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Standardizing clinical practice guidelines to manage infants with congenital diaphragmatic hernia can reduce ECMO need, improve clinical outcomes and survival at discharge: a single center experience

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Background: Infants with congenital diaphragmatic hernia (CDH) are critically ill with high risk of death and/or requiring extracorporeal membrane oxygenation (ECMO). In January 2012 our institution implemented standardized clinical practice guidelines (CPGs) to manage infants with CDH. We hypothesized that infants with CDH managed with CPGs have better outcomes, less ECMO need, and are more likely to survive.

Methods: Retrospective review of two cohorts of CDH patients admitted between January 2007 to July 2021 (N=133). Cohort 1 (management not standardized): January 2007 to December 2011 (N=53). Cohort 2 (management standardized): January 2012 to July 2021 (N=80). Descriptive statistics used to compare the cohorts including survivors vs non-survivors and ECMO vs non-ECMO patients.

Results: Patient demographics, antenatal diagnosis, hernia side, liver herniation, pre-operative pneumothorax, admission pCO2, timing and type of surgical repair were not statistically different. Cohort 1 had significantly more severe pulmonary hypertension (PH) with more vasopressor and iNO therapy ($p<0.001$). ECMO rates were significantly higher in Cohort 1 (51% vs 18%, OR 4.9; $p<0.001$) while survival at discharge was significantly lower compared to Cohort 2 (57% vs 85%, OR 4.3; $p<0.001$). Non-ECMO patient survival at discharge was not significantly different between cohorts. Survival for ECMO-treated patients in Cohort 1 was significantly lower than Cohort 2 (26% vs 71%, OR 7.1; $p<0.01$). Total ventilator and supplemental oxygen days were significantly lower for Cohort 2 ($p<0.001$). Cohort 1 patients were more likely to be discharged on gavage feeds, gastroesophageal reflux therapy and PH medications ($p<0.01$). Day of life at discharge, chronic lung disease diagnosis, and home oxygen or tracheostomy/ventilator at discharge were not significantly different.

Conclusions: Standardized CPG to treat patients with CDH reduced ECMO need, improved clinical outcomes and survival at discharge. Refinement of management strategies, implementing new interventions and meticulous care can improve outcomes in patients with severe disease.

Images

Variable	Cohort		p
	1 (N=53)	**2** (N=80)	
Pre-op Pneumothorax (n, %)	4 (8%)	2 (3%)	0.211
Post-op Chest Tube (n, %)	21 (45%)	31 (45%)	1.000
Admission pH (arterial) [median, IQR]	7.3 [7.2-7.4]	7.2 [7.2-7.3]	0.239
Admission PCO2 (arterial) [median, IQR]	51 [40.3-71.5]	57.5 [46-68.8]	0.281
Ventilator Days [median, IQR]	**19 [9-31.5]**	**9 [5-18]**	**<0.001**
Supplemental O2 Days [median, IQR]	**21 [11-36]**	**13 [6-25.5]**	**0.016**
DOL of Surgery [median, IQR]	8 [3-12]	6 [4-10]	0.339
Type of Surgical Repair (n, %)			0.253
TP	13 (28%)	32 (46%)	
OM	21 (46%)	24 (35%)	
TM	6 (13%)	8 (12%)	
OP	6 (13%)	5 (7%)	
Liver Up (n, %)	17 (33%)	21 (26%)	0.384
PH at Discharge (n, %)	8 (28%)	24 (39%)	0.300
Severity of PH on 1st Echo (n, %)			**<0.001**
SS	9 (18%)	26 (35%)	
S	30 (60%)	19 (25%)	
N	11 (22%)	30 (40%)	
Inotrope Use (n, %)	**40 (76%)**	**30 (38%)**	**<0.001**
iNO Use (n, %)	**41 (77%)**	**37 (46%)**	**<0.001**
Sildenafil Use (n, %)	**19 (38%)**	**5 (6%)**	**<0.001**
Hydrocortisone Use (n, %)	25 (47%)	30 (38%)	0.268
Prostin Use (n, %)	6 (11%)	4 (5%)	0.196
ECMO Use (n, %)	**27 (51%)**	**14 (18%)**	**<0.001**
Days on ECMO (n, %)	10.4 (6.2)	8.6 (5.3)	0.341
ECMO Survival (n, %)	19 (70%)	11 (61%)	0.519
Survived to Discharge (n, %)	**30 (57%)**	**68 (85%)**	**<0.001**
DOL at Discharge/Transfer/Death [median, IQR]	42 [18.5-57.5]	25.5 [15-45.5]	0.160
CLD Diagnosis (n, %)	7 (23%)	7 (10%)	0.122
Discharge/transfer on Home O2 (n, %)	7 (23%)	6 (9%)	0.061
PH Medication at Discharge (n, %)			**0.003**
N	22 (76%)	66 (97%)	
Y	7 (24%)	2 (3%)	
Pulmonary/CVS Medication at Discharge (n, %)			0.122
N	24 (83%)	64 (94%)	
Y	5 (17%)	4 (6%)	
GER Medication at Discharge (n, %)	**23 (77%)**	**17 (25%)**	**<0.001**
Trach/Home Ventilator (n, %)	2 (7%)	2 (3%)	0.584
GT/NG at Discharge (n, %)			**0.005**
N	13 (43%)	44 (65%)	
Y	17 (57%)	24 (35%)	

