



## **Congenital Pulmonary Airway Malformations**

Congenital pulmonary airway malformations (CPAM) refer to an unusual lesion of the pulmonary airways which combines features of hamartoma malformation and dysplastic proliferation. CPAM includes cystic pulmonary airway malformations, bronchopulmonary sequestration, bronchogenic cysts, hybrid lesions and lobar/segmental emphysema causing respiratory distress in 20-40% of affected babies in the postnatal period. The remaining cases continue asymptomatic or develop symptoms later in life such as chest infections. Most CPAM can be detected on the 20-week antenatal ultrasound increasing the diagnostic yield if MRI is utilized. Children with symptoms early in life are managed with surgery. The management of asymptomatic CPAM is a source of controversy in the literature. CPAM is classified 0 to IV. Type 0 is very rare described as acinar aplasia or agenesis and incompatible with life. Type I the most common is primarily macrocystic with large single or multiple cysts several centimeters in size. Type II is microcystic and associated with other anomalies. Type III appears more solid or with very small cysts similar to immature lungs without bronchi. Type IV originates from the acinus and present with small cysts on the periphery of the lung lobes. Once a cystic lesion is detected in antenatal ultrasound, the location, volume, size, macrocystic or microcystic classification and blood supply should be evaluated. CPAM volume to head circumference ratio (CVR) greater than 1.6 results in fetal demise in about 80% of cases without fetal intervention. CVR < 1.6 will often not continue to grow past the 28th week of gestation. The reasons used to remove asymptomatic lesions in the first year of life include the rate of empyema, abscess, recurrent pneumonia, air leak, pneumothorax and malignancy. Almost 25% of asymptomatic children show histologic evidence of infection. CPAM have a long-term risk of malignancy. Multiple courses of antenatal betamethasone for high-risk fetal CPAM often results in favorable short-term outcomes without the need for open fetal resection.

### **References:**

- 1- Baird R, Puligandla PS, Laberge JM: Congenital lung malformations: informing best practice. *Semin Pediatr Surg.* 23(5):270-7, 2014
- 2- Derderian SC, Coleman AM, Jeanty C, Lim FY, Shaaban AM, Farrell JA, Hirose S, MacKenzie TC, Lee H: Favorable outcomes in high-risk congenital pulmonary airway malformations treated with multiple courses of maternal betamethasone. *J Pediatr Surg.* 50(4):515-8, 2015
- 3- Gajewska-Knapik K, Impey L: Congenital lung lesions: Prenatal diagnosis and intervention. *Semin Pediatr Surg.* 24(4):156-9, 2015
- 4- Singh R, Davenport M: The argument for operative approach to asymptomatic lung lesions. *Semin Pediatr Surg.* 24(4):187-95, 2015
- 5- Macardle CA, Ehrenberg-Buchner S, Smith EA, Dillman JR, Mychaliska GB, Treadwell MC, Kunisaki SM: Surveillance of fetal lung lesions using the congenital pulmonary airway malformation volume ratio: Natural history and outcomes. *Prenat Diagn.* 2015 Dec 29

6- Durell J, Thakkar H, Gould S, Fowler D, Lakhoo K: Pathology of asymptomatic, prenatally diagnosed cystic lung malformations. *J Pediatr Surg.* 51(2):231-5, 2016

## **Cutaneous Schwannomas**

Schwannoma is a benign, expansile tumor that originates from any nerve sheath in the body that contains Schwann cell. Schwann cells insulate normal nerve fibers and enhance propagation of nerve impulses. Schwannomas can occur anywhere in the body along the course of a nerve such as cranial nerves, spinal nerves or peripheral nerves. As such most schwannomas appear intracranially, intraspinal or lying deep within soft-tissue. The most common schwannoma is the acoustic neuroma along the VIII cranial nerve. Cutaneous schwannomas present as a solitary deep-seated nodule in the deep dermis or subcutaneous tissue. Most cutaneous schwannomas are asymptomatic. Other times they may cause pain, tenderness or paresthesia depending on size, site and nerve involved. Males and females are equally affected. Histologically, schwannomas are encapsulated by perineurium and contain either cellular areas characterized by uniform spindle cell clustered in stack and arranged back to back, or a loose myxoid matrix. Immunohistochemistry of schwannomas reveals positive S100 and collagen type 4, and capsule is positive for epithelial membrane antigen. Schwannomas enlarge slowly and follow a benign course with very rare cases of malignant transformation. The preoperative diagnosis of a cutaneous schwannoma is seldom done as the nodule is usually confused with an epidermal, trichilemmal or dermoid cyst. Among the other types of schwannomas, plexiform variety constitute 5% of the presentation associated with neurofibromatosis. Few reports of malignant transformation in schwannomas have been reported in the literature. Management of schwannomas is surgical excision which is usually curative. Local recurrence can occur and long-term follow up is needed.

