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Retroperitoneal Sarcomas

Soft tissue sarcomas are a heterogenous group of rare tumors arising from embryonic mesoderm. Almost 15% of such sarcomas arise in the retroperitoneum. Rhabdomyosarcoma and fibrosarcoma are the two most common histologic variants in the retroperitoneum. The prognosis for patients with retroperitoneal sarcomas (RPS) is relatively poor characterized by late locoregional recurrence as principal cause of death. In the retroperitoneum tumor growth has a large capacity before causing overt symptoms reaching enormous size and invading adjacent vital vascular structures. At diagnosis RPS are the largest tumors found in the human body. Even with large size RPS rarely metastasize. The best potential curative treatment (a survival factor) is macroscopically complete, margin-negative gross surgical resection. The size and complexity of RPS tumors result in microscopically residual disease after surgery needing the use of adjuvant chemo- and radiotherapy. Rhabdomyosarcoma (RMS) is the most common soft tissue sarcoma arising in the retroperitoneum in children. Retroperitoneal RMS are quite large and seen at CT as a bulky mass with heterogenous attenuation equal to or slightly less than muscle. Areas of attenuation representing necrosis are common and calcifications are rare. The precise origin of the tumor is often difficult to determine because of infiltration of adjacent organs. Retroperitoneal and inguinal lymph node enlargement and bone and lung metastasis may be seen. RMS is more responsive to chemo- and radiotherapy in children than adults. Tumor histology and responsiveness to neoadjuvant therapy influence resectability. Debulking of RMS in combination with chemo- and radiotherapy induce tumor shrinkage and facilitate tumor resection improving survival. Children with low-grade tumors have better survival as compared to those with high-grade sarcomas. The efficacy of current chemotherapy is limited and there is a critical need to understand the molecular basis of sarcomas so that new drug therapies are developed.

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Renal Cell Carcinoma

Renal cell carcinoma (RCC) is an uncommon malignant tumor arising from an epithelial cell of the renal tubules accounting for 3% of all pediatric renal tumors. Median age is between 8 and 17 years with no gender predominance. Underlying associated conditions includes tuberous sclerosis and prior chemotherapy. Most RCC presents with symptoms such as flank pain, hematuria and abdominal mass with few cases diagnosed after incidental radiology studies, usually ultrasound. Children present with higher stage, higher grade and larger tumors when compared with older patients. Diagnosis is confirmed with CT-Scan and MRI. 30% of pediatric RCC presents with metastatic disease such as lymphadenopathy, vascular involvement, local and distant metastasis to liver, contralateral kidney or lungs. Differential diagnosis includes nephroblastoma. Calcifications are a single radiologic feature associated with 50% of RCC. Pathologic subtypes of RCC include the papillary histology most commonly (30-80%) followed by relative dearth of clear cell type (17-50%). In children RCC demonstrates translocation in the Xp11.2 (TFE3 gene) most commonly followed by the 6p21 loci (TFEB gene). Translocation tumors tend to have rather indolent disease with a good outcome even in the presence of advance disease. Children with Von Hippel Lindau syndrome typically develop clear cell RCC at a young age which can be multifocal or bilateral. Neuroblastoma survivors have a 300-fold increase risk of developing RCC. Surgical excision is the mainstay treatment of RCC and a significant prognostic factor. Radical nephrectomy is the most commonly used surgical procedure. Partial nephrectomy is performed in tumors less than 4 cm, location amenable to partial resection and Robson stage 1 or 2 lesions with and excellent five year survival. Children with associated syndromes and RCC should also under partial nephrectomy since they will require repeated resections. Laparoscopic nephrectomy has been proved equally effective to open surgery in RCC when the tumor does not cross the midline. Long-term survival of RCC is affected by tumor size, lymph node status and pathologic stage.

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Incisional Hernias

Incisional hernia (IH) is a frequent postoperative complication after abdominal surgery in children and adults. Incisional hernia occurs with greater incidence following open surgical procedures than with laparoscopic procedures. Emergency neonatal laparotomies are the mot common primary surgery associated with incisional hernias, with necrotizing enterocolitis comprising the major group. IH presents clinically as a reducible bulging in the scar area. Almost one-third of the patients who had an IH were unaware of the presence of the hernia. Ultrasound and CT-Scans increase the rate of detection of incisional hernias. Risk factors associated in the development of IH in children include age less than six months, wound infection, median incisions and emergency procedure. Most IH will developed in the next two years after the original abdominal procedure. Vertical incisions have a greater incidence of hernia development than transverse abdominal procedures in children. Guidelines to avoid incisional hernias include avoiding vertical incisions and closure using an absorbable monofilament suture in a single layer fascia closure technique without separate closure of the peritoneum. For laparoscopic surgery recommendations of closing the port defect whenever feasible, especially those of 10 mm. Indications for repair of incisional hernia should include symptoms of pain, limitation in daily activity and evident enlargement of the hernia defect. Methods of repair include primary closure whenever possible or mesh repair using open or laparoscopic technique. The most common group of pediatric patient who underwent an IH repair were those following closures of stomas.

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