The Prevalence and Impact of Congenital Diaphragmatic Hernia Among Patients Undergoing Surgery for Congenital Heart Disease

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There has not been a multicenter investigation to elucidate the prevalence and impact of congenital diaphragmatic hernia (CDH) on children undergoing cardiac surgery. We investigated the prevalence of CDH across congenital cardiac diagnostic and procedural groups and its impact on outcomes. The STS Congenital Heart Surgery Database was queried to identify children undergoing cardiac surgery who also had CDH. Baseline perioperative characteristics and postoperative outcomes were compared between groups. Subgroup analyses were performed based on case complexity as determined by STAT Categories. Overall 426 of 157,419 (0.27%) pediatric patients undergoing an index cardiac operation had a diagnosis of CDH including 89 neonates (0.25% of all neonatal index operations), 217 infants (0.39%), and 120 children (0.18%). The frequency of concomitant CDH varied across diagnostic groups and, in neonates and infants, was highest for tetralogy of Fallot (0.45%, $n = 41$), coarctation (0.39%, $n = 27$) and complete atrioventricular septal defects (0.31%, $n = 19$). For neonates and infants outcomes were generally worse in CDH vs no-CDH patients across the various procedural strata. For example, STAT ≥3 operative mortality was 34.4% vs 10.3% ($P < 0.001$) for neonates, and 24.2% vs 5.3% ($P < 0.001$) for infants with vs without CDH, respectively. In older children, outcomes disparities were less clear cut with no significant differences in mortality, and variable differences for complication and length of stay outcomes. CDH occurs in a small percentage of patients born with congenital heart disease. Concomitant diagnosis of CDH portends increased morbidity and mortality in infants and neonates undergoing cardiac surgery.

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Statistical analysis and design was performed in collaboration with the DCRI.

Central Message

A small percentage of patients born with congenital heart disease have a diagnosis of congenital diaphragmatic hernia which portends worse outcomes in infants and neonates undergoing cardiac surgery.

Perspective Statement

In this multicenter study of the prevalence and impact of diaphragmatic hernia onchildren undergoing cardiac surgery, concomitant diagnosis of CDH portendsincreased morbidity and mortality in infants and neonates. For older children, a diagnosis of CDH is not consistently predictive of outcomes. These data will provide clarity for risk stratification and operative management in the future.

Abbreviations: AVSD, atrioventricular septal defect; CDH, congenital diaphragmatic hernia; DCRI, Duke Clinical Research Institute; PDA, patent ductus arteriosus; PLOS, postoperative length of stay; STS-CHSD, Society of Thoracic Surgeons Congenital Heart Surgery Database; VSD, Ventricular septal defect

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INTRODUCTION

Congenital diaphragmatic hernia (CDH) has a prevalence of 2.5−5 of 10,000 births and is associated with significant morbidity and mortality. Reports of the prevalence of congenital heart disease among patients with CDH vary somewhat depending on the data source, but in the aggregate, they suggest that approximately 12−20% of children with CDH have a concomitant congenital cardiac anomaly.[1−3] Recent analysis of the Congenital Diaphragmatic Hernia Study Group database reported prevalence of cardiac defects among patients enrolled in the registry to be 18%.[2] The impact of concomitant cardiac malformations on the outcome of repair of CDH has also been explored.[1,4,5] Children with CDH and cardiac anomalies suffer worse survival when compared to those CDH patients with no evidence of congenital heart disease (41% vs 70%, respectively).[2]

Despite multiple investigations of the prevalence of congenital cardiac anomalies and their impact on patients with CDH, there has not yet been a large, multicenter data query to elucidate the frequency of CDH in children undergoing surgical repair of cardiac anomalies, and to assess the impact of CDH on postoperative outcomes. Morbidity and mortality of patients with the most prevalent congenital cardiac anomalies and a concomitant diagnosis of CDH has yet to be evaluated; nor has it been compared to patients without CDH. As such, we sought to define the frequency and to characterize the impact of CDH in patients with the most prevalent congenital cardiac anomalies requiring surgical repair in order to provide clarity for risk stratification, counseling, and operative management.

