Wed 27th April 13.30 – 13.55
Live Abstract Presentations in MAIN AUDITORIUM, live streamed with recording

6 Multiple breath washout measurement in patients with CDH at school age compared to Chest CT score and spirometry.

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39 Has Survival Improved for Congenital Diaphragmatic Hernia? A 25-Year Review of over 5000 Patients from the CDH Study Group

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57 Persistent health issues beyond childhood affect quality of life of CDH survivors

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86 Trends Over Time: What has 25 years of collaboration revealed in the Congenital Diaphragmatic Hernia Study Group?

Np Ashley Ebanks[1], MD Vikas Gupta[1], MD Pamela Lally[1], MD Kevin Lally[1], MD Krisa Van Meurs[2], MD Terry Buchmiller[3], MD Erin Perrone[4], MD Tim Jancelewicz[5], MD Matthew Harting[1]
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94 Surveillance endoscopy in CDH – is it indicated?

Miss Gerlin Naidoo[1], Mr David Colvin[1], Mr Carl Davis[1], Miss Rania Kronfli[1], Miss Gerlin Naidoo
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Multiple breath washout measurement in patients with CDH at school age compared to Chest CT score and spirometry.

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Data on long term structural and functional lung abnormalities in survivors of congenital diaphragmatic hernia (CDH) are scarce. The purpose of this follow-up program is to assess the structural lung sequelae with chest CT scoring at 1 year of age and at school age with lung function and exercise testing.

Methods: Prospective, clinical follow-up program of babies admitted to our NICU, with a of CDH between January 2007 and August 2015.
An unenhanced CT scan of the lungs was performed around the age of one year.
At the age of 7-10 years, spirometry, Nitrogen (N2) MBW as well as a six-minute walk test and a shuttle run are performed. Quality of life (Qol) questionnaires were filled out.

Results: 35 patients have been included so far of which 28 participated in the second follow-up. LCI was abnormal in 12/26 children and Fev1 was abnormal in 10/24. LCI Z-score correlates significantly with FEV1 Z-score ($r= - 0.748$, $p<0.01$) and with FVC Z-score ($r= -0.617$, $p<0.01$).
A significant correlation was found between the LCI Z-score and CT score at age 1 (Pearson $r= 0.464$, $p=0.02$), the FEV1 Z-score ($r=0.761$, $p<0.01$), the FVC Z-score ($r= -0.617$, $p<0.01$) and between LCI Z-score and the amount of days the patient was ventilated ($r= 0.734$, $p<0.01$). No correlation was found between LCI and the prenatal assessment of severity, exercise testing and Qol scoring.
Conclusion: In this small cohort of children with CDH, LCI is not more sensitive to detect lung disease compared to spirometry, but LCI correlates well with FEV1 and FVC. Both LCI and FEV1 correlate with chest CT performed at age 1. In our knowledge this is the first study to compare LCI and spirometry in a cohort of CDH survivors at this age. Data needs to be confirmed in a larger patient group.

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Objective: The Congenital Diaphragmatic Hernia Study Group (CDHSG) is a multicenter, international collaboration focused on evaluation of liveborn infants with congenital diaphragmatic hernia (CDH). We sought to determine if risk-adjusted survival in CDH patients has improved among centers with consistent involvement, and we hypothesized that there was no change in risk-adjusted survival over time.

Methods: We retrospectively studied outcomes at centers with 22-25 years of continuous participation in the CDHSG. We divided the CDHSG into five-year intervals starting with era 1 (E1) beginning in 1995 and analyzed multiple variables (minimally invasive repair rate, defect size, ECMO use, and mortality) over time to assess possible evolution of disease characteristics. For overall mortality analyses, patients were risk stratified using a validated CDH prediction score based on 5-minute Apgar and birth weight. An observed to expected (O:E) mortality model was created using Apgar, major cardiac anomaly, and prenatal diagnosis as risk-stratifiers and using 1995-2000 as a reference. Univariate and multiple regression analyses were performed.

