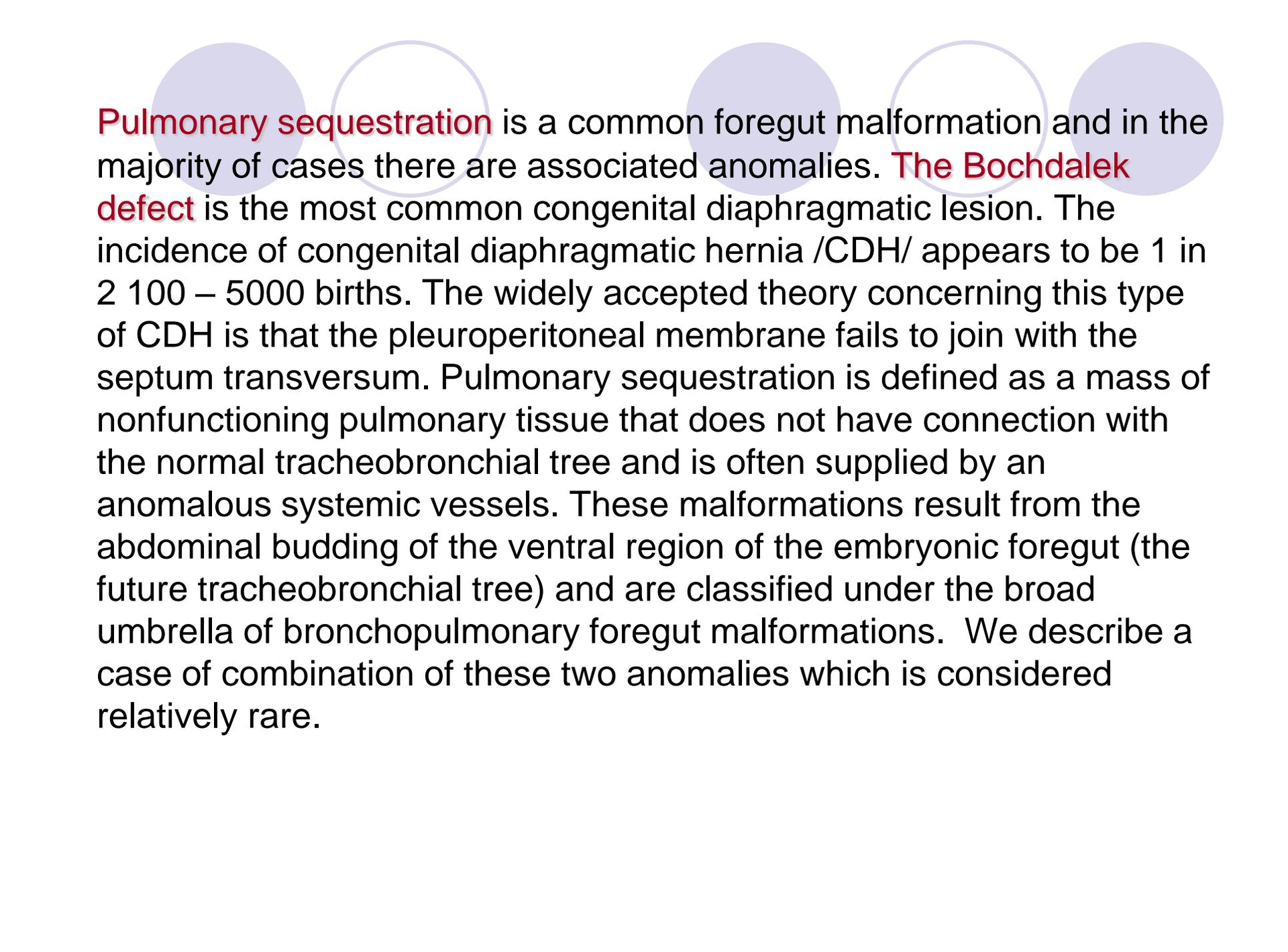


INTRAABDOMINAL EXTRALOBAR PULMONARY SEQUESTRATION MIGRATING THROUGH A BOCHDALEK DIAPHRAGMATIC HERNIA

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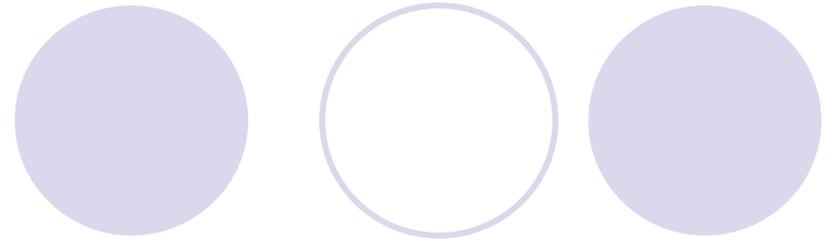
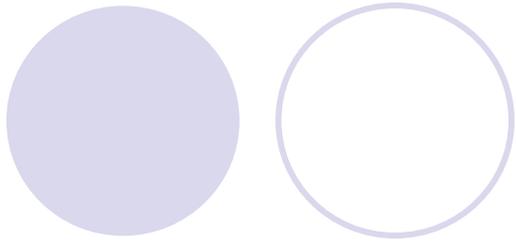
Pulmonary sequestration is a common foregut malformation and in the majority of cases there are associated anomalies. **The Bochdalek defect** is the most common congenital diaphragmatic lesion. The incidence of congenital diaphragmatic hernia /CDH/ appears to be 1 in 2 100 – 5000 births. The widely accepted theory concerning this type of CDH is that the pleuroperitoneal membrane fails to join with the septum transversum. Pulmonary sequestration is defined as a mass of nonfunctioning pulmonary tissue that does not have connection with the normal