

Left Bochdalek Hernia with Delayed Presentation: Report of Two Cases

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Left Bochdalek hernia is a serious and complex condition with high mortality. In most cases, it presents in the neonatal period and is seldom found later in life when symptomatology, usually after an asymptomatic period, is quite different and the prognosis excellent. The embryological development of left Bochdalek hernia suggests the presence of abdominal content in the left pleural cavity before birth. The type of clinical presentation and the prognosis depend on the time of visceral herniation. This study presents two cases of left Bochdalek hernia with delayed presentation. In both cases, after surgical reposition of the hernia, a small congenital diaphragmatic defect was found hidden between the rims of diaphragm. This indicates the possibility for the abdominal content to enter the left pleural cavity at the time of presentation.

Key words: diagnosis, differential; hernia, diaphragmatic; infant, newborn, diseases

Congenital diaphragmatic defect develops in the first 9 weeks of gestation. In this period, pulmonary, diaphragmatic, and intestinal developments are synchronous. The gut is rotated and pulled into the abdominal coelom, and pleuroperitoneal membrane closes (1). An unknown cause leads to premature return of the gut and/or late closure of the pleuroperitoneal membrane. When the abdominal organs enter the pleural cavity, the development of lung is disturbed. The degree of pulmonary hypoplasia depends on the stage of pulmonary development at the time of the accident (2,3). Earlier visceral herniation into the pleural cavity worsens pulmonary hypoplasia and its prognosis (4,5). When it happens later in gestation, there is no pulmonary hypoplasia and the symptomatology of the primary pulmonary hypertension predominates (6,7). If the clinical picture does not present within 8 hours after the birth, the symptoms that appear later are probably caused by simple compression of the lung (8,9). The prognosis is generally good. There is neither pulmonary hypoplasia nor primary pulmonary hypertension, because the abdominal organs enter the pleural cavity too late for the development of pulmonary hypertension. Visceral herniation through a congenital diaphragmatic defect can happen at any time, and that determinates the type of clinical presentation and prognosis.

In left Bochdalek hernia with delayed presentation respiratory symptoms are caused only by mechanical compression of the well-developed lung. After an asymptomatic period of various duration, only milder respiratory (cough and chest pain) or cardiac (murmur and extrasystoles) problems develop and gastrointestinal symptoms predominate (vomiting and abdominal pain) (10). Functioning of the lung at the side of hernia depends on the contralateral lung

only during the clinical presentation of Bochdalek hernia (11). The prognosis is excellent and mostly depends on the time of diagnosis (12).

In a 12-year period (1987-1998), two patients were treated for the left Bochdalek hernia with delayed presentation at the Split University Hospital. We think that there was no herniation during the asymptomatic period, probably due to a certain blocking mechanism that prevented the herniation, and that the symptoms appeared when abdominal organs entered the thorax through a persistent congenital diaphragmatic defect.

Case 1

A 4-month-old male infant born after a normal gestation with Apgar score 10 suddenly became febrile, started vomiting, and had frequent diarrheal stools. In the first three months of life, he thrived well. Gastroenterocolitis was suspected, but during the examination we found decreased breathing on the left side. A whole body radiogram ("babygram") was performed and showed air-fluid levels in the middle and lower parts of left hemithorax and empty abdomen. Left phrenicocostal sinus was obliterated and elevated the left diaphragmatic arch (Fig. 1). The finding of the bowel situated in the left pleural cavity and the absence of trauma history were pathognomonic for the diagnosis of congenital diaphragmatic hernia. At laparotomy we found left Bochdalek hernia with perisplenic abscess due to strangulated lienal artery and splenic necrosis. The stomach, small bowel, and parts of malrotated large bowel were herniated into the pleural cavity. After the reduction of the hernial content, a small congenital diaphragmatic defect was found between the flaps of the diaphragm and closed by simple sutures. The rest of the spleen was removed.



Figure 1. Left Bochdalek hernia. Deformation of left diaphragmatic arch contour by the mass of abdominal content (arrow).

The treatment of the abscess continued with antibiotic therapy (penicillin, gentamycin, and clindamycin) and the infant recovered. Benzathine benzyl-penicillin was administered for the prophylaxis of pneumococcal sepsis. No respiratory problems were noted. In a 5-year follow-up, the child had rare attacks of colic pain with abdominal distension.

Case 2

A 4-year-old male child was admitted to the hospital with acute abdominal colic pain. Before that, he had never been seriously ill, except for a minor respiratory infections. The patient was observed to exclude the acute abdomen. He complained about some discomfort on the left side of the chest. We found bowel sounds over the left hemithorax and made the final diagnosis by chest X-ray and gastrointestinal contrast study. The patient was in a good condition. At laparotomy we found Bochdalek hernia that contained stomach, small bowel, and spleen. There were adhesions around a small diaphragmatic defect. Intestinal malrotation was not found in the abdomen. We pulled the abdominal content back to the peritoneal cavity and simply closed the diaphragmatic defect with sutures. The patient recovered and today, four years later, he lives without any medical limitations.

