



PEDIATRIC SURGERY Update © **Vol. 38 No. 02 FEBRUARY 2012**

Middle Aortic Syndrome

Middle aortic syndrome (MAS) is a rare condition involving diffuse narrowing of the distal thoracic or abdominal aorta involving its visceral and renal branches. The etiology of MAS is unknown and occurs in young patients. MAS have been associated with neurofibromatosis, Alagille's and William syndrome. The mesenteric vascular stenosis is clinically silent, while the renal stenosis is responsible for the clinical picture of hypertension in these children. Absent femoral pulses and an abdominal bruit are also present. Bilateral involvement is common. Ultrasonography is the primary screening technique. Biplanar arteriography is the standard imaging to establish the diagnosis and involvement of the disease. Arteriography shows a smooth segmental narrowing of the abdominal aorta with concomitant stenosis at the origins of the renal arteries. Primary treatment goals are preservation of functional renal tissue and amelioration of the renal hypertension. Therapeutic approaches included medical management, percutaneous transluminal angioplasty and/or surgical intervention. Repair of the narrowing requires prosthetic material for bypass or patch reconstruction. Techniques used to revascularize the kidney included thoracoabdominal to infrarenal aortic bypass with renal artery reimplantation, splenorenal bypass, gastroduodenal to renal bypass, aortorenal bypass and autotransplantation.

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Ciliated Hepatic Foregut Cyst

Ciliated Hepatic Foregut Cyst (CHFC) is a rare cystic benign disease of the liver increasingly diagnosed. Most patients with CHFC are asymptomatic at the time of diagnosis. The most common symptom is right upper quadrant pain. In almost all cases

the cyst is located within the left liver segment IV or in close proximity. Average size of the cyst is 3.6 cm with most cyst unilocular, avascular and lying beneath the anterior surface of the liver. Cyst content is viscous or mucinous. It is the presence of shared histological features in the form of pseudostratified ciliated columnar epithelium that leads to label these cysts of foregut origin. The origin of CHFC is detachment and migration of foregut with subsequent entrapment in the liver. Infantile presentation is in the form of an abdominal mass due to the larger size. In infants the cyst may have bile duct communication and can develop squamous metaplasia, reason why excision and ligation of the biliary communication should be preferred to internal drainage. CT will show a complex hypodense cystic mass without enhancement. FNA cytology will show the ciliated pseudostratified columnar cells and establish the diagnosis. The presence of a tough fibrous outer layer in the cyst makes it amenable to mobilization and enucleation without spillage. This can be accomplished by laparoscopy or open surgery.

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ALTE

An apparent life-threatening event (ALTE) is defined as an episode that is frightening to the caretaker and is characterized by some combination of apnea (which can be central or occasionally obstructive), color change (usually cyanotic or pallid but occasionally erythematous or plethoric), marked change in muscle tone (usually limpness), choking or gagging. Although the natural history of ALTE is most often benign, there is a risk for subsequent morbidity and mortality. The most common causes of ALTE are gastroesophageal reflux, seizure activity and lower respiratory tract infections, specially respiratory syncytial virus infection. Other causes include otolaryngology conditions, inborn errors in metabolism and cardiac diseases. The incidence of sudden infant death syndrome is reported to be higher after ALTE episodes. Two mechanisms explain why reflux can result in ALTE: chemoreflex in the larynx prompted by acid fluid and stimulation of the distal esophagus by the gastric content. Children with ALTE and gastroesophageal reflux, other cardiac, respiratory and neurological causes appropriately excluded should be considered candidates for fundoplication since medical

management carries a high risk of recurrent ALTE. After fundoplication the incidence of ALTE is significantly reduced.

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