tracheobronchial tree and is often supplied by an anomalous systemic vessels. These malformations result from the abdominal budding of the ventral region of the embryonic foregut (the future tracheobronchial tree) and are classified under the broad umbrella of bronchopulmonary foregut malformations. We describe a case of combination of these two anomalies which is considered relatively rare.

Case report:

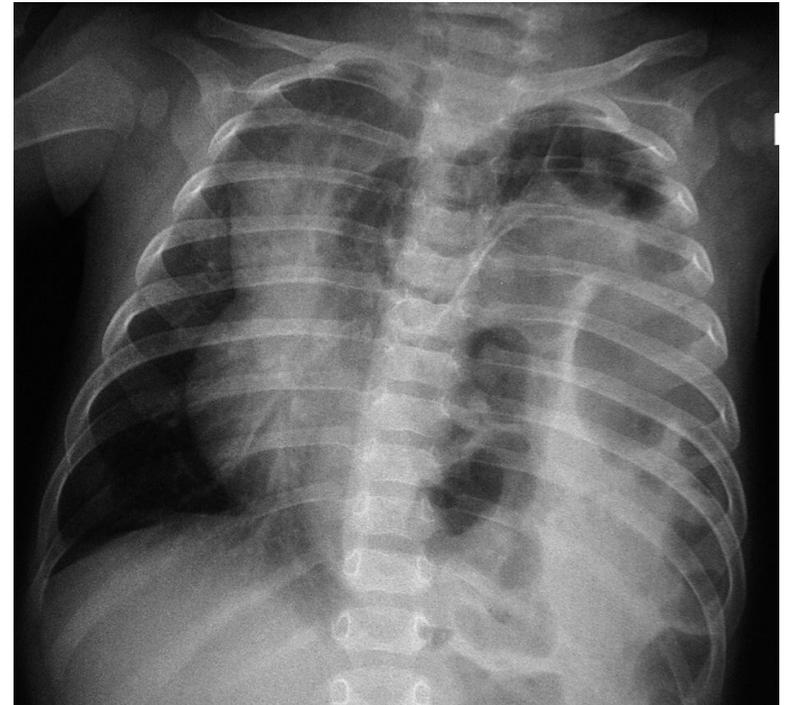
Our patient was a full-term infant, born after an uneventful pregnancy. At birth there were no signs of respiratory distress or pulmonary malformations. At the age of 7 months in the course of respiratory tract infection (right-sided pneumonia) the patient developed respiratory insufficiency and an X-ray was performed. It showed left-sided diaphragmatic hernia.

Physical findings:

