by the age of 30 years are complicated with chronic renal failure (CRF) or end-stage renal disease (ESRD).

It is the 2nd most common cause of CRF & ESRD in children after vesicoureteric reflux.

Renal function is affected by several factors as age at presentation, prenatal diagnosis, and initial renal function, presence of reflux at the time of diagnosis, upper tract obstruction, urinary tract infections (UTIs), treatment option and bladder dysfunction. It may develop early in infancy, but in the majority progresses slowly over years.

Several studies have identified predictive factors of future renal function in children with PUV.Positive predictive factors are serum creatinine less than 1 mg% at 1 year of age, unilateral vesicoureteral reflux (VUR), and late presentation.Negative predictors are early presentation, bilateral VUR, renal dysplasia, and delayed urinary continence.[7]

In many patients these valves are diagnosed in the neonatal period because of abdominal masses, infection and voiding difficulties.Two types of symptomatic presentation and clinical manifestations are seen in these children. The first group shows obstructive symptoms including straining during urination, intermittent voiding with reduced urinary flow rate, and a palpable midline bladder. The second group presents with infectious signs including septicemia, failure to thrive, and urinary tract infections, which are the most prominent features of this disease. In the neonatal and infantile periods, the primary obstructive symptoms are more prevalent.

Patients may have severe fluid and electrolyte disorders and renal insufficiency. Bladder outlet obstruction may be so severe as to cause trabeculation, detrusor hypertrophy, formation of diverticula

and secondary bladder neck hypertrophy. Chronic distension and elevated intravesical pressure leads to severe hydroureteronephrosis. Ultrasound findings are of unilateral or bilateral hydroureteronephrosis and a thick walled, poorly emptying or persistently full bladder.

Diagnosis of PUV improved a lot after maternal ultrasonography, as most of the cases detected prenatally. But, others are presented lately who are assumed to represent the milder clinical spectrum of the disease, with a lesser degree of obstruction but better outcome. Children who are not diagnosed prenatally may present with infection, hydronephrosis, ascites, and/or thick distended bladder.Generally, these patients present with voiding dysfunction and urinary tract infections. In severe cases, renal failure and respiratory distress may occur. [8]

The ‘gold-standard’ investigation for documenting PUV is the Voiding cystourethrography (VCUG) as it shows the anatomy and gives sufficient data about the function of the bladder (neurogenic, capacity, etc.) bladder neck, and urethra.

The boys are catheterized as soon as possible (generally a 6 Fr catheter is best) and fluid, electrolytes and acid/base balance are monitored and corrected aggressively (post obstructive dieresis), with fluid balance maintained by replacing urine output.

Images of the urethra during voiding are necessary to make the correct diagnosis and to differentiate from neuropathic bladder, urethral stricture, and anterior urethral obstruction. It is not necessary to remove the catheter during voiding to observe the valves. [9]

After successful initial bladder drainage and when the patient's medical condition has stabilized, the next step is to permanently destroy the valves.There are different management options which include primary valve fulgeration or vesicostomy followed by endoscopic valve fulgeration after the general condition has been improved depending on the age, serum chemistry, size of the urethra and availability of equipments.The issue of urinary diversion is somewhat controversial as endoscopic valve fulgeration is considered the treatment of choice.If the infant is too small for safe instrumentation for valve ablation, a cutaneous vesicostomy can be performed as a temporary measure. Temporary vesicostomy drainage allows the urologist to incise the valves later when the patient is older and healthier. The vesicostomy has proved to be a safe and efficient treatment, with long-term results in preserving renal function equal to those of primary valve ablation.

The aim of our study is to evaluate and compare the outcome of posterior urethral valve fulgeration versus vesicostomy in patients during first six months of life with serum creatinine above 0.5mg/dl in terms of renal functions and upper urinary tracts dilatation.

We need to find out the answers for several important questions:

Which of both management options has better outcome?

Is endoscopic valve fulgeration the treatment of choice?

Does vesicostomy have a role in improving renal function?

Will our choice change in cases with high serum creatinine level and severely affected upper tracts especially during first six months of life?

EMBRYOLOGY OF THE URETHRA

At week 6 of gestation male urethra arise to the surface of the genital tubercle with the urogenital sinus cavity. Then endoderm becomes a solid plate of cells, then tubularizes 1st proximally then distal to form the phallic urethra.At week 14 it completes its development. [10-12]

The urethra is divided into 4 parts differing in anatomy and predominant cell type contained within.Prostatic and membranous urethra, derived from the urogenital sinus and with transitional epithelium.Then bulbar urethra and pendulous urethra, derived from the urethral plate on the ventral aspect of the genital tubercle and lined with transitional epithelium proximal, changing to simple squamous epithelium distal.

