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ПРИКАЗ БОЛЕСНИКА / CASE REPORT
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# Late Presentation of Congenital Diaphragmatic Hernia – Case Report

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**SUMMARY**
**Introduction** Congenital diaphragmatic hernia (CDH) is a defect of the diaphragm with the penetration of organs of the abdominal cavity into thorax. Localization and size of the defect of the diaphragm condition the time and range of clinical manifestation. The most common is unilateral, without hernia sac, located on the left side, through posterolateral opening of the diaphragm and with clinical manifestation during the neonatal period. Smaller subgroup consists of patients with presentation outside the neonatal period with anatomic defect on the right side as is the case with our patient.
**Case Outline** Female infant aged seven months was hospitalized because of repeated episodes of shortness of breath and cough, with lack of progress in body weight. The discomforts had been present for the previous two months with an inadequate response to bronchodilator and antibiotic therapy used. After clinical, radiographic, ultrasound and computed tomography treatment, the right-sided diaphragmatic hernia was diagnosed, so surgical treatment followed. The defect of the diaphragm with hernia sac, through which the right lobe of the liver with visible line of entrapment, in the form of "hourglass", is pushed into thoracic cavity, was intraoperatively identified. The reposition of the right lobe of the liver in the abdominal cavity along with reconstruction of the diaphragm using interrupted mattress sutures was performed.
**Conclusion** The late manifestation of CDH should be suspected in cases of inexplicable acute or chronic respiratory or gastrointestinal symptoms with pathological radiography of the thorax. Accurate diagnosis and timely minimally invasive surgical intervention ensure an excellent prognosis.
**Keywords:** congenital diaphragmatic hernia; herniation; late presentation; defect of the diaphragm

## INTRODUCTION

Congenital diaphragmatic hernia (CDH) denotes a defect of the diaphragm with the penetration of organs of the abdominal cavity into thorax. Neonatal CDH was first described by Viktor Bochdalek in 1848 as migration of the spleen and intestines into the chest cavity. It occurs in one in 2,000 to one in 5,000 live-born infants, two times more often among males, and accounts for 8% of all congenital anomalies [1]. The most common is unilateral, without the hernia sac, in 90% of cases located on the left side, through a posterolateral opening of the diaphragm, which is explained by later closure of pleura-peritoneal canal on the left side [2]. Bilateral diaphragmatic hernias are very rare, accounting for less than 1%, with very poor prognosis and a survival rate of less than 35% [3]. Etiology of CDH is still unknown but it is increasingly associated with deficiency of vitamin A in the course of development of lungs and diaphragm [4]. Clinical features depend on location and size of the defect of the diaphragm and the presence of abdominal organs in pleural cavity. If the defect is large, symptoms of severe respiratory distress are manifested at birth: dyspnea, cyanosis and tachycardia get worse as the child swallows air, which leads to distension of the intrathoracic stomach and convolutions of intestines, increasing the compression of the pulmonary tissue, which is poorly developed on the side of hernia, with different degree of pulmonary hypoplasia and persistent pulmonary hypertension. [5]. Malrotation, pulmonary hypoplasia and open ductus arteriosus are present in all cases. If respiratory and circulatory disorders are minimal, the child may be without symptoms for a longer period and the diagnosis is made accidentally during the evaluation because of other, more frequent respiratory difficulties. Hernias on the right side are rare, they have poor symptomatology and are harder to detect. This is understandable because the liver blocks the defect of the diaphragm from the right side, preventing the penetration of other organs of the abdominal cavity, which is the reason for delayed diagnosis [6]. In 1701, Sir Charles Holt gave the first description of a late manifestation of the anomaly in the pediatric literature, and Batman Hess performed his first successful surgical intervention with a three-month-old girl in 1929 [7].

