Congenital diaphragmatic hernia

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It accounts for 80% of all congenital lung anomalies. Prevalence is 1:2000–4000, mortality 25.9%. In 95% it is left-sided (*Bochdalek's hernia* - lumbocostal left)[1]. In 5% it is right-sided (*Morgagni's hernia* - sternocostal right)[1]. In most cases, it is not covered by a hernia sac (false hernia). The defect in the diaphragm may be small or the entire diaphragm may be missing. The contents of the hernia can be part of the small and large intestine, stomach, sometimes liver, spleen or kidney.

Clinical picture

The main problems are **pulmonary parenchymal insufficiency** (pulmonary hypoplasia; abnormally small functional lung area) and **pulmonary hypertension**.[2]

Immediately after birth, a severe **respiratory distress syndrome** develops, accompanied by dyspnea, cyanosis and tachycardia. Dyspnea subsides in elevated chest position. There is a movement of the heart to the healthy side (the location of the echo can change - migration of echoes - the so-called "Peter's sign"). Breathing tends to be weak and there is a tympanic tap on the chest on the side of the hernia. We can hear twisting in the chest. The abdomen is sunken. The first critical 72 hours after birth are important for survival. After this period, it manifests itself in chronic respiratory and GIT difficulties.

Diagnosis

It states **trias: dyspnea, cyanosis, dextrocardia**. A native X-ray confirms the diagnosis – there is no air filling of the intestines in the abdomen, the mediastinum is displaced, in the left part of the chest there are circular clearings resembling loops of intestines (signet ring shape). It is possible to diagnose prenatally with ultrasound, which shows polyhydramnios.

Therapy

- Before surgery – application of oxygen, intubation, low-pressure ventilation, insertion of a gastric tube to suction the contents of the stomach, position on the affected side, elevation of the upper body.
- Inhalation of surfactant - lungs are immature.
- To reduce pulmonary hypertension - inhalation of NO (causes vasodilation).
- Surgical reposition of organs into the abdominal cavity and covering of the diaphragm defect is an urgent and life-saving procedure.

⚠ **Mask ventilation is contraindicated - it fills the stomach with air and thus worsens lung compression even more.**[1]

Prognosis

Prenatal ultrasound examination makes it possible to estimate the prognosis using the so-called o/e LHR (*observed/expected Lung area to Head circumference Ratio*), i.e. by comparing the circumference of the fetal head and the size of the lung on the opposite side to the congenital diaphragmatic hernia and correction for gestational age ("o/e"). The prognosis is also determined by the location of the fetal liver (intrathoracic vs. intra-abdominal), where the location of the liver in the chest worsens the prognosis. Severity of left-sided diaphragmatic hernia according to o/e LHR parameters – extremely severe: < 15%; severe: < 25%; moderate: 25-34.9% or 35-44.9% with liver in chest; mild: 35 – 44.9% with liver in abdominal cavity or o/e LHR > 45%.[3]

Links

Related articles

- Congenital developmental defects of the newborn requiring an urgent solution
- Congenital pneumopatias

External links

- website about congenital diaphragmatic hernia (http://www.branicni-kyla.cz/)

References

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Source

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