



PEDIATRIC SURGERY Update ©

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Trunk Rhabdomyosarcoma

Rhabdomyosarcoma (RMS) is the most common soft tissue sarcoma in children comprising one half of pediatric soft tissue sarcomas and 15% of all pediatric solid tumors. RMS can arise anywhere in the body and survival has been found highly dependent on such variables as tumor site, extent of disease and management. RMS arising in the trunk area is associated with a poor prognosis. The trunk includes chest wall, abdomen and paraspinal region. Trunk RMS presents as a painless, firm mass that increases rapidly in size. The diagnosis is made by incisional or excisional biopsy of the mass. Most patients with trunk RMS have embryonal histology. Adolescent patients with trunk RMS fare worse than younger children. This is due to the high incidence of unfavorable (alveolar) histology in adolescent cases. Factors associated with unfavorable outcomes after managing trunk RMS includes tumor size greater than 5 cm, advance stage at presentation, alveolar histology, lymph node involvement, and local and distant tumor recurrence. Complete surgical resection including the full-thickness of the abdominal or chest wall when feasible with clear tumor margins is the cornerstone of effective and curative management. The abdominal wall is a site at which tumors can be resected widely with minimal morbidity and the majority of wounds can be closed primarily or with substitution using a biologic mesh. When upfront surgery is not feasible, neoadjuvant chemotherapy followed by delayed surgical resection should be considered. Patient with bulk residual tumor remaining after surgery or metastatic disease requires adjuvant therapy (radiotherapy and chemotherapy).

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Gorham's Disease

Gorham's disease is a very rare lymphatic vascular malformation involving bone and leading to massive osteolysis. The disease is also known as vanishing bone syndrome or Gorham-Stout syndrome. The cause of the disease is not known but there is evidence that massive osteolysis is associated with increased osteoclast activity and increase sensitivity of osteoclast precursors to interleukin 6 which promotes bone resorption. There is no gender or race predilection. Gorham's disease affects more commonly adolescent and young adults. The disease mainly affects the shoulder, pelvis, and skull crossing joint boundaries. Mandible, ribs, scapula, humerus, pelvis and femur can be involved. The child presents with dull pain, swelling of the affected area and pathological fractures. Involvement of pleura and vertebrae is associated with poor prognosis since it leads to chylothorax and nerve root compression. Chylothorax occurs in one-fifth of all cases. Diagnosis of Gorham's disease is difficult. MRI shows evidence of massive osteolysis. The affected bone undergoes resorption replaced by hyper vascular fibrous tissue. Histology shows a non malignant proliferation of dilated thin walled vascular and lymphatic channels with associated loss of bone matrix. Splenic lesions and soft-tissue involvement are common in patients with Gorham's disease. Management of Gorham's disease includes sclerotherapy, immunotherapy, radiotherapy and surgery. Surgery to replace the affected bone with a graft or prosthesis has also been used successfully in early and localized disease. Death occurs due to malnutrition, lymphocytopenia or infection.

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Laparoscopic Peritoneal Cannula Placement

Peritoneal dialysis is preferred to hemodialysis to manage chronic renal failure in children because it has a lower incidence of serious complications, is more cost-effective and improves patient nutrition and independence. The peritoneal cannula for such purposes can be placed using an open conventional surgical technique, percutaneously or laparoscopically. Laparoscopic placement of peritoneal dialysis catheters or revision allows complete visualization of the peritoneal cavity placing the catheter under direct

vision, preferably in the pelvis. Suturing the catheter tip into the pelvis is associated with a low rate of catheter migration. In addition a more complete omentectomy can be performed and lysis of adhesions can be accomplished to increase the peritoneal absorptive surface. Omentectomy is beneficial during placement of peritoneal dialysis catheters to prevent blockage of the catheter. Laparoscopy can allow for the rescue of block catheters in cases of revisions due to malfunction. Others procedures that can be done concomitantly using the laparoscopic technique include repair of inguinal hernias, gastrostomy tube placement, kidney biopsy and cholecystectomy. Peritoneal dialysis is delayed as long as possible to allow for healing of the incision to prevent leakage. Other recognize benefits of the laparoscopic technique include better cosmetic results, less postoperative pain and shorter hospital stay.

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