## CASE REPORT

Female infant aged seven months was hospitalized because of repeated episodes of shortness of breath and cough, with lack of progress in body weight. Discomforts had been present for the previous two months with an inadequate response to bronchodilator and antibiotic therapy used. Body temperature was not elevated. From

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personal anamnesis it was known to be a child of the first pregnancy, which had a normal course, carried to term, with normal delivery and birth weight of 2,490 g and length of 48 cm, Apgar score 9. The child was on natural diet for 2.5 months, but due to poor advancement in the third month adapted formula was introduced and then from the sixth month nutrition without milk was introduced. Orderly performed immunization and prophylaxis AD.

On admission, the female infant aged seven months had body weight of 5,510 g, was with preserved sensorium, easily tachypneic, afebrile, of paler skin and visible mucous membranes, with small macular changes around the neck and less on the trunk, with preserved elasticity and turgor.

Percussion of the lungs: basal and in the back slightly duller percussion sound.

Auscultation of the lungs: sharpened respiratory sound, with polyphonic audible whistling on both sides. To the right basal and in the back slight decrease in breathing function. Other physical findings were within normal limits. Complete blood count values, laboratory parameters, urinalysis, urine culture and chlorine in sweat were within the physiological limits.

Lung radiography: paracardial and right basal tumorous shadow (Figure 1).

Ultrasound of the abdomen and the kidneys: the liver was under rib cage, homogeneous, with no focal infiltrations. On the right side, above the diaphragm, visible circular, clearly limited tumorous growth of about 3.2×2.5 cm in diameter, which, by echogenicity, corresponds to liver parenchyma – liver herniation. The size and shape of other abdominal organs appeared normal.

Computerized tomography of the thorax, lungs and abdomen: on soft and bony structures there were no pathological changes (Figure 2). On the left side in the lung parenchyma there were no active infiltrative lesions. Both hila were sharply and clearly differentiated, without the presence of expansive changes. In the middle mediastinum there was no increase in lymph glands. The ascending aorta, arch of the aorta and descending aorta had the appropriate size and lumen. To the right side dorsally, hyperdensic change was present posteriorly, and corresponded to the parenchyma of the liver which penetrates intrathoracically through the back opening of the diaphragm – Bochdalek. All blood vessels of the liver were intraabdominally continuously monitored, as was the liver parenchyma.

Intraoperative findings: right subcostal incision was used to access the abdominal cavity. The entrance of the peritoneal cavity was free. The normoposition of organs of the peritoneal cavity was verified with elevation of the left lobe of the liver. The liver was released from its ligaments and liver mobilization was performed. The posterolateral defect of the diaphragm, measuring about 4×3 cm with the hernia sac, through which the right lobe of the liver is suppressed into the thoracic cavity, was identified (Figures 3 and 4). There was visible line of entrapment in the form of hourglass on the liver parenchyma, but without change in its vitality. Hernia sac was adherent on the line of strangulation, it was prepared in its entirety and resected. Complete repositioning of the right lobe of the liver in the abdominal cavity was performed. With inspection of the right hemothorax, atelectasis of the lung parenchyma of the lower right lobe without significant hypoplasia was noticed. The front edge of the diaphragm was well developed, with visualized phrenic nerve positioned at about 1 cm from the edge. Back edge was prepared and separated, and the reconstruction of the diaphragm by interrupted mattress sutures and nonabsorbable suture TiCron™ 2.0 was done.