METHODS

Data Source

The Society of Thoracic Surgeons Congenital Heart Surgery Database (STS-CHSD) was used for this study. The STS-CHSD includes detailed information on more than 420,000 pediatric cardiac procedures performed at 133 centers in North America.[6] It is currently estimated that the database encompasses more than 95% of United States centers performing heart operations and approximately 98% of all pediatric cardiac operations performed annually in the United States.[7] Preoperative, operative, and outcomes data are collected on all patients undergoing pediatric and congenital operations at participating centers. Coding is accomplished by clinicians and ancillary support staff using the International Pediatric and Congenital Cardiac Code.[8] Data collected include demographic, anatomic diagnoses, noncardiac abnormalities, preoperative risk factors, intraoperative details, surgical procedures, postoperative complications, length of stay, and mortality.

The Duke Clinical Research Institute serves as the data warehouse and analysis center for all STS national databases.[6,7] For research projects that are approved by the STS Access and Publications Task Force, statistical analysis is performed at the Duke Clinical Research Institute. This investigation was proposed as a major data request to the STS-CHSD and was approved by the Access and Publications Task Force.

Patient Population

The STS-CHSD includes data pertaining to 157,419 index cardiovascular operations at 122 centers between January 1, 2010 and December 31, 2016. From this population, 427 patients undergoing repair of congenital cardiac anomalies with a concomitant diagnosis of CDH as a noncardiac congenital abnormality were identified and serve as the study cohort. Categorization was first performed by age groups: neonates (<30 days of life), infants (30 days to 1 year of life), and children (1 year to <18 years). Once divided by age, patients were categorized by their fundamental cardiac diagnosis, defined as the most complex cardiac anomaly or condition, and by the primary procedure of the index operation. For cases where a fundamental diagnosis had not been entered, the primary diagnosis of the admission was used. To further investigate the impact of CDH in relation to cardiac procedural complexity, patients were categorized on the basis of the STAT mortality category of their index cardiac operation.[8] Low complexity cases were defined as those with a STAT mortality category of 1−2, while high complexity cases were defined as cases with STAT mortality categories of 3−5. For each age group, outcomes were investigated based on the presence of CDH for each fundamental cardiac diagnosis, primary procedure, and STAT mortality category.

Outcomes

Outcomes of interest included operative mortality, any postoperative complication, any major postoperative complication[9], and postoperative length of stay. For this analysis, major complications were classified using standard STS-CHSD definitions[8] and include: postoperative renal failure requiring dialysis, neurologic deficit persisting at discharge, arrhythmias necessitating pacemaker insertion, post operative mechanical circulatory support, diaphragmatic paralysis, and any unplanned reintervention. Definitions of individual variables can be found in the STS-CHSD specifications at the STS web site.[8]

Statistical Analysis

Baseline demographic and clinical data as well as outcomes of interest were compared between neonates, infants, and children with and without a concomitant diagnosis of CDH. After age group stratification, baseline data and outcomes were compared based on cardiac diagnosis and case complexity of the primary cardiac procedure. Continuous parametric and nonparametric data were compared with Student's t test and the Wilcoxon's rank-sum test and are reported as mean ± standard deviation and median (interquartile range), respectively. Categorical variables were compared with chi-square analysis and are reported as number and percent. Statistical significance was established by a $P < 0.05$.

Table 1. Prevalence of CDH in Pediatric Patients Undergoing Cardiac Surgery

	No CDH	CDH
Neonates	99.8% (35,773)	0.2% (89)
Infants	99.6% (55,320)	0.4% (217)
Children	99.8% (65,900)	0.2% (120)
Total	99.7% (156,993)	0.3% (426)

RESULTS

Overall 427 of 157,419 (0.27%) patients undergoing cardiac surgery had a diagnosis of CDH including 89 neonates (0.25% of all neonatal index operations), 217 infants (0.39%), and 120 children (0.18%; Table 1). The frequency of concomitant CDH diagnoses varied across diagnostic groups and in neonates and infants was highest for tetralogy of Fallot (0.45%, $n = 41$), coarctation (0.39%, $n = 27$), and complete atrioventricular septal defects (AVSD) (0.31%, $n = 19$; Table 2). The most commonly performed primary procedures of the index cardiac operation in neonates and infants with CDH included ventricular septal defect (VSD) repair ($n = 49$), surgical patent ductus arteriosus (PDA) ligation ($n = 34$), Tetralogy of Fallot repair ($n = 30$), and coarctation repair ($n = 19$; Table 2).

