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ORIGINAL RESEARCH

Congenital diaphragmatic hernia: clinical and hospital aspects in a maternal and child reference hospital in the Amazon region

Hérnia diafragmática Congênita: aspectos clínico-hospitalares em um hospital de referência materno-infantil na região Amazônica

Hernia diafragmática congénita: aspectos clínicos y hospitalarios en un hospital de referencia materno-infantil de la Amazonía

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ABSTRACT | The aim of this study is to analyze the clinical and hospital profile of newborns diagnosed with Congenital Diaphragmatic Hernia (CDH) from 2008 to 2018 in a maternal and child reference hospital. It is a cross-sectional and descriptive analytical study, which mainly included data from medical records with the codes Q79.0, Q79.1, J98.6, K44 and K44.0 of the International Classification of Diseases (ICD-10). The exclusion criteria of medical records were the diagnosis of other types of diaphragmatic hernia, without the congenital aspect; hospitalization in units that were not Neonatal ICUs; and the age at admission equal to or greater than 29 days. A total of 25,602 records were analyzed, of which 14 corresponded to CDH. The gender involvement was 71.43% male (10 cases) and 28.57% female (4), with 21.34% of Bochdalek-type CDH located on the right (3) and 78.57% on the left (11). All newborns in this study underwent invasive mechanical ventilation (IMV) 9.21 ± 5.55 days. Surgery was indicated in 11 cases (78.57%), with thoracotomy access routes in four (36.36%) and subcostal laparotomy in seven (63.64%), all using a chest tube homolateral to the hemithorax. The total hospital stay was 19.42 ± 15.36 days. There was an improved discharge evolution in eight patients (57.14%) and the death of six (42.86%), with ages around 6.19 ± 4.79 days, with no follow-up of neuropsychomotor development afterwards. There were low incidences of cases per year, the gender involvement, the associated malformations and the IMV time were similar to other populations in the world.

Keywords | Congenital Diaphragmatic Hernias; Newborn; Physiotherapy.

RESUMO | O objetivo deste estudo é analisar o perfil clínico-hospitalar de neonatos com o diagnóstico de hérnia diafragmática congênita (HDC), no período de 2008 a 2018 em um hospital de referência materno-infantil. Trata-se de um estudo transversal e analítico descritivo, que incluiu principalmente dados de prontuários com os códigos Q79.0, Q79.1, J986, K44 e K44.0 da Classificação Internacional de Doenças (CID-10). Os critérios de exclusão de prontuários foram o diagnóstico de outros tipos de hérnia diafragmática, sem o aspecto congênito; a internação em unidades que não fossem UTI Neonatal; e a idade no ato de internação igual ou superior a 29 dias. Analisou-se um total de 25.602 prontuários, dos quais 14 corresponderam a HDC. O acometimento por gênero foi de 71,43% masculino (10 casos) e 28,57% feminino (4), com 21,34% das localizações de HDC tipo Bochdalek à direita (3) e 78,57% à esquerda

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(11). Todos os neonatos deste estudo passaram por ventilação mecânica invasiva (VMI) 9,21 ± 5,55 dias. A cirurgia foi indicada em 11 casos (78,57%), com vias de acesso por toracotomia em 4 (36,36%) e por laparotomia subcostal em 7 (63,64%), todos com uso de dreno torácico homolateral ao hemitórax. O tempo total de internação foi de 19,42 ± 15,36 dias. Observou-se a evolução de alta melhorada em oito pacientes (57,14%) e o óbito de seis (42,86%), com idade de 6,19 ± 4,79 dias, sem referência de acompanhamento do desenvolvimento neuropsicomotor posteriormente. Ocorreram baixas incidências de casos por ano, e o gênero de acometimento, as malformações associadas e o tempo de VMI foram semelhantes a outras populações no mundo.

Descritores | Hérnias Diafragmáticas Congênitas; Recém-Nascido; Fisioterapia.

