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Congenital Diaphragmatic Hernia Associated with Uncommon Abnormalities

CASE REPORT

Helson Freitas da Silveira[1], Howard Lopes Ribeiro Junior[2], Jalles Dantas De Lucena[1], Osvaldo Pereira da Costa Sobrinho[2], Luane Macedo de Sousa[1], Kalina Kelma Oliviera de Sousa[2], João Erivan Façanha Barreto[2], Roberta Silva Pessoa[2], Ariel Gustavo Scafuri[1,2], Delane Viana Gondim[1,2], Gilberto Santos Cerqueira[1,3]

1 Post-Graduate Program in Morphofunctional Sciences, Federal University of Ceara, Fortaleza, Ceara, Brazil.
2 Department of Morphology, Federal University of Ceará, Fortaleza, Ceara, Brazil.
3 Department of Medicine, Federal University of Piauí, Parnaíba, Piaui, Brazil.

Contact information:

Gilberto Santos Cerqueira.

Address: Av. Delmiro de Farias, s/n. Campus Porangabussu- Fortaleza/Ceará, Brazil. CEP: 64204-035.
Tel: +55 (85) 98648-9899.

giufarmacia@hotmail.com

Abstract

Introduction: Congenital diaphragmatic hernia is a poor formation of the diaphragm characterized by the presence of an intestinal malrotation, It is related to abnormal position of the intestine in the thorax.

Objective and Result: This case report a stillborn at 32 weeks, that was observe anatomical abnormalities associated with congenital diaphragmatic hernia, which occurred in the left antero-posterior region, such as intestinal malrotation, hepatomegaly and nephromegaly, with the presence of a hernial ring that occupied 80% of the left side of the diaphragm, besides a hypertrophied heart, deviated to the right, bilateral pulmonary hypotrophy.

Conclusion This case illustrates a rare case of diaphragmatic hernia with intestinal changes of clinical and surgical importance.

Keywords
Anatomy; Hernia; Medicine; Congenital Hernia.

Congenital diaphragmatic hernia (CDH) is a congenital malformation of diaphragm that occurs in approximately 1 in every 3500 live births in worldwide. [1] One of the main features of the CDH is the presence of Intestinal Malrotation (IM), and it is assumed that its presence is related to the abnormal prenatal position of the gut within a distorted thoracoabdominal space during the processes of reintegration from

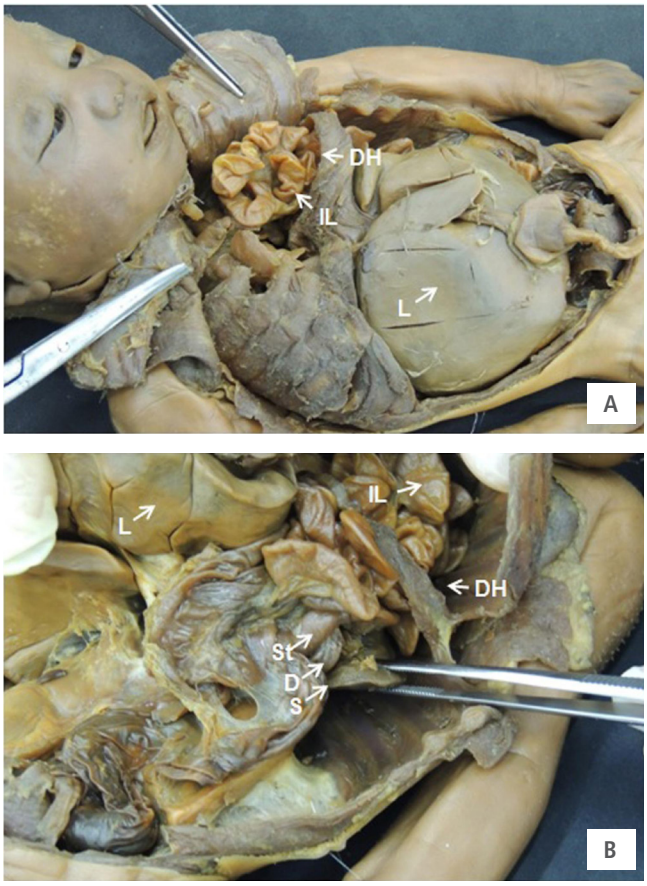
the umbilical cord hernia and definitive fixation. [2] Over 80% of reported cases are diagnosed as postero-lateral left-side CDH being reported that 40-50% of patients are affected with other congenital malformations. [3]

We report a stillbirth that presented unusual abnormalities (i.e. cardiac hypertrophy, several pulmonary hypoplasia, hepatomegaly, nephromagaly and cranial splenic torsion) associated to anteroposterior left-side CDH with IM.