Table 2. The Prevalence of CDH Among the Most Frequent Fundamental Cardiac Diagnoses and Primary Procedure of the Index Cardiac Operation in Neonates and Infants

	With CDH	Without CDH
Total, % (n)	0.33% (306)	99.67% (91,093)
Cardiac diagnosis, % (*n*)		
Transposition of the great arteries		
TGA with IVS	0.1% (3)	99.9% (3192)
TGA with VSD and DORV-TGA	0.1% (3)	99.9% (3539)
Coarctation	0.4% (27)	99.6% (6947)
Coarctation with VSD	0.3% (10)	99.7% (3062)
Single ventricle except HLHS/heterotaxia	0.2% (11)	99.8% (5612)
HLHS	0.1% (10)	99.9% (7861)
Complete AVSD	0.3% (19)	99.7% (6029)
TOF and DORV-TOF	0.5% (41)	99.5% (9032)
Aortic stenosis	0	100.0% (567)
Tricuspid valve disease	0.2% (1)	99.8% (480)
Pulmonary stenosis/ atresia with IVS	0	100.0% (2375)
TAPVC	0.0% (1)	100.0% (2649)
Other	0.5% (180)	99.5% (39,748)
Primary cardiac procedure, % (*n*)		
Norwood	0.2% (12)	99.8% (4900)
Pulmonary artery banding	0.5% (17)	99.5% (3238)
Surgical PDA ligation	0.3% (34)	99.7% (10,305)
Coarctation repair	0.3% (19)	99.7% (5429)
Aortic arch repair	0.4% (15)	99.6% (3430)
Systemic to pulmonary shunt	0.3% (17)	99.7% (5178)
TOF repair	0.3% (30)	99.7% (8693)
Complete AVSD repair	0.3% (13)	99.7% (4997)
VSD repair	0.5% (49)	99.5% (9588)
Glenn/Hemi-Fontan	0.2% (11)	99.8% (6656)
Hybrid procedure	0.9% (8)	873 (99.1%)
Other	0.3% (81)	99.7% (27806)

AVSD, atrioventricular septal defect; DORV, double outlet right ventricle; HLHS, hypoplastic left heart syndrome; IVS, intact ventricular septum; PDA, patent ductus arteriosus; TOF, tetralogy of Fallot; TAPVC, total anomalous pulmonary venous connection; TGA, transposition of the great arteries; VSD, ventricular septal defect.

Table 3 summarizes baseline demographic data for patients with vs those without CDH, stratified by age group and case complexity. Notable findings include a higher frequency of preoperative risk factors and genetic syndromes/chromosomal abnormalities amongst neonates, infants, and children with CHD regardless of case complexity. In particular patients with CDH were significantly more likely to require preoperative mechanical ventilation (Table 3).

Operative mortality, postoperative length of stay (PLOS) and major complication rates were all higher in CDH patients across each of the most common diagnostic and procedural groups when compared to patients without CDH (Table 4). Operative mortality associated with the index cardiac operation was significantly higher in patients with CDH and Tetralogy of Fallot (14.6% vs 4.2%, $P < 0.001$), coarctation (15.4% vs 2.1%, $P = 0.002$), complete AVSD (21.1% vs 4.5%, $P = 0.001$), and single ventricle (27.3% vs 6.3%, $P = 0.029$) than in those diagnostic groups without CDH. Operative mortality was also increased in children with CDH across the majority of the most common primary cardiac procedures (Table 4). Importantly, observed rates of operative mortality were higher in patients with CDH in all groups; however, statistical significance was not reached in all procedure and diagnostic categories, which is likely a reflection of under-powering related to the limited sample size of CDH patients undergoing cardiac surgery.