RESUMEN | El objetivo de este estudio es analizar el perfil clínico-hospitalario de neonatos diagnosticados con hernia diafragmática congénita (HDC) en el período de 2008 a 2018 en un hospital de referencia materno-infantil. Este es un estudio transversal, analítico-descriptivo, realizado con base en datos de historias clínicas con los códigos Q79.0, Q79.1, J986, K44 y K44.0 de la Clasificación Internacional de Enfermedades (CIE-10). Los criterios de exclusión de las historias clínicas fueron el diagnóstico de otros tipos de hernia diafragmática sin el aspecto congénito; hospitalización en unidades que no sean UCI neonatales; y edad de ingreso igual o mayor a 29 días. Se analizaron un total de 25.602 registros, de los cuales 14 correspondían a HDC. Entre el género afectado, el 71,43% fue el género masculino (10 casos) y el 28,57% el femenino (4), con un 21,34% de las ubicaciones de la HDC de tipo Bochdalek a la derecha (3) y un 78,57% a la izquierda (11). Todos los recién nacidos en este estudio estuvieron bajo ventilación mecánica invasiva (VMI) por 9,21±5,55 días. La cirugía estuvo indicada para 11 casos (78,57%), con vías de acceso por toracotomía para 4 (36,36%) y laparotomía subcostal para 7 (63,64%), todos con sonda torácica homolateral al hemitórax. La estancia hospitalaria total fue de 19,42±15,36 días. Se observó una mejor evolución del alta en ocho pacientes (57,14%) y muerte de seis (42,86%) con 6,19±4,79 días de edad, sin referencia de seguimiento del desarrollo neuropsicomotor posteriormente. Hubo baja incidencia de casos por año; y el género que acomete, las malformaciones asociadas y el tiempo de VMI fueron similares a otros estudios de la literatura.

Palabras clave | Hernias Diafragmáticas Congénitas; Recién Nacido; Fisioterapia.

INTRODUCTION

The congenital diaphragmatic hernia (CDH) is a defect characterized by the presence of abdominal organs within the fetal thoracic cavity, with a prevalence of 1:2,000 pregnancies and an incidence of 0.8-5:10,000 live births. CDH represents about 8% of the malformations seen in newborns (NB), and may be associated with other malformations, such as cardiac, neural tube, chromosomal or renal abnormalities[1,2].

The pathology comes from the non-evolution of the diaphragmatic membrane, which has its normal development initiated between the fourth and eighth week of gestation. This disorder has postnatal consequences, such as pulmonary hypoplasia and pulmonary hypertension, in addition to abnormal pulmonary vasculature morphology. CDH culminates in indicators of high morbidity and mortality around 50-60%, and individuals with this condition require intensive specialized care[3,4].

The treatments are marked by several aspects, prenatal and postnatal, such as the use of drugs, surgical approaches and ventilatory assistance strategies, for systemic and survival improvement[5].

Regarding national research, Brazil has few studies that address CDH, both in population and clinical characteristics, a fact that may be associated with the geographic dimension of the country and the disparity in the distribution of resources and technology. The Pará state, incorporated into the Amazonian scope, is made up of social, cultural and geographical particularities, whose representativeness is currently low compared to the national and international panorama. Furthermore, the characterization of the pathology in the region is important for the description of clinical approach processes in the north of the country, showing possibilities of change and adaptation to the context.

Given the above, the purpose of this research was to describe the clinical and hospital profile of newborns diagnosed with CDH in a reference hospital in the Pará state in maternal and child health care from 2008 to 2018.

METHODOLOGY

This is a cross-sectional and descriptive analytical study, with data from medical records with the diagnosis of CDH in the period from January 1st, 2008 to December 31st, 2018.

The sample consisted of medical records from the neonatology sector filed with the Patient Data Management of the Santa Casa de Misericórdia do Pará Foundation (FSCMP), being characterized as non-probabilistic for convenience. All Neonatal ICU (NICU) medical records with the following codes of the International Code of Diseases (ICD-10) were included for analysis: Q79.0 (congenital diaphragmatic hernia); Q79.1 (other congenital malformations of diaphragm); J98.6 (disorders of diaphragm); K44 (diaphragmatic hernia) e K44.0 (diaphragmatic hernia with obstruction, without gangrene). The codes not directly correlated with the disease were used to reduce the possibility of sample loss, since there was the use of secondary data over a long period of time. The medical records of newborns hospitalized in other hospital units that were not NICUs, ages between 29 days or older and/or incomplete description of data, were excluded.

A clinical and hospital characteristics sheet was used, organized by the authors, according to: alphanumeric identification code, need for ICU admission, gestational age, corrected age, gender, ethnicity, origin, complications during childbirth, associated malformations, systemic pulmonary hypertension, appearance of complications, need for invasive ventilatory support (IMV), non-invasive mechanical ventilation (NIMV), supplemental oxygen support, as well as the duration of use of each therapy.

