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Seminar article

Current surgical standards of care in Wilms tumor

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Abstract

Background: Wilms tumor (WT) is the second most common abdominal tumor in children.

Methods: This chapter discusses surgical considerations for the management of unilateral and bilateral WT.

Results: Currently, survival exceeds 90%, owing to multicenter studies under the auspices of the Children's Oncology Group and Société Internationale d'Oncologie Pédiatrique. Surgical excision remains the mainstay of oncologic control and is also crucial for proper staging of disease in order to direct adjuvant therapy and limit treatment-related morbidity.

Conclusions: Careful attention must be paid to proper disease staging, upfront and adjuvant therapy, and surgical technique in order to optimize oncologic outcomes while minimizing short- and long-term morbidity.

Keywords: Wilms tumor; Surgery; Pediatric

Introduction

Wilms tumor (WT) represents one of the greatest success stories in modern medicine, with survival rates increasing from 5% in 1900 to more than 90% currently [1–4]. This increase reflects in large part the systematic data collection and analysis facilitated by 2 multinational consortiums—the National Wilms Tumor Study Group (NWTSG), now part of the Children's Oncology Group (COG), and the Société Internationale d'Oncologie Pédiatrique (SIOP)—which supported several large controlled trials in children with WT. Current survival and late effects outcomes of children with differential WT histologies are shown in Table 1.

WT in Brief

WT is the second most frequent solid organ abdominal tumor (after neuroblastoma) in children. The annual incidence is 8.1 per million children. The mean age at diagnosis is 3 years, with most children between the ages of 1 and 4 years. Most children with WT present with an asymptomatic abdominal mass usually detected by their parents or the physician. A child may also present with hematuria, abdominal pain due to intratumor or extratumor capsular rupture. Overall, 20% of children with WT have hematuria, 10% have coagulopathy, and 20% to 25% present with hypertension owing to activation of the renin-angiotensin system. Fever, anorexia, and weight loss occur in 10%. In rare instances, tumor rupture and bleeding can cause an acute abdomen. WT can extend through the renal vein, inferior vena cava (IVC), and right atrium, or down through the ureter. The common sites of metastatic spread include the lungs and the liver. Ultrasound (US) is a good screening examination to determine if a mass is renal or extrarenal in origin. Computed tomography (CT) scan of the abdomen or magnetic resonance imaging would further define the tumor. A color Doppler US or properly phased CT scan should be used routinely at diagnosis to identify tumor in the renal vein, IVC, or right atrium [1,2]. A CT of the chest would diagnose the presence of pulmonary disease. Approximately 4% of WTs present with IVC or atrial involvement and 11% present with renal vein involvement. Caval thrombosis can cause mortality when an unrecognized thrombus embolizes during nephrectomy.

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Table 1
Relapse-free survival and late effects for children with Wilms tumor

Relapse-Free Survival	Potential for Late Effects	
	Low	Moderate to High
Excellent (≥85%)	Stage I/II FHWT, LOH-	Stage III FHWT, LOH−
Good (75%–84%)		Stage IV FHWT, LOH−
		Stage II AHWT
Unsatisfactory (<75%)	Stage I/II FHWT, LOH+	Stage III/IV FHWT, LOH+
	Stage I AHWT	Stage III/IV AHWT
		Stage V WT
		Relapsed FHWT

AHWT = anaplastic-histology Wilms tumor; FHWT = favorable-histology Wilms tumor; LOH = loss of heterozygosity.

WTs are assigned both an abdominal stage and a disease stage. Treatment is different based on the age, stage, pathology (favorable or anaplastic), response to therapy, and genetic status. The COG staging system is based on the initial pathology (in most cases a primary nephrectomy). The SIOP staging is determined after upfront chemotherapy. Table 2 lists the current COG and SIOP staging systems.

Surgery for children with a WT

There are several important considerations to keep in mind when performing operations on children with WT. The first is to do a safe operation. The second is to thoroughly understand what constitutes a complete procedure. Third, the surgeon plays an important role in accurately staging the disease, which is essential to determine the chemotherapy regimen and the need for radiation therapy. Intraoperative events that negatively affect patient survival include tumor spill, deficient operations, incomplete tumor removal, not assessing for extrarenal tumor extension, and surgical complications.

Unilateral WTs

The current standard surgical procedure for a unilateral WT in a child is a unilateral radical ureteronephrectomy with lymph node sampling [3,4]. Although the kidney can be approached through numerous incisions, a transperitoneal approach first described approximately 75 years ago remains the standard today. Other incisions (flank or paramedian) have been associated with an increased number of complications as well as poorer oncologic outcomes [3–5]. For very large tumors or those that come off the superior pole and extend up to the diaphragm, a thoracic extension of the incision through the eighth or ninth rib helps with exposure.

Complete exploration of the abdomen is mandated, specifically examining the liver, renal vein, and IVC for tumor extension and peritoneal surfaces. Routine exploration of the contralateral kidney was originally mandated. Owing to improvement in imaging, especially spiral CT scans, this is not necessary if imaging is satisfactory and does not suggest a bilateral process [5,6]. If the initial imaging studies were suggestive of a possible lesion on the contralateral kidney, the contralateral kidney should be formally explored before a nephrectomy. Early ligation of the vascular pedicle is ideal; however, WTs can be very large, and it may not be possible or safe to ligate the vessels at first. WTs can be hypervascular and have large areas of necrosis that may predispose to rupture; thus, careful handling of the tumor is needed. Intraoperative tumor rupture occurs in approximately 9% of procedures and is associated with tumors more than 15 cm [7].

Ipsilateral adrenalectomy was once considered a component of radical nephrectomy. However, the need for routine resection of the adrenal gland has been reconsidered in both adult and pediatric renal neoplasms. A review of nephrectomy for nonmetastatic disease in NWTS-4 and NWTS-5 [8] found adrenal involvement in 4.4% of patients, but similar overall and event-free survival (EFS) in patients who had undergone adrenalectomy or adrenal preservation. There were no cases of adrenal insufficiency is any child with a unilateral WT. Other reports support these observations [9]. The current recommendation is that the adrenal gland should remain in situ if possible, but not at the risk of rupturing the tumor.

Once the tumor is removed, lymph node sampling is required. Visual inspection and radiographic determination of nodal status has poor sensitivity and specificity. Lymph nodes should be sampled from the renal hilum and great vessels. Although there are no formal recommendations for the number of lymph nodes to sample, recent retrospective data suggest that a minimum of 7 increases the chances of detecting a metastasis [10]. Failure to sample lymph nodes is the most common error made by surgeons. Lymph node status is important for staging, therapy, and long-term outcome [11]. Failure to sample lymph nodes may result in understaging and increase the risk of recurrence and a poor outcome.

Vascular extension

Extension of tumor into either the extrarenal vascular or the collecting system has been reported in approximately 1