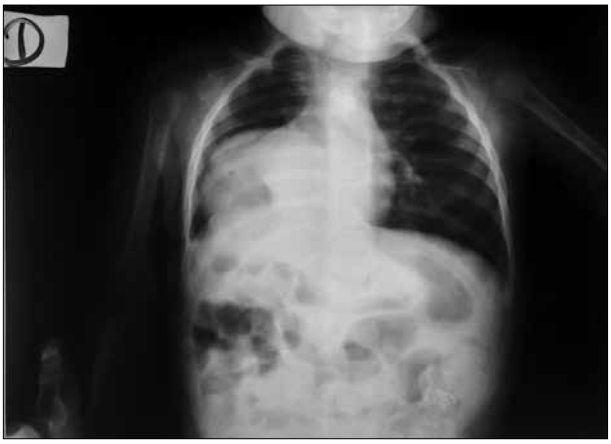

**Figure 1.** Chest X-ray: paracardial and right basal tumorous shading

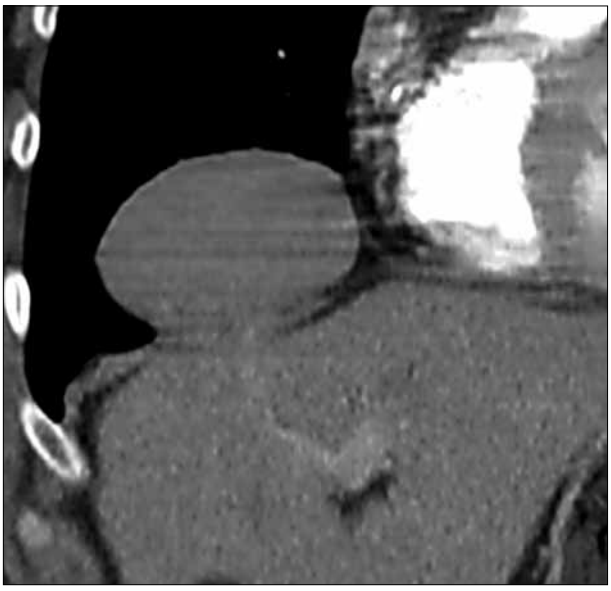

**Figure 2.** CT of the chest and abdomen: hyperdense lesion corresponding to the parenchyma of the liver penetrating into the chest cavity through the postero-lateral aperture of the diaphragm – Bochdalek

Control X-ray of the thorax on the fifth postoperative day: the presence of small amounts of air between the diaphragm and parietal pleura. The child was in good overall state three months after surgery, with proper advancement in body mass (7,800 g) and regular chest radiography.

## DISCUSSION

Late presentation of CDH outside the neonatal period is present in 2.6–20% of all cases, between the ages of one month to 18 years [8]. Even if the anatomical substrate of

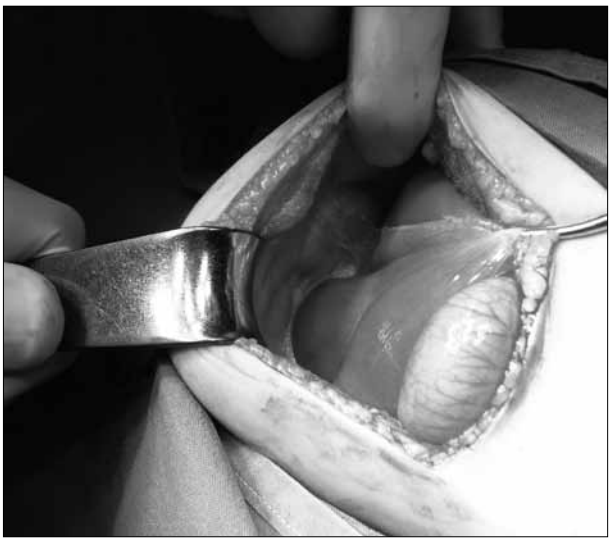

**Figure 3.** Intraoperative report: herniation of the liver into the form of hourglass with the visible border of the hernia sac

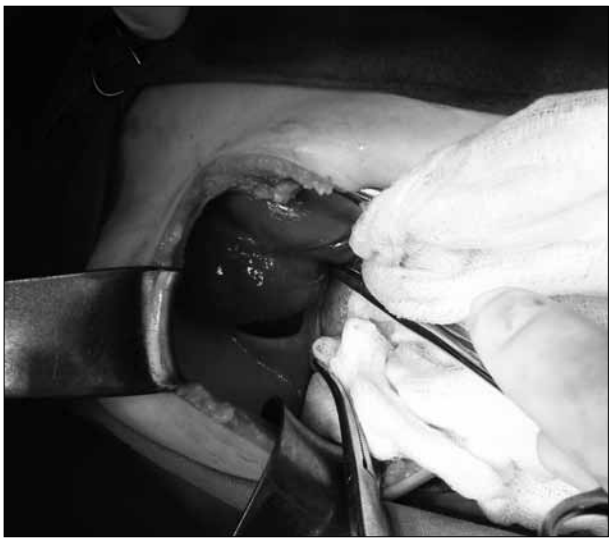

**Figure 4.** Intraoperative report: mobilized lobe of the liver with a visible herniation path and visualization of the diaphragmatic posterior defect