The fossa navicularis originate from the urethral plate as it traverses the glans penis,with stratified squamous epithelium. [13]

EMBRYOLOGY OF PUVS

Tolmatschew described the valves as “overgrowths of the normally present anatomical folds and ridges in the urethra.” . It is pathological only after attaining sufficient height and size. [14]

Wilckens differed in the origin of these valves. He said they were remnants of the cloacal membrane after division into the urogenital sinus, perineum and rectum. [15]

Bazy said its site corresponded to the site of the urogenital membrane that separated the anterior and posterior urethra during development. [16]

Lowsley suggested a theory of development depending on microscopic examination before any instrumentation.It showed a membrane arising from the distal verumontanum dividing into 2 leaflets, closing all circumference of the urethra except a small posterior opening in the membrane.After opening up into the prostatic lumen the sheath normally continued distal, its fibers dissipating inferior on the urethral floor or lateral on the urethral walls & valves came from the same connective tissue sheath[17]

Lowsley thought it was due to an anomaly of wolffian or müllerian duct origin.

In 1918 Watson supposed etiology was an arrested phase of normal development instead of a phase of anomalous development. [18]

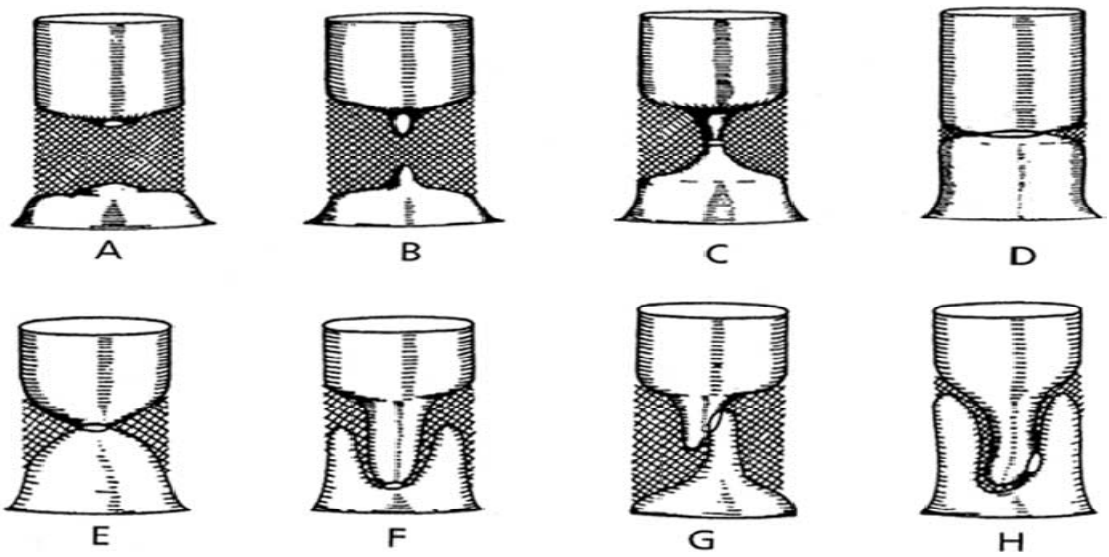

Fig (1) PUV formation due to persistent urogenital membrane (hashed markings), as described by Stephens [19]

A to D, normal canalization. E to H, variations in valvular appearance.

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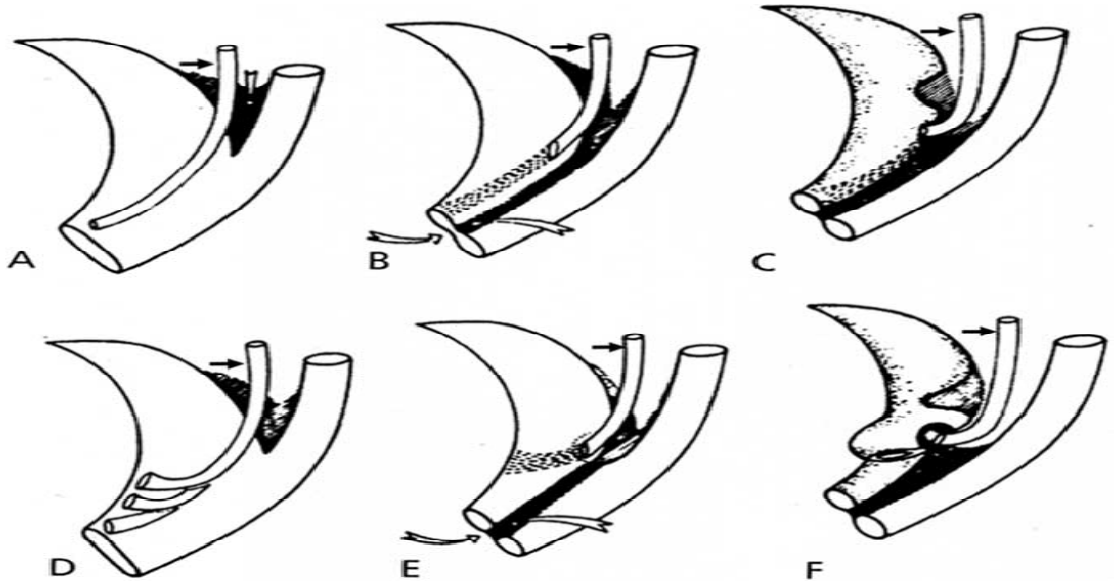

Fig. (2) PUVs as result of abnormal integration of mesonephric ducts into posterior urethra, as described by Stephens [19]

A to C, normal development. D to F, anomalous integration of mesonephric ducts (arrow) and subsequent valve formation.

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