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Living-related kidney transplantation with catheterizable urinary conduit in prune belly syndrome: A case report

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ABSTRACT

INTRODUCTION: Prune belly syndrome (PBS) presents with both renal dysplasia and urinary tract abnormalities. When performing kidney transplantation in PBS patients with kidney failure, extensive pretransplant urinary tract preparation may be necessary.**PRESENTATION OF CASE:** We report the case of a 36-year-old man with PBS who underwent living-related kidney transplantation with urinary diversion using the Mitrofanoff principle. The patient had a bilateral loop ureterostomy for a urethral obstruction. Cystourethrography before the operation showed tortuous and dilated ureters with vesicoureteral reflux (VUR), and complete occlusion of the urethra. Before transplantation, we created a catheterizable urinary conduit with the patient's own malformed left ureter in accordance with the Mitrofanoff principle. The folding procedure was selected for ureteroplasty to preserve good blood supply. Extravesical detrusorhaphy was performed as an antireflux procedure. V-quadrilateral-Z ureterostomy for catheterization was performed. Then, we performed living-related kidney transplantation from the patient's mother. Postoperative cystourethrography did not show left VUR. The patient performed clean intermittent self-catheterization without complications, and had good graft function.**DISCUSSION:** The appendix and ileum are currently the most commonly used options for urinary conduits based on the Mitrofanoff principle. However, the patient had complications of diarrhoea and constipation, so we used the patient's own malformed ureter. We performed a folding procedure to avoid ureteral stenosis and VUR, and used the V-quadrilateral-Z flap technique to avoid stoma stenosis.**CONCLUSION:** When performing kidney transplantation in patients with PBS, urinary tract anomalies should be thoroughly evaluated. Our surgical procedure will help to reduce complications after kidney transplantation.© 2018 The Author(s). Published by Elsevier Ltd on behalf of IJS Publishing Group Ltd. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

1. Introduction

Prune belly syndrome (PBS) is a congenital disorder with a characteristic clinical triad, including abdominal muscle deficiency, severe urinary tract abnormalities, and bilateral cryptorchidism in males. PBS patients also have both renal dysplasia and urinary tract abnormalities that cause pyelonephritis and obstruction, resulting in end-stage renal disease (ESRD) in childhood or adolescence [1,2]. Kidney transplantation is one option for renal replacement therapy in PBS, as in other causes of ESRD. However, extensive pretransplant urinary tract preparation may be necessary to reduce the postvoid residual volume to a minimum [3,4]. The complication

rate with use of ureteral conduits is higher than that with use of the appendix or ileum. Thus, the appendix and ileum are currently the most common options for creation of urinary conduits.

In this case report, we created a catheterizable urinary conduit before kidney transplantation, using the malformed ureter with a folding procedure and V-quadrilateral-Z ureterostomy, based on the Mitrofanoff principle, without complications in a patient with PBS who had undergone bilateral ureterocutaneostomy.

This work was reported according to the SCARE guidelines [5].

2. Presentation of case

A 36-year-old man with ESRD secondary to PBS who had been receiving hemodialysis for 15 years underwent ABO-compatible and donor-specific antibody-negative living-related kidney transplantation. The donor was his 62-year-old mother, who had a

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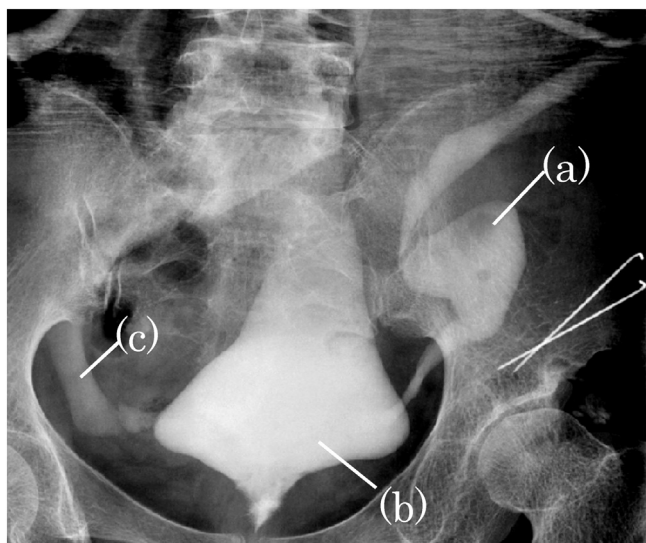


Fig. 1. Preoperative cystography. Cystography was performed via the right ureteral ureterostomy. Left vesicoureteral reflux appeared when 40-ml contrast agent was injected. The bladder capacity was 180 ml. (a) left dilated ureter, (b) bladder, (c) right ureter.

creatinine clearance of 96.6 ml/min. The recipient had undergone bilateral loop ureterostomy for urethral obstruction as a neonate.

