



PEDIATRIC SURGERY Update ©

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Desmoplastic Small Round Cell Tumor

Within the non-rhabdomyosarcoma soft tissue tumors in children, the Desmoplastic Small Round Cell Tumor (DSRCT) is an extremely locally aggressive and rare tumor with predilection for children and adolescent patients and predominant or exclusive intra-abdominal location. DSRCT is characterized by a generally diffuse pattern of growth of small round cells with hyperchromatic nuclei, scanty cytoplasm, patchy epithelial differentiation, immunohistochemical co-expression of keratin and desmin intermediate filaments and a focal but pronounced desmoplastic stromal component. This neoplasm exhibits a predominantly intra-abdominal serosal pattern with frequent pelvic extension, less frequent retroperitoneal involvement and very rarely pulmonary and mediastinal spread. The child usually present with a palpable, painful mass which may cause abdominal distension, constipation, bowel or ureteral obstruction. On imaging bulky peritoneal soft-tissue masses without an apparent organ-based primary site are characteristic of intra-abdominal DSRCT. DSRCT can be mistaken for a rhabdomyosarcoma as it contains cells with rhabdoid features. Cytogenetic studies showed a t(11;22) translocation that differs from the Ewing's tumor translocation and seems to be specific to this entity. Management consists of dose intensive multimodal chemotherapy, aggressive surgery to resect visible disease, local radiotherapy, and myeloablative chemotherapy with stem-cell rescue in selected cases. Unfortunately the prognosis is very poor and most children succumb to widespread metastasis disease.

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Juvenile Fibroadenoma

Juvenile or giant fibroadenoma of the breast is a benign lesion that can obtain a large

formidable proportional size during breast development in female adolescent patients. Most cases in children are seen between the ages of 10 and 15 years. The tumor is solitary in most affected children with a diameter of 4-6 centimeters. Multiple and bilateral involvement has been reported in a few cases. Differential diagnosis includes cystosarcoma phylloides, benign virginal hypertrophy (juvenile gigantomastia) or rhabdomyosarcoma. FNA or Tru-cut needle biopsy can establish a precise histological diagnosis. Growth is so fast that it can cause non-tender cellulitis of the skin by way of stretching. Microscopically the tumor is characterized by a rich cellular stroma and a prominent glandular epithelium. Juvenile adenofibromas regardless of size, should be excised so as to preserve as much breast tissue as possible. Management options include local excision with reconstruction, reduction mammoplasty, or simple mastectomy with reconstruction.

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Laparoscopic Bowel Resection

The enthusiasm brought by laparoscopic surgery results has expanded its capabilities to other intra-abdominal surgical procedures such as isolated or limited bowel resection in children. So far reported indications have included: resection of a Meckel's diverticulum, post-enterocolitis small bowel or colonic stricture, localized mesenteric cyst, inflammatory bowel disease (IBD), intussusception, intestinal lymphangioma and duplication. Technically the approach can include mobilization with extracorporeal resection and anastomosis (video-assisted), resection with intracorporeal stapled anastomosis or resection with hand-sewn end-to-end anastomosis depending on the lesion, mobility, size of the bowel and ability of the surgeon. Largest experience with lap bowel resection and anastomosis has been for inflammatory bowel disease, specially Crohn's disease children. Laparoscopically assisted colectomies can be performed safely in treating IBD. Laparoscopic-assisted ileocolic resection has also been found a safe alternative to open surgery in adolescent patients with Crohn disease. Lap bowel resection has offered a faster recovery of pulmonary function, fewer complications, and shorter length of stay compared with conventional surgery for selected patients undergoing ileocolic resection for Crohn's disease.

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* Edited by: **Humberto Lugo-Vicente, MD, FACS, FAAP**

Professor / Academic Director of Pediatric Surgery, University of Puerto Rico School of Medicine and
University Pediatric Hospital, Rio Piedras, Puerto Rico.

Address: P.O. Box 10426, Caparra Heights Station, San Juan, Puerto Rico USA 00922-0426.

Tel (787)-786-3495 Fax (787)-720-6103 E-mail: *titolugo@coqui.net*

Internet: <http://home.coqui.net/titolugo>

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