# Laparoscopic Retroperitoneal Lymph Node Dissection for High-Risk Pediatric Patients with Paratesticular Rhabdomyosarcoma

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#### Abstract

*Background and Purpose:* Retroperitoneal lymph node dissection (RPLND) is recommended in children 10 years or older with paratesticular rhabdomyosarcoma (PTRMS). Primary tumors >5 cm are an additional risk factor for disease recurrence in the retroperitoneum. We report our experience with laparoscopic RPLND (LRPLND) in high-risk pediatric patients with PTRMS.

*Patients and Methods:* Three patients, mean age 13.6 years (range 10–16 yrs), underwent modified template LRPLND after radical orchiectomy for preoperative rhabdomyosarcoma stage  $T_{1a}N_0M_0$ ,  $T_{1b}N_0M_0$ , and  $T_{2b}N_0M_0$ , respectively. Primary paratesticular masses measured a mean 7.5 cm (range 4–10 cm). LRPLND was performed a mean of 8.6 days (range 7–12 d) after radical orchiectomy using four trocars that were placed equidistant in the midline.

*Results:* Average operative time was 382 minutes (range 245–656 minutes). Mean estimated blood loss was 53 mL (range 10–75 mL), and mean postoperative hospital stay was 2.5 days (range 2–3 d). There were no post-operative complications. Retroperitoneal nodes had negative findings for microscopic disease in two patients and positive findings in one patient. All patients received adjuvant chemotherapy with vincristine, actinomycin, and cyclophosphamide.

*Conclusion:* LRPLND for high-risk pediatric patients with PTRMS is a safe diagnostic and therapeutic procedure with the benefit of rapid convalescence, enabling early commencement of adjuvant chemotherapy.

#### Introduction

**P**ARATESTICULAR RHABDOMYOSARCOMA (PTRMS) occurs infrequently in the pediatric population, with an incidence of 2 to 4 per million.<sup>1</sup> Although the current standard of care for PTRMS is multimodal, a retroperitoneal lymph node dissection (RPLND) is recommended in those with "high-risk status," including all children 10 years of age or older, regardless of radiologic evidence of retroperitoneal disease.<sup>2</sup> Additional risk factors for disease recurrence in the retroperitoneum include primary tumors >5 cm, unfavorable tumor histology, and group. We report on our experience with laparoscopic RPLND (LRPLND) in high-risk pediatric patients with PTRMS.

#### **Representative Case**

A 16-year-old male underwent a right inguinal orchiectomy for a large painless paratesticular mass. Pathologic evaluation revealed an 8.5 cm embryonal rhabdomyosarcoma with spindle cell components. CT of the chest, abdomen, and pelvis, which was performed before the orchiectomy, had negative findings for nodal disease. Twelve days later, a right modified template LRPLND was performed for preoperative rhabdomyosarcoma stage  $T_{2b}N_XM_X$ .

With the patient in a modified prone position, four 12-mm trocars were placed equidistant in the midline. After medial mobilization of the right colon and kocherization of the duodenum, the ureter and gonadal vessels were identified and separated. The ureter was tented up anteriorly to allow for dissection posterior to the kidney, and the dissection was carried to the superior renal hilum.

Adventitia and lymph nodes that were surrounding the renal vein were identified and removed. The gonadal vessels and surrounding lymph nodes were dissected from the level of insertion into the vena cava to the internal inguinal ring, and removed *en bloc*. Silk sutures placed at the time of radical

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Lumba

FIG. 1. Intraoperative photograph taken during left modified template laparoscopic retroperitoneal lymph node dissection illustrates visualization and preservation of sympathetic chain ganglion (black arrow).

orchiectomy and remnant cord structures were identified, mobilized, and removed, along with surrounding lymph nodes. Iliac and interaortocaval lymphadenectomy was performed using a split-roll technique up to the level of the renal vein. Sympathetic chain ganglia and paraspinal nerves were visualized and spared (Fig. 1). The retrocaval and posterior caval lymph node packets were mobilized and removed. Lymphadenectomy was bounded by the ureter laterally, the renal vein superiorly, the aorta medially, and the inguinal vessels inferiorly to the level of the inguinal canal.

