Continent urinary reconstruction in rhabdomyosarcoma: A new approach

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Continent Urinary Reconstruction in Rhabdomyosarcoma: A New Approach

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São Paulo, Brazil

Purpose: The authors present here 5 cases of continent urinary diversion in rhabdomyosarcoma applying a recently described technique for the Mitrofanoff Principle devised by the authors.

Methods: Two previously irradiated rhabdomyosarcoma patients presenting with residual bladder disease and massive sensitive urinary urgency underwent a transverse colonic reservoir with catheterizable stoma. Two other patients presenting with a Bricker conduit underwent conversion into an ileal reservoir. One patient underwent reconstruction after a cystectomy.

Results: All patients were continent and able to perform continent intermittent catheterization.

Conclusions: The technique proved to be feasible for this group of patients. The authors believe that because of its simplicity, it should be an option of continent urinary diversion when the Mitrofanoff Principle is considered.

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INDEX WORDS: Rhabdomyosarcoma, continent diversion, Mitrofanoff principle.

UROGENITAL rhabdomyosarcomas (RMS) constitute from 4% to 8% of the total malignant solid tumors of childhood, second only to lymphomas, central nervous system tumors, neuroblastomas, and Wilms’ tumors. In the past, patients were treated with mutilating surgeries that created humid ostomies as a way of reconstructing the urinary tract after a cystectomy. With the development of better radiotherapeutic and chemotherapeutic techniques, radical surgery was reserved only for the nonresponsive forms of RMS.

Over the years, however, great technical advances occurred within the realm of continent urinary reservoirs. The objective of this work is to present our experience with a new technique of reconstruction of the urinary tract that allows the emptying of the urinary reservoir by means of abdominal catheterization.

This technique was developed in our institution and has the advantage of exempting the surgeon from the use of the appendix or substitutes for making the efferent conduit, thus, making the surgical procedure much simpler and reducing time under anesthesia.

MATERIALS AND METHODS

We analyzed 5 patients with RMS submitted to urinary reconstruction with a reservoir using the Macedo technique (Fig 1). Their age varied from 2 to 21 years (mean, 7.1 years) Three were boys and 2 were girls. In 2 cases, the reconstruction was performed to convert a Bricker surgery. In 2 other cases the continent reconstruction was performed immediately after a cystectomy, all in one surgical stage. One patient responded completely to treatment with chemotherapy and radiotherapy but showed sensitive urgency and disabling dysuria forcing us to perform a urinary diversion. The intestinal segment used in 3 cases was the ileum and, in 2 cases, the transverse colon.

CASE REPORTS

Case 1

A 2-year-old girl had embryonic RMS of the bladder diagnosed in 1982 by endoscopic biopsy. She was sent for radiotherapy (3,000 rads) and chemotherapy (T6-2 cycles protocol). She remained with a nodular image in the bladder after 6 months and was submitted to a partial cystectomy, the anatomicopathologic results of which showed botryoid RMS. She then was submitted to chemotherapy (until June 1983) and radiotherapy (2,000 rads).

After 6 months of the last chemotherapy, she presented with dysuria and an ultrasound image suggestive of tumoral recurrence. She then was sent for radical cystectomy and Bricker’s urinary diversion (February 1984), the anatomicopathologic results of which showed pleomorphic RMS. She presented with various episodes of urinary tract infections and, having passed 14 years of age, she demanded a continent diversion. We chose to convert the Bricker to a continent ileum reservoir in 1998. She now presents clinically well, with continence and without any signs of tumoral recurrence in a 5-year follow-up.

Case 2

A boy had botryoid RMS of the bladder diagnosed at the age of 2 years, 4 months by endoscopic biopsy. He then was sent to chemotherapy (vincristine, actinomycin, cyclophosphamide [VAC]) and radiotherapy. He presented with partial remission and vesical dysfunction (75%) after the primary treatment and then was submitted to a radical cystoprostatectomy (the anatomicopathologic results of which showed...
embryonic RMS and free margins) and to a Bricker urinary diversion within 6 months of diagnosis.

After 4 years and 9 months he underwent conversion to a continent ileum reservoir. Today he is asymptomatic with continence and showing no signs of recurrence in a 4-year follow-up.

