



PEDIATRIC SURGERY Update*

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Rapidly Involuting Congenital Hemangioma

Congenital hemangiomas are benign vascular tumors that have proliferated in utero, grown to their maximum size at birth and do not exhibit accelerated postnatal growth. They are further subdivided into: rapidly involuting congenital hemangioma, non-involuting congenital hemangiomas and partially involuting congenital hemangioma. Congenital hemangiomas are usually cutaneous but can be found in the viscera like the liver. Complications include hemorrhage, transient heart failure and transient coagulopathy. Rapidly involuting congenital hemangiomas (RICH) are large high-flow lesions that are completely formed at birth but rapidly involute by 12 to 15 months of age. RICH lesions are rare compared with other types of hemangiomas and tend to be located on the head, trunk and arms. The diagnosis of RICH is based on its clinical history of rapid involution. MRI will show high intensity flow with areas of heterogeneity. Angiography reveals an arterial aneurysm with large and irregular feeding arteries in a disorganized pattern, large dilated draining veins direct arteriovenous shunts and intravascular thrombi. Biopsy histopathology will show that the RICH lesions do not express glucose transporter-1 protein. RICH usually resolves by the age of 14 months while non-involuting congenital hemangiomas continue to grow and often need to be surgically excised. Any rapidly growing mass that is firm on palpation, show signs of ulceration and has an atypical appearance should be considered for biopsy to rule out malignancy. A singular hepatic vascular lesion in infancy is almost always a rapidly involuting congenital hemangioma. RICH lesions do not respond to propranolol or any other medication. Thrombocytopenia is typical of RICH not due to Kasabach-Merritt phenomenon but due to central thrombosis in the lesion occurring after birth with transition from fetal to postnatal portomesenteric circulation. It is self-limited within the first few weeks. Management of RICH lesions consist of observation during the involuting phase. Surgical excision or embolization is indicated for persistent ulceration, hemodynamic instability, thrombocytopenia or bleeding that does not respond to medical therapy.

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Axillary Lymphangiomas

Lymphangiomas are congenital malformations of the lymphatic system composed of single or multiple macro- or microcystic lesions with almost no communication with lymphatic system or drainage to the venous system. They are classified as capillary, cavernous or cystic. Microcystic lymphangiomas measure less than 2 mm in size, while macrocystic are more than 2 cm. The most common anatomic locations of lymphangiomas are cervicofacial (75%), followed by axilla (20%), mediastinum, groin and abdominal cavity. Lymphangiomas are painless cystic masses. Lymphangiomas usually present early in life or develop later, even in adult years. Axillary lymphangiomas present as painless swelling, which is soft, compressible, nontender and transilluminant. Differential diagnosis includes lipoma, neurofibroma, hematoma, and dermoid cyst. It is difficult to get complete excision of axillary lymphangiomas without disrupting breast tissue in females. Axillary lymphangiomas can be complicated by infection associated to a respiratory tract infection. The lesion develops redness, pain and tenderness sometimes requiring incision and drainage. They can also develop spontaneous bleeding into the cystic cavity. Ultrasound is the initial imaging performed for diagnosing axillary lymphangiomas as they are seen as anechoic cavities with septa and debris. CT or MRI are better diagnostic tools in determining the relation of vessels and nerves with the axillary lymphangioma if surgical excision is planned. Complete surgical excision is the gold standard for management of all types of lymphangiomas if the anatomic location permit. The association with vital vascular and neural structures may preclude complete excision of lymphangiomas in neck, chest, axilla and retroperitoneum. Lymphangiomas can also be managed with sclerotherapy (OK-432, Bleomycin, doxycycline, acetic acid, alcohol and hypertonic saline), especially if they are macrocystic. Propranolol downregulate the Raf mitogen activated protein kinase signaling pathway reducing expression of vascular endothelial growth factor and decreasing lymphangiomas size in 30% of patients.

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Ectopic Breast

Ectopic (accessory) breast tissue can occur anywhere along what is known as the milk line from the axilla to the groin. May occurs unilaterally or bilaterally. The incidence of ectopic breast tissue is 1-6% in the general population. Axillary breast tissue is a subtype of ectopic breast that is found on 2-5% of women. Ectopic breast tissue has also been reported on the face, perineum and vulva. The same diseases that affect the normal positioned breast tissue occur in ectopic breast tissue, namely cysts, benign tumors such as fibroadenomas, inflammatory (mastitis), and neoplastic conditions (phyllodes and carcinoma). Breast tissue normally develops from the embryonic ectodermal thickening extending from the axilla to the groin region. It develops in the pectoral region and the rest of the milk line undergoing regression. Failure of such regression give rise to supernumerary breast (polythelia or polymastia). Ectopic breast tissue increases in size during puberty, pregnancy, birth and lactation due to hormonal stimulation. Native American women have a higher incidence of accessory breast compared with non-native American. The differential diagnosis in the axillary region would include lymphadenopathy, epidermal cyst or lipoma. Malignant transformation can occur in ectopic breast tissue mentioned above. The possibility of future complications in ectopic breast and axillary breast tissue along with the aesthetic and psychological involvement are the main reason for offering surgical excision of the accessory gland to affected patients. Axillary tail of Spence is axillary accessory breast tissue that is connected with outer part of normal thoracic breast tissue and is located deep. Diagnosis can be suggested using ultrasound and confirmed with fine-needle aspiration biopsy. Conservatory management (observation) is an option, though not commonly used among affected women.

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

Professor of Pediatric Surgery, University of Puerto Rico - School of Medicine,
Rio Piedras, Puerto Rico. Director - Pediatric Surgery, San Jorge Children's & Woman
Hospital.

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

Tel (787)-340-1868 E-mail: titolugo@coqui.net

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

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