Total operative time was 656 minutes, with an estimated blood loss of 10 mL and a hospital stay of 3 days. Specimens removed included the right iliac, interaortocaval, and retrocaval lymph node packets as well as the right spermatic cord remnant. Metastatic rhabdomyosarcoma was present in 1 of 10 retrocaval periaortic lymph nodes, and in the right iliac nodes. There were no postoperative or intraoperative complications or sequelae. At 1-year follow-up, the patient had completed adjunctive chemotherapy with vincristine, actinomycin, and cyclophosphamide (VAC) with external beam radiation and had no evidence of recurrent disease on CT of the chest, abdomen, and pelvis. He also denied any history of retrograde ejaculation.

### Results

For our three patients, mean patient age was 13.6 years (range 10-16 yrs), with preoperative rhabdomyosarcoma stage T<sub>1a</sub>N<sub>X</sub>M<sub>X</sub>, T<sub>1b</sub>N<sub>X</sub>M<sub>X</sub>, and T<sub>2b</sub>N<sub>X</sub>M<sub>X</sub>, respectively. Primary paratesticular masses measured a mean 7.5 cm (range 4-10 cm). LRPLND was performed a mean of 8.6 days (range 7-12 d) after radical orchiectomy, with an average operative time of 382 minutes (range 245-656 min). Mean estimated blood loss was 53 mL (range 10-75 mL), with a mean postoperative hospital stay of 2.5 days (range 2–3 d). There were no postoperative complications. Retroperitoneal lymph nodes were negative for microscopic disease in two patients and positive in the remaining patient. All patients received adjuvant chemotherapy with VAC. When evaluated at last followup, none of the patients have encountered significant morbidity secondary to surgery, including leg edema, retrograde ejaculation, lymphedema, and hydrocele formation. At 2 years of follow-up, all patients remain negative for recurrent disease.

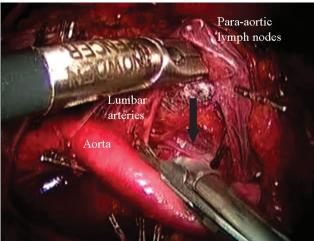
#### Discussion

PTRMS occurs infrequently and presents a challenging problem for pediatric urologists. Although current multimodal therapy using surgery, chemotherapy, and radiotherapy can yield 5-year survival rates of 90%, extent of disease is one of the most important prognostic predictors of which patients are likely to be cured.<sup>3–6</sup> A CT scan performed at the time of diagnosis can detect extension to retroperitoneal lymph nodes, which occurs in up to 20% of patients.<sup>6</sup>

The role of RPLND in PTRMS among children with favorable prognostic factors remains controversial.<sup>4,7,8</sup> The Intergroup Rhabdomyosarcoma Study Group (IRSG) recommended in the early Intergroup Rhabdomyosarcoma Study (IRS) trials that ipsilateral RPLND (IRPLND) be performed for all patients with PTRMS, secondary to microscopic retroperitoneal nodal involvement occurring in 30% to 40% of patients.<sup>2</sup> The paradigm shifted in IRS-IV, however, when chemotherapy was found to be effective for microscopic nodal disease. During this trial, IRPLND was only recommended for those patients with positive lymph nodes on CT of the abdomen and pelvis.<sup>2</sup> Ultimately, this led to understaging of the disease and a decrease in overall disease-free survival, particularly in the adolescent population. Thus, the current recommendations from the Children's Oncology Group Soft Tissue Sarcoma Committee (formerly the IRSG) are that all patients over the age of 10 undergo RPLND, regardless of CT findings.7,9,10