Results: 5015 posterolateral hernia patients from 23 centers were included. Birth weight, 5-minute Apgar, incidence of agenesis, ECMO use, and rate of repair did not vary over time (all $p>0.05$). Minimally invasive repair and patch repair were more prevalent, and timing of diaphragmatic repair was later in E5 compared to E1 (all $p<0.01$). Overall, mortality decreased over time: E1 (30.7%), E2 (30.4%), E3 (28.7%), E4 (26.0%), E5 (26.0%) ($p=0.04$, Chi2) (Figure 1a). Risk-adjusted mortality showed a trend favoring significant improvement in E5 compared to E1 (OR 0.79, 95% CI 0.63-1.01; $p=0.06$). O:E mortality improved over time (Slope: -0.02, $p<0.01$), with the most significant improvement occurring in the last five years (Figure 1b).

Conclusions: Among consistent participating CDHSG centers, risk-adjusted CDH mortality has improved over time, with the most significant improvements occurring in the recent eras.

Images

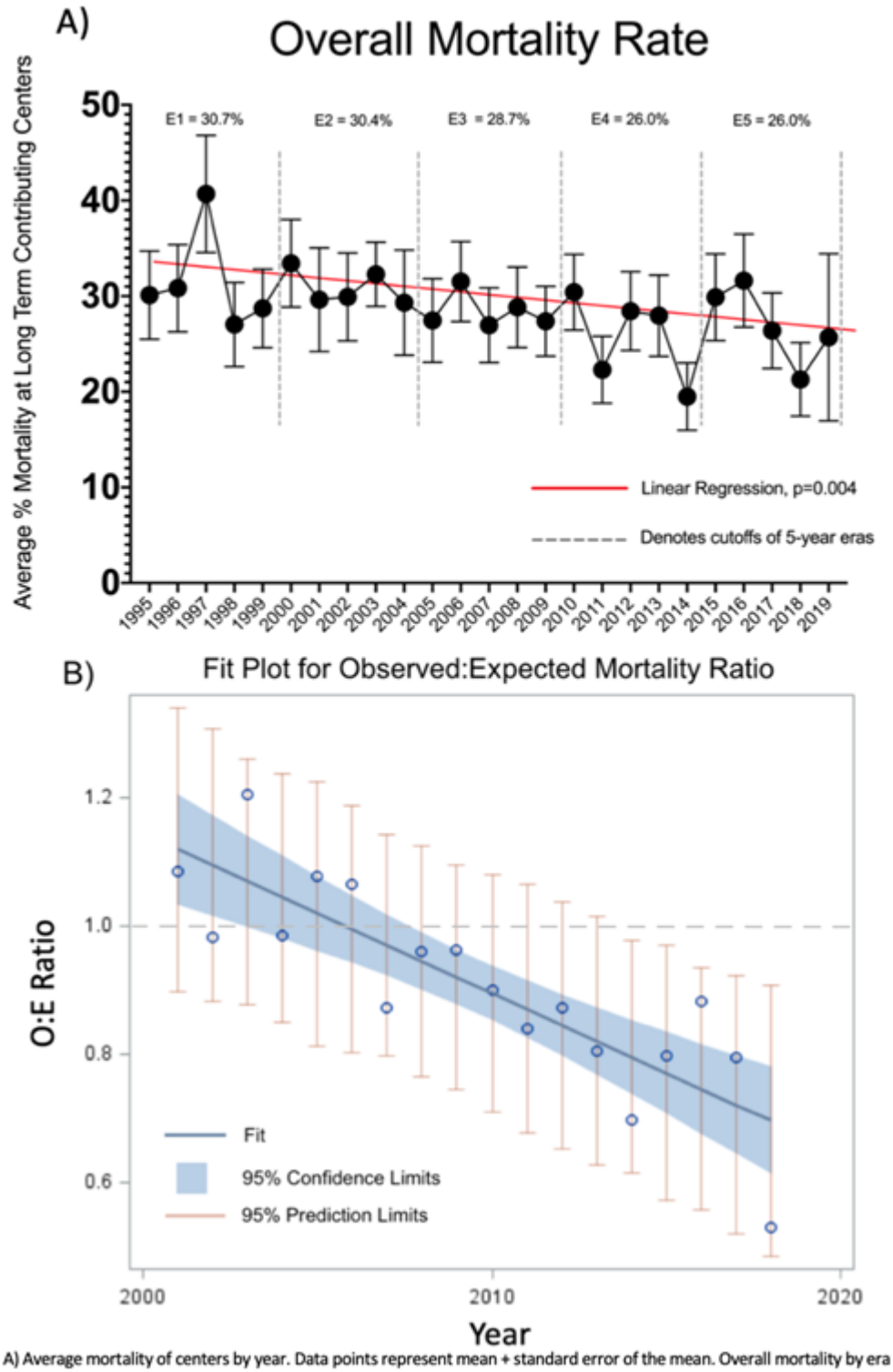

Figure 1: A) Average mortality of centers by year. Data points represent mean + standard error of the mean. Overall mortality by era (E1-E5) is listed and separated by grey hash lines. B) Fit plot of O:E mortality ratio using 1995-2000 as a reference and adjusted for birth weight, Apgar at 5 minutes, presence of a major cardiac anomaly, prenatal diagnosis, and center volume. O:E ratio of 1 is denoted by the grey hash line.

