Transpl Int (1996) 9 [Suppl 1]: S86–S89

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Kidney transplantation to patients with congenital malformations of the distal urinary tract

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Abstract Fourteen of 1000 consecutive kidney transplant patients had congenital malformations affecting the bladder or urethra: six had congenital valvulus of the urethra, two congenital sclerosis of the bladder outlet, and six a neurogenic bladder. Pretransplant surgey had been performed in all patients: reimplantation of ureter ($n = 11$), resection of congenital valvulus ($n = 7$), and nephrectomy ($n = 6$). Four patients had an intestinal bladder. Age was 0–17 (median 1) years at diagnosis. Follow-up time was 3–10 (median 5) years. Special transplant surgery techniques were required in five patients. Patient survival after 2 years was 100 % and graft survival 93 %. No graft was lost due to outflow obstruction, infection or other causes related to the underlying disorder. Late technical problems were seen in two patients. Urinary tract infections were reported in 13 patients before transplantation and in eight after. Results of transplantation were excellent. Infections and surgical problems had a minor impact on outcome.

Key words Kidney transplantation · Congenital malformation · Distal urinary tract

Introduction

Almost irrespective of the original disease, kidney transplantation is the superior treatment for patients with end-stage renal disease. However, the various renal diseases may influence the transplant procedure and outcome in different ways. In patients with distal urinary tract malformations, problems such as abnormal anatomy, bladder dysfunction, and urinary tract infection (UTI) might cause complications and thus have an impact on the outcome of transplantation. This report presents our experience of kidney transplantation in patients with congenital malformations affecting the distal urinary tract.

Patients and methods

We have reevaluated the underlying disease in 1000 consecutive patients who received 1095 kidney transplants in Göteborg between 1985 and 1993 [10]. This was done by retrieval of data from the patients' records in the Transplant Unit and the Renal Unit and/or Pediatric Units where they were first investigated. In this population, 14 patients had well-defined distal urinary tract malformations: congenital valvulus of the urethra, sclerosis of the bladder outlet, and neurogenic bladder. In the neurogenic bladder group, two patients had myelomeningocele, two spina bifida, one megaloureters with neurogenic bladder, and one patient had a neurogenic bladder with concomitant absence of perineal reflex and reduced sphincter tone in combination with moderate mental retardation. Hydronephrosis had been found in eight of the patients. Patients with hydronephrosis without defined malformation in the distal urinary tract are not included in this study. Demographic data are shown in Table 1.

The patients were 14–44 (median 25) years of age at the time of transplantation. Twelve transplants were first transplants and two, retransplants. Kidneys from living related donors were used in six recipients of primary transplants. Two patients had preemptive transplantations. All patients were supervised for UTI after the

Table 1 Demographic data for kidney transplanted patients with congenital malformations of the distal urinary tract ($n = 14$) separated according to type of malformation

Diagnosis	Number of patients	Male: female ratio	Diagnosis [age years, median (range)]	Start of renal replacement therapy [age years, median (range)]
Congenital valvulus of the urethra	6	6/0	1 (0–17)	22 (12–44)
Congenital sclerosis of the bladder outlet	2	2/0	2 (2–2)	25 (22–28)
Neurogenic bladder	6	4/2	3 (0–7)	30 (14–36)

Table 2 Pretransplant operations on the urinary tract in kidney transplanted patients with congenital malformations of the distal urinary tract ($n = 14$)

Type of operation	Number
Kidney	
Nephro- and/or ureterectomy, uni/bilateral	6
Ureter	
Cutaneous ureterostomy	3
Disconnection of cutaneous ureterostomies	1
Bladder	
Reimplantation of ureter(s)	12
Construction of intestinal bladder	
Bricker	2
Kock	2
Disconnection of Bricker bladder	1
Implantation of Scott's sphincter	1
Plastic operation of the bladder	1
Extirpation of bladder diverticle	1
Bladder neck incision/resection	4
Urethra	
Urethral valvulus operation	7
Exstirpation of urethral diverticle	1
Total	42

transplantation with a urine culture test at every out-patient control. Treatment was according to bacterial resistance patterns for short periods or with low-dose long-term prophylaxis.

Routinely, the ureter was anastomosed to the urinary bladder using an extravesical ureteroneocystotomy, as described by Röhl and Ziegler [12]. The immunosuppressive protocol was a combination of cyclosporine, prednisolone, and azathioprine [10]. Follow-up was until September 1995, which gives 3–10 (median 5) years of observation time.

Contemporary non-diabetic controls, two per patient, were picked from the consecutive file of patients, one by moving forwards, one backwards. Controls were matched for age ± 5 years, sex, kidney source (cadaveric donor or living donor), and transplant number.

Results

Pretransplant operations

Surgical intervention, aimed at correcting the congenital deformity or problem, had been performed before transplantation in all 14 patients on a total of 42 occasions: reimplantation of ureters, resection of congenital valves or bladder neck, construction of an intestinal bladder, and nephrectomy. These procedures are specified in Table 2. Operations were distributed in the whole pretransplant life-span with some procedures in infancy, others just prior to transplantation.

Special procedures at the transplantation

At the time of transplantation, three patients had ileal conduits: one a Bricker bladder, two a Kock's pouch with continent nipple. A fourth patient had previously had a Bricker bladder but at the time of transplantation this had been disconnected and the patient practised triple voiding and manual, suprapubical pressure.

As a consequence of the original disease, special procedures were required during the transplant operation in five patients (Table 3). A cutaneous ureterostomy was performed in one patient and ureteroileoanastomosis in the three patients with ileal conduits. The patient who had previously had a Bricker bladder used a suprapubical catheter to the bladder in the first postoperative period.

