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Incidence of Congenital Diaphragmatic Hernia in Olmsted County, Minnesota: A Population-Based Study

Jason M. Woodbury, MD, Katarina Bojani , MD, Ruža Grizelj, MD, PhD, Alexandre N. Cavalcante, MD, Vinay K. Donempudi, MD, Toby N. Weingarten, MD, Darrell R. Schroeder, MS, and Juraj Sprung, MD, PhD

Department of Anesthesiology and Perioperative Medicine (Drs Woodbury, Cavalcante, Weingarten, and Sprung, and Dr Donempudi), and Division of Biomedical Statistics and Informatics (Mr Schroeder), Mayo Clinic, Rochester, Minnesota; Division of Neonatology, Department of Obstetrics and Gynecology (Dr Bojani), University Hospital Merkur, Zagreb, Croatia; and Department of Pediatrics (Dr Grizelj), University of Zagreb, School of Medicine, University Hospital Centre Zagreb, Croatia.

Abstract

Objective—The rate of congenital diaphragmatic hernia (CDH) varies, but most reports estimate the incidence to be less than 3 per 10,000 births. Our objective was to document the incidence of CDH in a geographically well-defined population using available resources for highly accurate incident case ascertainment.

Methods—We ascertained CDH cases in Olmsted County, Minnesota, United States, from 1981 through 2014 using resources of the Rochester Epidemiology Project. Overall and sex-specific incidence rates were calculated, and hospital survival was assessed.

Results—A total of 26 incident CDH cases were identified; the overall incidence of 3.6 (95% CI, 2.2–5.0) per 10,000 did not differ significantly over the 34-year study period ($P=.28$). The estimated incidence was 4.3 (95% CI, 2.2–6.5) for male infants and 2.9 (95% CI, 1.1–4.6) for female infants. The percentage of cases diagnosed prenatally was 33% from 1981 through 2000 and 50% from 2001 through 2014. The rate of survival to discharge in the 2 periods was 50% and 88%, respectively.

Conclusion—The incidence of CDH in Olmsted County exceeds the majority of published rates, which most likely can be attributed to our comprehensive case ascertainment.

Keywords

antenatal diagnosis; congenital diaphragmatic hernia; incidence; survival

Reprints: Juraj Sprung, MD, PhD, Department of Anesthesiology and Perioperative Medicine, Mayo Clinic, 200 First St SW, Rochester, MN 55905 (sprung.juraj@mayo.edu).

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Introduction

Congenital diaphragmatic hernia (CDH) is a rare anomaly with a high associated mortality rate (1). According to the US Centers for Disease Control and Prevention, the incidence is 2.6 per 10,000 births (2). Data from the European Surveillance for Congenital Anomalies registry, a large study that includes 31 European registries, showed the average CDH birth prevalence to be 2.3 per 10,000 births, with substantial variation in total prevalence among the registries (3). Registry-based epidemiologic studies may be affected by reporting compliance and by differences in case ascertainment, which may in part explain the resulting variable estimates of birth prevalence (3). However, geographic distinctions, as well as changes in incidence over time, may reflect true variations that are related to differences in exposure (3). In addition, estimates depend on study design and differ because of inclusion criteria: live births only (4), live births and stillbirths (5), or miscarriages and termination of pregnancy (6–9). It is generally accepted that population-based studies, which include more comprehensive case inclusion (stillbirths, terminations of pregnancy), provide more precise estimates of incidence (10,11).

Debate is ongoing regarding whether CDH survival rates have improved over the last 3 decades. One of the most important prerequisites for assessing accurate CDH survival rates is precise ascertainment of incidence. While some have suggested that survival rates have improved (12–14), others have argued that the apparent gains were attributed to biases in case selection (10,11).

