

# Contents

<b>Foreword: Pediatric Cancer</b>	<b>xiii</b>
Timothy M. Pawlik	
<b>Preface: An Introduction to Pediatric Surgical Oncology</b>	<b>xv</b>
Roshni Dasgupta	
<b>Pediatric Gastrointestinal Stromal Tumors and Neuroendocrine Tumors: Advances in Surgical Management</b>	<b>219</b>
Hanna Garnier, Caitlyn Loo, Piotr Czauderna, and Sanjeev A. Vasudevan	
<p>Gastrointestinal stromal tumors and neuroendocrine tumors in adult and pediatric populations differ immensely. Despite these established differences, the extreme rarity of gastrointestinal stromal tumors and neuroendocrine tumors in the pediatric population has resulted in the lack of consensus management guidelines, making optimal surgical approaches unclear. Comprehensive management principles to guide surgical approaches in adult literature are extensive. However, these are still lacking for pediatric patients. International cooperation to develop standardized pediatric-specific guidelines is urgently warranted in the future. This article highlights the vast differences between adult and pediatric parameters and provides recommendations on optimal and novel surgical approaches in children.</p>	
<b>Management of Differentiated Thyroid Carcinoma in Pediatric Patients</b>	<b>235</b>
Emily Christison-Lagay and Reto M. Baertschiger	
<p>Differentiated thyroid carcinomas are rare in young children but represent almost 10% of all malignancies diagnosed in older adolescents. Differentiated thyroid carcinoma in children is more likely to demonstrate nodal involvement and is associated with higher recurrence rates than seen in adults. Decisions regarding extent of surgical resection are based on clinical and radiologic features, cytology, and risk assessment. Total thyroidectomy and compartment-based resection of involved lymph node basins form the cornerstone of treatment. The use of molecular genetics to inform treatment strategies and the use of targeted therapies to unresectable progressive disease is evolving.</p>	
<b>Liver Tumors in Pediatric Patients</b>	<b>253</b>
Rebecka Meyers, Eiso Hiyama, Piotr Czauderna, and Greg M. Tiao	
<p>The most recent advance in the care of children diagnosed with hepatoblastoma and hepatocellular carcinoma is the Pediatric Hepatic International Tumor Trial, which opened to international enrollment in 2018. It is being conducted as a collaborative effort by the pediatric multicenter trial groups in North America, Europe, and the Far East. This international effort was catalyzed by a new unified global risk stratification system for</p>	

hepatoblastoma, an international histopathologic consensus classification for pediatric liver tumors, and a revised 2017 collaborative update of the PRE-Treatment EXTent of disease radiographic based staging system.

**Management of Adrenal Tumors in Pediatric Patients** **275**

Simone de Campos Vieira Abib and Christopher B. Weldon

Pediatric adrenal tumors are uncommon entities that are frequently occult and identified incidentally or by recognizing symptoms related to hormone overproduction. They often have a genetic underpinning, arise from the medulla or cortex, can be malignant or benign, and require precise diagnostic algorithms. However, pseudotumors must also be a diagnostic consideration. Therapeutic interventions and plans are tumor dependent, but surgery is a cornerstone of treatment. Ongoing surveillance after treatment, regardless of malignant determination, is of utmost importance as well.

**Management of Neuroblastoma in Pediatric Patients** **291**

Nikke Croteau, Jed Nuchtern, and Michael P. LaQuaglia

Surgeons caring for patients with neuroblastoma must be familiar with recent developments in assessing risk. In particular, the Children's Oncology Group, along with major international groups, uses the International Neuroblastoma Risk Group Staging System as a risk assessment tool. Accurate risk determination is essential for optimal surgical therapy. Some tumors like neonatal adrenal neuroblastomas and those in the metastatic category can be observed. Very-low-risk and low-risk neuroblastomas can be treated with surgery alone. Intermediate-risk tumors also often require systemic chemotherapy

**Surgical Management of Wilms Tumor (Nephroblastoma) and Renal Cell Carcinoma in Children and Young Adults** **305**

Natalie M. Lopyan and Peter F. Ehrlich

This article reviews the epidemiology, pathophysiology, clinical presentation, and multimodality management of Wilms tumors and renal cell carcinoma in pediatric and young adults. Key renal Société Internationale d'Oncologie Pédiatrique and Children Oncology Group studies are presented. The article reviews the common staging systems and risk-adapted treatment strategies with particular attention to the surgical management.

**Management of Germ Cell Tumors in Pediatric Patients** **325**

Brent R. Weil and Deborah F. Billmire

Germ cell tumors arise from primordial germ cells. Most develop in the gonads or along midline structures of the body. Genetic aberrations leading to disruption in the molecular signaling responsible for primordial germ cell migration early in development may provide rationale for why germ cell tumors originate in extragonadal locations. Establishing best practices for treating pediatric germ cell tumors remains an area of active investigation. Recent advances focused on limiting toxicities of therapy, identifying new therapies for relapsed and refractory tumors, defining best practices for

surgical staging and resection, and developing novel methods to monitor for disease relapse.

