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Evaluation and Management of a Patient with Neuroblastoma or Wilms Tumor

Description of the Activity	Neuroblastoma and Wilms tumor are the most common intra-abdominal solid tumors in children. The pediatric surgeon needs to identify the child at risk for a malignant abdominal mass, initiate a timely workup, and participate in the surgical care of the patient in the context of the larger oncologic care strategy.
Functions	❖ Nonoperative/Preoperative ➢ Neuroblastoma ▪ Perform prenatal counseling for a congenital adrenal mass, recognizing that the differential diagnosis includes adrenal hemorrhage, congenital neuroblastoma, and intra-abdominal extralobar sequestration and that postnatal investigations will include an abdominal ultrasound (US) and urine vanillylmandelic acid (VMA) and homovanillic acid (HVA) levels. Emphasize that it is safe to deliver at term and vaginally. ▪ Perform postnatal management of congenital neuroblastoma. Check catecholamines and abdominal US immediately. Discuss an imaging strategy when proceeding with observation. Discuss the criteria for operative treatment if the patient "fails" observation. ▪ Perform a focused abdominal US and obtain urine VMA and HVA levels at diagnosis and every 3 months for 12 months, and then at 18, 30, 42, 66, and 90 months. ▪ Terminate observation, and refer the patient for surgical resection if the volume of the mass increased by 50% compared with the initial US, if the metabolites increased by more than 50% of the initial baseline value, or if the tumor is still present at the end of the 90-week observation period. ❖ Preoperative ➢ Neuroblastoma ▪ Initiate diagnostic and staging workup, including laboratory tests (complete blood count, comprehensive metabolic panel, coagulation studies, VMA, HVA, ferritin), US, and cross-sectional imaging (computed tomography or magnetic resonance imaging). ▪ Include a bone marrow biopsy, metaiodobenzylguanidine (MIBG) scan, and bone scan in the workup when available. ▪ Understand the International Neuroblastoma Risk Group (INRG) classification system (L1, L2, M, MS) and the importance of image-defined risk factors (IDRFs). Determine resectability based on a review of imaging and stage. ▪ Recognize atypical presentations of neuroblastoma, including retro-orbital metastasis (raccoon eyes) and paraneoplastic syndromes such as diarrhea (vasoactive intestinal peptide [VIP]-secreting tumors) and opsoclonus myoclonus syndrome (OMS). ▪ Discuss the pros and cons of the 2 main approaches to biopsy: open and minimally invasive (MIS). ▪ Discuss the timing of and approach to surgery, recognizing that surgery may not be indicated initially. ▪ Recognize the importance of multidisciplinary care and the possible need for transfer to a higher level of care. ▪ Obtain informed consent, describing the indications, risks, benefits, alternatives, and potential complications of the planned operation, including nuances relevant to the patient's individual condition and comorbidities, and ensure familial understanding. Document the informed consent discussion in the medical record. ▪ Devise an operative plan, and communicate it to the operative team (anesthesia, nursing, techs, assistants), including patient position, anesthesia needs, special instrumentation, and postoperative planning.

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- Wilms tumor
  - Discuss how to stage a Wilms tumor.
  - Recognize that there is a local stage and a disease stage.
  - Verbalize the importance of recognizing preoperative tumor rupture, the role of preoperative biopsy, the need for bowel preparation, and avoiding intraoperative rupture.
  - Assess for vascular invasion into the renal vein and the inferior vena cava (IVC) using US, Doppler, echocardiography, magnetic resonance angiography, or a combination of these.
  - Recognize the presence of and risk factors for syndromic Wilms tumor, and consider nephron-sparing surgery (NSS) if feasible. Identify conditions that predispose to syndromic Wilms tumor, including patients with *WT1* deletions or mutations; **W**ilms tumor/**A**niridia/**G**enitourinary anomalies/**R**ange of developmental delays [WAGR]; Denys-Drash and Frasier syndromes; overgrowth syndromes such as Beckwith-Wiedemann syndrome (BWS); diffuse hyperplastic perilobar nephroblastomatosis (DHPLN); and syndromes associated with other chromosomal anomalies.
  - Discuss the role of neoadjuvant chemotherapy, local radiation, and whole lung radiation in stage III/IV disease.
  - Obtain informed consent, describing the indications, risks, benefits, alternatives, and potential complications of the planned operation, including nuances relevant to the patient's individual condition and comorbidities, and ensure familial understanding. Document the informed consent discussion in the medical record.
  - Devise an operative plan, and communicate it to the operative team (anesthesia, nursing, techs, assistants), including patient position, anesthesia needs, special instrumentation, and postoperative planning.

