IV. PEDIATRIC SURGERY

CONGENITAL DYAPHRAGMATIC HERNIA - CASE REPORT

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Abstract

Congenital diaphragmatic hernia (CDH) constitutes a major surgical emergency in the newborn, and the key to survival lies is prompt diagnosis and treatment. CDH is characterized by a variable degree of pulmonary hyperplasia associated with a decrease in cross-sectional area of the pulmonary vasculature and dysfunction of the surfactant system. The lungs have a small alveolar capillary membrane for gas exchange, which may be further decreased by surfactant dysfunction. In addition to parenchyma disease, increased muscularization of the intraacinar pulmonary arteries appears to occur. In very severe cases, left ventricular hypoplasia is observed. We report a case of a 3 year old girl presented with congental diaphragmatic hernia to which we applied surgical treatment.

Key words: congenital diaphragmatic hernia, pulmonary hypoplasia, posterolateral Bochdalek hernia

Abbreviations

CDH- congenital diaphragmatic hernia

PPHN- persistent pulmonary hypoplasia in the newborn

Introduction

The diaphragm is the major muscle of respiration and the second most important muscle within the body after the heart. Then a decrease in diaphragmatic function occurs, a concomitant respiratory dysfunction occurs. However, no compensatory mechanisms are in place to prevent respiratory compromise in the setting of decreased diaphragmatic excursion. Congenital diaphragmatic hernias (CDH) occur through embryologic defects in the diaphragm, and most patients present early in life rather than later[1]. However, a subset of adults may present with a smaller congenital hernia that was undetected during childhood[2]. The diaphragm initially develops as a septum between the heart and liver, progresses posterolaterally, and closes at the left Bochdalek foramen at approximately 8-10 weeks' gestation. The herniation of viscera in CDH usually occurs during the pseudoglandular stage of lung development. Lung compression results in pulmonary hypoplasia that is most severe on the ipsilateral side, although both lungs may be abnormal. Pulmonary hypoplasia is associated with fewer bronchial, alveolar and arterial generations. Pulmonary capillary blood flow is decreased because of the small cross-sectional area of the pulmonary vascular bed, and flow may be further decreased by abnormal pulmonary vasoconstriction. Pulmonary hypertension and pulmonary hypoplasia have been recognized as the 2 cornerstones of the pathophysiology of CDH. In recent years, evidence suggests that cardiac maldevelopment may further complicate the pathophysiology of CDH[3,4,5].

Etiology

Genetic factors: The initiating factor responsible for the development of CDH is unknown. Wide variations (7-31%) have been noted in the reported prevalence of chromosomal abnormalities (trisomy 13, trisomy 18, and tetrasomy 12P mosaicism) in patients with CDH. The prevalence is higher in cases of CDH associated with other defects. Familial occurrence has been noted in fewer than 2% of CDH cases[6,7,8].

The role of *drugs and environmental chemicals* in the development of CDH is uncertain, but quinine, thalidomide, phenmetrazine, and polybrominated diphenyls have been used to induce CDH in various species.

Frequency

In the US, CDH occurs in 1 of every 2000-4000 live births and accounts for 8% of all major congenital anomalies. Worldwide, the frequency is the same as that in the United States.

Classification of CDH

The 3 basic types of CDH are the posterolateral Bochdalek hernia (occurring in utero at approximately 8-10 weeks of gestation), the anterior Morgagni hernia, and the less-common hiatus hernia. The left-sided Bochdalek hernia is seen in approximately 90% of cases. The major problem in a Bochdalek hernia is the posterolateral defect of the diaphragm, which results in either the failure of the pleuroperitoneal folds to develop or the improper or absent

migration of the diaphragmatic musculature. Bilateral Bochdalek hernias are rare.

Positiv diagnosis

Prenatal: The diagnosis of CDH is frequently made prenatally prior to 25 weeks' gestation. CDH is usually detected in the antenatal period (46-97%), depending on the use of level II ultrasonography techniques. Ultrasonography reveals polyhydramnios, an absent intra-abdominal gastric air bubble, mediastinal shift, and hydrops fetalis. Ultrasonography demonstrates the dynamic nature of the visceral herniation observed with CDH. The visceral hernia has moved in and out of the chest in several fetuses[9].

