



PEDIATRIC SURGERY Update*

Vol. 54 No. 04 APRIL 2020

Thoracic Duct Obstruction

Congenital anomalies of the thoracic duct is very rare, poorly characterized and difficult to manage. Lymphatics from the lower extremity and trunk join those of the bowel mesentery in the cisterna chyli located in the lumbar prevertebral plane posterior to the aorta and inferior vena cava. The thoracic duct forms from here and ascends in the thoracic prevertebral plane emptying into the central venous system near the junction of the left internal jugular vein and subclavian vein. An accessory thoracic duct occurs on the right side also. Thoracic duct obstruction, slow flow, reflux or leak of lymph or chyle into the pleural, peritoneal and pericardial space cavity causing respiratory compromise, chylothorax, chylous ascites, malnutrition, hypoproteinemia, lymphopenia, immunosuppression and bony erosion. Chyle in any other body cavity or tissue implies leakage from the central lymphatic channels. Chronic management with medications (octreotide, sirolimus), diuretics, pleurocenteses, peritoneocenteses, shunting procedures, sclerotherapy, embolization, radiation therapy and TPN can provide short-term palliation. Anatomic delineation of the problem using lymphangiography is essential and the best technique. Dynamic contrast magnetic resonance lymphangiography can also help delineate the obstructing problem. Peristaltic activity and negative intrathoracic pressure cause lymph to move in the thoracic duct. Lymphatic embolization and lymphovenous anastomotic techniques are relatively new techniques of management of thoracic duct obstruction with limited long-term results. Thoracic duct bypass (lymphovenous) procedures in the neck appear to be a safe treatment option for children with central conducting lymphatic obstruction whose lymphatic imaging indicates possible dysfunction or blockage of the thoracic duct at its terminus draining point into the central system. Thoracic duct to vein anastomosis in the neck can only be performed if the child has a central obstructed thoracic duct present. Utilization of high-power microscope (10x magnification) for the anastomosis is mandatory. Children with high central venous pressure do not respond to the anastomosis. Lymphovenous anastomosis can restore normal lymphatic circulation within two weeks, liberate patients from mechanical ventilation, and enable expeditious return to enteral feeding.

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Stridor in Children

Stridor refers to a variable, high-pitched respiratory sound that can occur during breathing. The sound is produced by abnormal flow of air in the upper airways most prominently heard during inspiration, though it can be present during both inspiration and expiration. The resulting turbulent airflow causes abnormal vibrations of the surrounding tissues causing the characteristic noise. Stridor occurs due to congenital malformations and anomalies as well as in an acute life-threatening obstruction or infection of the airway. In infants and young children, a small closure of the windpipe due to inflammation or infection can result in rapid airway obstruction. In children the most common cause of a stridor is acquired, such as croup, foreign body aspiration, airways burn, bacterial tracheitis, epiglottitis, peritonsillar abscess and vocal cord dysfunction. Congenital causes of stridor include choanal atresia, septum deformities, vestibular atresia, macroglossia, laryngomalacia, webs, clefts, tracheomalacia, tracheal stenosis and subglottic stenosis. The subglottic region is the narrowest portion of the infant airway and normally contains the only complete cartilaginous ring (cricoid cartilage). Stridor secondary to tracheal narrowing is typically expiratory because increased intrathoracic pressure with expiration reduces the tracheal diameter. Stridor is more common in children than adults. Inspiratory stridor is caused by an obstruction in the extrathoracic region, while an expiratory stridor is caused by an airway obstruction in the intrathoracic region. Diagnostic workup includes simple chest films, neck and chest CT or MRI, and laryngotracheobronchoscopy. Laryngo-bronchoscopy visualizing the airways establish the diagnosis. Should the child appear critically ill, then endotracheal intubation should be performed. Specific management of the stridor depends on the underlying etiology. In general, with a child with stridor avoid agitating him further, monitor for rapid respiratory deterioration, secure the airway and oxygenation, examine in controlled environment such as operating room and avoid beta agonist therapy in children with croup as it worsens airway obstruction.

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Biliary Cysts after Kasai Procedure

Biliary atresia (BA) is an inflammatory, progressive cholangiopathy affecting the intra- and extrahepatic bile duct system typical of the neonatal period and manifesting with cholestatic jaundice, acholia and hepatomegaly. Without management biliary atresia progress to hepatic cirrhosis and portal hypertension in need of a liver transplant. The Kasai procedure, namely an hepaticoportenterostomy, is the initial procedure of choice for infants with biliary atresia. It is a definitive procedure for 20-40% of children with biliary atresia and a bridge to transplant to the rest. Biliary atresia is the leading cause of liver transplant in children in the world. Bile lakes also known as parenchymal cystic dilatations of the intrahepatic bile ducts have been described in up to one-third of the patients after the Kasai procedure. They have been described histologically as fibrocystic wall damage bile ducts associated with invasion by inflammatory cells. Bile lakes are almost always preceded to an episode of postoperative cholangitis after the Kasai procedure. Clinical symptoms include fever, jaundice, leukocytosis and acholic stool. Diagnosis is established with ultrasonography. Intrahepatic biliary cysts are divided into two types: solitary simple cystic lesions or multiple continuous oval or beaded lesions. In both categories of lesions prognosis has been established as poor. Intrahepatic biliary cysts without cholangitis are not a source of infection and require no treatment. More than the presence of the cyst is the associated cholangitis and onset of cyst development which is associated with a poor prognosis. Biliary cysts seen with cholangitis and occurring early after the Kasai procedure (less than six months) do carry a poor prognosis. Percutaneous transhepatic cholangiodrainage can help drain the solitary cyst while children with cholangitis and multiple cysts should be managed with intravenous antibiotics. Patients with multiple cysts and intractable cholangitis can develop hepatic deterioration in need of liver transplantation.

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* *PSU 1993-2020*
ISSN 1089-7739