Transpl Int (2004) 17: 549–552
DOI 10.1007/s00147-004-0759-7

BRIEF REPORT

Fabio Fusaro
Giovanni F. Zanon
Anna M. Ferreli
Stefano Giuliani
Graziella Zacchello
Giacomo Passerini-Glazel
Waifro Rigamonti

Renal transplantation in prune-belly syndrome

Received: 13 June 2003
Revised: 1 December 2003
Accepted: 18 March 2004
Published online: 30 September 2004

F. Fusaro (✉) · G. F. Zanon
A. M. Ferreli · S. Giuliani
Division of Pediatric Surgery,
University of Padova, via Giustiniani 3,
35128 Padova, Italy
E-mail: fabiofusaro@libero.it
Tel.: +39-049-8213681
Fax: +39-049-8211781

G. Passerini-Glazel · W. Rigamonti
Institute of Urology, University of Padova,
Padova, Italy

G. Zacchello
Department of Pediatrics,
University of Padova, Padova, Italy

Abstract We assess the effect of the prune-belly syndrome (PBS) on renal transplantation outcome. Six renal transplantations were performed in five boys affected by PBS (median age 5.8 ± 2.1 years, median weight 13.6 ± 2.4kg). Renal graft survival, graft function, lower urinary tract dysfunction, urinary tract infections (UTIs), associated complications and patients' survival after 1 and 5 years of follow-up were analysed. The rate for 1–5-year graft survival was 66.7% (mean serum creatinine 98–103 μmol/l). The surgical treatment of the documented bladder obstruction (two patients) and the severe abdominal wall deficit (one patient) led to a reduction of UTI: the patients maintained their native urinary tract and none received prophylactic antibiotics. The lack of abdominal wall musculature led to severe mechanical complication in one patient, but Monfort's abdominal wall reconstruction was able to restore the graft's function. The outcome of patients with PBS who undergo renal transplantation is good. Before the transplant, an accurate assessment of urinary tract anomalies and deficiency of the abdominal wall musculature is mandatory, to program the appropriate treatment and obtain a good long-term prognosis for the renal graft.

Keywords Kidney transplantation · Paediatrics · Prune-belly syndrome · Urinary tract infection · Graft outcome · Abdominal wall defect

Introduction

The congenital deficiency or hypoplasia of the abdominal wall muscles, associated with various genitourinary anomalies of different grades, is the characteristic feature of the prune-belly syndrome (PBS). Only 3% of the affected patients are girls. Approximately 20% of newborn infants die in the perinatal period, and renal failure develops in about 30% of the survivors. The causes are congenital renal hypoplasia, urinary stasis or infection. These patients become candidates for renal transplantation [1, 2, 3]. In two retrospective studies, Reinberg et al. and Fontaine and colleagues analysed the long-term outcome of renal transplantation in two groups of eight and nine children affected by PBS and concluded that this syndrome did not adversely affect the outcome of the transplantation [4, 5]. On the basis of those observations, we assessed the possible effect of lower urinary tract dysfunction and abdominal wall defect on the outcome of renal transplantation after 1 and 5 years of follow-up in our patients affected by PBS.

Material and methods

Between October 1987 and February 2002, 216 paediatric renal transplantations were performed, including six in five boys affected by PBS. At the time of

transplantation, the median age of the children was 5.8 ± 2.1 years, and the median weight was 13.6 ± 2.4 kg. All patients had undergone urological surgical interventions before transplantation. The procedures included a left nephro-ureterectomy due to infectious complications in one patient, a bilateral cutaneous ureterostomy in one, a left cutaneous ureterostomy in one, tapered re-implants (unilateral in three, bilateral in two), cystoscopy with optical internal urethrotomy in two (urethral ring), orchidopexy in five (bilateral in three, unilateral in two) and orchiectomy in one patient. Reduction cystoplasty procedures were not performed. Before the transplantation all cutaneous ureterostomies were undiverted at different time. All grafts were obtained from cadavers and transplanted into the iliac fossa, through an extra-peritoneal access. End-to-side vascular and uretero-vesical Lich-Gregoire anastomoses were performed. The immunosuppressive drug protocols consisted of a double-drugs association (steroids with cyclosporine or azathioprine). Examination of the lower urinary tract included ultrasound and urodynamics studies, urinalysis and urine culture. Urodynamics consisted of cystometry, uroflowmetry, and measurement of post-void residuals. The capacity was measured during the cystometrogram and then recorded in a fashion relative to the normal capacity for age in patients with normal bladders. After transplantation, ultrasound scan (US) was preformed routinely to monitor the graft, native urinary tract and bladder function (post-void residuals). We defined UTI as a urine culture positive for bacteria in a quantity greater than 10^5 colonies with or without symptoms (i.e. fever, dysuria, and kidney tenderness). Serum creatinine (Cr) level was used to indicate graft function. Since the number of children was small, a statistical analysis was not possible.