Case 3

A 21-years old man had alveolar RMS of the prostate diagnosed after recurring episodes of urinary retention. He was submitted to chemotherapy and radiotherapy obtaining no adequate response. The choice was then made for the performance of a radical prostatectomy (the anatomicopathologic results of which showed margins compromised by neoplasia) with a continent ileum urinary reservoir being sent to adjuvant chemotherapy and radiotherapy. The patient died in September 2001 of postchemotherapy sepsis. He showed continence and no complications related to the urinary reconstruction.

Case 4

An 8-year boy presented with hematuria, dysuria, urinary retention, and an ultrasound image of intravesical vegetation on the left lateral wall and vesical trigone. The diagnosis of botryoid RMS was made after an endoscopic biopsy, and the patient received chemotherapy and radiotherapy obtaining no adequate response to the primary treatment (Fig 2). The choice was made for a radical cystoprostatectomy and urinary diversion with a continent reservoir using the transverse colon. Currently, at 11 months of follow-up, the patient presents with continence and no signs of tumoral recurrence and is taking citrate.

Case 5

A 2-year, 6-month girl presented with dysuria, abdominal pain, and urinary retention, which was diagnosed as botryoid RMS of the urethra, vagina, and bladder. She was submitted to chemotherapy (VAC) and radiotherapy (4,500 rads) obtaining an adequate response. After 7 months of primary treatment, the patient presented with disabling dysuria and sensitive urgency. Vesical biopsy results were negative to neoplasia, and imaging examinations showed no evidence of tumoral recurrence. The choice to perform a continent urinary diversion with transverse colon was made because of the urinary dysfunction of actinic origin.

RESULTS

The most frequent histologic types of RMS were the botryoid followed by the pleomorphic and alveolar. In these series, 4 patients remain alive, continent, and performing intermittent catheterization (4 every 4 hours). One patient died of febrile neutropenia, months after the urinary reconstruction, having gained continence and having obtained satisfactory surgical results (Table 1).

The follow-up concerning the treatment of neoplasia...
<table>
<thead>
<tr>
<th>Case No.</th>
<th>Sex/Age at Diagnosis</th>
<th>Staging/Location of Tumor</th>
<th>Histologic Type</th>
<th>Complications After Primary Treatment</th>
<th>First Surgical Approach</th>
<th>Complications After Surgery</th>
<th>Second Surgical Approach/Reconstruction</th>
<th>Urodynamic Study</th>
<th>Current Status</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>F/2 yr</td>
<td>III/Bladder</td>
<td>Embryonic (botryoid)/pleomorphic</td>
<td>—</td>
<td>Partial cystectomy</td>
<td>Tumoral recurrence</td>
<td>Radical cystectomy + Bricker/Conversion to Continent ileal diversion</td>
<td>—</td>
<td>No recurrence; with continence; light metabolic acidosis; 5-year follow-up</td>
</tr>
<tr>
<td>2</td>
<td>M/2 yr</td>
<td>II/Bladder</td>
<td>Embryonic (botryoid)</td>
<td>Vesical dysfunction</td>
<td>Radical cystoprostatectomy + Bricker</td>
<td>Obstructive acute abdomen and UTI</td>
<td>Conversion to continent ileal diversion</td>
<td>400 ml, low pressure, no leakage</td>
<td>No recurrence; with continence; light metabolic acidosis; 4-year follow-up</td>
</tr>
<tr>
<td>3</td>
<td>M/21 yr</td>
<td>III/Prostate</td>
<td>Alveolar</td>
<td>—</td>
<td>Radical cystoprostatectomy + continent ileal reservoir</td>
<td>Partial intestinal obstruction</td>
<td>—</td>
<td>—</td>
<td>Died of postchemotherapy sepsis; 2-year and 7-month follow-up</td>
</tr>
<tr>
<td>4</td>
<td>M/8 yr</td>
<td>III/Bladder and prostate</td>
<td>Embryonic (botryoid)</td>
<td>Febrile neutropenia, herpes zoster, actinic dermatitis</td>
<td>Radical cystoprostatectomy + continent ileal reservoir</td>
<td>—</td>
<td>—</td>
<td>BC 300 mL, involuntary colonic contractions 30-40 cm H2O without leakage</td>
<td>No recurrence; with continence; 13-month follow-up</td>
</tr>
<tr>
<td>5</td>
<td>F/2 yr, 7 mo</td>
<td>III/Bladder and urethra</td>
<td>Embryonic (botryoid)</td>
<td>Sensitive urinary urgency</td>
<td>Continent reservoir of transverse colon</td>
<td>Febrile UTI</td>
<td>—</td>
<td>BC 220 mL; involuntary, colonic contractions 15-20 cm H2O Without leakage</td>
<td>No recurrence; with continence; 5-month follow-up</td>
</tr>
</tbody>
</table>

MP, Macedo’s pouch; UTI, urinary tract infections; VAC, vincristine, actinomycin, cyclophosphamide; BC, bladder capacity.
was 7.2 years, and the follow-up after the continent urinary reconstructions was 2.7 years.

