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MAIN TOPIC

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Long-term results of surgery for posterior urethral valves: a review

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Abstract Although the techniques for valve resection have been refined and the short-term management of patients with posterior valves has improved remarkably, there is growing concern about long-term outcome. Prenatal diagnosis has added to the urgency of predicting long-term outcome. This paper reviews all the important long-term data available with the aim of providing a concise picture for the reader, vis-a-vis long-term renal function, the role of proximal diversion in long-term outcome, bladder function and incontinence, the place of renal transplantation, prenatal diagnosis, intervention and prediction of renal function and sexual function and fertility.

Keywords Posterior urethral valves · Prenatal diagnosis · Incontinence · Fertility

Introduction

Over the decades, the techniques for posterior urethral valve (PUV) fulguration have undergone several refinements, and the problems associated with resection of the valves per se have been overcome. Earlier, when blind and inaccurate techniques were used, the results of PUV were much worse than they are today. At present, the short-term prognosis is good. However, there is growing concern that the long-term prognosis may not be so good. Moreover, the possibility of prenatal diagnosis of PUV has made it necessary to be able offer a prognosis for long-term renal and bladder function and fertility and to counsel parents accordingly.

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Renal function

Due to the rarity of PUV and the difficulty of following children into adulthood, there are very few medical units in the world (especially the Western world) with sufficient experience to provide long-term data on large numbers of boys. In a 1990 issue of the *Urological Clinics of North America*, Parkhouse and Woodhouse presented an excellent review of the long-term status of patients with PUV [1]. Up to 1990 only three important series were published: Krueger et al. [2] reported on 74 boys followed for a mean of 7.4 years in 1980 and Egami and Smith [3] followed 93 boys for a mean of 6 years and published their findings in 1982.

The only study to include many postpubertal boys was published in 1988 from the Great Ormond Street Hospital for Sick Children (GOS) [4], which included 98 boys followed for a mean period of 15 years. In the GOS series 50 boys were more than 18 years old at follow-up, and 26% of these postpubertal boys were in chronic renal failure (CRF) or end-stage renal disease (ESRD). In 1996, Smith et al. [5] published data on the long-term outcome of PUV treated with primary valve ablation and observation. These studies identified three major risk factors for late development of renal failure: (1) bilateral vesicoureteric reflux (VUR); (2) presentation in the 1st year of life; and (3) diurnal incontinence at the age of 5 years.

Pathogenesis of late-onset renal failure

Initially, late onset CRF was thought to be secondary to the increased metabolic demands of puberty, but this is probably not true, as renal deterioration precedes maximum somatic growth. The currently accepted theories of late-onset CRF are: (1) increased single-nephron filtration rates (hyperfiltration); (2) reflux nephropathy; and (3) persistent abnormal bladder function.

The role of proximal urinary diversion in the long-term results of PUV

In infants with PUV in whom renal function fails to normalize following decompression of the lower urinary tract, it has been assumed to be due to relative ureterovesical junction (UVJ) obstruction because of a thick-walled bladder, and supravesical diversion (SVD) has been customarily recommended. Recently, opponents of upper-tract urinary diversion have argued that persistent elevation of serum creatinine after bladder decompression reflects renal dysplasia and that these patients do not benefit from high urinary diversion.

Tietjen et al. [6] from the Mayo Clinic recently evaluated 26 patients with PUV to determine the true incidence of fixed or permanent UVJ obstruction and the renal prognosis for infants treated with proximal urinary diversion. They concluded that in neonates with PUV who undergo proximal diversion fixed UVJ obstruction is rare, a renal biopsy invariably demonstrates dysplasia, and ESRD frequently develops despite proximal diversion. They question the necessity of SVD in PUV.

There is also growing concern about recovery of bladder function after SVD, especially if bilateral. Persistent bladder dysfunction prevents renal function from returning to normal. Thus, many centers are adopting a ''no-diversion'' policy. On the other hand, several centers in the developing world, where follow-up is difficult and patient compliance for long-term urinary chemoprophylaxis is doubtful, are too liberal in performing SVD. In our unit, we do not subscribe to either the ''no-diversion'' view, which seems to be emerging in the West, or the traditional ''over-diversion'' view. We believe in selected diversion, and prefer to perform SVD as infrequently as possible.