The objectives of this population-based study were 2-fold. The first aim was to examine the incidence of CDH in the geographically defined population of Olmsted County, Minnesota, United States. The incidence rate of CDH was determined through the resources of the Rochester Epidemiology Project (REP) (15,16). The REP provides access to all medical records for county residents, and this comprehensive health database allows study of virtually an entire geographic population to identify incident and prevalent cases of disease and trends in disease development and to explore the risk factors associated with illness (15). The structure of the REP provides a reliable means for capturing incident cases of disease or congenital malformations because of the direct link to the health database of the entire population. Our second aim was to examine survival to discharge rates over the past several decades.

Methods

Study Design and Settings

We conducted a population-based study of neonates with CDH who were born between January 1, 1981, and December 31, 2014, in Olmsted County, MN, in order to describe the incidence of CDH and survival to discharge of the neonates. After approval from the Mayo Clinic Institutional Review Board and the Olmsted Medical Center Institutional Review Board, we conducted a community-based historical cohort study of CDH by ascertaining all cases of CDH through the records-linkage system of the REP (15).

The REP health records–linkage system was established in 1966 to capture health care information for the population of Olmsted County. This system provides the infrastructure for indexing and linking practically the entire data set of health information for the Olmsted County population (Supplemental Digital Content) (15). Medical care is essentially self-contained within this county community of approximately 150,000 residents (July 1, 2015). The current providers participating in the REP are all Mayo Clinic hospitals, freestanding Mayo Clinic–associated clinics and branch offices, the Olmsted Medical Center with its branch offices and affiliated hospitals, and the Rochester Family Medicine Clinic, as well as various private dental practices. Regarding the management of CDH, there are no other medical facilities in the county with the capability to treat patients with this type of malformation.

The electronic REP indexes include data on demographic characteristics, medical diagnostic codes, surgical procedure codes, and causes of death. The system also includes the full text of all records (paper, electronic, and scanned) that are available for review. All diagnoses, surgical procedures, radiographic reports, and pathology reports, including autopsies, are indexed, and all medical records are retrievable for review (15). All health information is abstracted and entered into computerized indexes using the Hospital Adaptation of the International Classification of Diseases, Eighth Revision, or the International Classification of Diseases, Ninth Revision.

To be included as an incident case in our study, the mother of the patient had to be an Olmsted County resident at the time of the infant's birth. Consistent with Minnesota Statute 144.295, we included only patients who had provided authorization for research use of their health records; traditionally, 98% of the Olmsted County population provides written authorization (15). We ascertained cases of CDH through computerized screening, and all identified cases from the REP were subsequently confirmed for the mother's Olmsted County residency by reviewing individual health records. Health records for all identified cases were thoroughly reviewed by 2 of the authors (K.B., J.M.W.). In addition, to confirm the accuracy of the cases we identified as CDH in the REP, we crosschecked all cases against the Mayo Clinic CDH Registry, which was established in January 1990 and is maintained by our neonatology department.

Collection of Data

We recorded the infants' demographic characteristics, the timing of CDH detection (prenatal vs postnatal), sex, race, gestational age at birth, birth weight, and Apgar scores. The diaphragmatic defect was noted as left-sided congenital diaphragmatic hernia (L-CDH), right-sided congenital diaphragmatic hernia (R-CDH), liver-down (liver not herniated in the chest), liver-up (liver herniated into the chest), and bilateral. The main outcome was survival to discharge.

Statistical Analysis

All infants diagnosed with CDH during the study period whose mothers were residents of Olmsted County at the time of childbirth were included as incident cases. During the study period, in 2001, extracorporeal membrane oxygenation (ECMO) was introduced at Mayo

Clinic and represented a major advance in the management of CDH in neonates. Therefore, all data summaries are presented for both the overall study period and for 2 distinct calendar periods: before ECMO (1981 through 2000) and after ECMO (2001 through 2014). We calculated overall and sex-specific incidence rates using the number of incident cases as the numerator and newborn (age=0) population counts from the REP census as the denominator (16). Incidence rates are presented using point estimates and 95% CIs, assuming a Poisson distribution. Overall incidence rates were standardized to the sex distribution of newborns in the total US white population from the 2010 decennial census. To assess whether the incidence of CDH changed over calendar time or differed between male and female infants, a Poisson regression analysis was performed using year-specific incidence counts. For this analysis, calendar year was treated as a continuous variable and $P<.05$ denoted statistical significance. Data were analyzed with SAS 9.3 software (SAS Institute Inc).