Postnatal: History and clinical findings vary with the presence of associated anomalies and the degree of pulmonary hypoplasia and visceral herniation. In the infant presenting in the neonatal period without prenatal diagnosis, variable respiratory distress and cyanosis, feeding intolerance, and tachycardia are noted. In the physical examination, the abdomen is scaphoid if significant visceral herniation is present. On auscultation, breath sounds are diminished, bowel sounds may be heard in the chest, and heart sounds are distant or displaced.A chest radiography confirms the diagnosis of CDH.Findings include loops of bowel in the chest, mediastinal shift, paucity of bowel gas in the abdomen, and presence of the tip of a nasogastric tube in the thoracic stomach . Repeated chest radiographs may reveal a change in the intrathoracic gas pattern. Right-sided lesions are difficult to differentiate from diaphragmatic eventration and lobar consolidation. Early echocardiography may reveal cardiac defects, decreased left ventricular mass, poor ventricular contractility, pulmonary and tricuspid valve regurgitation, and right-to-left shunting.Repeated echocardiography is recommended to measure changes in the pulmonary artery pressure, left-to-right shunt, and flow across the ductus arteriosus. MRI clearly depicts diaphragmatic discontinuity, the fetal compressed lung, connecting bowel segments between the abdomen and chest. MRI findings can be used to differentiate CDH from other chest masses, and it is superior to ultrasonography in demonstrating the position of the fetal liver above or below the diaphragm.In childhood or adult period, Undiagnosed Bochdalek hernias are most frequently identified when the patients undergo CT for reasons that appear to be unrelated to the hernia. In adults, Bochdalek hernias usually contain retroperitoneal fat or a kidney[10,11].

Differential diagnoses are as follows:

Congenital cystic adenomatoid malformation

Pulmonary sequestration

Mediastinal cystic processes (cystic teratoma, thymic cysts, foregut duplication cysts)

Neurogenic tumors

Treatment

Medical Care: Because of associated PPHN (persistent pulmonary hypertension in the newborn) and pulmonary hypoplasia, medical therapy is directed toward optimizing oxygenation while avoiding barotrauma. In the delivery room, if the infant is known or suspected to have CDH, immediately place a nasogastric tube and connect it to continuous suction to prevent bowel distension and further lung compression. For the same reason, avoid mask ventilation and immediately intubate the trachea. Avoid high peak inspiratory pressures and be alert to the possibility of early pneumothorax if the infant does not stabilize[12,13].

Surgical Care:

Fetal surgery: Harrison et al reported the first human fetal surgery for CDH in 1990[14]. However, a randomized trial published in 1998 showed that in utero repair did not improve survival compared with standard therapy. Currently, fetal intervention is not indicated in CDH.

Postnatal surgical care: until recently, specialists believed that reduction of the herniated visceras and closure of the diaphragmatic defect should be emergently performed following birth. However, a delayed surgical approach that enables preoperative stabilization decreases morbidity and mortality. This change in protocol is due to the recent understanding that the medical problems of pulmonary hypoplasia and PPHN(persistent pulmonary hypertension in the newborn) are largely responsible for the outcome of CDH and that the severity of these pathophysiologies is largely predetermined in utero. Herniated viscera in the chest does not appear to exacerbate the pathophysiology as long as bowel decompression with a nasogastric tube is continuous. Several reports indicate that circulatory stability, respiratory mechanics, and gas exchange deteriorate after surgical repair. The ideal time to repair a CDH is unknown. Some suggest that repair 24 hours after stabilization is ideal, but delays of up to 7-10 days are typically well tolerated, and many surgeons now adopt this approach. Some surgeons prefer to operate on these neonates when normal pulmonary artery pressure is maintained for at least 24-48 hours based on echocardiography[15,16].

