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Bilateral congenital diaphragmatic hernia: prognostic evaluation of a large international cohort☆

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ABSTRACT

Background: Congenital diaphragmatic hernia (CDH) is a lethal birth defect, which occurs in 1:2000–3000 live births. Bilateral CDH is a rare form (1%), with a high mortality. This study presents the outcomes of the largest cohort of bilateral CDH patients.
Methods: The records of patients with bilateral CDH from the Congenital Diaphragmatic Hernia Registry born between 1995 and 2015 were retrospectively analyzed to identify parameters associated with mortality.
Results: Eighty patients with a bilateral CDH were identified. Overall mortality was 74% (n = 59). Apgar scores at 1 and 5 min were statistically lower in the non-survivors compared to the survivors (median 3.0 and 5.0, versus 6.5 and 8.0, respectively, $p < 0.001$). All survivors were repaired (n = 21), compared to 22% of the non-survivors (n = 17). The type of repair was equally divided in the survivors (52% primary versus 48% patch), while non-survivors were mainly patch repaired (82% versus 12%). Nineteen were treated with extracorporeal membrane oxygenation (ECMO) (24%), only three of them survived. When calculating the risk on mortality for the patients who lived until repair, ECMO had an adjusted odds ratio for mortality of 10.8 (95% CI: 2.0–57.7) and patch repair 5.2 (95% CI: 0.8–34.9).
Conclusions: The treatment of bilateral CDH patients remains challenging with a high mortality rate. Lower Apgar-scores, ECMO (probably as a surrogate for the severity of disease), and patch repair were negatively associated with outcome.
Level of evidence: Level IV study

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Congenital diaphragmatic hernia (CDH) is a major birth defect, which occurs in approximately 1 in 2000–3000 births [1,2]. Bilateral CDH is a variation of CDH which is extremely rare and accounts for approximately 1–2% of all CDH patients [2–4]. The prenatal diagnosis of bilateral CDH poses a challenging task. First of all, the mediastinal shift is often minimal, which, is in unilateral CDH, often the first manifestation on prenatal ultrasound [5,6]. A further challenge in prenatal diagnosis is that on the right side of the bilateral CDH, the liver and lung have a similar echogenicity [3].

Pulmonary hypoplasia combined with abnormal morphology of the pulmonary vasculature leads to severe pulmonary hypertension and consequently respiratory insufficiency in more than 90% of the CDH patients in the first hours after birth [7]. Despite advances in the postnatal care of patients with CDH, pulmonary hypoplasia and subsequent pulmonary hypertension remain a major cause of neonatal death [6]. In bilateral CDH the unpredictable nature of the pulmonary hypoplasia and pulmonary hypertension is expected to be more detrimental compared to unilateral CDH. Furthermore, a higher prevalence of major congenital anomalies has been reported for bilateral compared to unilateral CDH and could play a role in the higher mortality in bilateral CDH [4,6,8]. The mortality rate of bilateral CDH patients reported in literature is 65%, compared to 20–35% for unilateral CDH [4,8].

Unfortunately, literature on bilateral CDH is limited, and only some case reports and small retrospective series have been published [2–4,6,9,10]. The aim of this study was to present the outcomes of the largest cohort of bilateral CDH patients, from the international Congenital Diaphragmatic Hernia Registry.

Abbreviations: CDH, congenital diaphragmatic hernia, ECMO: Extracorporeal membrane oxygenation; CDHSG, Congenital Diaphragmatic Hernia Study Group; OR, Odds ratio, 95% CI: 95% confidence interval.

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1. Methods

1.1. Patients

All patients with a bilateral CDH in the Congenital Diaphragmatic Hernia Registry (CDHR) from January 1995 to July 2015 were included. The international CDH registry holds records from centers participating in the registry of all live born patients born with CDH from 1995 onwards [11]. Patients in the registry with a unilateral CDH were excluded. This cohort study is a retrospective review of data that has been collected using the structured Congenital Diaphragmatic Hernia Study Group (CDHSG) forms, which are completed per patient by each of the participating centers.

The international CDHSG registry is a voluntary registry of more than 8800 live born infants with CDH from more than 60 international institutions. Institutional review board approval has been obtained from the University of Texas Health Science Center at Houston (HSC-MS-03-223). Informed consent was not required as all data in the CDHSG registry are de-identified.

1.2. Data collection

Patients were evaluated on a number of parameters including demographics, comorbidity, medical treatment, surgical treatment, and outcome. The primary outcome measure was survival, with the need for oxygen at 30 days and duration of hospital stay as secondary outcome measures.