the defect in the diaphragm is the same, the clinical presentation of neonatal and late CDH is nevertheless different. It is clinically manifested as acute or chronic obstruction of the digestive signs (nausea, vomiting, abdominal pain, constipation) or as recurrent respiratory symptoms (cough, wheezing, tachypnea, infections), or symptoms of both systems [9]. Asymptomatic CDH is present in 10% of cases [10]. In two thirds of cases, right-sided CDH manifests itself only in the form of respiratory symptoms, which is in accordance with the theory that the liver prevents herniation of abdominal organs and thus the gastrointestinal symptomatology. It has been shown that the children with gastrointestinal symptomatology are older when compared to children where the respiratory symptomatology dominates. Generally, when the child is older, the gastrointestinal symptomatology of CDH as a result of incarceration of parts of digestive tube is a more probable way of manifestation [11]. Lack of progress as a clinical sign is present in one third of cases [12]. Migrating organs are the spleen, liver, stomach, small and large intestine, kidney and pancreas. Cases of the liver fusion with lower lung lobe are described, in a way that connection from the fibrovascular ribbon to complete fusion of parenchyma of two organs is presented. In that case, thoracic approach provides adequate surgical intervention [13]. Very rare possibility of intrapericardial diaphragmatic hernia, with herniation of the intestine into the pericardial sac, should be noted [14]. It is paradoxical that the CDH of newborns is a major therapeutic challenge, but clinically and radiologically easy to diagnose. On the other hand, late presentation of CDH is very frequently a difficult diagnostic problem, due to the fact that when the patient is older, the CDH is not even thought of, which all leads to delays in diagnosis and the possibility of iatrogenic complications. Clinical examination and auscultation of lungs is the first step. Intensified or weakened percutory finding of sonority and auscultation decrease in breathing function require careful evaluation. The diversity and size of migrating organs result in different radiographic image. Radiography of the thorax, according to the data of Bagłaj [6], enables accurate diagnosis in approximately 49%. X-ray image of the thorax with intestines filled with air, sometimes with levels of liquids with the mediastinal shift, is a typical sign [12]. Wrong diagnosis is present in approximately 25% of cases as pneumonia, pneumothorax, congenital cystic adenomatoid malformation of the lungs, pleural effusion or eventration of the diaphragm, which can result in unnecessary thoracic drainage and thoracentesis and consequent perforations of the abdominal organs or bleedings of the liver or spleen [15]. Wrong interpretation can be eliminated by inserting a nasogastric tube before scanning [16]. Even normal chest radiography finding does not exclude the diagnosis of CDH, since the herniation of visceral organs in thorax may be periodic and radiologically provable only in Trendelenburg position [17]. Using ultrasonography of abdomen and thorax, the relationship between intraabdominal organs and the dome of the diaphragm may be visualized, and comparison of density of intrathoracic finding with pulmonary tissue can be performed. Respiratory mobility of the diaphragm, verified by ultrasonography, allows differential diagnosis of CDH and eventration.

Computerized tomography of the thorax and abdomen is a diagnostic option that provides precious information especially in cases of the right-sided CDH, since in 80% of cases there is a herniation of the liver and the possibility of misdiagnosis. It allows clear visualization of the defect and the possibility of solving differential diagnostic dilemmas toward other tumorous shadows in the thorax [18].

The therapy of choice is surgical intervention. In cases of acute manifestation, an urgent intervention is of key importance to prevent volvulus of the stomach, perforation, strangulation and progression of respiratory distress [19]. Other patients require postponed surgical intervention within a few days, with the prevention of pulmonary infection, respiratory and metabolic acidosis [12].

