



# **PEDIATRIC SURGERY UPDATE ©**

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### **TFS**

Testicular feminization syndrome (TFS) is a genetic form of male pseudohermaphroditism (patient who is genetically 46 XY but has deficient masculinization of external genitalia) caused by complete or partial resistance of end organs to the peripheral effects of androgens. This androgenic insensitivity is caused by a mutation of the gene for androgenic receptor inherited as an X-linked recessive trait. In the complete form the external genitalia appear to be female with a rudimentary vagina, absent uterus and ovaries. The infant may present with inguinal hernias that at surgery may contain testes. Axillary/pubis hair is sparse and primary amenorrhea is present. The incomplete form may represent undervirilized infertile men. Evaluation should include: karyotype, hormonal assays, pelvic ultrasound, urethrovaginogram, gonadal biopsy and labial skin bx for androgen receptor assay. These patients will never menstruate or bear children. Malignant degeneration (germ cell tumors) of the gonads is increased (22-33%). Early gonadectomy is advised to: decrease the possible development of malignancy, avoid the latter psychological trauma to the older child, and eliminate risk of losing the pt during follow-up. Vaginal reconstruction is planned when the patient wishes to be sexually active.

### **NEC and PPD**

Complicated Necrotizing Enterocolitis (NEC) is the most common neonatal surgical emergency of modern times, has diverse etiologies, significant mortality and affects mostly premature babies. The use of primary peritoneal drain (PPD) in the management of NEC dates from 1977. The technique is used in the very low birth weight premature infant (<1500 gm) with pneumoperitoneum, metabolic and hemodynamic instability. Consists of a right lower quadrant incision and placement of a drainage (penrose or catheter) under local anesthesia with subsequent irrigation performed bedside at the NICU. Initially used as a temporizing measure before formal laparotomy, some patients went to improvement without the need for further surgery (almost one-third). They either had an immature (fetal type) healing process or a focal perforation (not associated to NEC?) which healed spontaneously. Those

babies not improved by PPD either die (20%), go on to laparotomy and half die (20%) or develop complications (24%). Some suggestion made are: PPD should be an adjunct to preop stabilization, before placing drain be sure pt has NEC by X-rays, persistent metabolic acidosis means uncontrolled peritoneal sepsis, do not place drain in pts with inflammatory mass or rapid development of intraperitoneal fluid, the longer the drainage the higher the need for laparotomy.

### **Pancreatic Pseudocysts**

Pancreatic pseudocyst formation is an uncommon complication of pancreatic inflammatory disease (pancreatitis) or trauma in children. More than half cases are caused by blunt abdominal trauma. Ultrasound is the most effective and non-invasive way of diagnosing pancreatic pseudocysts. Acute pseudocysts are managed expectantly for 4-6 wk. until spontaneous resolution occurs. 25-50% will undergo spontaneous resolution. Medical therapy consists of decreasing pancreatic stimulation and giving nutritional support. Rupture is the major complication of conservative management. Chronic pseudocysts (> three mo.) will benefit from prompt operation and internal drainage since resolution is rare. Percutaneous catheter drainage under local anesthesia using Ultrasound or CT guided technique is an appropriate method of first-line therapy for non-resolving (chronic) or enlarging pancreatic pseudocysts. The approach is transgastric or transcutaneous. Those cysts that fail to resolve with percutaneous drainage should go investigation of ductal anatomy to rule out disruption of the main pancreatic duct. The need for further surgery (drainage or resectional) will depend on the status of the duct of Wirsung.

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