Congenital Diaphragmatic Hernia Associated with Uncommon Abnormalities

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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 pos-
tero-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 ab-
normalities (i.e. cardiac hypertrophy, several pulmo-
mary hypoplasia, hepatomegaly, nephromegaly 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 ante-
roposterior 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 her-
nia 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, presen-
ting cardiac hypertrophy when compared to fetal
age (Figure 1B).

Additionally, laterally to mediastinum, it is possi-
ble to visualize the pulmonary hypoplasia, charac-
terized more sternly in the left lung, and being still
possible to identify the oblique fissure and the pul-
monary 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 ab-
dominal 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 me-
dial region of the abdominal cavity being possible to
identify the descending and sigmoid colon in anato-
mical position. It is possible to identify the umbilical
vein moving toward the umbilicus, was continued
as the umbilical cord (Figure 2D).

To the best of our knowledge, this is the first
case report which identifies the presence of cardiac
hypertrophy, several pulmonary hypoplasia, hepa-
tomegaly, nephromegaly and cranial splenic torsion
associated to anteroposterior left-side CDH with IM
in a stillborn. In this report we note that the con-
genital diaphragmatic hernia can be seen that the
intrathoracic displacement of the intestinal loops im-
pairs 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 develop-
ment of the fetus. It is important to mention that
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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References


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