Results

During the study period, 26 CDH incident cases were identified. All identified CDH cases had written authorization for research on file; therefore, all of the cases were included in the review. The infants' birth characteristics are summarized in Table 1, both overall and according to calendar period (pre-ECMO and post-ECMO). Overall and sex-specific incidence rates are summarized in Table 2. The overall incidence of CDH was 3.6 (95% CI, 2.2–5.0) per 10,000 births and did not differ significantly over the 34-year period of this study ($P=.28$). The incidence of CDH was 4.3 (95% CI, 2.2–6.5) for male infants and 2.9 (95% CI, 1.1–4.6) for female infants, with a rate ratio of 1.5 (95% CI, 0.7–3.3). During the pre-ECMO time period (1981–2000), 33% of cases were diagnosed prenatally; in the more recent time period (2001–2014), 50% of cases were diagnosed prenatally. The rate of survival to discharge in the CDH cases was 50% before ECMO and 88% thereafter. Table 3 summarizes the clinical characteristics of patients who did not survive hospitalization. There were 3 pregnancy terminations (all before 21 weeks of gestation) after CDH incompatible with life was diagnosed.

Discussion

The incidence of CDH in Olmsted County was 3.6 per 10,000 births and did not differ significantly over the period of the study. The rate of survival to discharge was 50% from 1981 through 2000 and 88% from 2001 through 2014 (after ECMO).

Incidence of CDH at Birth

A meta-analysis of 16 population-based studies showed the average CDH incidence to be 2.5 per 10,000 births (17), which agrees with the estimate by the US Centers for Disease Control and Prevention of 2.6 per 10,000 births (2). Subsequent studies worldwide also reported incidence rates of less than 3 per 10,000 births (3,5,7,9,18–21). However, variations have been reported in CDH incidence from 1.7 (22) to 5.7 per 10,000 births (23), which is most likely related to specific inclusion and exclusion criteria, as well as to variable case reporting among registry studies. The variation in incidence is less likely due to environmental factors affecting the pathogenesis of CDH (24).

Using multilevel analysis, McGivern et al (3) identified a small but statistically significant increase in CDH birth prevalence over time. Because this trend has never before been reported, the authors commented that it most likely resulted from improvements in ascertainment rather than from a true increase in prevalence (3). For our study, we had consistent ascertainment tools available for decades in the REP, and we did not find a change in the incidence rate of CDH over time. Compared with registries, the REP is less affected by flaws in administrative reporting. All records for patients using the health system are systematically entered into the county database to build a single health record for each patient and a comprehensive database for the entire county. Therefore, we believe that the REP allowed us to consistently capture all incident CDH cases in Olmsted County over the long period of our study.

An earlier population-based survey from Minnesota (1988–1990) claimed high rates of ascertainment and “low hidden mortality” (25). In this study, the CDH rate was 3.6 per 10,000 live births (25), identical to our present rate for a single Minnesota county. Mohamed and Aly (24) suggested that race may impact the prevalence of CDH, with blacks having a lower risk than whites (odds ratio, 0.78). Although most characteristics of Olmsted County residents are similar to those of persons living elsewhere in Minnesota, the prevalence of black individuals is low (white, 85.5%; black, 5.8%) compared with the average prevalence of the black population in the United States. Although this fact may in part explain our relatively higher than average CDH incidence, the most plausible explanation remains the comprehensive ascertainment of cases.