We performed cystourethrography via right ureterostomy to accurately evaluate bladder capacity and other coexisting urinary abnormalities. Cystourethrography revealed tortuous and dilated ureters with vesicoureteral reflux (VUR) and complete urethral occlusion (Fig. 1). His bladder volume was 180 ml.

Post-transplantation immunosuppression included induction with tacrolimus, mycophenolate mofetil, methylprednisolone, and basiliximab.

We created a catheterizable urinary conduit before kidney transplantation with the patient’s own left ureter, in accordance with the Mitrofanoff principle. A Gibson incision was performed from the existing left ureterocutaneous fistula to above the pubis. As it was difficult to identify his left ureter with an extravesical approach, we incised the suprapubic area of the bladder wall and inserted a catheter from the left ureteral orifice. A folding procedure was selected for ureteroplasty to preserve good blood supply. For smooth catheterization, intermittent sutures were placed over the 14-Fr catheter before folding the dilated ureter, and the ureter was fixed to the abdominal wall with several stay sutures. Then, extravesical detrusorrrhaphy was performed as an antireflux procedure. V-quadrilateral-Z ureterostomy for catheterization was performed. A vesicostomy was also created.

After creating a urinary conduit, we performed right nephroureterectomy with closure of the right ureterocutaneous fistula. The graft was transplanted into the right iliac fossa through the extraperitoneal access. End-to-side vascular and ureterovesical Lich-Gregoir anastomoses were performed. Diuresis began 3 min after declamping.

The serum creatinine level decreased after the operation. We removed the vesicostomy, and the patient started clean intermittent self-catheterization on the ninth postoperative day. Postoperative cystourethrography did not show left VUR (Fig. 2). He performed clean intermittent self-catheterization without complications, and had good graft function at 6-month follow-up.

3. Discussion

PBS is characterized by hypoplasia of the abdominal wall muscles, urinary tract malformations, and cryptorchidism in males. Approximately 30% of patients who survive infancy will develop renal failure due to renal dysplasia, recurrent pyelonephritis, or obstructive nephropathy [6]. Kidney transplantation is a well-accepted treatment, but urinary diversion is essential, depending on the severity of urinary tract malformations. In this case report, we describe urinary diversion during kidney transplantation, in which a catheterizable conduit was created with the patient’s own malformed ureter in accordance with the Mitrofanoff principle for a PBS patient who had complete urethral occlusion and bilateral ureterocutaneostomy (Fig. 3).

The transappendicular continent cystostomy was first described by Mitrofanoff in 1980, with use of the appendix as a conduit between the bladder and skin [7]. This procedure allowed the bladder to be emptied by a route other than the urethra and was a further revolutionary step in the field of urinary incontinence management following the introduction of clean intermittent self-catheterization [8]. Various segments have been used for the creation of a catheterizable conduit, including the cecum, small bowel, stomach, ureter, and bladder [7]. The ureter remains a good catheterizable channel and was indeed used partly in Mitrofanoff’s initial report; however, complication rates were higher. In patients for whom the ureter was used as conduit, a 40% major complication rate was reported, including urine leak and the need for complete revision [9]. A further study of conduits made with the appendix or ureter suggested a higher stenosis rate in patients where the ureter was used [10]. The high stenosis rate of ureteral conduits, as compared with those of the appendix and ileum, may result from low blood supply. Thus, the appendix and ileum are currently the most common options for urinary conduits. In this case, the patient often had abdominal symptoms of diarrhoea and constipation before the operation. Hence, we decided to use his ureter rather than his appendix or ileum to avoid postoperative complications such as bowel obstruction. Some reports on ureteroplasty for