- Intraoperative
  - Neuroblastoma
    - Discuss surgical approach and exposure, recognizing that MIS is safe for most L1 tumors. Open is standard and offers superior exposure, particularly for L2 tumors.
    - Identify key principles, including:
      - Obtain wide exposure and proximal and distal control of the aorta or encased vessels when possible.
      - Bivalve the tumor, and remove it piecemeal if indicated to dissect out encased vasculature.
      - Do not perform en bloc resection of other organs.
      - Subtotal resection is the goal but is not mandatory.
      - Do not dissect into the neural foramina.
      - Consider intraoperative nerve monitoring for cervical and pelvic tumors.
    - Perform a thoracoabdominal incision for large L2 tumors traversing the thorax and abdomen.
  - Wilms tumor
    - Obtain safe initial exposure, and identify anatomic distortion in large Wilms tumors.
    - Maintain the integrity of the kidney capsule, recognizing the importance of avoiding intraoperative rupture.
    - Spare the adrenal gland when able; remove the adrenal gland en bloc with an upper pole Wilms tumor.
    - Palpate the renal vein to assess for tumor thrombus; perform venotomy and intimal dissection to remove the tumor.

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- Recognize that lymph node (LN) sampling is mandatory, with a minimum of 5 nodes:
  - Left side: aortocaval window, periaortic, left hilar
  - Right side: right hilar, pericaval, aortocaval window
- Discuss the indications for partial nephrectomy, including patients with bilateral Wilms tumors and those at risk for developing additional tumors in the future.

❖ Postoperative

➢ Neuroblastoma

- Anticipate, recognize, and treat potential complications based on the primary tumor site, including:
  - Abdomen: hypertension, chyle leak, diarrhea, small bowel obstruction (SBO), bowel ischemia, renal ischemia/atrophy, diaphragm injury
  - Cervical: brachial plexus injury, chyle leak
  - Pelvic/paraspinal: paralysis, retrograde ejaculation
  - Thorax: Horner syndrome, chyle leak, paralysis
- Demonstrate understanding of the importance of INRG risk stratification, tumor biology, segmental chromosomal abnormalities, and multimodal treatment strategies and their effect on oncologic outcome.
- Demonstrate understanding that neuroblastoma is unique in that biology is critical to staging, treatment, and prognosis. List common genetic aberrations and the effect of each on outcomes and novel treatments.

➢ Wilms tumor

- Anticipate, recognize, and treat potential complications, such as urine leak, intussusception, SBO, chyle leak, and renal failure.
- Recognize that tumor anaplasia and genetic aberrations, such as loss of heterozygosity at chromosome 1p and 16p and gain of function at 1q, have implications in terms of prognosis and risk groups.
- Describe risk-adjusted multimodal therapy and the potential of treatment de-escalation
  - Avoiding flank radiation if the tumor is completely resected and not ruptured,
  - Avoiding whole lung irradiation if there is a complete response to lung nodules in tumors with metastasis,
  - Decreasing the doxorubicin dose for patients with low-risk features.

Scope

❖ In scope

- Diagnoses
  - Renal tumor
    - Clear cell tumor kidney
    - Mesoblastic nephroma
    - Rhabdoid tumor kidney
    - Wilms tumor

➢ Neuroblastoma

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- Anatomic location: head/neck, chest, intra-abdominal, pelvic
- Ganglioneuroma, ganglioneuroblastoma

❖ Procedures:

- ➢ Biopsy for neuroblastoma
- ➢ Neuroblastoma resection and LN sampling
- ➢ Partial nephrectomy with LN dissection
- ➢ Thoracic exposure for neuroblastoma resection
- ➢ Unilateral radical nephroureterectomy with LN dissection

- Special populations
  - Patients with:
    - Bilateral Wilms tumor
    - Syndromic Wilms tumor, Denys-Drash syndrome, WAGR
    - Syndromic neuroblastoma, OMS, invading foramina with paralysis
    - Initial nonoperative treatment:
      - Patients with L2 neuroblastoma encircling vasculature
      - Newborn with an adrenal mass
      - Patients with Wilms tumor invading into the IVC

❖ Out of scope

- Diagnoses/procedures
  - ➢ Neurofibromatosis
  - ➢ Other adrenal tumors: adrenal cortical carcinoma, pheochromocytoma, adrenal hyperplasia, and adrenal hemorrhage
  - ➢ Other renal tumors