TP = thoracoscopic primary, OM = open mesh/patch, TM = thoracoscopic mesh/patch, OP = open primary, SS = suprasystemic, S = systemic, N = no/none, Y = yes, CLD = chronic lung disease, DOL = day of life, GER = gastroesophageal reflux, GT = gastrostomy tube, NG = nasogastric tube

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Continuous oxygen saturation index measurements as early predictor of outcomes in congenital diaphragmatic hernia

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Background: Early identification of infants with a congenital diaphragmatic hernia (CDH) at risk for a complicated postnatal course could alert physicians, refine individualized treatment strategies, and improve parental counselling. The oxygen saturation index (OSI), a ratio that continuously reflects the infant's respiratory status, has the potential for real-time guidance and thereby detection of early signs of deterioration. We aimed at evaluating the OSI as such an early predictor.
Methods: A single-center retrospective cohort study in consecutive infants born with an isolated CDH between June 2017-July 2021. Continuous OSI measurements were collected for all infants in the first 24 hours after birth. Outcomes of interest were pulmonary hypertension, extracorporeal membrane oxygenation (ECMO) therapy, and mortality before discharge. We evaluated the predictive values of (1) the area under the curve (AUC) in the graph with OSI versus time in hours and (2) the highest OSI value.
Results: In 42 infants with 49,473 OSI measurements, the median OSI was 5.0 [interquartile range 3.1-10.6]. The AUC for the first 3 hours (AUC3) and the highest OSI values in the first 12 hours (OSI12) and 24 hours (OSI24) after birth were significantly higher in infants with adverse outcomes (Figure 1). Pulmonary hypertension was predicted by AUC3≥11.6 (sensitivity 62%; specificity 75%) and highest OSI12 and OSI24 ≥16.3 (sensitivity 75-78%; specificity 93-100%). Need for ECMO therapy was predicted by AUC3≥26.8 (sensitivity 67%; specificity 96%), OSI12≥20.9 (sensitivity 100%; specificity 89%), and OSI24≥21.4 (sensitivity 93%; specificity 86%). Mortality was predicted by AUC3≥46.0 (sensitivity 75%; specificity 97%) and highest OSI12 and OSI24 ≥21.9 (sensitivity 100%; specificity 75-80%).
Conclusions: Continuous OSI evaluation in the first hours is a promising modality to identify CDH infants that are at highest risk for clinical deterioration. This provides an opportunity to tailor postnatal management based on the individual patient's needs.

