How to Deal With Children With End-Stage Renal Disease and Severe Bladder Dysfunction

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ADEQUATE urinary storage in a low-pressure reservoir and regular and complete emptying are essential features of bladder physiology. The need for a normal lower urinary tract has been recognized since the beginning of renal transplant programs. If the pressure inside the bladder is high or if does not completely empty, graft survival may be affected.[1] Children with lower urinary tract dysfunction due to functional or congenital anomalies leading to end-stage renal disease should have the anomaly treated before renal transplantation. This study summarizes our experience with kidney transplantation in children with lower urinary tract dysfunction and chronic renal failure.

MATERIALS AND METHODS

Over the last 10 years 992 kidney transplants have been performed at the University of São Paulo, including 185 cases in patients under 18 years of age, with 115 recipients being younger than 15 years old. Seventeen patients (10 boys and 7 girls) with severe lower urinary abnormalities underwent 19 kidney transplants. The mean age at transplantation was 13.3 years (range: 3 to 18 years). The etiology of bladder dysfunction was a neurogenic bladder (n = 5), posterior urethral valve (n = 5) and vesicoureteral reflux (n = 4). Other causes were responsible for three additional cases: one with exstrophy submitted to a primary closure; one with an imperforate anus, ectopic ureterocele and bladder hyploplasia; and the last one with prune-belly syndrome and a hypoplastic urethra. All patients were submitted to cystourethrography and urodynamic evaluation. Patients with primary reflux had previously undergone more than one antireflux procedure; they presented with poor bladder compliance.

The incontinent child with an ectopic ureterocele had her bladder removed and underwent construction of an external continent reservoir using an ileal segment, and a Monti procedure. The child with prune belly and difficulty draining the bladder due to a urethral disorder underwent a Mitrofanoff procedure to permit easier clean intermittent catheterization (CIC). One child with posterior urethral valve underwent auto-augmentation, and a dilated nonrefluxing ureter was placed in the iliac fossa for easier CIC. The other 14 children underwent augmentation: 11 with a bowel segment (ileum in 8; ileocecum in 2; sigmoid in 1): Two of these patients had been initially enlarged, with the ureter leading to inadequate results. The last three cases successfully underwent ureterocystoplasty.

The lower urinary tract was repaired at a minimum of 12 weeks before kidney transplantation. Nineteen kidney transplants were performed in 17 children: 18 grafts from living and one from a cadaveric donor. The graft was always placed extraperitoneally, with the ureter implanted into the bladder in 14 cases, into the intestinal portion of the reservoir in 4 cases (ileum in 3, sigmoid in 1), and reconstructed by an uretero-uretero anastomosis with the native ureter in one case.

RESULTS

Mean follow-up is 55.2 months (3 to 192 months). Bladder capacity and compliance were markedly improved in all augmented children. Eleven children empty their bladder by CIC. Ten children are maintained on prophylactic antibiotic therapy. Mean creatinine level is 1.6 mg/dL (0.8 to 3.1 mg/dL).

One child developed a urinary fistula, and another had a ureteral stenosis at the site of implantation into the sigmoid. The first was treated by catheterization of the reser-

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voir through the Mitrofanoff; the other underwent ureteral reimplantation. Urinary tract infection was the most frequent complication, occurring in 10 children, including five who presented with more than five episodes. The child who was transplanted into a urinary reservoir is partially continent, awaiting a revision of the Monti procedure.

Three grafts were lost: one was due to chronic interstitial nephritis and episodes of urinary tract infection after 39 months, and the other two were due to chronic rejection at 48 and 69 months posttransplantation. Two patients received a second graft and are doing well at follow-up. The actuarial graft survivals were 100% and 90% at 36 and 60 months, respectively.

DISCUSSION

The lower urinary tract must show a good capacity, compliance, and drainage. Children displaying findings suspicious for bladder dysfunction should undergo urethrocystography and urodynamic evaluation. Whenever possible, the lower urinary tract should be preserved and provided with adequate capacity and compliance. Bladder enlargement is the solution for this problem, even for the group of transplanted patients who will need to perform CIC for bladder drainage.[2,3]

Cystoplasty was performed before transplantation in all cases except in one child whose bladder dysfunction was not recognized until hydronephrosis and impaired graft function developed. The adequate timing for the cystoplasty is at least 12 weeks before transplantation and before the initiation of immunosuppression.

There is no doubt that the ureter is the best biological material for bladder enlargement for it does not pose the risk of cancer development or the inconvenience of reabsorption and production of mucus. Among the five patients who underwent ureterocystoplasty, the procedure failed in two, both having undergone previous ureteral reimplantations.

When the ureter is not dilated, available, or has been previously manipulated, the use of an intestinal segment is the best option. Bowel detubularization, is important, for it interrupts the peristaltic activity, abolishes the massive contractions, and enhances the capacity. In combination with CIC, the success rate of augmentation cystoplasty has improved.[4,5] Detubularized ileum is the most frequently used segment, owing to its abundance and reduced complication rates.

One incontinent patient was transplanted into a continent reservoir, and one child with a hypoplastic urethra and inadequate bladder drainage underwent a Mitrofanoff procedure for easier bladder drainage, with subsequent successful transplantation.

Whenever possible the ureter should be reimplanted into the bladder portion of the reservoir. Two urological complications were successfully addressed: a ureteral stenosis treated by reimplantation, and a urinary leak managed by catheterization of the Mitrofanoff.

The most frequent complication was urinary tract infection, mainly in the group of patients maintained on CIC; some patients had fever. Recognition and prompt treatment of the complications are both important for graft preservation.

CONCLUSION

The ureter, when available, is the most attractive source for bladder augmentation. If it is not disposable, an intestinal segment can be used. When the urinary tract cannot be restored, kidney transplantation into a urinary diversion is the sole solution. Augmentation cystoplasty and urinary diversion are safe and effective methods for the treatment of severe lower urinary tract abnormalities in children who are candidates for kidney transplantation.

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