Complications of CDH

Pulmonary hypoplasia

The main problem lies in the presence of pulmonary hypoplasia, which may be unilateral or bilateral. Pulmonary hypoplasia is thought to result from long-standing intrauterine (embryonic) compression of the lungs by the hernia. Mortality in babies with CDH is largely confined to those with bilateral pulmonary hypoplasia, but hypoplasia is always more severe in the lung ipsilateral to the hernia. The pulmonary vasculature is also affected to a greater degree than the bronchial tree[17]. Infants with the largest and longest-standing hernias have the most-hypoplastic lungs and are less likely to survive after birth. If a diaphragmatic hernia develops toward the end of pregnancy or after birth, pulmonary hypoplasia does not occur.

Gastric volvulus

Gastric volvulus can occur in early infancy as a complication of CDH, and it usually produces acute gastric

obstruction. Radiographic findings usually consist of an inverted distended stomach[11,18].

Rotational abnormalities and midgut volvulus

Intestinal malrotation is commonly observed in children with CDH (30-62%), and it also occurs in 37-40% of the cases of right-sided CDH. Volvulus is a complication in a small minority of these cases[19].

Gastric or other intestinal perforations

Gastric or other intestinal perforations occur rarely.

Hypoplasia of the left ventricle with a left-sided hernia or pleural effusions due to the right-sided involvement

The pleural effusion is believed to be the result of lymphatic obstruction secondary to the compressive effects of the hernia.

Bilateral renal hypertrophy

The kidneys are often enlarged and hyperplastic[20]. Some have suggested that an embryonic liaison exists between the kidneys and the lungs wherein the kidney produces a pulmonary growth factor (Proline) that influences normal lung development. Conversely when the lung is hypoplastic, it produces a renotropic substance and causes the kidneys to hypertrophy.

Case report

We present a 3-year old girl, 10 kilograms weight who was admissed to our Department of Pediatric Surgery with bilious emesis, signs of respiratory distress (retractions, cyanosis, grunting respirations),colicky abdominal pain and lack of stools for 5 days. Physical examinations revealed asymetric chest and also asymmetric abdominal distension (more on the left side). The auscultation of the lungs revealed no air entry on the left side with bowel movements on the left pulmonary aria, with a shift of cardiac sounds over the right chest. Chest-abdominal plain films with contrast substance showed dilated stomach and also some dilated loops of small bowel in left-side of the thoraces, and a few air-fluids levels in the left chest and abdomen.

Blood analyses revealed nutritional anemia, dehidratation with hypovolemia, mixed acidosis and intestinal parasitosis.We diagnosted congenital diaphragmatic hernia with acute bowel obstruction. First time we applied non-operative treatment for acute bowel obstruction (gastric aspiration, hydro-electrolitic and metabolic equilibrated and also total parentheral nutrition). After this period we continued the investigations. A CT-scan of chest and abdomen confirmed our diagnosis and excluded a possible diaphragmatic relaxation. (Figure no 1 and 2).