Graph

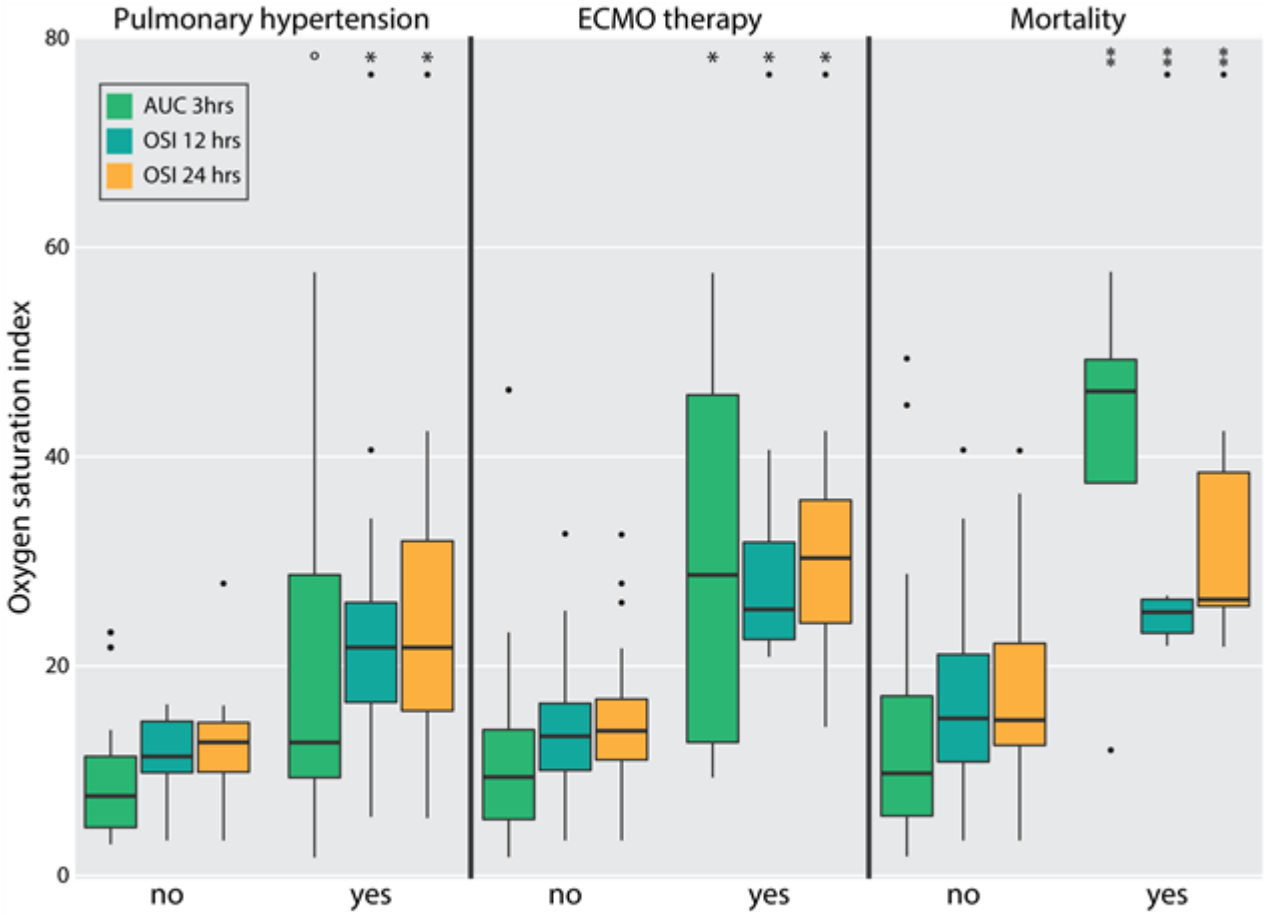

Figure 1 Area under the curve (AUC) in the first three hours after birth and highest oxygen saturation index (OSI) in the first 12 and 24 hours after birth. * $p<0.001$; ⁑ $p<0.01$; ° $p<0.05$. ECMO: extracorporeal membrane oxygenation.

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A Review of COVID-19 Infections Identified in a CDH Multidisciplinary Clinic

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Background

The Boston Children's Hospital CDH Clinic follows almost 500 patients. During the COVID-19 pandemic, our patients have reached out for guidance, especially if they have a positive test. This is a summary of our experience.

Methods

Between March 2020 and January 2021, we identified 20 CDH patients with COVID-19. Patients were identified by cross-referencing CDH and COVID-19 diagnosis codes, provider identification, and a review of the institutional CDH database. A retrospective review was performed to characterize clinical course.

