



# **PEDIATRIC SURGERY UPDATE ©**

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### **Spherocytosis**

Hereditary spherocytosis is a clinically heterogeneous autosomal dominant red blood cell membrane disorder that causes anemia. The genetic defect results in deficiency of spectrin, the largest and most abundant structural protein of the erythrocyte membrane skeleton. The affected RBC loses its biconcave shape, strength and flexibility to the stress of circulation, becomes round and is trapped and destructs early in the spleen. Initial symptoms are those of pallor, jaundice and chronic anemia, followed by splenomegaly. Hemolytic crises are triggered by intercurrent infections. Pigment gallstones are common after the first decade of life. Labs' findings are those of many spherocytes in the peripheral smear, 8-10 mg% hemoglobin, elevated reticulocyte count, increase erythropoiesis in the bone marrow, and negative Coomb's test. Erythrocytes' shows increased osmotic fragility with autohemolysis in hypotonic solutions. Definitive therapy consists of splenectomy. This condition is the most common indication for elective splenectomy in children. The risk of overwhelming postsplenectomy sepsis makes it advisable to delay splenectomy until after six years of age unless the child becomes transfusion dependent. At the time of splenectomy, it is important to identify and remove accessory spleens. If gallstones are present, cholecystectomy should be done. A low content of spectrin and high percentage of microcyte has been used as determinants of early splenectomy as judge by the clinical severity of the disease process. Howell-Jolly bodies in erythrocyte are identified after total splenectomy.

### **CSP**

First described by Muller in 1838, cystosarcoma phylloides (CSP) is a rare breast neoplasm of fibroepithelial origin rarely seen in adolescent females. Phylloides tumors have been histologically classified as benign, borderline, or malignant. Classification of malignancy is based on stromal findings of cellular atypia, anaplasia, and degree of mitotic activity. Malignancy is similar to sarcomas of the breast. Presentation of CSP is that of a relative large, movable, painless mass sought to be a fibroadenoma. Lack complete encapsulation and extends into surrounding tissue in multiple projections of

different sizes. Young patients elicited a short history with rapid growth of the lesion. FNA cytology is often diagnostic of CSP. Management depends on histopathology. Benign lesions can be managed with local excision (enucleation) using a cosmetic incision. Borderline or malignant CSP should be treated with wide local excision (simple mastectomy) or re-excision to negative margins. Failure of complete excision results in local recurrence. Metastasis are bloodborn involving most frequently the lung and bones. Axillary dissection is recommended only if nodes are palpable. Adjuvant radiation is necessary, if wide local spread of the tumor is present and a resection of the lesion with a 2-cm tumorfree zone is not possible. Close followup is mandatory.

### **Vaginal Agenesis**

It is estimated that vaginal atresia (or agenesis) occurs in every one to 5-10,000 birth, the result of the Mayer-Rokitansky syndrome (partial or complete absence of the uterus with an absent or hypoplastic vagina), or an intersexual condition. In either case the vagina may be represented by a solid epithelial cord. Usually there is concomitant aplasia of the upper part of the paramesonephric ducts so that the uterus and tubes are rudimentary. The ovaries are usually normal. Associated anomalies of the urinary tract are frequent. Those cases not diagnosed in early infancy will eventually present during adolescence with primary amenorrhea and cyclic pelvic abdominal pain. US, CT, and recently MRI is extremely useful in delineating the anatomic malformation. Many ingenious procedures have been devised for creation of a neovagina in children: skin-flaps and bowel (colon) replacement vaginoplasty (see figure). They are accomplished during puberty.

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

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

Tel (787)-786-3495 Fax (787)-720-6103

E-mail: [titolugo@coqui.net](mailto:titolugo@coqui.net) Internet: <http://home.coqui.net/titolugo>

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