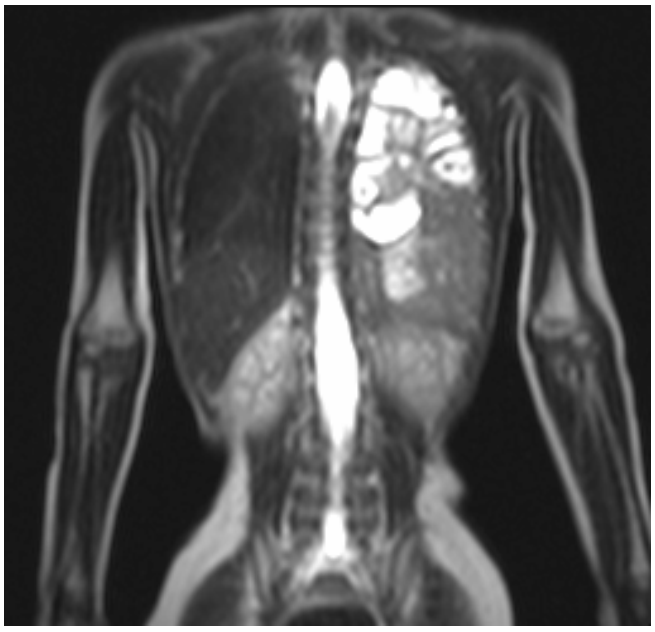

FIG. 1 CT scan image

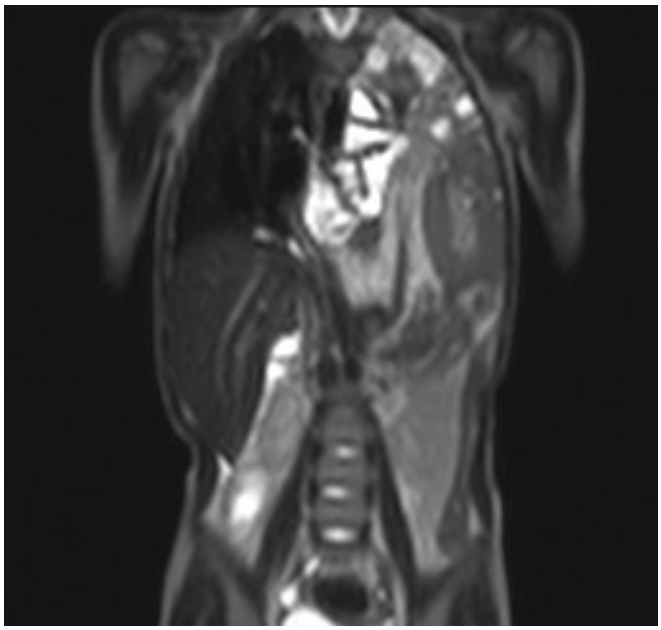

FIG. 2 CT scan image

After these completed investigations and the treatment of upper respiratory tract infection and intestinal parasitosis we decided that this patient fullfield conditions for surgical treatment. The surgical procedure was made under general endotracheal anesthesia. (figure 3). We approached with laparotomy so that abdominal contents could be inspected adequately. The hole in the diaphragm is found (figure no 4) and a tube is placed in the chest to equilibrate the pressures. We gently reduced the herniated visceras: stomach, spleen, complete small bowel, ascending and transverse colon (hepatic flexure) (figures no 5 and 6).

Then we approximated the edges of the diaphragm with nonabsorbable suture (figure no 7).

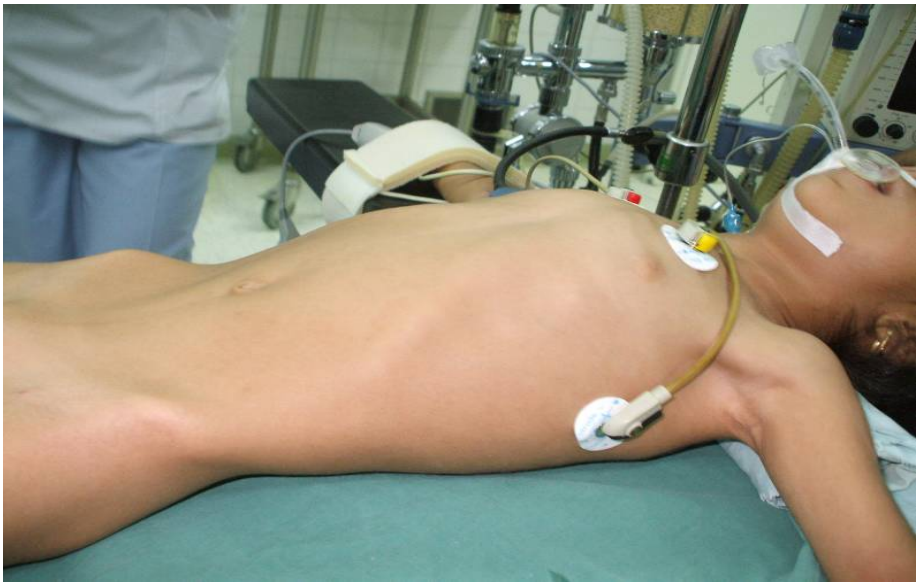

FIG. 3 Pre-surgical aspect.