Results

Patients and graft outcome: The survival rate at 1 and 5 years was 100% for the patients and 66.7% for the grafts. Two grafts in a single patient were lost: of these, the first (en-bloc graft) was lost because of a venous thrombosis in the first post-operative day, the second because of intractable acute rejection18 days after the operation. The remaining four grafts have good function with a mean serum Cr value of 98 μmol/l and 103 μmol/l at 1 and 5 years, respectively.

Abdominal wall defect: None of the patients had undergone correction of the abdominal wall before transplantation. One patient suffered from a mechanical complication related to the lack of the abdominal wall musculature: this boy presented, 10 months after transplantation, intermittent anuria and rise in serum Cr in orthostatism. The colour Doppler US showed good blood perfusion of the graft, and a MAG-3 renal scan demonstrated an intermittent orthostatic uereterohydronephrosis. At the physical examination a hypogastric hernia was seen, and ultrasound confirmed herniation of the bladder with strangulation of the transplanted ureter and an intermittent hydronephrosis. A Monfort abdominoplasty was able to restore and stabilize the graft function (Cr 116 μmol/l at 24 months' follow-up).

Lower urinary tract function: Before transplantation, all patients were continent and voided without clean intermittent catheterization. In only one patient, the Crede manoeuvre was used to improve voiding. At the urodynamic examination, the bladders of all five patients had large capacity, normal or high compliance (range 80–480 ml/cm water; average 234), the maximal detrusor pressures ranged from 30 to 97 cm water (mean 65.4), and the post-void residuals ranged from 4.3% to 80.3% of individual capacity (mean 50.5%). Two patients underwent internal urethrotomy because of urethral ring and high post-void residual at the urodynamic study (80.3% and 78.6%). After this treatment, the urodynamics showed reduction in residual volume (12% and 30.2%). After transplantation, deterioration in bladder function and dilatations of the graft and native urinary tract were not observed. The patient who underwent abdominoplasty showed reduction of residual volume (Table 1).

Table 1 Post-void residuals (*PVRs*) (*Tx* transplantation)

Patient	Bladder capacity (ml)	Surgical procedure	PVRs		
			Pre-procedure (%)	Post-procedure (%)	Five years after Tx (%)
1	420	Urethrotomy[a]	80.3	12	12.3
2	540	Urethrotomy[a]	78.6	30.2	29
3	650	Abdominoplasty[b]	58.1	5.6	5.6
4	380	None	4.3	–	4.4
5	400	None	38.4	–	–

[a]Bladder capacity was measured during a cystometrogram and compared with the normal capacity for age in patients with normal bladders

[b]Post-void residuals were recorded as a percentage of individual bladder capacity

UTIs: Before transplantation, all patients except one had one or more documented UTIs per year and two had shown a decrease in UTIs after optical urethrotomy. After transplantation, among the four patients with functioning grafts, three did not presented UTI: two patients who had undergone urethrotomy, and one in whom no infections before transplantation were reported; one patient presented infection episodes until an abdominoplasty was carried out. The introduction of immunosuppression was not related to any increase in incidence of UTI after transplantation (Table 2). In fact, UTIs were not observed, and none of these patients received prophylactic antibiotics or reduction in immunosuppressive drugs.

Discussion

Prune-belly syndrome is characterized by hypoplasia of the abdominal wall muscles, urinary tract malformations and, in boys, cryptorchidism. Approximately 30% of those patients develop renal failure as a result of renal dysplasia, recurrent pyelonephritis or obstructive nephropathy and become candidates for renal transplantation. In our experience, we report a graft survival rate of 66.7% and stable graft function (mean Cr 103 μmol/l), after a follow-up period of 5 years. These data are similar to those reported in previous studies [4, 5, 6]. In our study, besides graft survival rate and graft function, in particular we analysed the impact on the outcome of lower urinary tract function and the abdominal wall defect.

Theoretically, in PBS patients, the urinary stasis due to the lower urinary tract anomalies associated with the immunosuppression could predispose the patient to the development of severe UTI, with detrimental effect on transplant outcome [7, 8]. Our results, in accordance with others', suggest the importance of treating the documented bladder obstruction and severe abdominal wall defect in order to obtain a good graft outcome [9, 10]. The importance of obtaining improvements in micturition and consequent voiding efficiency is underlined by the following observations: after internal urethrotomy, our two patients showed simultaneous improvement in voiding efficiency and decline in the incidence of UTI. At the US performed before and after micturition, dilatation and urinary stasis in the native urinary tract decreased. The improvement in bladder emptying, and the reduction in voiding pressure, most likely reduced the infection rate by decreasing the post-void residuals and, thereby, diminishing the urinary stasis and storage in the ureter with the associated risk of bacterial overgrowth. As a consequence of this, our patients, contrary to those described in previous reports, have maintained their native upper urinary tract without an increase in UTIs even in the presence of immunosuppressive drugs.