The existence and type of surgical approach, the age of the newborn in the approach, surgical and care complications, the vital status of hospital discharge, the total length of hospital stay, age and weight during hospital discharge and the monitoring of development was also evaluated.

For statistical analysis, categorical variables were presented as frequencies and numerical variables were presented using measures of central tendency (mean) and dispersion (standard deviation). In the inferential statistical analysis, the chi-squared test ($\chi 2$) was used to analyze a sample. For the analysis of two independent samples, the G-test (Contingency) was used. All statistical processing was performed using both Epi Info 3.5.1 and BioEstat 5.3 software, considering an alpha significance level of 5% (p-value ≤ 0.05).

RESULTS

From the timeline, a total of 25,602 records were obtained from the FSCMP Patient Data Management, with an average of 2,327 per year. Of this total, 14 records complied with the research requirements, 12 (85.7%) presenting the ICD-10 code Q79.0 and two (14.3%) presenting the K44.0 code. There were no medical records with codes Q79.1, J98.6 and K44 in neonatal hospitalization units, nor were their ages more than 28 days at the time of admission or incomplete medical records. In such manner, the research followed without exclusions.

Table 1. Main neonatal characteristics from 2008 to 2018, (N=14). Belém, Pará, 2019

Neonatal characteristics	Mean and standard deviation
Age at admission	4,50 ± 5,22 days
Gestational age (Capurro method)	37,78 ± 2,19 weeks
Weight (grams)	2.894,64 ± 516,30
APGAR 1 min	5,5 ± 2,3
APGAR 5 min	7,9 ± 1,4
Region of origin	N (%)
Metropolitan mesoregion of Belém	7 (50%)
Northeast mesoregion of Pará state	4 (28,57%)
Southeast mesoregion of Pará state	2 (14,28%)
Mesoregion of Marajó	1 (7,14%)

Approximately 50% of the medical records came from the metropolitan region of Belém, with an admission age of 4.50 ± 5.22 days and a gestational age of 37.78 ± 2.19 weeks using the Capurro method (Table 1). The proportions of involvement and the characteristics of gender were: 10 (71.43%) males and four (28.57%) females, corresponding to 2.5:1, without statistical significance (p=0.18). The hernia locations were reported to be of the Bochdalek-type, with three (21.43%) on the right and 11 (78.57%) on the left (Figure 1).

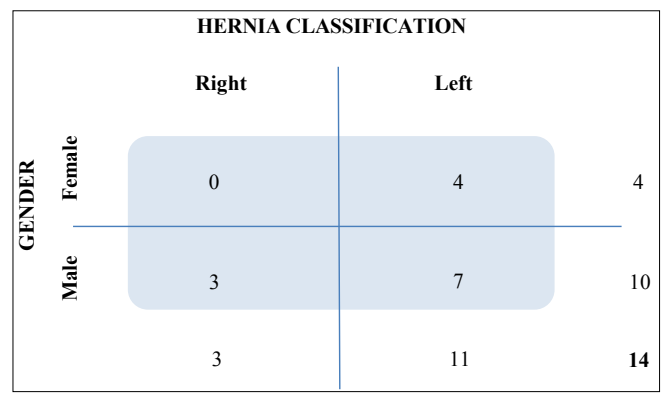

Figure 1. Classification of congenital diaphragmatic hernia according to gender and involvement characteristic, (N=14). Belém, Pará, 2019

The associated malformations found were three cases of pulmonary hypoplasia, one case of congenital heart disease and two cases of suspected Down syndrome (DS), being described in this study as chromosomal abnormalities. All newborns in this study underwent orotracheal intubation with an average exposure of 9.21 ± 5.55 days. When the association between IMV time and extubation using the G-test was tested, the result was not statistically significant (p=0.09). Therefore, there is no association between the variables analyzed.

Treatment with NIMV as part of post-surgical ventilatory weaning occurred in six cases (42.86%), accounting for 0.92 ± 1.32 days of therapy. Oxygen therapy was used in 10 cases (71.43%), with an average day of 3.38 ± 3.98.

In this series of cases (Table 2), three died (21.43%) in the group of 14 newborns during pre-surgical stabilization, all of whom had CDH on the left, two were male and one was female. Corrective surgery was indicated in 11 cases (78.57%), with ages between 7.35 ± 6.23 days at the time of repair.