Renal transplantation (RT) and post-transplant renal function in PUV

RT is required in many patients with PUV who develop CRF due to renal dysplasia, VUR, persistent bladder-outlet obstruction, or vesical dysfunction. In PUV the rates of graft loss after RT have been reported to be higher than in age-matched controls [7], presumably due to persistent bladder dysfunction [7–9]. However, Solomon et al. [10] recently assessed the long-term efficacy of RT in PUV and concluded that it is not associated with a high failure rate.

Prenatal diagnosis, intervention, and prediction of renal function

Prenatal diagnosis and intervention offer promises and pitfalls. Prenatal diagnosis has increased with the widespread use of ultrasound (US), so that up to two-thirds of infants with PUV were identified in this way in the 1990s. The early prognosis for renal function in prenatally-detected PUV is associated with the gestational age at which dilatation first becomes apparent [11]. Recently, further reports on the qualitative aspects of antenatal renal scans have also enhanced the predictive value of US [12]. US findings that predict poor postnatal outcome are: (1) early detection of upper-tract dilatation; (2) moderate to severe upper-tract dilatation, defined as a renal-pelvic anteroposterior diameter of 10 mm or greater; (3) increased echogenicity of the renal parenchyma; and (4) cystic changes in the renal parenchyma.

When second-trimester US predicts a poor functional outcome, parents and counseling physicians have three potential options including nonintervention, fetal intervention (vesico-amniotic shunting), and termination of pregnancy. When pregnancy is allowed to continue (nonintervention) spontaneous abortion or intrauterine death ensues in some cases. In pregnancies that proceed to term the neonate may have respiratory distress due to pulmonary hypoplasia. Those that survive the neonatal period will have CRF in infancy or early childhood, requiring frequent hospitalization, dialysis, and RT.

To date, the results of in-utero treatment of fetuses with PUV have been universally disappointing. In addition to US findings, identification of suitable candidates for prenatal intervention has centered on the use of fetal urinary electrolyte concentrations. Initially, it was shown that urinary sodium of less than 100 mmol/l, chloride below 90 mmol/l, and osmolality below 210 mmol/kg were favorable factors for shunting. Recently, it has been shown that sequential urine analysis is more accurate in determining which fetuses will benefit from prenatal intervention [13, 14]. Attitudes to termination of pregnancy for fetal anomalies vary from country to country, reflecting the prevailing legal, moral, and religious climate.

Urinary incontinence after valve ablation

Urinary incontinence has been a common problem in boys after resection of PUV; this observation has been reported by many authors [15–17]. It was assumed to be due to external-sphincter weakness as a result of primary surgery or to bladder-neck incompetence after bladder-neck resection, which was widely practiced in the past. Although numerous related causes exist, post-obstructive bladder dysfunction has gained recognition as the principal factor in the etiology of incontinence after valve surgery [18, 19]. The reported incidence of significant voiding dysfunction in boys after surgery for PUV varies from 13% to 38% [2, 16–18, 20].

A GOS study of 42 patients with PUV investigated for a mean period of 5 years after their initial presentation by videourodynamics suggested that despite adequate relief of urethral obstruction, urodynamic abnormalities were identifiable in approximately 75% of cases [1].

Bauer et al. first reported the specific urodynamic findings in boys with PUV and voiding disturbances in 1979. This concept has now been accepted and supported by the recognition of the "valve-bladder syndrome" a term used specifically for the situation of persistent hydronephrosis after valve ablation in the absence of UVJ obstruction. The reason for this persistent hydronephrosis is a small-capacity, high-pressure bladder.

This persistent bladder dysfunction has now been identified as a preventable cause of late-onset RF, the implication being that all boys with urinary incontinence after resection of PUV must be investigated urodynamically. Three major categories of bladder dysfunction may cause voiding symptoms: myogenic failure, detrusor hyperreflexia, and bladder hypertonia, and must be properly treated medically or surgically.

Sexual function and fertility

There are five important reasons why patients with PUV may have impaired sexual and reproductive function: (1) cryptorchidism, which is more common than in the general population; (2) vasal reflux as a result of high pressure in the posterior urethra proximal to the valve, which affects various components of semen; (3) retrograde ejaculation; (4) decreased libido and sexual function due to CRF; and (5) damage to the nerves of erection around the prostate.

In 1989, Woodhouse et al. reviewed 21 adults who were previously treated for PUV; 18 had normal erections [21]. Semen and urine analyses suggested that these patients did not have retrograde ejaculation. The data did not allow any definite conclusions, but it is certainly possible for patients with a history of PUV to father children.

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