Results

COVID-19-positive patients ranged in age from 2 months to 27 years, with median age 8 years. 3 recent cases have been in infants < 1 year. Of 20 positive patients, 2 cases were asymptomatic; 7 had mild, non-respiratory symptoms; 8 had mild respiratory symptoms; and 3 patients had a severe presentations. There were no fatalities.

Severe cases included a 2 month old infant on weaning ventilator support after patch repair and VA-ECMO who developed bacteremia, worsening pulmonary hypertension, and was COVID-19 positive. An adult with pulmonary hypertension presented with respiratory distress, an increase from baseline oxygen requirement, and was treated with monoclonal antibody. Another adult with chronic lung disease developed significant hypoxemia weeks after COVID diagnosis.

Of our COVID cases, 7 patients were under age 5 and not eligible for vaccination. One patient who contracted COVID was unvaccinated, and 3 recent breakthrough cases were in patients who had received 2 vaccinations, but no booster. The remaining cases were not eligible for vaccine at the time of their infection. 3 additional patients received monoclonal antibody.

Conclusions

The incidence of COVID-19 has been low, and most CDH patients have not manifested severe disease, with no fatalities or use of ECMO to report. Risk factors may include pulmonary hypertension, severe respiratory compromise, and recent CDH surgery. Milder cases may not have been reported.

Graph

Table 1. Characteristics of CDH patients with COVID infections.

COVID symptoms	# cases	Age range	Left CDH	ECMO	Pulmonary HTN infancy	O2 at ICU d/c infancy	Current Inhaled steroids
No symptoms	2	17 mo-10 yr	1	0	0	0	0
*Mild non-respiratory	7	2 mo-17 yr	7	2	0	0	2
**Mild respiratory	8	4 mo-17 yr	8	2	0	0	4
***Severe	3	2 mo-27 yr	2	3	2#	n/a #	2#