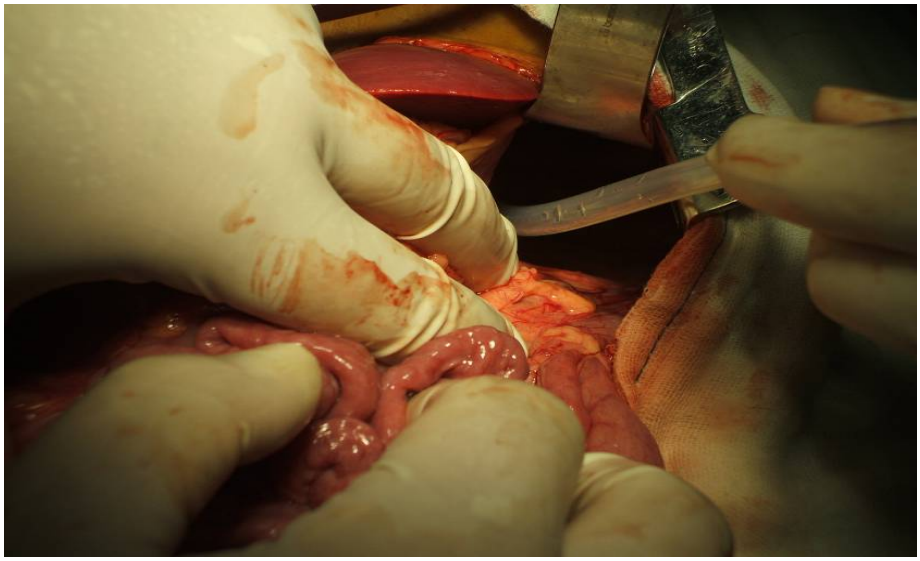

FIG. 4 Intraoperator aspect (the hole in the diaphragm).

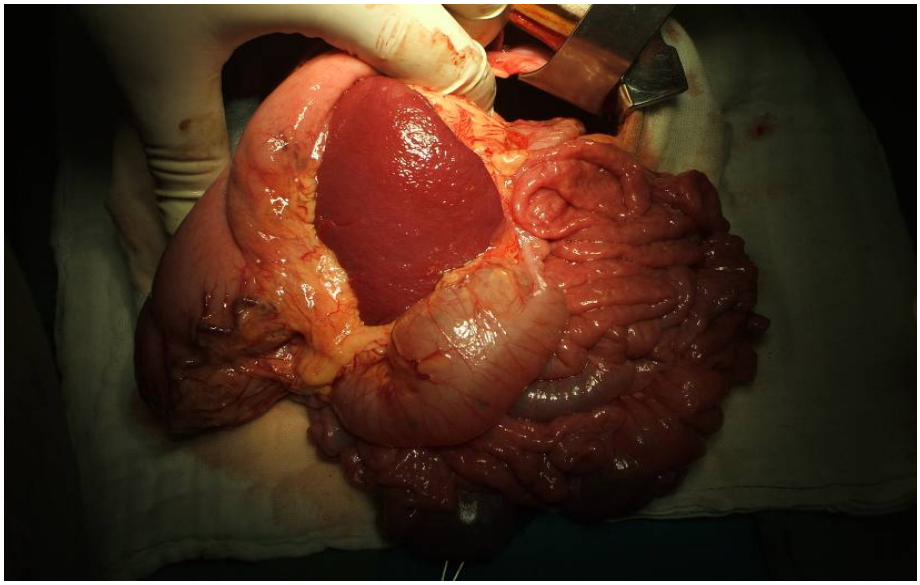

FIG. 5 Intraoperator aspect (herniated visceras).