### **Management of Rhabdomyosarcoma in Pediatric Patients**

339

Timothy N. Rogers and Roshni Dasgupta

Rhabdomyosarcoma is the commonest soft tissue sarcoma in children. Clinicians need vigilance to recognize the different signs and symptoms this tumor can present with because of variable sites of origin. Diagnosis requires a safe biopsy that obtains sufficient tissue for pathologic, genetic, and biological characterization of the tumor. Treatment depends on accurate staging with imaging and surgical sampling of draining lymph nodes. A multidisciplinary team assigns patients to risk-based therapy. Patients require chemotherapy and usually a combination of complex, site-specific surgery and/or radiotherapy. Outcomes for localized rhabdomyosarcoma continue to improve but new treatments are required for metastatic and relapsed disease.

### **Treatment Concepts and Challenges in Nonrhabdomyosarcoma Soft Tissue Sarcomas**

355

Joerg Fuchs, Andreas Schmidt, Steven W. Warmann, and David A. Rodeberg

Pediatric nonrhabdomyosarcoma soft tissue sarcomas (NRSTSs) encompass a heterogeneous group of mesenchymal tumors with more than 50 histologic variants. The incidence of NRSTS is greater than rhabdomyosarcoma; however, each histologic type is rare. The treatment schema for all NRSTSs is largely surgical. The treatment is a risk-adapted approach based on tumor size, localization, tumor grade, and presence of metastases. Low-grade tumors are mainly managed by surgery alone, whereas for high-grade tumors a multimodal treatment concept is necessary. The multimodal treatment consists of tumor biopsy, chemotherapy, local treatment (surgery  $\pm$  radiotherapy), and immunotherapy in selected conditions.

### **Pediatric Melanoma—Diagnosis, Management, and Anticipated Outcomes**

373

Jennifer H. Aldrink, Stephanie F. Polites, and Mary Austin

Melanoma is the most common skin cancer in children, often presenting in an atypical fashion. The incidence of melanoma in children has been declining. The mainstay of therapy is surgical resection. Sentinel lymph node biopsy often is indicated to guide therapy and determine prognosis. Completion lymph node dissection is recommended in selective cases after positive sentinel lymph node biopsy. Those with advanced disease receive adjuvant systemic treatment. Because children are excluded from melanoma clinical trials, management is based on pediatric retrospective data and adult clinical trials. This review focuses on epidemiology, presentation, surgical management, adjuvant therapy, and outcomes of pediatric melanoma.

### **A Surgical Approach to Pulmonary Metastasis in Children**

389

Jonathan Karpelowksy and Guido Seitz

Decisions regarding the role of surgery in pulmonary metastasis need to take into account histology and biology of the cancer. Response to

chemotherapy and radiotherapy, balanced with toxicities, factors into decisions about metastasectomy. The less sensitive the tumor is to adjuvant therapy, the more likely that metastasectomy may be beneficial. Broad principles include the following: the aims of resection are localized resections with clear margins, with the aim of preserving adequate lung volume; unnecessary toxic therapy sometimes is avoided with accurate diagnosis; tumor type is of utmost importance; and number of metastases and the disease-free interval are not contraindications to metastasectomy.

### **Fertility Considerations in Pediatric and Adolescent Patients Undergoing Cancer Therapy**

401

Timothy B. Lautz, Karen Burns, and Erin E. Rowell

Survivors of pediatric cancer are at increased risk for infertility and premature hormonal failure. Surgeons caring for children with cancer have an important role to play in understanding this risk, as well as advocating for and performing appropriate fertility preservation procedures. Fertility preservation options in males and females vary by pubertal status and include nonexperimental (oocyte harvest, ovarian tissue cryopreservation, sperm cryopreservation) and experimental (testicular tissue cryopreservation) options. This review summarizes the basics of risk assessment and fertility preservation options and explores unique considerations in pediatric fertility preservation.

### **Minimally Invasive Techniques in Pediatric Surgical Oncology**

417

Marc W.H. Wijnen and Andrew M. Davidoff

Minimally invasive approaches to pediatric cancer surgery are increasingly used, not only for the benefits of smaller incisions, but also for better field visualization and precise dissection. Advances in technology and surgeon experience have facilitated this trend. However, the appropriate indications for its use remain to be determined, and oncologic principles should not be compromised. We discuss the current and potential future uses, and new technologies that are being developed and introduced to assist with and enhance the role of minimally invasive surgery in the management of children with cancer.