ARCHIVES OF HELLENIC MEDICINE: ISSN 11-05-3992

SHORT COMMUNICATION
ΒΡΑΧΕΙΑ ΔΗΜΟΣΙΕΥΣΗ

ARCHIVES OF HELLENIC MEDICINE 2025, 42(3):422–424
ΑΡΧΕΙΑ ΕΛΛΗΝΙΚΗΣ ΙΑΤΡΙΚΗΣ 2025, 42(3):422–424

Diaphragmatic hernia
Challenges of diverse levels

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Διαφραγματοκήλη: Προκλήσεις διαφόρων επιπέδων

Περίληψη στο τέλος του άρθρου

Key words: Diagnosis, Diaphragmatic hernia, Management, Types

Diaphragmatic hernia (DH) is an infrequent condition with a congenital or acquired origin and is characterized by the presence of abdominal structures into the thoracic cavity, such as the stomach, liver, right colon, appendix, gallbladder, omentum, and mesentery; while the hiatal hernia (HH) type is due to gastric prolapse through the hiatal orifice.[1–9] The DHs have been classified as (a) congenital, (b) Bochdalek (on the posterior left side), (c) Morgagni (anterior, retrosternal, or parasternal), (d) eventration (sited in anteromedial portion of the right hemidiaphragm), (e) central tendon defect, (f) acquired, (g) hiatal (with the types I to IV, sliding, and paraesophageal), (h) traumatic, and (i) iatrogenic.[1–9] The estimated incidence of congenital DH (CDH) is up to 5 per 10,000 births; the mortality rate in pregnancy is up to 12.4%,[2] and may be 31% in a DH acquired type.[1] Symptomatic paraesophageal hernias, volvulus, and obstructions require urgent surgery; surgical option is the best choice for severe symptoms unresponsive to other treatments, risk of strangulation, evolution with hemorrhagic ulcers, or esophageal strictures.[1,2,6] Complicated DH often involves large herniation of abdominal organs, incarceration, perforation, strangulation, pulmonary or cardiac compression, and the heart tamponade; whereas laparoscopic management can constitute the best option for the stable patients. Minimally invasive method must be done as early as possible for the newborn with DH.[9] Aiming to highlight the importance of the DH inclusion among differential diagnosis of chest and or high digestive disturbances, data of recent literature are commented on.[1–9]

An 80-year-old male was reported with three months of dysphagia and weight loss, and his chest X-ray besides images of the upper digestive tract showed the stomach within the thorax, characterizing a HH; the endoscopy ruled out esophagitis and cancer.[1] He was promptly referred to another hospital with better technical resources for the adequate specialized management, which is recommended in order to get successful outcomes.[1] A review conducted from 1911 to 2020, including 158 cases of maternal DH in pregnancy, found similar number of reports in the last two decades, with an average maternal age of 28.4 years, an increased rate of CDH (82.0%), and the majority of DH in the left-side (n=124).[2] Most of cases (44.0%) had one herniated organ, followed by three (22.0%), two (21.3%), and four or more (12.7%), while the majority of DHs had received surgical treatment (75.3%).[2] The authors stressed that manifestations of DH as abdominal pain, with dyspnea and chest pain, may be mistaken for common thoracic disorders, constituting diagnostic challenges that enhance the maternal and fetal mortalities, mainly when are followed by a collapse.[2] A 36-year-old man with antecedent of accidental trauma 18 years before was evaluated because of abdominal pain, and the imaging studies revealed a large right-sided DH, with liver, right colon, appendix, gallbladder, omentum, and the mesentery within the thorax.[3] He underwent a transabdominal robotic procedure to correct the voluminous DH with success, and persisted without symptoms during close clinical and imaging follow-up.[3] The authors cited the small number of reports about robotic repairs to correct CDHs, besides diaphragm eversion and Valsalva maneuver for reductions of herniated organs.[3] An 18-year-old man with non-well controlled type 1 diabetes mellitus and mild asthma seeked emergency care because