### **References:**

- 1- Agaram NP, Prakash S, Antonescu CR: Deep-seated plexiform schwannoma: a pathologic study of 16 cases and comparative analysis with the superficial variety. *Am J Surg Pathol.* 29(8):1042-8, 2005
- 2- Attia EA, Yassin M, Lasheen MA, Salem SA, Khafagy NH: Multiple isolated cutaneous plexiform schwannomas. *Indian J Dermatol Venereol Leprol.* 77(5):594-6, 2011
- 3- Elwood H, Taube J: Dermal and Subcutaneous Plexiform Soft Tissue Neoplasms. *Surg Pathol Clin.* 4(3):819-42, 2011
- 4- Noh S, Do JE, Park JM, Jee H, Oh SH: Cutaneous schwannoma presented as a pedunculated protruding mass. *Ann Dermatol.* 23(Suppl 2):S264-6, 2011
- 5- Mohan Kh, Manjunath H: Cutaneous schwannoma masquerading as trichilemmal cyst over scalp in a young male. *Indian J Dermatol.* 58(5):407, 2013
- 6- Kacerovska D, Michal M, Kazakov DV: Hybrid Epithelioid Schwannoma/Perineurioma. *Am J Dermatopathol.* 2016 Feb 9.

## **Enteral Refeeding**

Many neonates undergo intestinal resections for a variety of conditions losing a significant segment of bowel including the construction of proximal enterotomies with distal mucous fistulas. They rely on parenteral nutrition for a significant period of time

after surgery. Parenteral nutrition is associated with a significant risk of catheter-related blood stream infection, thrombosis, and neonatal cholestasis. The absence of food in the GI tract produces mucosal and villous atrophy with reduction of enzymes necessary for digestion and substrate absorption. Promoting early enteral nutrition is beneficial even in the face of a proximal enterostomy. The presence of a proximal enterostomy can produce high stomal losses with associated fluid and electrolytes imbalances, metabolic acidosis and impaired absorption of nutrition. Refeeding of stomal losses into the distal mucous fistula has been used to minimize fluid and electrolytes losses as well as dependence on parenteral nutrition. Mucous fistula refeeding (MFR) stimulates mucosal growth and intestinal adaptation preventing atrophy of the distal gut. Enteral refeeding requires substantial nursing expertise, time and commitment to the process. MFR has also been associated with distal perforation with bacterial overgrowth in the stomal output if there is a delay between collection and refeeding of the stoma effluent. Fluoroscopy can be used to insert the distal refeeding tube to be utilized taking care to determine if there is thin intestinal wall prone to perforation or distal strictures in the case of NEC babies. Neonates with enteral refeeding have a better gain in body weight than those without it, regardless of the gestational age. Only the MFR procedure and birth weight were significant independent predictors of a good weight gain. Babies using MFR have a high rate of central venous catheter removal because of nutritional improvement. The longer the distal bowel to be fed the better the rate of body weight gain.

**References:**

- 1- Hay WW Jr: Strategies for feeding the preterm infant. *Neonatology*. 94(4):245-54, 2008
- 2- Morgan J, Bombell S, McGuire W: Early trophic feeding versus enteral fasting for very preterm or very low birth weight infants. *Cochrane Database Syst Rev*. Mar 28;3:CD000504, 2013
- 3- Haddock CA, Stanger JD, Albersheim SG, Casey LM, Butterworth SA: Mucous fistula refeeding in neonates with enterostomies. *J Pediatr Surg*. 50(5):779-82, 2015
- 4- Wong KK, Lan LC, Lin SC, Chan AW, Tam PK: Mucous fistula refeeding in premature neonates with enterostomies. *J Pediatr Gastroenterol Nutr*. 39(1):43-5, 2004
- 5- Al-Harbi K, Walton JM, Gardner V, Chessell L, Fitzgerald PG: Mucous fistula refeeding in neonates with short bowel syndrome. *J Pediatr Surg*. 34(7):1100-3, 1999
- 6- Koike Y, Uchida K, Nagano Y, Matsushita K, Otake K, Inoue M, Kusunoki M: Enteral refeeding is useful for promoting growth in neonates with enterostomy before stoma closure. *J Pediatr Surg*. 51(3):390-4, 2016

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