Significant morbidity can be associated with RPLND. Extended follow-up of 86 adolescents and children from IRS I and II revealed numerous complications related to RPLND, including bowel obstruction in nine patients, loss of normal ejaculatory function in eight, development of hydrocele in five, and lymphedema of the leg in five.<sup>11</sup> Review of the surgical morbidity in 478 patients who underwent primary open RPLND for testicular cancer at Indiana University revealed an overall complication rate of 10.6% (28% minor, 72% major), with most major complications related to small bowel obstruction, atelectasis, retrograde ejaculation, and wound infections.<sup>12</sup> Contemporary series of open RPLND performed for testicular carcinoma, however, demonstrate improved short-term morbidity compared with historical controls, with decreased operative time, blood loss, and hospital stay.13

The desire to improve on the morbidity of RPLND has prompted the development and advancement of minimally invasive techniques, such as LRPLND. Proposed benefits of the laparoscopic approach include decreased morbidity and complications, improved intraoperative visualization, cosmesis, and quicker convalescence, resulting in a higher postoperative quality of life.<sup>14,15</sup> Laparoscopy also provides greater magnification, which may facilitate nerve identification and preservation without template modification.



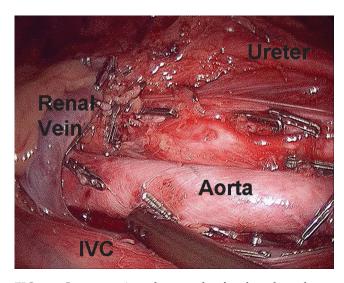


FIG. 2. Intraoperative photograph after lymph node removal illustrates the boundaries of left modified-template retroperitoneal lymph node dissection. IVC = inferior vena cava.

When LRPLND is performed for primary testis cancer, vascular injury is the most common complication, occurring in 2.2% to 20% of reported cases,<sup>16</sup> while retrograde ejaculation occurs in less than 5%; serious complications, such as bowel and nerve injury, retroperitoneal hematoma, and ureteral injury, are rarely reported.<sup>16</sup> Modified template LRPLND may improve postoperative morbidity (Fig. 2). More long-term data are needed to confirm the oncologic efficacy of LRPLND.<sup>17</sup>

In our series, mean operative time, blood loss, and hospital stay for LRPLND were comparable to those reported for open RPLND. There were no operative complications among our patients, and LRPLND did not delay initiation of adjuvant chemotherapy. Pathologic findings at LRPLND were negative in two of three patients, while metastatic embryonal rhabdomyosarcoma was present in 1 of 10 retrocaval periaortic lymph nodes, and in the right iliac nodes. At the most recent follow-up, all patients are negative for disease.

These results are comparable to what has been reported in the literature for the open series, as well as the only other reported series of staging LRPLND for pediatric PTRMS.<sup>16</sup> Although not directly studied here, there appears to be a subjective benefit in convalescence, cosmesis, and morbidity, especially in older children and adolescents.

Disadvantages to the procedure are related to the technical difficulty of the laparoscopic technique. There is a fairly steep learning curve that is associated early on with long operative times, and the need for advanced laparoscopic skills.

#### Conclusion

LRPLND for PTRMS in the pediatric population is technically challenging; however, with experience, excellent success rates with few complications and reasonable operative times can be expected. Results are consistent with those for open RPLND with potentially less postoperative incisional discomfort, a quicker convalescence, and an excellent cosmetic outcome. In experienced hands, LRPLND is a safe staging and potentially therapeutic procedure with the benefit of rapid convalescence, enabling early commencement of adjuvant chemotherapy.

#### **Disclosure Statement**

No competing financial interests exist.

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## Abbreviations Used

- CT = computed tomography
- IRS = Intergroup Rhabdomyosarcoma Study
- IRSG = Intergroup Rhabdomyosarcoma Study Group
- PTRMS = paratesticular rhabdomyosarcoma
- IRPLND = ipsilateral retroperitoneal lymph node dissection
- LRPLND = laparoscopic retroperitoneal lymph node dissection
- RPLND = retroperitoneal lymph node dissection
  - VAC = vincristine, actinomycin, and cyclophosphamide