Table 2 UTIs (mean number/year) before and after transplantation (*TX*)

Patient	Immunosuppression	UTIs	
		pre-TX	Post-TX
1	Prednisone, cyclosporine A	5.2–0[a]	0
2	Prednisone, azathioprine	4.4–0*	0
3	Prednisone, cyclosporine A	3.2	3–0[b]
4	Prednisone, cyclosporine A	0	0
5	Prednisone, azathioprine	4	–

[a]Number of UTIs before and after urethrotomy
[b]Number of UTIs before and after abdominoplasty

As other authors point out, the reconstruction of abdominal wall musculature may also be important to improve bladder function and to reduce the risk of UTI [10]. Our patient, after abdominoplasty, showed improvement in post-void residual, reduction in UTI and interruption of the antibiotics. As reported by Smith et al, the Monfort abdominoplasty probably reduce the potential surface area to which an applied Valsalva force is distributed. As illustrated by Pascal's principle (pressure equals force over area), when a steady force is applied over a reduced area, an increased pressure will result. Therefore, after the abdominoplasty, the bladder received an increased pressure when a similar Valsalva force was exerted. This increased pressure may assist urination by more the complete emptying of urine.

The congenital deficiency or hypoplasia of the abdominal wall muscles not only might affect bladder function but it might also be the cause of several complications that may lead to graft loss. In one of our patients we observed a particular complication related to the lack of musculature. The boy presented acute graft failure due to bladder herniation with strangulation of the graft ureter. In the literature two other cases, which presented a complication related to the abdominal wall defect, have been described [11, 12]. In those two cases, the grafts were transplanted intraperitoneally and presented torsion of the vascular pedicle, which led to the graft loss. In our patient the Monfort abdominoplasty stabilized the graft ureter and bladder and restored the graft's function. These three severe complications related to the abdominal wall defect, in a total of 32 cases of transplantation for PBS (3/32, 9.3%) described in the literature, suggest that this anomaly in PBS probably needs to be taken into account as a prognostic factor for the graft's outcome.

In conclusion, we agree with previous reports that renal transplantation in PBS is associated with a good graft survival rate. Before the transplantation, it is mandatory to treat lower urinary tract anomalies in order to improve voiding efficiency, because it reduces urinary stasis, protects from severe UTI and permits the

maintenance of the native urinary tract with any antibiotic prophylactic regimen. From an analysis of the literature and from our experience, it seems that the associated abdominal wall defects can expose the graft to severe complications. Therefore, because the syndrome is a spectrum disorder, before the transplantation an accurate assessment of the deficiency of the abdominal wall musculature and urinary tract anomalies is mandatory, to program the appropriate treatment and obtain a good long-term prognosis for the renal graft.

References

1. Burbige KA, Amodio J, Berdon WE, Hensle TW, Blanc W, Lattimer JK. Prune-belly syndrome: 35 years of experience. J Urol 1987; 137:86.
2. Reinberg Y, Manivel JC, Pettinato G, Gonzales R. Development of renal failure in children with the prune-belly syndrome. J Urol 1991; 145:1017.
3. Shenasky JH 2nd, Whelchel JD. Renal transplantation in prune belly syndrome. J Urol 1976; 115:112.
4. Reinberg Y, Manivel JC, Fryd D, Najarian JS, Gonzales R. The outcome of renal transplantation in children with the prune-belly syndrome. J Urol 1989; 142:1541.
5. Fontaine E, Salomon L, Gagnadoux MF, Niaudet P, Broyer M, Beurton D. Long-term results of renal transplantation in children with the prune-belly syndrome. J Urol 1997; 158:892.
6. Koo HP, Bunchman TE, Flynn JT, Punch JD, Schwartz AC, Bloom DA. Renal transplantation in children with severe lower urinary tract dysfunction. J Urol 1999; 161:240.
7. Woodard JR. The bladder in prune belly syndrome. Dialog Pediatr Urol 1990; 13:6.
8. Woodard JR. Lesson learned in 3 decades of managing the prune-belly syndrome. J Urol 1998; 159:1680.
9. Kinahan TJ, Churchill BM, McLorie GA, Gilmour RF, Khoury AE. The efficiency of bladder emptying in the prune belly syndrome. J Urol 1992; 148:600.
10. Smith CA, Smith EA, Parrott TS, Broecker BH, Woodard JR. Voiding function in patients with the prune-belly syndrome after Monfort abdominoplasty. J Urol 1998; 159:1675.
11. Abbitt PL, Chevalier RL, Rodgers BM, Howard SS. Acute torsion of renal transplant: cause of organ loss. Pediatr Nephrol 1990; 4:174.
12. Marvin RG, Halff GA, Elshihabi I. Renal allograft torsion associated with prune-belly syndrome. Pediatr Nephrol 1995; 9:81.