**DISCUSSION**

Bearing in mind the IRS-IV (Intergroup Rhabdomyosarcoma Study) protocol, the general approach in treatment is achieved with chemotherapy and radiotherapy as soon as the diagnosis is established, leaving to radical surgery only those cases without an adequate response to the primary treatment or those with recurrence or progression of the neoplasia or radiotherapy-related complications.7

There are numerous options to reconstruction, and the reconstruction process and timing should be considered individually. Currently, the continent diversion should be preferably chosen in spite of the fact that there is argument over the usefulness of the noncontinent diversion in cases of reconstruction in young children.4 In this case, the priority is to preserve renal function achieving maximum survival rates and trying to, if possible, facilitate the conversion to a continent ostomy of abdominal catheterization.5

When analyzing the options for continent diversions, one should note the tendency for the use of intestinal segments located outside the irradiated area (transverse colon, proximal ileum), which is justified by the complications that occur in previously irradiated intestinal segments, like actinic obliterating endarteritis.6

When the appendix remains intact, its use as an efferent, abdominally catheterizable conduit is well established (the Mitrofanoff Principle).7 In the absence of this, the Yang-Monti Principle made use of an ileum segment of 2 to 3 cm that, once detubularized, constitutes an alternative to the appendix, demanding, however, an enterectomy.8 Recently Macedo and Srougi2 published a technique with the use of an ileal urinai reservoir that presents an efferent, continent, catheterizable segment adnaxal to the reservoir. The advantage of this technique is to exempt us from the use of the appendix and of the tube of Monti, thus, reducing surgical time and morbidity.2,3

We were able to gather a lot of knowledge from the learning curve of the technique.9 The incision that delineates the transverse flap should also reach the posterior aspect of the intestinal wall, allowing the construction of a channel that would be long enough to be invaginated to the valve and anastomosed to the skin or umbilicus. The ostomy, in cases of amplification, should be placed midway between the umbilicus and the pubis, in such a way as to make the acute angle between the conduit and the anterior wall of the reservoir more stable. In a recent experimental study, our group of researchers found that maintaining such slope is even more important than the length of the valve itself.10 In this RMS case series, the ostomy was placed at the umbilicus. This is because in every patient in this series the surgery performed was for vesical replacement and therefore the reservoir reached that location without complications. The placement of the anterior wall of the new bladder fixed to the peritoneal portion of the anterior abdominal wall strengthens the continent mechanisms, pressing the conduit also against the abdomen.

The authors agree that the number of cases so far is insufficient to let us make assumptions on cancer cure criteria or on the effectiveness of the new technique. However, when reviewing articles on surgical treatment for RMS with continent cutaneous reservoirs, we find that the published series also have limited numbers of cases. Merguerian et al11 reported 13 patients with rhabdomyosarcomas, of which, only 3 received a continent diversion, 2 an incontinent diversion, and 1 an ileocystoplasty for a small noncompliant bladder.

In spite of the fact that the Macedo principle has been developed primarily for ileal reservoirs, we applied the same principle to the transverse colon obtaining excellent results. The advantage of the application of this principle to the transverse colon rests on the fact that this segment is located outside the irradiation field, therefore, being less subject to actinic complications.

The use of continent urinary reservoirs in cases of lack of adequate response to an initial treatment of chemotherapy and radiotherapy in patients with RMS should be fostered. As a rule, one should always opt for the diversion technique with which one’s team is most familiar. In this context, the application of the Macedo Principle comes as an attractive option because not only can it be performed along with the use of the ileum and the transverse colon, but it also exempts the surgeon from the use of the appendix and substitutes. Our current tendency is to perform this technique while reconstructing the urinary tract in patients with RMS, particularly when using the transverse colon.

We, therefore, feel that by performing an alternative surgical option in a complex group of patients, obtaining excellent results, we are opening a world of possibilities so that new surgical teams can apply the same principle and, especially, use the transverse colon because of previous radiotherapy.

**REFERENCES**


3. Adams MC, Joseph DB: Urinary tract reconstruction in chil-