Table 2. Main clinical and hospital characteristics from 2008 to 2018 (N=14), Belém, Pará, 2019

Variables	
Surgical characteristics	Mean and standard deviation (N=11)
Age at surgery	7,35 ± 6,23 days
Age at death	8,66 ± 6,11 days
Operation type	N=11 (100%)
Subcostal laparotomy	7 (63,64%)
Thoracotomy	4 (36,36%)
Deaths	N=11 (100%)
Pre-surgical	3 (21,43%)
Post-surgical	3 (21,43%)
Hospital discharge characteristics	N=14 (100%)
Improved hospital discharge	8 (57,14%)
Death	6 (42,86%)
Total hospitalization time	19,42 ± 15,36 days

The surgical access routes were four (36.36%) by thoracotomy and seven (63.64%) by subcostal laparotomy, with a chest tube homolateral to the hemithorax inserted with the diaphragmatic defect. In addition, there were three (21.43%) post-surgical deaths, all male newborns, ages between 8.66 ± 6.11 days, two with CDH on the right and one on the left.

The total hospitalization time for these newborns was 19.42 ± 15.36 days. An improved discharge evolution was observed in eight (57.14%) patients and death in six (42.86%), ages between 6.19 ± 4.79 days. There was no reference in the medical records for monitoring of neuropsychomotor development after surgical intervention and/or hospital discharge.

DISCUSSION

The gestational age and weight of the group were equal to other populations around the world, such as those found in a study with 119 newborns in Sydney (Australia), which obtained a gestational age of 37.9 ± 2.5 weeks and a weight of 2.990 ± 650g[6].

In another study, with 330 patients in the city of Philadelphia (United States of America), aiming at a comparative analysis of the CDH characteristics on the right versus the left side, a gestational age of 37 weeks was obtained for the group with defect on the left and 36 weeks for the group on the right. In this study, it was highlighted that the birth weight of newborns with a defect on the left was significantly higher[7].

As for the characterization with the involvement frequency of the defect to gender, greater expressiveness was obtained in males (71.43%), without a statistical significance standard (p=0.18) expected (p-value≤0.05). The data corroborates the characterization of live children with CDH from 28 pediatric surgical centers in the United Kingdom and Ireland, where 1.5 times more births of males with CDH were observed in relation to the females[8].

These data are also equivalent to those obtained by Brazilian authors[9] who, through analysis of CDH deaths in a reference service in pediatric surgery in the southern region of Brazil, obtained 42 (60%) male cases, in a total of 70 patients.

The prevalence of posterolateral hernia (Bochdalek) – which is verified in this study from the occurrence in 100% of the analyzed newborns, with three (21.43%) on the right and 11 (78.57%) on the left, with a predominance of CDH on the right in males – it is in the same manner as other studies, in which there is a greater incidence of hernias with posterolateral aspect of 70-75%, with a proportion of 85% left, 13% on the right and 2% bilateral[10]. This fact is reiterated with the finding of 56 (17%) of CDH on the right, 31 (51%) of whom are males[7].

Regarding pulmonary hypertension (PH) and pulmonary hypoplasia, these are intricately linked to the prognosis of morbidity and mortality. Based on a multicenter risk scale assessment for mortality in newborns with PH with seven hospitals in six Asian countries, it was concluded that PH and hypoplasia are significant aggravating factors, which increase the risk of neonatal mortality[11].

PH and the 0-24-month-old respiratory clinic in children with CDH concluded that PH complications and breathing difficulties tend to decrease with time and the child's development. There is a low frequency of worsening of PH after 6 months of age, with no significant association with the need for new hospitalizations[12].

Cardiac malformations and pulmonary hypoplasia are common findings in CDH, arising from characteristics of the pulmonary vasculature in this injury, such as the thickening of the vessels in the middle and adventitial layers, the presence of a hypoplastic vascular bed with reduced arborization and the unconventional response of the pulmonary artery/arteriole to physiological and pharmacological signs, soon culminating in cardiac overload[13,14].

In this study, we have the result of two cases of suspected DS associated with CDH, both of which evolved to death. From a fourteen-year survey to characterize the risks of neonatal mortality in the population with DS[15], it was concluded that greater deaths occurred in the group of DS with CDH. Furthermore, the study highlights that the diagnosis of CDH alone is already a predictor of high mortality.