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1 **Framework:** The attending will show and tell or the learner acts as first assistant. **Entrustment:** The learner demonstrates understanding of information and has basic skills. What a new pediatric surgery fellow should know.	• With active guidance, initiates the workup and staging of a patient with suspected neuroblastoma or Wilms tumor, including US, cross-sectional imaging (CT/MRI), and biochemical markers, and demonstrates basic knowledge of typical/pathognomonic findings • With active guidance, verbalizes indications/contraindications for biopsy when working up a patient with suspected neuroblastoma or Wilms tumor • With active guidance, reviews prenatal diagnostic imaging during a prenatal consultation for a fetus with an adrenal mass • With active guidance, integrates results of imaging studies, biochemical markers, and biopsies to accurately assign a stage and risk group in a patient with neuroblastoma or Wilms tumor • Recognizes the importance of multidisciplinary communication for a patient with neuroblastoma or Wilms tumor • Establishes a culturally sensitive rapport with the patient's family and demonstrates empathy during the initial consultation	• With active assistance, prepares the patient for surgery in collaboration with anesthesiology and the care team, including ensuring the availability of blood products and all special equipment • Accesses the body cavity and identifies the tumor mass and distorted anatomy • Understand the importance of assessing for preoperative rupture and avoiding intraoperative rupture in Wilms tumor • Demonstrates basic knowledge of the role of lymph node assessment or resection during resection of neuroblastoma or Wilms tumor • With active assistance, makes intraop decisions regarding the extent of resection/role of en bloc resection of critical surrounding structures in the surgical management of neuroblastoma or Wilms tumor • With active guidance, identifies intraop complications	• With active assistance, guides the postop phase of care, including evaluation and management of pain and simple postop problems such as wound complications, ileus, and SBO • Demonstrates understanding of how to access and use available evidence and incorporate patient/family preferences and values into the postop oncologic treatment plan • With active guidance, articulates operative findings, postop expectations, and next steps with the patient and family immediately following the operation • With active assistance, communicates operative findings and postop expectations with members of the multidisciplinary team

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2 **Framework:** The learner demonstrates understanding of the steps of the operation but requires direction through principles and does not know the nuances of a basic case. **Entrustment:** The learner can use the tools but may not know exactly what, where, or how to do it. The attending gives active help throughout the case to maintain forward progression or may need to take over the case at a certain point.	• Under direct supervision, interprets preoperative lab results and atypical/complex imaging findings for a patient with neuroblastoma or Wilms tumor • Under direct supervision, verbalizes the indications/contraindications for biopsy during the workup of a patient with suspected neuroblastoma or Wilms tumor • Under direct supervision, interprets prenatal imaging and discusses the differential during a prenatal consultation for a fetus with an adrenal mass • With passive guidance, integrates results of imaging studies, biochemical markers, and biopsies to accurately assign a stage and risk group in a patient with neuroblastoma or Wilms tumor • Identifies key consultants and seeks out multidisciplinary preoperative conferences for a patient with suspected neuroblastoma or Wilms tumor • Establishes a therapeutic relationship with the family of a straightforward patient with neuroblastoma or Wilms tumor and obtains informed consent, incorporating the family's values	• With direct supervision, prepares the patient for surgery in collaboration with anesthesiology and the care team, including ensuring availability of blood products and all special equipment • Mobilizes surrounding organs to safely expose and initiate dissection of the tumor mass, requiring assistance to determine the extent of resection • Assesses patient with a Wilms tumor for preoperative rupture, actively avoids intraop rupture, and demonstrates delicate tissue handling • Demonstrates detailed understanding of the role of lymph node assessment or resection during resection of a neuroblastoma or Wilms tumor • With passive assistance, makes intraop decisions regarding the extent of resection/role of en bloc resection of critical surrounding structures in the surgical management of neuroblastoma or Wilms tumor • With direct supervision, addresses intraop complications	• With passive assistance, guides the postop phase of care, including evaluation and management of pain and simple postop problems such as wound complications, ileus, or SBO • Integrates available evidence and family preferences/values to guide the timing of postop oncologic interventions and elicits the patient's and family's preferences and values to guide evidence-based care • With passive guidance, articulates operative findings, postop expectations, and next steps with the patient and family immediately following the operation • With passive assistance, communicates operative findings and postop expectations with members of the multidisciplinary team
3 **Framework:**	• With indirect supervision, interprets distorted anatomy and atypical imaging	• With indirect supervision, positions the patient and collaborates with the	• With passive assistance, manages the postop care of a complex patient