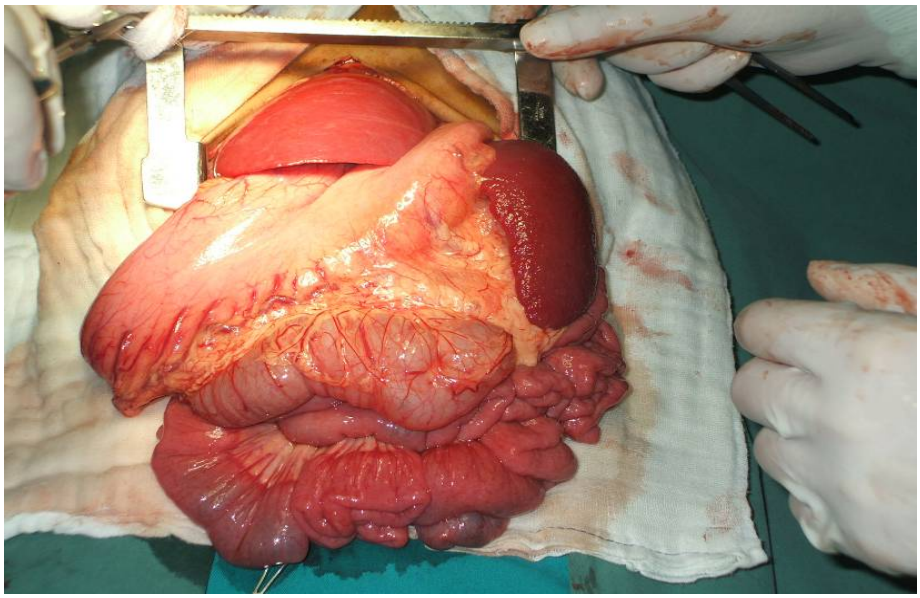

FIG. 6 Intraoperator aspect (herniated visceras).

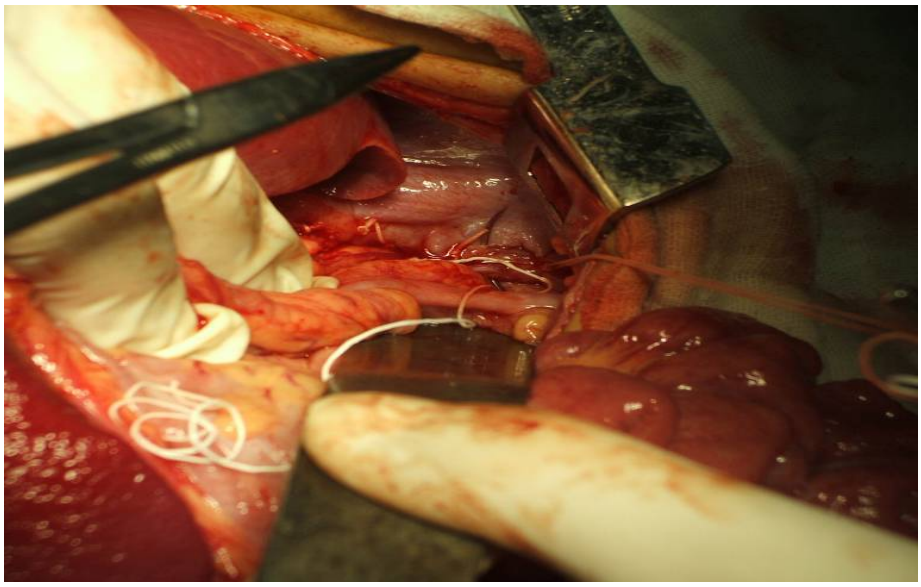

FIG. 7 Intraoperator aspect (suture of diaphragm).

The defect was large (approximately 8 cm in diameter) but prosthetic mesh was not necessary. A tube is placed in the left chest to allow air, blood, and fluid to drain so the lung can re-expand. In our case, the left lung was not functionally, but the mediastin took the normally placed.Pain management required intravenous narcotics and non-steroidal anti-inflammatory drugs (NSAIDs) seven days. Protective anthybiotherapy and parentheral nutrition was necessary for 3 days. The patient left the hospital 10 days after surgery.

Follow-up care

Once an anatomic defect has been corrected, periodically assessing pulmonary function and obtaining chest radiographs is important. Although spontaneous recurrence of a repaired diaphragmatic hernia is low, small defects in the repair site have been reported, so surveillance is essential.

Conclusions

The prognosis in CDH is different.For the baby who is born with respiratory impairment and is immediately symptomatic, the chance of survival is poor unless an almost perfect resuscitative effort is accomplish without delay. Other baby who presents with symptoms after 24 hours of life and becomes compromised more slowly has an excelent prognosis. Our case confirms the hypothesis that CDH is a major medical emergency but a delayed surgical emergency.