*MILD non-respiratory -fever, fatigue, congestion

**MILD with respiratory cough or wheeze in addition to above. 7/8 were on bronchodilator, ICS or oral steroids.

***SEVERE -increase in baseline supplemental oxygen or new oxygen requirement with hypoxia.

one patient remains hospitalized at time of data collection

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SEX DIFFERENCES AND MORTALITY IN CONGENITAL DIAPHRAGMATIC HERNIA

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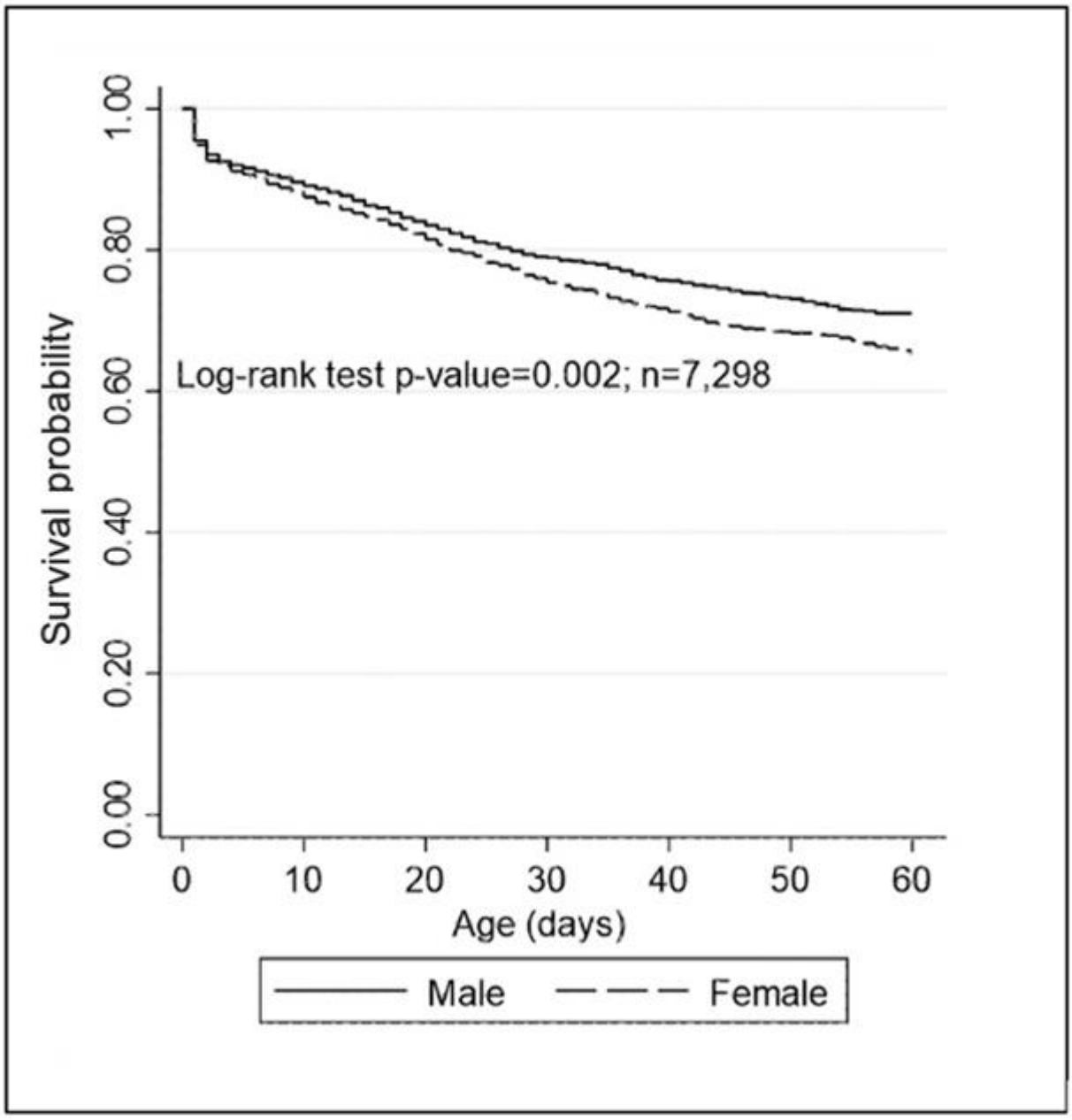
Background: Previous studies have suggested that female neonates with congenital diaphragmatic hernia (CDH) may have worse outcomes compared to their male counterparts. The purpose of this study was to assess the differences in mortality between female and male patients with CDH.

Methods: After Institutional Review Board approval (IRB00127299), we queried the Congenital Diaphragmatic Hernia Study Group (CDHSG) database for female and male CDH patients from 2007-2018. Statistical analyses were performed with t-tests, Chi-square tests, Kaplan-Meier estimates, and Cox regression ($p \leq 0.05$).

Results: There were 7,288 CDH patients managed by 105 institutions. On univariate analysis, female patients weighed less at birth than males (2.84 vs. 2.97 kg, $p<0.0001$) despite having comparable gestational ages and Apgar scores. Female patients had a higher rate of overall cardiac anomalies (22.5 vs. 20.5%, $p=0.03$), but similar rates of major cardiac anomalies (8.7% vs. 7.8%, $p=0.16$) and extracorporeal membrane oxygenation (ECMO) utilization (27.8% vs. 27.3%, $p=0.65$). Although both cohorts had equivalent defect size, laterality, and rates of patch repair, females had increased incidence of intrathoracic liver herniation (49.2% vs. 45.9%, $p=0.01$) and pulmonary hypertension (86.6% vs. 81.1%, $p=0.0001$). Females had lower survival rates at two months of age compared to males (72.3% vs. 75.3%, $p=0.002$, Figure), specifically in the subgroup who underwent repair and were never started on ECMO ($p=0.002$). On Cox regression analysis, there was an increased risk of mortality with female gender (Hazard Ratio 1.26, 95% CI 1.05-1.53, $p=0.01$), when controlling for available markers of disease severity.

Conclusions: Despite comparable defect size and ECMO utilization rates, short-term mortality risk is 26% higher among female neonates with CDH even after adjustment for potential clinical confounders. Results also suggest females with CDH have higher incidence of liver herniation and pulmonary hypertension, which may help guide future research on the etiologies of this excess mortality risk.

Images


Survival probability
1.00
0.80
0.60
0.40
0.20
0.00
Log-rank test p-value=0.002; n=7,298
0
10
20
30
40
50
60
Age (days)
Male
Female