Male-to-Female Ratio

One epidemiological survey recorded a higher incidence of CDH in female infants, albeit only among whites in the southern United States (24). In contrast, several studies cite a higher risk for CDH in male infants (3,9,26,27). Although we did find statistically significant differences in CDH rates between male and female infants, the overall point estimate ratio rate (1.5 [95% CI, 0.7–3.3]) was in the same direction and of similar magnitude as the rates reported by Tennant et al (1.5 [95%, CI 1.1–2.1]) (26) and Lisi et al (1.4 [95% CI, 1.3–1.5]) (27).

Survival Rate

The survival rates for CDH are difficult to compare among studies because the rate depends on study design (3,6,9). Specifically, higher rates have been reported in studies of neonates referred to a treatment center than in estimates from population-based studies that typically include early (ie, immediate perinatal) death (10,28). Up to 60% of newborns with CDH are estimated to die before they reach a tertiary referral center (6,11). Stege et al (8) suggested that the mortality of CDH, when complete case ascertainment is achieved, was unaffected by new therapies and was principally determined by the rate of antenatal termination and the incidence of associated anomalies. Their study contrasts with a registry-based study from France (1986–2003) that showed improvement in CDH survival over time, albeit only for isolated, less complex cases (9). In the first period of our study (1981–2000), the survival rate was 50% and included 3 pregnancy terminations (all between 1984 and 1986) that were based on ultrasonographic assessment that the fetal condition was incompatible with life.

After 2000, survival was 88%. Although this assessment was based on a small number of cases, it defies the notion that more recent improvements in survival are a result of case selection artifact and, instead, indicates improved perinatal emergency care to be the reason.

Prenatal Diagnosis and Survival

Prenatal diagnosis of CDH was done for 33% of our cases in the first period (1981–2000) and 50% in the later period (2001–2014). In countries with established prenatal screening programs, the rates of prenatal diagnosis range from 53% to 73% (6,9,29). A 13-study meta-analysis (30) that compared mortality related to the presence or absence of prenatal diagnosis showed significantly higher mortality for those infants with a prenatal diagnosis (odds ratio, 1.9 [95% CI, 1.4–2.8]) (ie, when pooled analysis was considered), even though most individual studies did not find a significant association. Prenatal diagnosis is believed to be a marker of more severe CDH because lower birth weights and higher rates of additional congenital anomalies are prognosticating features that are frequently seen and detected earlier in neonates who undergo ultrasonography (6,30). Despite that similarly to a previous report from Minnesota (25), our study did not confirm an association between prenatal diagnosis and survival.

In conclusion, the incidence of CDH in our study is higher than that reported by others, which is likely attributable to the meticulous case ascertainment rather than to environmental factors associated with the pathogenesis of CDH. Our CDH rate remained unchanged over the study period and agrees with previous reports for the incidence of CDH in Minnesota. Rates of survival to discharge were very high for our patients with CDH in the more recent period of our study, which coincides with the period during which ECMO was available.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

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Abbreviations

CDH	congenital diaphragmatic hernia
ECMO	extracorporeal membrane oxygenation
L-CDH	left-sided congenital diaphragmatic hernia
R-CDH	right-sided congenital diaphragmatic hernia

REP

Rochester Epidemiology Project

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Table 1

Birth Characteristics of Incident Cases of Congenital Diaphragmatic Hernia

Characteristic	Overall (1981–2014) N=26	Period	
		Pre-ECMO (1981–2000) n=18	Post-ECMO (2001–2014) n=8
Prenatal diagnosis	10 (38)	6 (33)	4 (50)
Sex, No. (%)			
Male	16 (62)	10 (56)	6 (75)
Female	10 (38)	8 (44)	2 (25)
Race			
White	24 (92)	17 (94)	7 (88)
Asian	1 (4)	1 (6)	1 (12)
Black	1 (4)	0 (0)	0 (0)
Birth weight, g			
Mean (SD)	2662 (1093)	2485 (1225)	3060 (601)
Median (min, max)	3046 (375, 3800)	3046 (375, 3600)	3100 (1860, 3800)
Gestational age, wk			
Mean (SD)	35.9 (6.5)	34.7 (7.4)	38.4 (2.8)
Median (min, max)	39 (20, 42)	38 (20, 42)	39 (33, 41)
CDH side ^a			
Right	5 (21)	2 (13)	3 (38)
Left	17 (71)	13 (81)	4 (50)
Bilateral	2 (8)	1 (6)	1 (12)
Liver position ^a			
Up	10 (48)	7 (54)	3 (38)
Down	11 (52)	6 (46)	5 (62)
Survived to discharge	16 (62) ^b	9 (50)	7 (88)