The most commonly used surgical approaches in the period before the development of minimally invasive sur-

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gery were subcostal laparotomy or thoracotomy. Laparotomy has been ensuring a good visualization of the defect and easier repositioning of migrating organs with reduced possibility of injury of parenchymatous organs and uncontrolled increase in vascular pressure.

Progress in endosurgical instrumentarium and the acquisition of relevant experience have permitted minimally invasive surgery to be a recommended treatment standard for both neonatal and CDH with late presentation.

Thoracoscopic approach provides clear visualization, minimal trauma, short duration of the procedure with quick postoperative recovery and good pulmonary function. Esthetic results are excellent, but the intervention must be carried out in specialized centers [20].

The percentage of relapses in case of minimally invasive surgery is higher but it decreases with surgical teams' obtainment of experience and with long-term monitoring of patients [21, 22].

Recurrence of CDH after minimally invasive surgery does not involve open procedure, but it can be also taken care of with thoracoscopic or laparoscopic approach [23].

Limiting factor for thoracoscopy is persistent pulmonary hypertension [24].

The late manifestation of CDH should be suspected in cases of inexplicable acute or chronic respiratory or gastrointestinal symptoms with pathological radiography of the thorax. Accurate diagnosis and timely minimally invasive surgical intervention ensure an excellent prognosis.

## REFERENCES


1. Stolar CJ, Dillon P. Congenital diaphragmatic hernia. In: Grosfeld JL, O'Neill JA, Fonkalsrud EW, editors. Pediatric Surgery. 6th ed. Philadelphia: Mosby-Elsevier; 2006. p.932.
2. Isselstijn H, Tibboel D. The lungs in congenital diaphragmatic hernia: Do we understand? Pediatric Pulmonol. 1998; 26:204-18.
3. The Congenital Diaphragmatic Hernia Study Group. Bilateral congenital diaphragmatic hernia. J Pediatr Surg 2003; 38(3):522-4.
4. Greer JJ. Etiology of congenital diaphragmatic hernia: the retinoid hypothesis. Pediatr Res. 2003; 53(5):726-30.
5. Krstić Z, et al. Patologija dijafragme. In: Osnovi dečije hirurgije. Beograd: BMG; 1993. p.104-105.
6. Bagłaj M. Late-presenting congenital diaphragmatic hernia in children: a clinical spectrum. Pediatr Surg Int. 2004; 20(9):658-69.
7. Irish M, Holm B, Glick P. Congenital diaphragmatic hernia. A historical review. Clin Perinatol. 1996; 23:625-53.
8. Kitano Y, Lally KP, Lally PA; Congenital Diaphragmatic Hernia Study Group. Late-presenting congenital diaphragmatic hernia. J Pediatr Surg. 2005; 40(12):1839-43.
9. Ozturk H, Karnak I, Sakarya M, Cetinkursun S. Late presentation of Bochdalek hernia: clinical and radiologicalaspects. Pediatr Pulmonol. 2001; 31:306-10.
10. Bagłaj M, Dorobisz U. Late-presenting congenital diaphragmatic hernia in children: a literature review. Pediatr Radiol. 2005; 35:478-88.
11. Manning PB, Murphy JP, Raynor SC, Asch KW. Congenital diaphragmatic hernia presenting due gastrointestinal complications. J Pediatr Surg. 1992; 27(9):1225-8.
12. Chao PH, Chuang JH, Lee SY, Huang HC. Late-presenting congenital diaphragmatic hernia in childhood. Acta Paediatr. 2011; 100(3):425-8.
13. Taide DV, Bendre PS, Kirtane JM, Mukunda R. Hepatic pulmonary fusion: a rare case. Afr J Paediatr Surg. 2010; 7(1):28-9.
14. Jain V, Roy Choudhury S, Chadha R, Puri A. Intrapericardial diaphragmatic hernia: a rare type of congenital diaphragmatic hernia. J Pediatr Surg. 2011; 46(5):e29-31.
15. Elhalaby EA, Abo Sikeena MH. Delayed presentation of congenital diaphragmatic hernia. Pediatr Surg Int. 2002; 18:480-5.
16. Schimpl G, Fotter R, Sauer H. Congenital diaphragmatic hernia presenting after the newborn period. Eur J Pediatr. 1993; 152:765-8.
17. Hight DW, Hixson SD, Reed JO, Wats FB, Herler JH. Intermittent diaphragmatic hernia of Bochdale: report of a case and literature review. Pediatrics. 1982; 34:601-4.
18. Fotter R, Schimpl G, Sorantin E, Fritz K, Landler U. Delayed presentation of congenital diaphragmatic hernia. Pediatr Radiol. 1992; 22:187-91.
19. Cigdem MK, Onen A, Otcu S, Okur H. Late presentation of Bochdalek-type congenital diaphragmatic hernia in children: a 23-year experience at a single center. Surg Today. 2007; 37:642-5.
20. Huang JS, Lau CT, Wong WY, Tao Q, Wong KK, Tam PK. Thoracoscopic repair of congenital diaphragmatic hernia: two centres' experience with 60 patients. Pediatr Surg Int. 2015; 31(2):191-5.
21. Tanaka T, Okazaki T, Fukatsu Y, Okawada M, Koga H, Miyano G, et al. Surgical intervention for congenital diaphragmatic hernia: open versus thoracoscopic surgery. Pediatr Surg Int. 2013; 29(11):1183-6.
22. Liem NT, Nhat LQ, Tuan TM, Dung le A, Ung NQ, Dien TM. Thoracoscopic repair for congenital diaphragmatic hernia: experience with 139 cases. J Laparoendosc Adv Surg Tech A. 2011; 21(3):267-70.
23. Cantone N, Destro F, Libri M, Pavia S, Lima M. Laparoscopic patch repair of recurrent anterior diaphragmatic hernia in a child with SMA. Springerplus. 2014; 3:165.
24. Gomes Ferreira C, Kuhn P, Lacreuse I, Kasleas C, Philippe P, Podevin G, et al. Congenital diaphragmatic hernia: an evaluation of risk factors for failure of thoracoscopic primary repair in neonates. J Pediatr Surg. 2013; 48(3):488-95.