Submitted 29.1.2024
Accepted 24.2.2024

of copious vomiting and diffuse abdominal pain.[4] Three years before, he had suffered an accident causing a femur fracture that was surgically managed, and current review of the chest X-ray images obtained eight months prior this admission revealed an undiagnosed small DH sited in the left pulmonary base.[4] The authors commented on the challenges of diagnostic hypothesis involving a DH in this adolescent with manifestations of diabetic ketoacidosis, who might have a unsuspected CDH, besides the antecedent of a vehicle accident eventually causing a traumatic DH; therefore, the diagnosis can be missed or delayed, enhancing the morbidity and mortality.[4] Retrospective study about survival of 225 infants with diagnosis of CDH in England from January 2000 to December 2020 focused on the side of defect, the hemodynamic and ventilatory procedures, prenatal diagnosis, related anomalies, birth weight, and gestation.[5] In 66% of cases the diagnosis was prenatal; the survival rate was 60% (134/225), with postnatal survival of 68% (134/198 liveborn), and post repair survival of 84% (134/159). The survival rate improved in comparison with the number of deaths in a prior decade, and the need for complex ventilatory procedures was the major predictor of the deaths.[5] A study in Korea about data from 1994 to 2018 of 26 patients with thoracic (n=15) or abdominal DH repairs, while 5 of them underwent video-assisted thoracic surgery (VATS).[6] The thoracic procedures were related with a longer duration of DH than the abdominal ones (2 versus 0.1 months), with herniation of right-sided abdominal and retroperitoneal organs; the patients presented no DH recurrence during the median follow-up of 23 months, and the authors commented on the role of the VATS as a good option for the DH repairing.[6] Isolated and complex CDH types were compared in England from 2002 to 2018 about the birth prevalence, maternal age, sociodemographic factors, and the one-year mortality rate.[7] It is noteworthy that the prevalence of isolated and complex CDHs were 1.4 and 1.2 per 10,000 livebirths, respectively; compared with maternal ages between 25–34 years, the complex CDH risk was higher for maternal ages <20 years; and the one-year mortality rate for the complex CDHs was slightly higher than for the isolated CDHs (33.1 versus 29.7%).[7] A Brazilian evaluation was done in São Paulo from 2004–2015 concerning the prevalence, neonatal mortality and lethality of CDH, and time to CDH-associated neonatal death included all live births with the gestational age ≥22 weeks, and birthweight ≥400 g.[8]

The CDH prevalence was 1.67 per 10,000 live births, with a significant increase in the period, as occurred with the neonatal mortality, but not with the lethality of near 79.0%; for CDH of all subgroups, half of these deaths occurred during the first day after birth.[8] The authors commented on the significant increase of CDH prevalence and neonatal mortality with an elevated lethality in comparison with data from high income countries.[8] A 20-year-old female presented abdominal pain during 10 days and the imaging studies revealed part of intestinal cavity with gas, besides the spleen, stomach, and portion of the bowel within the left thoracic cavity due to a large posterolateral diaphragmatic defect.[9] Although with the established diagnosis of a congenital DH, she was not submitted to the surgical repair of the diaphragmatic defect during two years of a regular follow-up; but this procedure was indicated to be done before her decision about future pregnancies.[9] The authors highlighted that CDH is among the most ominous birth defects and associated with elevated neonatal mortality rate, and the repair should be performed with surgical methods and techniques more adequate to avoid the elevate frequency of recurrences.[9]

Concluding, the variety of DH types and unspecific clinical manifestations since early infancy to elderly groups of patients pose diverse challenges to establish prompt diagnosis and adequate procedure for repair and prevention of the common recurrences. Additional concern is on the insufficiency of necessary resources in low-income regions.

ΠΕΡΙΛΗΨΗ

Διαφραγματοκήλη: Προκλήσεις διαφόρων επιπέδων

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Η διαφραγματοκήλη είναι μια ασυνήθιστη κατάσταση που χαρακτηρίζεται από την παρουσία κοιλιακών δομών στη θωρακική κοιλότητα λόγω συγγενούς ή επίκτητου ελαττώματος. Η εκτιμώμενη επίπτωση της συγγενούς διαφραγματοκήλης είναι έως και 5 ανά 10.000 γεννήσεις, ενώ το ποσοστό θνησιμότητας ποικίλλει σύμφωνα με την ταξινόμηση και μπορεί να φθάνει έως και το 31% στην επίκτητη διαφραγματοκήλη. Η διάγνωση εξαρτάται από το επίπεδο κλινικής ευαισθητοποίησης και τις απεικονιστικές μελέτες. Οι επιπλοκές της διαφραγματοκήλης μπορεί να περιλαμβάνουν την κήλη των κοιλιακών οργάνων, τον εγκλεισμό, τη διάτρηση, τον στραγγαλισμό, την πνευμονική ή την καρδιακή συμπίεση και τον επιπωματισμό της καρδιάς. Συνιστώνται ελάχιστα επεμβατικές διαδικασίες για τη θεραπεία νεογνών με

διαφραγματοκήλη και η λαπαροσκοπική αντιμετώπιση θεωρείται επαρκής επιλογή για ασθενείς κλινικά σταθερούς. Στόχος είναι η ενίσχυση της υποψίας των εργαζομένων στον τομέα της υγείας για την παρουσία διαφραγματοκήλης.

Λέξεις ευρετηρίου: Αντιμετώπιση, Διάγνωση, Διαφραγματοκήλη, Τύποι

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