Abbreviations: CDH, congenital diaphragmatic hernia; ECMO, extracorporeal membrane oxygenation.

^aThe side of the CDH was missing for 2 neonates, and liver position was missing for 5.^bDuring this period, there were 3 terminations of pregnancy.

Table 2

Incidence (Per 10,000 Births) of Congenital Diaphragmatic Hernia in Olmsted County, MN, From 1981 Through 2014

Incident Rate	Overall (1981–2014)	Period	
		Pre-ECMO (1981–2000)	Post-ECMO (2001–2014)
Overall (95% CI)	3.6 (2.2–5.0)	4.6 (2.5–6.7)	2.4 (0.8–4.1)
Male sex (95% CI)	4.3 (2.2–6.5)	5.0 (1.9–8.0)	3.6 (0.7–6.5)
Female sex (95% CI)	2.9 (1.1–4.6)	4.2 (1.3–7.1)	1.3 (0.0–3.0)
M/F rate ratio (95% CI)	1.5 (0.7–3.3)	1.2 (0.5–3.0)	2.9 (0.6–14.2)

Abbreviation: ECMO, extracorporeal membrane oxygenation; F, female; M, male.

In-Hospital Deaths and Pregnancy Terminations for Cases of Congenital Diaphragmatic Hernia

Table 3

Case, Sex	BW, kg	GA, wk	AS 1/5 min	CDH type	Associated Anomalies	Hospital Course	Death, Time
1, M	0.9	28.0	0/N/A	Right	None noted, liver position unknown	CDH repair; died (<1 h) after surgery	<4 h
2, F	3.0	39.6	6/7	Left	None noted, liver position unknown	CDH repair	16 h
3, M	3.5	40.0	3/5	LLU	None noted	CDH repair; ECMO; laparotomy for liver infarction; multiorgan failure	12 d
4, M	3.3	36.0	1/N/A	Left	None noted, liver position unknown	ECMO; oxyhemoglobin desaturation to 40–50 mm Hg	Same day
5, M	0.4	20.0	NA	LLU	Quadracuspid pulmonary valve	Prenatal diagnosis of CDH incompatible with life	TOP, 20 wk
6, F	1.1	31.0	1/1	Unknown	Trisomy 18; ASD	Comfort care only	Same day
7, F	0.4	20.9	NA	Unknown	Unbalanced X chromosome translocation	Prenatal diagnosis of CDH incompatible with life	TOP, 20.9 wk
8, M	2.4	34.7	0/0	Bilateral LU	Absent diaphragm	No CDH repair; cardiac arrest at delivery	1 h
9, M	0.35	20.4	NA	LLU	Bilateral renal pelvis dilatation; micrognathia; low-set ears; absence of thymus lobe	Prenatal diagnosis of CDH incompatible with life	TOP, 20.4 wk
10, M	1.8	32.9	0/6	RLD	Omphalocele ^a	Third triplet	2 d

Abbreviations: AS, Apgar score; ASD, arial septal defect; BW, body weight; CDH, congenital diaphragmatic hernia; ECMO, extracorporeal membrane oxygenation; F, female; GA, gestational age; LLU, left liver up; LU, liver up; M, male; NA, not applicable; RLD, right liver down; RLU, right liver up; TOP, termination of pregnancy.

^aThe liver was contained within the omphalocele sac.