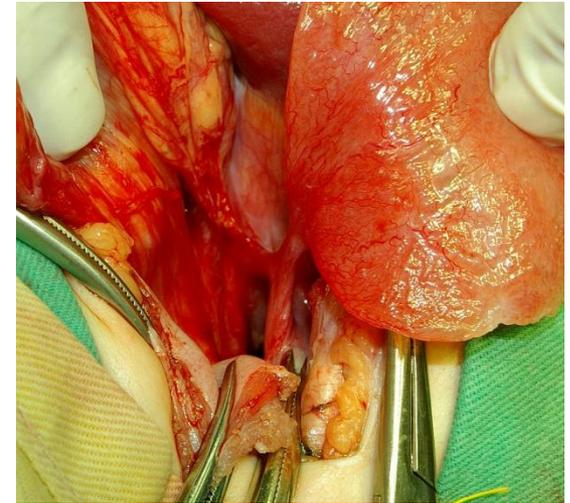
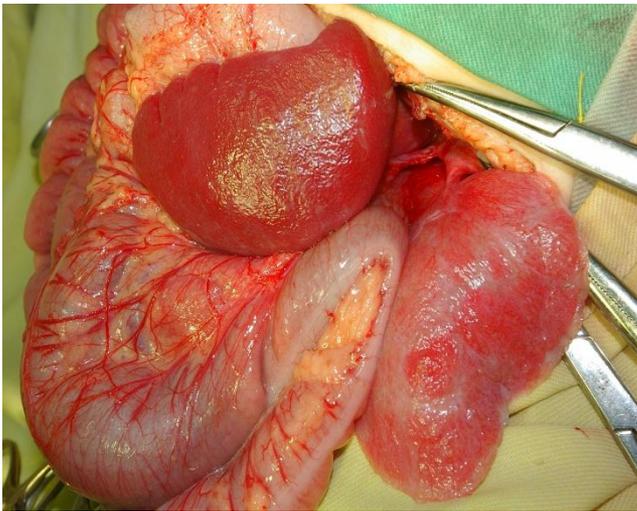
- Acute respiratory distress
- High temperature
- Tachypnea
- Absent breathing sounds to the left
- Rhonchi to the right
- Heart sounds to the right
- Non-distended abdomen
- Absent peristalsis
- Non-palpable liver.



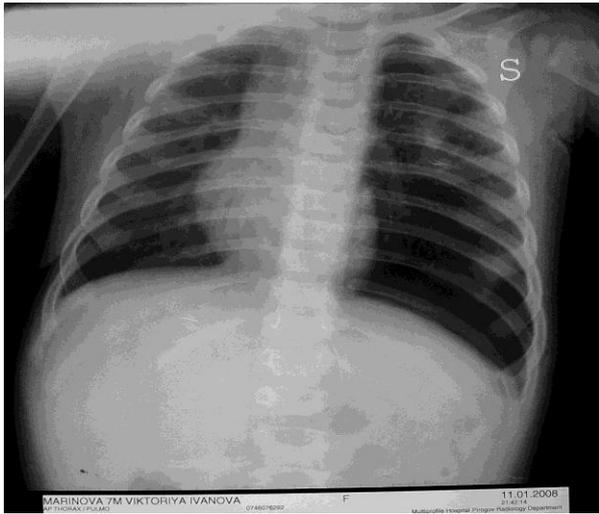
- Lab study – mild hypoproteinemia
- Plain radiograph – absence of lung marking and diaphragmatic dome to the left, presence of intestinal loops, shifting of the mediastinal structures to the right
- Echocardiography – structurally normal heart and large blood vessels



Surgery was performed on the second day after admission when the child was stable. We used abdominal approach. There was a large defect in the left diaphragm, spleen, small and large intestines were dislocated to the left hemithorax. After reposition of the abdominal organs in the abdominal cavity, a pink-colored mass appeared, hanging on an anomalous blood vessel arising from the celiac trunk. The blood vessel was ligated and the sequestered tissue removed



The left lung was hypoplastic but functional. Suture of the diaphragmatic defect without a patch. Prophylactic appendectomy. Histological findings were pulmonary tissue, covered with pleura, dilated bronchioles and alveoli and dilated subpleural lymph vessels which is consistent with extralobar pulmonary sequestration.



In the postoperative period a left-sided pneumothorax developed. Eight days after operation the patient was discharged. The X-ray showed well expanded left lung

The presence of sequestered pulmonary tissue in CDH possibly interferes mechanically with normal closure of the pleuroperitoneal channel and diaphragmatic muscularization and may result in a posterolateral and anterolateral defects of the diaphragm. The combined associations indicate an embryonic defect occurring at the same time early in gestation before closure of the diaphragm (6 weeks) or earlier. Delayed clinical presentation of associated malformations has different clinical manifestation from the more common newborn entity and these cases should be evaluated carefully.

In our case intraoperatively we clearly defined the intrathoracic EPS and abdominal blood supply of the EPS through the Bochdalek defect. The mobility of the EPS and the location of the vascular pedicle suggested that the EPS first developed intraabdominally. Later, when the bowels shifted up in the chest they compressed and dislocated the EPS into the thoracic cavity. Our case illustrates the diagnostic and surgical challenges in cases of association of EPS and CDH.

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