Bladder function

There was no immediate change of bladder status or capacity following kidney transplantation. Micturition was satisfactory in seven patients but seven had bladder dysfunction. Four of these had urinary diversion, namely the two patients with a Kock's pouch, the one with a Bricker conduit, and the one with a cutaneous ureterostomy before and after transplantation. Two patients applied triple voiding and/or external pressure. One patient with incontinence had used napkins and initially continued with them.

During the continued follow-up, the patient with napkins, 5 years after transplantation, received, to his own satisfaction, a suprapubical catheter. Another female patient with a neurogenic bladder and native voiding before transplantation started clean intermittent catheterization 3 years after transplantation.

Two patients had late technical problems necessitating additional surgery. Due to signs of outlet obstruction, one patient had a revision of a ureterostomy and one a transposition of ureter in an intestinal bladder and simultaneous augmentation of the bladder.

Table 3 Special surgical procedures during the transplant operation in kidney transplanted patients with congenital malformations

Type of operative procedure	Number	Reason for procedure
Ureteroileoanastomosis	2	Kock's bladder
	1	Bricker bladder
Cutaneous ureterostomy	1	Bladder incontinence
Suprapubical catheter	1	Neurovesical dysfunction

Outcome of transplantation

All patients were alive at the end of the study period. In the control group, one patient was dead due to malignant lymphoma and sepsis.

Graft survival at 2 years was 93 % in the patient group and 82 % in the control group (not significant). During the whole study period, five patients lost their grafts, two due to chronic vascular rejection and three due to chronic glomerulopathy, all confirmed by transplant biopsies. The underlying disease in four of these cases was congenital urethral valve disease. No graft was lost due to outflow obstruction, UTI or other causes related to the underlying disorder. Four of five patients have undergone retransplantation. These grafts function, as at September 1995, which means that 13/14 patients have functioning kidney transplants.

In the control patient group, nine grafts were lost during the study period: one acute rejection and arterial thrombosis, four chronic rejection, three recurrence of the original disease. One graft was never biopsied. Four of nine patients had a retransplant performed, and three patients are on the transplant waiting list.

UTI

In the pretransplant period, 13 patients had urinary infections: ten had a history of acute pyelonephritis or urosepsis and three had recurrent distal urinary infections with symptoms. Six patients were reported with previous vesico-ureteral reflux uremia (VUR). All of them had had acute pyelonephritis.

In the posttransplant period, 3–10 (median 5) years, eight patients have had infection problems. Six patients had prophylactic treatment with antibiotics, permanently or intermittently. Two patients had acute pyelonephritis and in one of them nephrectomy of the native kidneys was performed.

Discussion

Patients with distal urinary tract malformations constitute 1.4 % of our kidney transplanted population [10]. Only two of the patients reported in this paper was pediatric in the sense that they received transplants before the age of 18 years. Although congenital malformation of the distal urinary tract is "a childhood disease", our patients are adult at the time of kidney transplantation. This indicates that good medical treatment and surgical therapy had delayed and reduced the need for dialysis and kidney transplantation.

For comparison of incidences of congenital malformation of the distal urinary tract in different renal replacement therapy populations, the National Registries from United States of America, Canada, Japan, and Europe [5] are no help because they do not classify the patients into subgroups, only into groups headed, for example, chronic pyelonephritis or interstitial nephritis. In the registry from the North American Pediatric Renal Transplant Cooperative Study, where 2037 children (< 17 years) received kidney transplants from 1987 to 1992, 17 % had "obstructive uropathy" [9]. In the United States Renal Data System, 4.7 % of the patients had "congenital obstructive uropathy" as the cause of pediatric (< 20 years of age) end-stage renal disease (ESRD) [13]. These figures are also difficult to compare with ours due to ambiguities of definition.

There was no mortality in the patient group. This is probably related to their young age. Graft survival at 2 years was satisfactory and not different from the controls but superior to overall results of 75.6 % for the 1095 transplantations performed in the same period. The good graft survival is probably also an effect of youth. No graft loss was related to underlying disease. All retransplantations turned out successfully. A small series also reporting excellent results of kidney transplantations to patients with problems in the lower urinary tract has previously been published [3, 4, 6, 8].

In accordance with our results, Reinberg et al. [11] found that, in children who received kidney transplants because of posterior urethral valve uremia, failed grafts showed rejection, not obstructive lesions, when analyzed histologically. He also found a 5-year graft survival rate in these children comparable to that for children with VUR. Kalicinski et al. [7] found the same graft survival for transplanted children with "urologic" and "nonurologic" ESRD.

All patients had a history of repeated surgery from infancy to the day of transplantation. Correcting operations were performed with the hope of abolishing mechanical obstruction, preventing further VUR, and putting an end to recurrent infectious foci. Today, the trend is towards less surgey but early and intensive treatment of infections and hypertension [1, 2]. Due to the original disease and related problems, all patients were urologi-

cally extensively evaluated and treated before transplantation. This might be the reason why UTIs caused less problems after transplantation than before and no transplant-threatening infections were recorded.

A characteristic of this population is the bladder emptying problems, which are the cause of disease and uremia, the reason for several operations before transplantation, and an Achilles heel after transplantation. Our patients with Kock's bladder are regularly seen by urologist. Patients with native bladders are checked for the presence of residual urine volumes and the strategy for voiding is changed as indicated. In our patients, the bladder capacity was unchanged after transplantation, but in the long-term one patient was able to start clean intermittent catheterization and a second received a suprapubical catheter.

Today's young patients with congenital malformations of the distal urinary tract will develop ESRD later in life, thanks to a greater awareness of the problem among pediatricians, physicians and surgeons, but also better and more effective treatment, especially handling of infections. In the case of ESRD there should be no hesitation about kidney transplantation because results are gratifying and, despite the previous history, the actual condition of these patients is of minor importance for the outcome.

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