1.3. Statistical analysis

The results are presented in the tables as median values with 5–95% range, because of the skewness of most data. The outcome parameters were compared between survivors and non-survivors, using the Mann–Whitney *U* test for continuous variables. For comparison of categorical data between the patient's cohorts, the chi-square or Fisher exact test were used as appropriate. A p-value of <0.05 was considered statistically significant. The odds ratio (OR) for mortality was estimated for repair type and treatment with extracorporeal membrane oxygenation (EMCO) using logistic regression analyses. Independent ORs were derived by adjusting the mortality risk for repair type for ECMO treatment and vice versa. All statistical analyses were performed using IBM SPSS Statistics 22.

2. Results

Between 1995 and 2015, 8800 patients were included in the CDH registry, of which 80 patients with a bilateral CDH (0.9% of the total CDH patients). The mortality of this cohort was 74% (n = 59). The prenatal characteristics and basic demographics are presented in Table 1. The non-survivors were significantly more often prenatally diagnosed (78% versus 50%, p = 0.02), however it was not stated whether the diagnosis of CDH was unilateral or bilateral. Corresponding with the higher prenatal diagnosis, there were also more inborn patients in the non-surviving group (p = 0.003). The Apgar scores at 1 and 5 min differed statistically significant between the two groups ($p < 0.001$), with lower Apgar scores in the non-survivors.

Nine patients (11%) were diagnosed with a chromosomal abnormality, of which three had Fryns syndrome and none of them survived. The two main cardiac anomalies were coarctation of the aorta and pentalogy of Cantrell, which were each reported in 7 patients. Ventricle septum defect was present in 16% and atrium septum defect in 13%, with 8% having a combination of both. No statistical significance between types of other congenital anomaly and mortality were found, although there were more than twice as many omphalocele in the survivors group compared to the non-survivors.

The method of repair of the diaphragmatic defect can be primary or with patch depending on the defect size. Just more than half of the patients in the surviving group had a primary repair, suggesting a smaller defect, while this was only possible in 11% of the non-survivor group (p = 0.009) (Table 2). Defect size according to the CDHSG scale [12] was only stated in three patients, therefore no comparisons could be made based on defect size. The median DOL at repair with a patch (and therefore a larger defect) of the survivors (n = 10) was more than eleven days later than for the non-survivors (n = 12), however this was not statistically significant (median (5–95% range): 15.5 (1–37) versus 4.0 (0–18), p = 0.12 Mann–Whitney *U* test).

The majority of non-surviving patients died soon after birth, during the postnatal attempt to stabilization prior to delay surgery (27/59 in first 24 h and another 8/59 in second 24 h). Those who survived that first critical period of three days were generally repaired. Five patients (8.5%) died after three months, which shows that late mortality is rather

Table 1
Baseline perinatal characteristics of patients with bilateral CDH in the CDH registry.

	Total (N = 80)	Survivors (N = 21)	Non-survivors (N = 59)	P-value
Prenatal diagnosis	71% (55/78)	50% (10/20)	78% (45/58)	0.02
In hospital born	56% (45/80)	29% (6/21)	66% (39/59)	0.003
Male	58% (46/80)	62% (13/21)	56% (33/59)	0.63
GA at birth (weeks)				
Median (5–95% range)	37 (30–41)	38 (32–41)	37 (29–40)	0.06
Birth weight (kilogram)				
Median (5–95% range)	2.72 (1.45–3.48)	2.92 (1.53–3.48)	2.55 (1.33–3.52)	0.03
Apgar- 1				<0.001
Median (5–95% range)	3.0 (1.0–8.0)	6.5 (2.0–8.0)	3.0 (1.0–8.0)	
Apgar- 5				<0.001
Median (5–95% range)	6.0 (1.0–9.0)	8.0 (3.1–9.0)	5.0 (1.0–8.0)	
Chromosomal abnormality	11% (9/80)	10% (2/21)	12% (7/59)	1.00
Congenital cardiac anomalies	46% (37/80)	48% (10/21)	46% (27/59)	0.88
Minor	11% (9/80)	14% (3/21)	10% (6/59)	0.88*
Major	35% (28/80)	33% (7/21)	36% (21/59)	
Other congenital anomalies	44% (35/80)	57% (12/21)	39% (23/59)	0.15
Omphalocele	16% (13/80)	29% (6/21)	12% (7/59)	0.09
Intubation timing				0.001
At day 0	91% (72/79)	70% (14/20))	98% (58/59)	
At day 1 or later	9% (7/79)	30% (6/20)	2% (1/59)	

All continuous variables are displayed as median (5–95% range) and differences tested with the Mann–Whitney *U* test. Categorical data are reported as percentage (n) and using Pearson's chi square test or Fisher exact test when appropriate for calculation of statistical differences.
GA = gestational age.
* When comparing the three groups regarding major, minor or no congenital cardiac abnormalities.

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