Congenital chylothorax

Chylothorax is an accumulation of lymph (sap) in the chest cavity. Chylothorax is the most common cause of pleural effusion in newborns. This is a relatively rare, but in more severe cases, a very serious diagnosis (if hydrops, is present at the same time, the mortality rate is up to 98 %)[1].

Chylothorax can develop spontaneously or together with lymphedema in congenital abnormality of lymphatic vessels, such as in Turner's or Noonan's syndrome or in congenital lymphangiectasia (diffuse dilatation of interlobular and subpleural lymphatic vessels). In most cases, the cause of chylothorax is unknown.[2]

The most serious complications of fetal chylothorax include pulmonary hypoplasia, congestive heart failure and hydrops.

Clinical picture

In about 50% of cases, chylothorax manifests itself in the first week of life as an isolated pleural effusion, typically right-sided. Chronic chylothorax can be accompanied by hypovolemia, hypoalbuminemia, hyponatremia and weight loss. These children are immunocompromised due to loss of lymphocytes and antibodies. Rarely, chylothorax can lead to the development of fetal hydrops, due to impaired venous return due to compression of the vena cava and heart and/or due to loss of proteins into the pleural cavity.

Chylothorax with superior vena cava obstruction will cause swelling of the face, neck, and upper extremities.[2]

Diagnostics

Fluid aspirated during thoracentesis (chest puncture) is sent for cytological (determines the number of lymphocytes), biochemical and microbiological examination.

In an unfed newborn, this fluid is clear, yellowish, contains a large number of lymphocytes, a large amount of triglycerides and little cholesterol. After starting milk feeding, the fluid is chylous, and after switching to infant formula with MCT oils, it changes to clear.

Fluid in the pleural cavity can be confirmed by sonography or X-ray.[2]

Treatment

Fetal chylothorax may require prenatal drainage (thoracocentesis or insertion of a thoracoamniotic shunt for continuous drainage). An indication for prenatal drainage is the development of hydrops and displacement of the mediastinum during unilateral effusion.

Treatment after birth depends on the severity/extent of the finding. Sometimes, a one-time chest drainage is sufficient to allow expansion of the lungs and thus tamponade of the defect, which prevents further formation of pleural effusion. If the chylothorax reaccumulates, chest drainage is performed and the child is switched to a special milk formula with lipids only in the form of MCTs (medium chain triglycerides). MCT fatty acids pass from the intestine straight into the vena portae, while long-chain fatty acids pass in the form of chylomicrons into the lymph and from there into the venous system.

Neonates with extensive pleural effusions require active resuscitation with intubation and mechanical ventilation.[2]

Links

Related Articles

- Pleural effusion • Pleural effusion (internal propaedutics) • Chylothorax
- Pneumotorax (neonatology)

External links

- T. Jimramovský: Chylothorax u novorozence (https://www.pediatriepropraxi.cz/pdfs/ped/2010/06/11.pdf)

Reference

1. Al-Tawil K, Ahmed G, Al-Hathal M, Al-Jarallah Y, Campbell N. Congenital chylothorax. Am J Perinatol. 2000;17(3):121–126.
2. RENNIE, JM. *Textbook of Neonatology.* 5. edition. Churchill Livingstone Elsevier, 2012. pp. 515. ISBN 978-0-7020-3479-4.