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Persistent health issues beyond childhood affect quality of life of CDH survivors

Dr Louise Montalva[1], Lina Antounians[1], Dawn Ireland[2], Jason Miller[2], Tracy Meats[2], Dr Augusto Zani[1]
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Background:
The quality of life (QOL) of adults with congenital diaphragmatic hernia (CDH) remains understudied. Herein, we aimed to evaluate the long-term morbidity of CDH and its impact on QOL in adult survivors.

Methods:
An online survey was sent to adult CDH survivors worldwide through CDH International, a global association of CDH patients and families. The survey included questions on medical history, current health issues and a validated SF-12 QOL questionnaire reporting on physical (PCS) and mental (MCS) component scores. Statistics: Mann-Whitney test.

Results
The survey was completed by 99 CDH survivors (72 left CDH, 50 patch repair), from 9 different countries. The median age was 22 years (17-30). At least one redo-surgery was required in 31 respondents. Reintervention for CDH was associated with altered QOL with a decreased PCS (48.1 vs 55.2, p=0.0178). Current health issues were reported by 67% (n=66) of respondents (respiratory, 37%; digestive, 24%; neurologic/psychosocial, 18%) and affected QOL by decreasing both PCS (47.81 vs 55.91, p<0.0001) and MCS (50.17 vs 54.01, p=0.0045). Respiratory symptoms were reported in the last month in 41 CDH survivors affecting QOL by decreasing both PCS (44.5 vs. 55.5, p=0.0004) and MCS (46.2 vs 53.3, p=0.02). Gastroesophageal reflux was reported in the last month in 36 respondents and was associated with a decreased MCS (39.6 vs 53.5, p=0.0007). Persistent scoliosis or chest wall deformities were reported by 46 CDH survivors, without QOL deterioration (PCS: 52.8 vs 54.8, p=0.5; MCS: 51.3 vs 52.7, p=0.8). Of 34 CDH survivors who had a pregnancy, 21% reported having issues related to CDH during pregnancy.

Conclusions
Two-thirds of CDH adult survivors suffer from health issues that affect their QOL. A standardized multidisciplinary follow-up of CDH survivors should be pursued beyond childhood to monitor active symptoms.