Regarding corrective surgery, its indication was observed in 11 (78.57%) cases, ages between 7.35 ± 6.23 days at the time of repair. This time for correction is contrary to what it was concluded with the population of Belgrade, in southeastern Europe, where 29 (69%) newborns underwent correction, mainly on the second, third and fourth day of life[16]. In a study with a population in the southern region of Brazil, the data also differs from the findings in this study: 26 (45.6%) newborns were operated on the first day and 31 (54.4%) from the second to the fourth day of life[9].

When assessing the influence of time for surgical correction of CDH with 477 patients, it was concluded that the time of repair does not seem to affect survival in 90 days, regardless of the severity of the defect's involvement. However, it is noteworthy that patients with "moderate" severity can benefit from surgery in 48 hours, reducing the duration of ventilation, oxygen and hospitalization[17].

As for the surgical technique used, it is clear that in this study, all individuals underwent interventions for correction with open access route via thoracotomy (n=4, 36.36%) and subcostal laparotomy (n=6, 63.64%), these being the most common operations[10,18].

The use of a chest tube homolateral to the hemithorax with the diaphragmatic defect was observed in all individuals participating in this study. This practice is contraindicated as a postoperative routine by the European Consensus, since the pleural effusion after the repair is quick to fill, in which the drain promotes contamination of the pleural space without benefits and, also, culminates in the acceleration of the ipsilateral lung expansion. Nevertheless, its evaluation should be careful and indicated only after the surgical procedure in cases of impaired pulmonary function and ventilation[19].

The clinical outcomes obtained with a prevalence of eight (57.14%) patients with improved hospital discharge evolution and six (42.86%) deaths, similar to the population observed in the southern region of Brazil, with 38 (54.6%) survivors and 36 (45.7%) deaths[9]. Internationally, there are studies that oppose this average, with a variation of deaths of 20.1% in six Neonatal ICU centers[11]. It can be inferred that the possibilities of diagnosis, prenatal care, quality of the reception structure and immediate assistance in these cases are decisive points for the numerical differences.

There was no monitoring of child development after hospital discharge, a fact that is alarming in terms of long-term surveillance, since they present risks of respiratory complications arising both from pulmonary hypoplasia and PH, as well as from injuries resulting from surgery, aggressive IMV, gastroesophageal reflux, risks of motor and osteomyoarticular deficit.

As already mentioned, there is a close embryological relationship between the lungs, the rib cage and the diaphragm. Thus, within the pathophysiology of the injury, it is reasonable to expect deformities of the chest wall and the thoracic spine[20].

The characterization regarding the frequency and severity of scoliosis, chest and abdominal wall deformities after surgical repair was carried out in a study from 1989 to 2012. It concluded that scoliosis and chest deformity were common in children with large CDH, and that the surgical technique did not appear to affect the incidence of skeletal deformities later[21]. Thus, this is a point of observance for health professionals when in contact with this audience for evaluation.

In reference to the total length of hospital stay, the result was a total of 19.42 ± 15.36 days, different from

what was found in other studies, of 12 days[11] and 33 ± 24 days in the group without the use of nitric oxide (iNO), 67 ± 40 days in the iNO-responsive group and 32 ± 32 days in the non-iNO-responsive group[22].

The authors point out that the need for advanced stabilization supports, such as iNO, and the use of a mesh for the repair of the diaphragmatic defect influence the increase in ICU stay, being a factor of the worst expressions of the child's global development[23].

This study had important limitations regarding the search by ICD-10 for a small quantitative sample, even with the inclusion of other ICD-10 that did not correspond directly to the pathology in its congenital form. The fact raises the assumption of the existence of registration errors during the birth and hospitalization of patients, such as the absence of alteration of the ICD-10 even after the correct identification of the underlying and/or associated pathology.

It is also exposed, as a limiting factor, the survey carried out in a single maternal and child care center, stimulating the possibility of reaching new research in other reference centers for the same public, both in the north and in other regions of Brazil.

CONCLUSION

Low frequencies of cases registered per year were observed, as well as involved gender aspects and associated malformations remarkably similar to other studies around the world.

Regarding the approach therapy for treatment, the majority use of IMV was observed. The clinical outcomes of the cases were along the lines of those observed in the southern region of Brazil, and the total length of hospital stay differed from all studies found – it is assumed that the main reasons for this fact are the differences in care support and the severity of the pathology involvement.

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