Oncologic Outcome and Urinary Function after Radical Cystectomy for Rhabdomyosarcoma in Children: Role of the Orthotopic Ileal Neobladder Based on 15-Year Experience at a Single Center

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Abbreviations and Acronyms

CIC = clean intermittent catheterization

fUTI = febrile urinary tract infection

RC = radical cystectomy

RMS = rhabdomyosarcoma

RT = radiotherapy

SFU = Society for Fetal Urology

VIP = Padua ileal neobladder

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Purpose: We determined the oncologic and urological outcomes in patients with bladder/prostate rhabdomyosarcoma according to the type and timing of urinary tract surgery, with emphasis on the role of the Padua orthotopic ileal neobladder.

Materials and Methods: We retrospectively analyzed oncologic and urological outcomes of 11 consecutive patients treated at our institution between 1998 and 2012.

Results: Two patients underwent urethrectomy and placement of a heterotopic catheterizable ileal neobladder. The membranous urethra was preserved in 9 patients, 6 underwent primary Padua ileal neobladder at radical cystectomy, 2 underwent delayed Padua ileal neobladder and 1 underwent bilateral cutaneous ureterostomy. Four of these 9 patients experienced disease recurrence, including local recurrence in 2 despite negative intraoperative biopsies. Survivors undergoing heterotopic catheterizable ileal neobladder or primary Padua ileal neobladder learned to empty the bladder to completion without long-term upper tract deterioration. Both cases managed by delayed Padua ileal neobladder required clean intermittent catheterization eventually. Erections were reported in 5 of 6 surviving males.

Conclusions: The Padua ileal neobladder allowed preservation of volitional urethral voiding in all survivors in whom it was placed at radical cystectomy. Nevertheless, local recurrence was noted in 2 of the 9 cases where the membranous urethra was preserved. By comparison, patients undergoing delayed Padua ileal neobladder after attaining disease-free status never achieved voiding per urethra. Therefore, a heterotopic reservoir might be a more reliable choice under these circumstances. Erectile function is preserved in the majority of cases.

Key Words: cystectomy, prostate, rhabdomyosarcoma, urinary bladder, urinary diversion

Rhabdomyosarcomas arising from the bladder/prostate account for approximately 25% of all such tumors in children.[1] Multidisciplinary protocols including chemotherapy and radiotherapy have improved patient survival and increase the chances of bladder preservation.[1,2] Radical cystectomy reportedly is required because of failure of medical treatments and inability to accomplish partial resection in about 40% of

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cases.[3] After radical cystectomy a heterotopic intestinal catheterizable reservoir seems to be a viable option.[4–6] However, if the membranous urethra is spared, an orthotopic neobladder joined to it might preserve volitional urethral voiding.[5,7] Since 1998, we have incorporated the Padua ileal neobladder into our armamentarium for bladder substitution after radical cystectomy in patients with rhabdomyosarcoma.[7] This procedure was initially devised at our institution, where it is extensively used in adults requiring radical cystectomy for transitional cell carcinoma of the bladder.[8]

Despite the putative advantages of orthotopic neobladder, few instances have been reported thus far in patients with RMS, making information about the functional outcomes of this diversion scarce.[5,7] A reason for such a small number of cases is also that evidence suggests that the involvement of the membranous urethra could not be ruled out reliably in RMS cases preoperatively or intraoperatively.[4] Therefore, few surgeons recommend extensive urethral excision during RC, and some recommend that urinary tract reconstruction be delayed until after the patient has achieved a durable disease-free status.[4,9,10] We reviewed our 15-year experience with urinary tract management in cases of RMS requiring RC to determine the oncologic and urological outcomes according to the type and timing of urinary tract surgery.

MATERIALS AND METHODS

Inclusion/Exclusion Criteria

A total of 64 patients with bladder/prostate RMS were registered in 2 consecutive protocols (RMS96 and RMS2005) coordinated by the AIEOP (Associazione Italiana di Ematologia e Oncologia Pediatrica) Soft Tissue Sarcoma Committee between January 1998 and December 2012. Of the patients 6 were diagnosed and treated elsewhere and 11 were referred to our institution for surgery. We retrospectively reviewed the records of the latter, which included 9 patients undergoing RC at our institution and 2 referred for continent diversion after RC performed elsewhere. Patients with RMS confined to the genitalia or undergoing other kinds of urinary tract surgery for RMS were excluded.

Case Management

Protocol guidelines recommended initial biopsy, normally endoscopic, to establish the histological diagnosis. Biopsy was followed by intensive chemotherapy according to the risk profile and preoperative or postoperative RT generally to a total dose of 45 Gy. RC was considered in instances of persistent disease after RT or to avoid pelvic irradiation in young children.[11,12]

In general, our policy regarding urinary tract management in individuals undergoing RC was to favor VIP whenever the membranous urethra was deemed salvageable based on preoperative assessment, intraoperative findings and the presence of free margins on intraoperative histology on frozen sections. Otherwise, urethrectomy was performed (up to the bulbar segment in males and total in females), and a heterotopic continent catheterizable ileal neobladder was placed. VIP was created as described previously.[7] A catheterizable conduit was associated with delayed reconstruction. Heterotopic catheterizable ileal neobladders were fashioned by reshaping a 30 to 40 cm long ileal segment in a spherical configuration and placing the catheterizable conduit at the umbilicus.

Postoperatively serial imaging (usually magnetic resonance) was performed to rule out disease recurrence every 3 months in the first year and at increasing intervals thereafter. Followup endoscopy was not systematically performed. Ultrasonography was used to monitor bladder emptying and upper tract status. Post-void catheterization was not recommended. Surveillance cystoscopies plus biopsies of the reservoir were recommended starting 10 years after reconstruction.

Outcome Parameters

We evaluated patient clinical characteristics, treatment administered, and oncologic and urological functional outcomes. The latter was assessed in patients surviving at least 1 year after urinary tract reconstruction and included the presence of recurrent fUTIs, urinary continence, upper tract dilatation, reservoir capacity, post-void residual, need for CIC and reported erectile, ejaculatory and sexual function. Hydronephrosis was graded according to the SFU system. Upper tract dilatation was defined as severe in cases of SFU grade III or IV hydronephrosis and concomitant ureteral dilatation. In patients with upper tract dilatation obstruction was ruled out by dynamic diuretic renography.

Capacity of the reservoir was assessed on a voiding diary or on videourodynamics. VIP emptying as assessed by ultrasonography or videourodynamics was considered complete when the residual was less than 20 ml. Erectile function was assessed by patient or parent inquiry about the presence of erections. Oncologic and urological outcomes were assessed according to the type and timing of urinary tract surgery, ie if urethrectomy was performed at RC and if the reconstruction was simultaneous to RC (primary) or delayed.

Given the small sample size, only descriptive statistics were used. Categorical variables were expressed as ratios and continuous variables as medians (ranges).

RESULTS

Patients

Patient characteristics are summarized in table 1. Nine patients (7 males) were primarily treated at our institution and the membranous urethra was preserved during RC in 7 (6 males). Of the 2 patients undergoing urethrectomy 1 was a female with neurofibromatosis type 1 with disease extending to the vagina and 1 was a male with positive intraoperative biopsies. Two cases were secondary referrals after RC performed elsewhere 3 and 7 years