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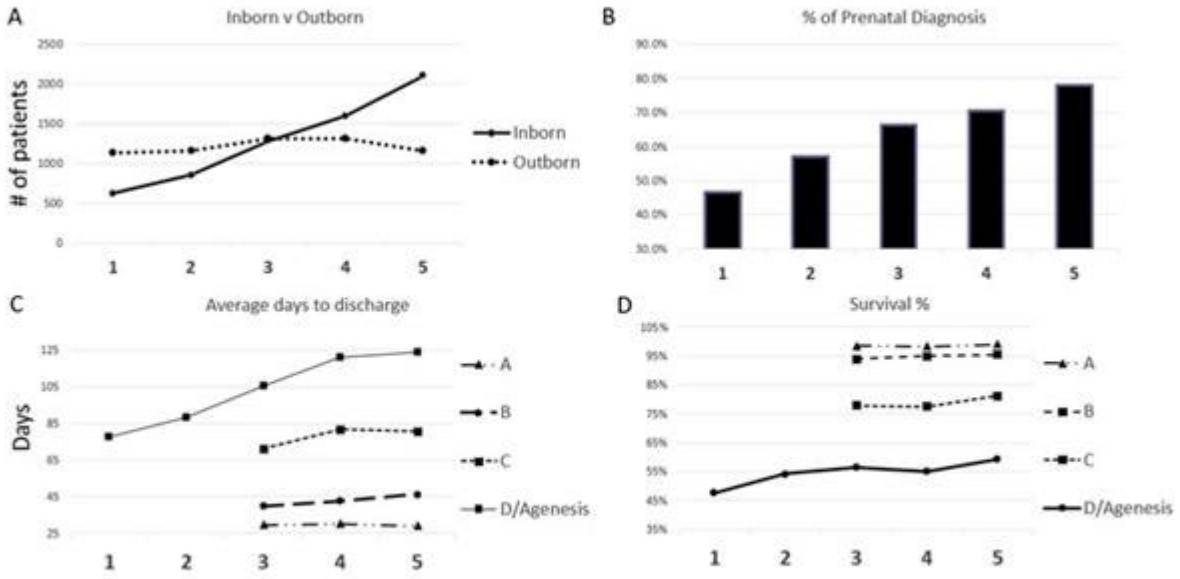
Background The Congenital Diaphragmatic Hernia (CDH) Study Group (CDHSG) is an international collaborative registry focused on liveborn neonates born with Bochdalek hernias. Data trends over 25 years in a consortium focused on CDH infants are unknown.
Methods This is a retrospective study of the CDHSG data collected 1995-2019. Registry data were divided into five-year eras (E1-E5) beginning in 1995 and variables analyzed to assess changes over time. All liveborn neonatal Bochdalek patients were analyzed.
Results 12,542 patients from 140 centers were included. There was an increase in the annual number of patients over time (E1: 1771, E5: 3258). There was an increase of inborn patients with no significant change in number of outborn infants. The rate of prenatal diagnosis increased over time (E1: 46.5%, E5: 78.0%). The rate of extracorporeal life support (ECLS) utilization decreased from 36.3% in E1 to 26.6% in E5 with no significant change in ECLS survival. High risk CDH, defined as agenesis or type D defect, showed little change in prevalence over time, though survival has improved from 47.7% in E1 to 59.3% in E5. Survival for non-agenesis, A, B, and C type defects has not changed. The use of minimally invasive surgery (MIS) has increased from 0% in E1to 14.3% in E5. There has been an increase in mean length of stay for B defects (E3: 39.4, E5: 46.1) and agenesis/D defects (E1: 77.7, E5: 123.9) without significant change in other defects.
Conclusion Our analyses show an increase in inborn infants, prenatal diagnosis, MIS surgical approach, and length of stay over time. There has been a decreasing trend in ECLS use, with no change in ECLS survival. The survival for the high risk CDH patients has improved. The CDHSG continues to strive to optimize the care and outcome of neonates born with CDH.

Images

Figure



Legend: X Axis shows Eras 1-5, A. Inborn versus Outborn B. Percentage of Prenatal Diagnosis C. Average Days to Discharge D. Survival Percentage

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Surveillance endoscopy in CDH – is it indicated?

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Background: Gastroesophageal reflux disease (GORD) is a known consequence of congenital diaphragmatic hernia (CDH). This can present with feeding issues resulting in the need for anti-reflux medication or surgical intervention. It is also accepted that chronic, asymptomatic or "silent" reflux may occur throughout childhood. Surveillance endoscopy is often performed before transition to adult services. A review of practice in our tertiary institution was performed to assess for the incidence of GORD symptoms, use of anti-reflux therapy, and surveillance endoscopy for children following CDH repair.

Methods: Retrospective cohort study of patients undergoing CDH repair between 1999-2011 at a single tertiary centre (n = 171). Data on reflux symptoms, anti-reflux medication use, surgical intervention and endoscopy findings were collected.

Results: 93/171 had complete clinical data (M:F 2:1). Mortality was 32% (30/93) in the first 5 years of life. At 5 year follow-up, 29% (6/21) had GORD symptoms, 19% (4/21) patients were on anti-reflux medication and 48% (10/21) had undergone fundoplication. At 10 year follow-up, 20% (7/34) had GORD symptoms, with only 3% (1) on anti-reflux medication. At 15yr follow-up, 32% (7/22) had GORD symptoms, with 9% (2/22) on anti-reflux medication. 68% (15/22) of the cohort had endoscopy prior to transition to adult services. 53% (8/15) were asymptomatic and had normal histological features on biopsy. Of the 46% (7/15) symptomatic patients, 3 had pathological evidence of oesophagitis, one of whom had Barrett's oesophagitis.

Conclusions: During the study timeframe there was a wide variation in practice. There is now a multi-disciplinary clinic with a standard schedule for follow-up that mandates endoscopy prior to transition. In our subset of patients, however, endoscopy for asymptomatic patients at transition to adult services did not elicit evidence of GORD.