# Касна презентација конгениталне дијафрагмалне киле – приказ болесника

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**КРАТАК САДРЖАЈ**

**Увод** Конгенитална дијафрагмална кила је дефект дијафрагме с продором органа трбушне дупље у грудни кош. Локализација и величина оштећења на дијафрагми условљавају време и спектар клиничког испољавања. Најчешће је једнострана, без килне кесе, лоцирана на левој страни, кроз постеролатерални отвор дијафрагме, и клиничком манифестацијом током неонаталног периода. Јавља се и код мање подгрупе нешто старије деце с анатомским дефектом на десној страни, што је забележено и код болесника којег приказујемо.

**Приказ болесника** Женско одојче узраста од седам месеци примљено је на болничко лечење због поновљене епизоде отежаног дисања и кашља и јер није добијало на тежини. Тегобе су се јавиле два месеца раније, а одговор на примењену бронходилататорну и антибиотску терапију није био одговарајући. Након клиничког, радиографског, ултразвучног и прегледа компјутеризованом томографијом постављена је дијагноза десностране дијафрагмалне хернije, те је приступљено хируршком лечењу. Током операције установљен је дефект дијафрагме с килном кесом кроз који је у грудну дупљу потиснут десни режањ јетре, на којем је била видљива линија уклештења у виду пешчаног сата. Учињена је репозиција десног режња јетре у трбушну дупљу уз пластику дијафрагме појединачним мадрац-шавовима.

**Закључак** На касну манифестацију конгениталне дијафрагмалне киле треба мислити када постоје необјашњиви акутни или хронични респираторни или гастроинтестинални симптоми и патолошки налаз радиографије грудног коша. Тачна дијагноза и благовремена минимално инвазивна хируршка интервенција обезбеђују одличну прогнозу стања болесника.

**Кључне речи:** конгенитална дијафрагмална кила; херниjација; касна презентација; дефект дијафрагме

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