A 32-weeks-old boy stillborn, at autopsy, was diagnosed with a clinical condition of large anteroposterior left-side CDH associated with IM and the presence of five unusual abnormalities. First, after dissection of the thoracic cage, was possible to visualize the intestinal loops displaced from the abdomen to the thorax as from the congenital hernia in the diaphragm muscle **(Figure 1A)**. The right diaphragmatic cupula is normal. In the mediastinum is possible to visualize the heart slightly deflected to the right, later covered by the pericardium, presenting cardiac hypertrophy when compared to fetal age **(Figure 1B)**.

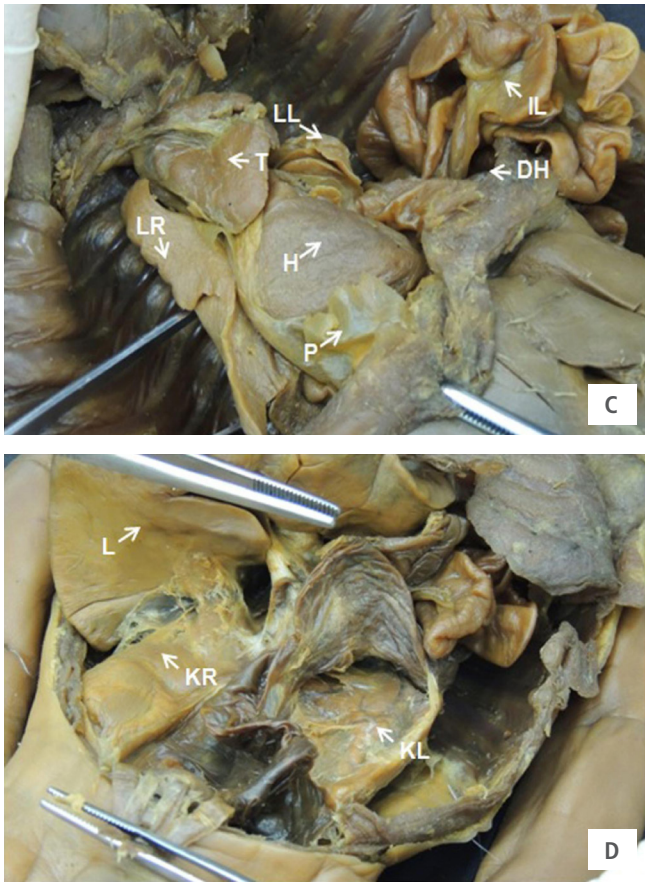
Additionally, laterally to mediastinum, it is possible to visualize the pulmonary hypoplasia, characterized more sternly in the left lung, and being still possible to identify the oblique fissure and the pulmonary lingula (Figure 1B). After dissection of the abdomen, was possible to identify a hepatomegaly and nephromegaly severe condition and having the liver and kidneys almost totally occupying the abdominal cavity (**Figure 1A, 1B** and **2C**, respectively). Additionally, we verified the presence of a cranial splenic torsion toward the CDH **(Figure 2D)**. It was also possible to identify the ileocecal junction in the upper left quadrant of the abdominal cavity **(Figure 2D)**. The large intestine was located in the left medial region of the abdominal cavity being possible to identify the descending and sigmoid colon in anatomical position. It is possible to identify the umbilical vein moving toward the umbilicus, was continued as the umbilical cord **(Figure 2D)**.

Figure 1: A) Dissection of a stillborn presenting congenital hernia in the muscle of the diaphragm associated with hepatomegaly. B) Presentation of cardiac hypertrophy and pulmonary hypoplasia.



To the best of our knowledge, this is the first case report which identifies the presence of cardiac hypertrophy, several pulmonary hypoplasia, hepatomegaly, nephromagaly and cranial splenic torsion associated to anteroposterior left-side CDH with IM in a stillborn. In this report we note that the congenital diaphragmatic hernia can be seen that the intrathoracic displacement of the intestinal loops impairs cardiac development and lung often resulting in neonatal death by respiratory insufficiency due to pulmonary hypoplasia and hypertension. Additional comorbidities, such as those cited in this study, may have further aggravated the gestational development of the fetus. It is important to mention that

Figure 2: C) Presentation of Nephromegaly. D) Presentation of cranial splenic torsion. The arrow shows the anatomy structures.



recent advances in fetal surgery have developed new surgical methods for increasing the survival of the newborn especially in relation to pulmonary hypoplasia. [4]

This case illustrates a rare case of diaphragmatic hernia with intestinal changes of clinical and surgical importance. Thus, this report demonstrates the importance of knowledge of diaphragmatic congenital malformations for the surgical planning of perinatal urgency, describing parameters to aid in the repair of acute diaphragmatic lesions using laparoscopy or conventional surgeries, as well as an anatomic-topographic description.

Abbreviations

L: Liver; DH: Diaphragmatic Hernia; IL: Intestinal Loop; S: Spleen; St: Stomach; D: Duodenum; H: Heart; P: Pericardium; LL: Lung Left; LR: Lung Right; T: Thyme; DC - Descending Colon; SC: Sigmoid Colon

Declarations of Interests

The authors declare that they have no competing interests.

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