Discussion

Both cases share some important, common characteristics. The asymptomatic period in both patients finished with acute attack of the disease. The whole left pleural cavity was filled with the abdominal content, but the symptomatology was dominated by gastrointestinal symptoms. The diaphragmatic defect was small and easy to close.

Bochdalek hernia is congenital diaphragmatic prolapse and usually, when left-sided, presents with a

clinical picture that demands urgent medical attention. The serious respiratory distress is related to pulmonary hypoplasia, often bilateral, combined with persistent fetal circulation and mechanical respiratory disorders (13). Overall survival of about 60% makes left Bochdalek hernia a serious neonatal problem associated with high mortality (14).

In 5-10% of patients, the presentation of Bochdalek hernia is delayed, whereas 7-10% of patients have no symptoms (10,15). Most asymptomatic Bochdalek hernias are right-sided with diaphragmatic defect covered by the liver. Left-sided hernias, however, seem impossible to be asymptomatic (10). In our two patients we found small hidden diaphragmatic defects between the rims of the diaphragm. If there was no herniation during the asymptomatic period, as we suspected, it might have been due to the size of the defect. There was a chance for a small defect to be covered by the spleen or left lobe of liver, or the rims of the diaphragmatic defect overlapped.

The breaking events that turn asymptomatic period into an evident condition could have been the same in both cases. Vomiting in Case 1 could be explained by sudden increase in abdominal pressure. Golladay et al (16) noted the same event in their 13-month-old female patient. The information about the cough, constipation, or some other kind of physical effort in the beginning of the disease was negative. There was also no information about the child's unusual posture at that time. An "accident" as such can happen at any point in patient's lifetime (17,18). Osebold et al (10) concluded that the asymptomatic period might be due to the persistence of a pleuroperitoneal hernial sac, and that it finishes with its rupture. In our patients we did not find the remnants of the ruptured sac. Golladay et al (16) presumed that spleen or liver could block the diaphragmatic defect like a cork in a bottle. In Case 1 the spleen was necrotic. In left Bochdalek hernia with classic presentation the adhesions are never mentioned and we never found them. Congenital diaphragmatic defect is usually large and open for the entrance of the abdominal content, and the technical difficulties to close it are not rare (19).

Chest X-ray findings in left Bochdalek hernia are usually pathognomonic for the disease. It is almost unbelievable that children or adults with the abdominal organs in the left pleural cavity never needed chest X-ray during their life, never had a chest X-ray for other reasons, and that, even if they had it, the anomaly was not recognized. It is also hard to believe that bowel or altered pulmonary sounds over the chest were not heard on general medical examinations in childhood. The embryological development of this anomaly suggests the presence of abdominal contents in the left pleural cavity before birth (20). Glasson et al (21) found a normal chest radiogram before the onset of the symptoms in their patient with delayed left Bochdalek hernia. There are reports on cases of the left Bochdalek hernia with delayed presentation in which the patients were asymptomatic for months and years (10,12,15,16). Patients with right congenital diaphragmatic defect often live a

long, asymptomatic, and normal life (18,22). Usually, there is no foreign content in the right pleural cavity, so maybe the asymptomatic period of left Bochdalek hernia means the absence of foreign content in the left pleural cavity. This suggests that the asymptomatic period finishes with the entrance of abdominal content through the left diaphragmatic defect. Glasson et al (21) agreed that in their patient the reason was delayed herniation, not delayed onset of symptoms, and that the acute onset of the disease suggested the acute accident which began with the entrance of bowel into the pleural cavity. The prompt return of chest X-ray to normal following correction of the diaphragmatic defect indicated that the bowel was not chronically displaced in the left hemithorax (16). However, partial postoperative atelectasis of ipsilateral and contralateral lung was also noted after corrective operation by thoracotomy (23).

Clinical presentation of delayed left Bochdalek hernia is not specific and the diagnosis is often overlooked. There is also a very good respiratory compensatory mechanism, which is why the serious respiratory symptoms are not expected. Therefore, the asymptomatic period should be studied in the light of presence or absence of the gastrointestinal symptoms (10,16). However, Vazquez Rueda et al (12) noted recurrent respiratory symptoms during the asymptomatic period of hernia. A surgeon usually does not expect the anomaly in the postneonatal period, although it is easy to diagnose. Bochdalek hernia should be considered in cases of tension pneumothorax (24), pneumonia, pleuritis, and pulmonary tuberculosis (15,25). Not recognized, the condition can be complicated with incarceration and strangulation, as every other type of hernia (17). Intrapleural position makes the complications only more serious.

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