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The learner has a good understanding of surgical options and techniques but does not recognize abnormalities and does not understand the nuances of a complicated case. **Entrustment:** The learner can perform the operation/task independently in the uncomplicated patient or The attending provides passive/indirect supervision/suggestions in the complicated patient but still allows the learner to perform the operation/task themselves.	findings in a patient with neuroblastoma or Wilms tumor • With indirect supervision, recognizes atypical presentations of neuroblastoma or Wilms tumor, including paraneoplastic syndromes and cancer predisposition syndromes, and modifies the diagnostic and surgical plan accordingly • With indirect supervision, interprets diagnostic information and discusses postnatal expectations, workup, and the treatment plan, including observation vs operative treatment during a prenatal consultation for a fetus with an adrenal mass • With passive guidance, integrates image-defined risk factors (IDRF) in neuroblastoma staging and displays understanding of how they affect the timing of surgical intervention • In a multidisciplinary setting, articulates the diagnostic findings and surgical plan for a straightforward patient with a neuroblastoma or Wilms tumor • With passive assistance, establishes a therapeutic relationship with the family of a complex patient with neuroblastoma or Wilms tumor and obtains informed consent, incorporating the family's values and using shared decision-making	anesthesia team regarding invasive monitoring and anesthetic strategy • Resects an uncomplicated neuroblastoma or Wilms tumor • With passive assistance, gains vascular control in a complex patient with neuroblastoma or abdominal Wilms tumor • With passive assistance, performs resection of involved lymph nodes in a patient with neuroblastoma or formal lymphadenectomy in a patient with Wilms tumor • With passive assistance, performs complex operative maneuvers in resection of a neuroblastoma or Wilms tumor, such as safe tumor bivalving over encased vascular structures in neuroblastoma or partial nephrectomy in bilateral Wilms tumor • With indirect supervision, addresses intraop complications	undergoing tumor resection, including anticipating, diagnosing, and treating a rare or complex complication such as HTN, Horner syndrome, chyle leak, or urine/bowel leak • Uses best available evidence to determine the optimal postop oncology treatment plan while integrating the patient's medical status, parental preferences, and family resources • Customizes emotionally difficult news such as unanticipated surgical findings or changes to the postop plan to the family in an empathetic and culturally sensitive manner • With passive assistance, implements recommendations from the multidisciplinary care team and uses shared decision-making to align the postop care plan with the patient's and family's values and preferences

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4 **Framework** The learner has a strong and indepth understanding of surgical options and techniques. **Entrustment:** Can perform the operation/task independently in complicated cases or The attending may need to provide indirect supervision or suggestions in the context of extremely rare or severely complicated cases	• Independently modifies the surgical plan for a patient with neuroblastoma or Wilms tumor based on complex or atypical imaging findings such as bilateral tumors, IVC tumor thrombus, or image defined risk-factors (IDRFs), specifically addressing the timing of surgical intervention • Independently recognizes atypical presentations of neuroblastoma or Wilms tumor, including paraneoplastic syndromes and cancer predisposition syndromes, and modifies the diagnostic and surgical plan based on initial biopsy results, disease staging, and unique imaging findings • Independently interprets diagnostic information during a prenatal consultation for a fetus with an adrenal mass and discusses the criteria to change the postnatal management plan from observation to surgery • Independently integrates IDRFs in neuroblastoma staging and displays understanding of how it affects the timing of surgical intervention • In a multidisciplinary setting, independently articulates the diagnostic findings and surgical plan for a complex patient with neuroblastoma or Wilms tumor and critically appraises and applies evidence from up-to-date treatment protocols, including COG and other guidelines	• Independently positions the patient and collaborates with the anesthesia team regarding invasive monitoring and anesthetic strategy • Performs radical nephroureterectomy and adjunct procedures such as IVC thrombectomy in a patient with complex Wilms tumor • Independently obtains vascular control in a complex patient undergoing neuroblastoma or Wilms tumor resection • Independently performs resection of involved lymph nodes in neuroblastoma or formal lymphadenectomy in Wilms tumor • Independently performs complex operative maneuvers in resection of neuroblastoma or Wilms tumor, such as safe tumor bivalving over encased vascular structures in neuroblastoma or partial nephrectomy in bilateral Wilms tumor • Independently addresses intraop complications	• Independently leads the team in managing postop care, including anticipating, diagnosing, and treating postop complications • Critically appraises and applies evidence, including the effect of pathology and genetic results on risk stratification and subsequent oncologic treatment strategy, tailored to the patient and family • Communicates complex surgical findings and plans to the family, including treatment-specific side effects and long-term complications of cancer therapy such as recurrence/relapse and secondary malignancies • Independently implements recommendations from the multidisciplinary care team and uses shared decision-making to align the postop care plan with the patient's and family's values and preferences

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	• Establishes a therapeutic relationship with the family of a complex patient with neuroblastoma or Wilms tumor and independently obtains informed consent, incorporating the family's values and using shared decision-making		