References

1. Gross RE: Congenital hernia of the diaphragm. Am J Dis Child 1946; 71: 579-592.
2. Mullins ME, Stein J, Saini SS: Prevalence of incidental Bochdalek's hernia in a large adult population. AJR Am J Roentgenol 2001 Aug; 177(2): 363-6[Medline].
3. Fauza DO, Wilson JM: Congenital diaphragmatic hernia and associated anomalies: their incidence, identification, and impact on prognosis. J Pediatr Surg 1994 Aug; 29(8): 1113-7[Medline].
4. Areechon W, Reid L: Hypoplasia of the lung associated with congenital diaphragmatic hernia. Br Med J 1963; i: 230-233.
5. Wilcox DT, Irish MS, Holm BA, Glick PL: Pulmonary parenchymal abnormalities in congenital diaphragmatic hernia. Clin Perinatol 1996 Dec; 23(4): 771-9[Medline].
6. Tibboel D, Gaag AV: Etiologic and genetic factors in congenital diaphragmatic hernia. Clin Perinatol 1996 Dec; 23(4): 689-99[Medline].
7. Torfs CP, Curry CJ, Bateson TF, Honore LH: A population-based study of congenital diaphragmatic hernia. Teratology 1992 Dec; 46(6): 555-65[Medline].
8. Slavotinek AM: The genetics of congenital diaphragmatic hernia. Semin Perinatol 2005 Apr; 29(2): 77-85[Medline].
9. Wilcox DT, Irish MS, Holm BA, Glick PL: Prenatal diagnosis of congenital diaphragmatic hernia with predictors of mortality. Clin Perinatol 1996 Dec; 23(4): Bagolan P, Casaccia G, Crescenzi F, et al: Impact of current treatment protocol on outcome of high risk congenital diaphragmatic hernia. Journal of Pediatric surgery 2004; 39: 313-318[Medline].
10. Gale ME: Bochdalek hernia: prevalence and CT characteristics. Radiology 1985 Aug; 156(2): 449-52[Medline].
11. Pelizzo G, Lembo MA, Franchella A: Gastric volvulus associated with congenital diaphragmatic hernia, wandering spleen, and intrathoracic left kidney: CT findings. Abdom Imaging 2001 May-Jun; 26(3): 306-8[Medline].
12. Bohn DJ, Pearl R, Irish MS, Glick PL: Postnatal management of congenital diaphragmatic hernia. Clin Perinatol 1996 Dec; 23(4): 843-72[Medline].
13. Chess PR: The effects of gentle ventilation on survival in congenital diaphragmatic hernia. Pediatrics 2004; 113: 917[Medline].
14. Harrison MR, Keller RL, Hawgood SB, et al: A randomized trial of fetal endoscopic tracheal occlusion for severe fetalcongenital diaphragmatic hernia. N Engl J Med 2003 Nov 13; 349(20): 1916-24[Medline].
15. Clark RH, Hardin WD Jr, Hirschl RB, et al: Current surgical management of congenital diaphragmatic hernia: a report from the Congenital Diaphragmatic Hernia Study Group. J Pediatr Surg 1998 Jul; 33(7): 1004-9[Medline].
16. Wung JT, Sahni R, Moffitt ST, et al: Congenital diaphragmatic hernia: survival treated with very delayed surgery, spontaneous respiration, and no chest tube. J Pediatr Surg 1995 Mar; 30(3): 406-9[Medline].
17. Saifuddin A, Arthur RJ: Congenital diaphragmatic hernia--a review of pre- and postoperative chest radiology. Clin Radiol 1993 Feb; 47(2): 104-10[Medline].
18. Reed JO, Long EF: Diaphragmatic hernia in infancy. AJR Am J Roentgenol 1959; 82: 437.
19. Rescorla FJ, Shedd FJ, Grosfeld JL, et al: Anomalies of intestinal rotation in childhood: analysis of 447 cases. Surgery 1990 Oct; 108(4): 710-5; discussion 715-6[Medline].
20. Swischuk LE: Imaging of the Newborn, Infant and Young Child. 4th ed. Lippincott Williams & Wilkins; 1997:68-72, 398-99, 412-23.

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