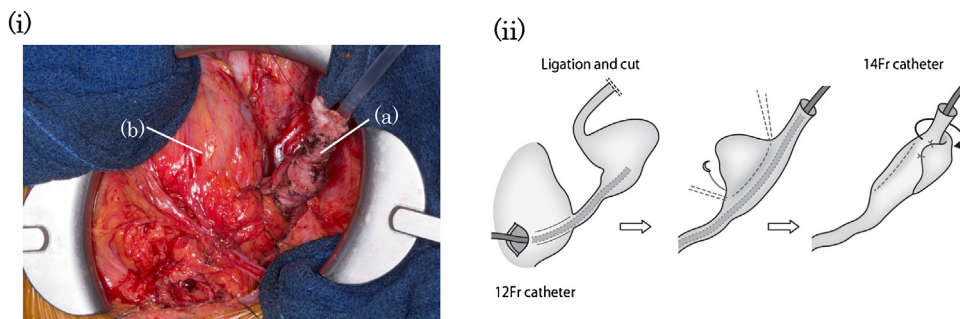


Fig. 2. Left ureteroplasty using the folding procedure. (i) (a) left ureter, (b) bladder. (ii) For smooth catheterization, intermittent sutures were placed over the 14-Fr catheter before folding the dilated ureter.

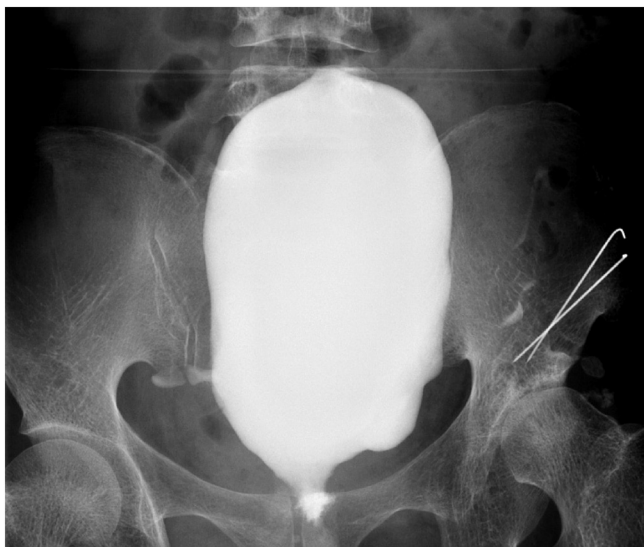


Fig. 3. Postoperative cystography. Cystography was performed via the urinary conduit. Left vesicoureteral reflux was not observed. Self-catheterization allowed bladder emptying.

megaureter suggested that the folding procedure had fewer complications such as ureteral stenosis and VUR owing to good blood flow [11,12].

Generally, the conduit is attached to the abdominal wall and a stoma is created. Four main techniques for stoma creation have been described, including direct anastomosis, umbilical stoma, tubular skin flap, and the V-quadrilateral-Z flap technique. Some studies reported complication rates for the umbilical stoma, tubular skin flap, and V-quadrilateral-Z flap technique. The revision or dilatation rate for stoma stenosis was 45% for the tubular skin flap technique, 25% for the umbilical stoma, and 0% for the V-quadrilateral-Z flap technique [13]. For this reason, the folding procedure and V-quadrilateral-Z flap technique were selected in our case to prevent stenosis of the catheterizable conduit.

The prognosis of kidney transplantation for PBS is usually excellent, as in other causes of ESRD [14,15]. Yalcinkaya et al. recently reported that the 5- and 10-year graft survival rates were 86.2% and 78.4%, respectively, in patients with PBS after kidney transplantation [16]. Fusaro et al. suggested that it is mandatory to treat lower urinary tract anomalies before kidney transplantation to improve voiding efficiency, which reduces urinary stasis and protects from severe urinary tract infection, thereby contributing to graft survival [14].

4. Conclusion

When performing kidney transplantation for urinary tract dysfunction, anomalies should be thoroughly evaluated and the method used to drain urine from the transplanted kidney should be chosen. Our surgical procedure will help to reduce complications after kidney transplantation.

Conflicts of interest

This manuscript did not receive any conflicts of interest.

Sources of funding

This manuscript did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Ethical approval

The Japan Academic Consortium of Kidney Transplantation (JACK), approved by the Institutional Ethics Committee of Tokyo Women's Medical University (Identifier: 3336-R), is a multicentre observational cohort study of kidney transplant patients in Japan. The case described in this article is included in the JACK observational cohort.

Consent

Written informed consent was obtained from the patient for publication of this case report. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contributions

Taro Banno and Yoichi Kakuta contributed to study design, data collection, data interpretation, and writing the paper.

Kohei Unagami, Akiko Sakoda, Masayoshi Okumi, Hideki Ishida, and Kazunari Tanabe contributed to study design, data collection, data interpretation.

Registration of research studies

This manuscript is not a human study, but case report.

Guarantor

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Provenance and peer review

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