Outcomes for each age group were then analyzed based on case complexity (Figs. 1–3 and Supplementary Tables E1–E3). Among neonates undergoing low complexity cardiac procedures, operative mortality was twice as high (11.8% vs 5.7%) in neonates with CDH than in those without. That this difference was not statistically significant ($P = 0.252$), is likely a reflection of the relatively small numbers. For STAT 3–5 procedures performed in neonates, operative mortality was significantly higher in CDH patients (34.4% vs 10.3%, $P < 0.001$) (Fig. 1 and Table E1). In both low and high complexity cases, neonates with CDH were more likely to experience any postoperative complication as well as any major postoperative complication. Furthermore, neonates with CDH were more likely to undergo an unplanned reintervention after their index cardiac procedure compared to neonates without CDH (Fig. 1 and Table E1). Among infants, operative mortality was significantly higher for the CDH group irrespective of STAT category (Fig. 2 and

Table 3. Baseline Demographic Data for Patients Undergoing Surgical Repair of Congenital Heart Disease With and Without CDH Stratified by Index Case Complexity

	Low Complexity Cases (STAT 1-2)								
	Neonates			Infants			Children		
Variable	No CDH	CDH	*P* Value	No CDH	CDH	*P* Value	No CDH	CDH	*P* Value
Total, % (*n*)	99.85% (11,061)	0.15% (17)		99.6% (36,398)	0.4% (145)		99.8% (47,663)	0.2% (95)	
Age, median (IQR)	14 (7–22)*	17 (12–22)*	0.15	141 (87–201)*	130 (80–179)*	0.09	5.1 (2.8–10.8)†	4.3 (1.9–9.3)†	0.04
Male gender, % (*n*)	55.4% (6129)	64.7% (11)	0.48	53.6% (19,510)	53.1% (77)	1.00	54.6% (26,006)	52.6% (50)	
Weight at operation, kilograms, mean ± SD	2.3 ± 3.0	2.8 ± 1.8	0.22	5.6 ± 3.3	5.1 ± 1.6	0.01	27.2 ± 21.4	22.0 ± 17.1	0.003
Gestational age, wk, median (IQR)	32 (25–38)	37.5 (34–38.5)	0.06	38 (36–39)	37 (35–39)	0.01	39 (37–40)	38 (35–39)	<0.001
Prematurity (<37 wk), % (*n*)	56.3% (6223)	35.2% (6)	0.14	26.3% (9577)	34.4% (50)	0.03	12.3% (5875)	29.5% (28)	<0.001
Any syndrome/genetic abnormality, % (*n*)	13.2% (1460)	52.9% (9)	<0.001	25.3% (9214)	38.6% (56)	<0.001	18.9% (9025)	31.6% (30)	0.004
Preoperative mechanical ventilation, % (*n*)	45.8% (5065)	70.6% (12)	0.05	9.6% (3,511)	33.8% (49)	<0.001	1.0% (492)	3.2% (3)	0.08
Preoperative mechanical circulatory support, % (*n*)	0.4% (44)	5.9% (1)	0.07	2.2% (80)	3.4% (5)	<0.001	0.14% (70)	0	1.00
Single lung, % (*n*)	0.02% (2)	5.9% (1)	0.005	0.04% (14)	2.8% (4)	<0.001	0.06 (29)	2.1% (2)	0.002
	High Complexity Cases (STAT 3-5)								
	Neonates			Infants			Children		
Variable	No CDH	CDH	*P* Value	No CDH	CDH	*P* Value	No CDH	CDH	*P* Value
Total, % (*n*)	99.7% (23,768)	0.3% (66)		99.6% (17,042)	0.4% (66)		99.8% (12,996)	0.2% (23)	
Age, median (IQR)	7 (4–11)*	10.5 (7–16)*	<0.001	136 (81–196)*	138.5 (63–204)*	0.61	6.2 (2.4–12.2)†	4.9 (1.6–8.11)†	0.33
Male gender, % (*n*)	59.6% (14,163)	54.5% (36)	<0.001	51.7% (8804)	54.5% (36)	0.62	54.4% (7066)	52.2% (12)	0.84
Weight at operation, kilograms, mean ± SD	3.2 ± 1.2	3.0 ± 0.7	0.05	5.4 ± 2.8	5.0 ± 1.8	0.10	28.2 ± 22.2	21.7 ± 16.5	0.13
Gestational age, wk, median (IQR)	39 (37–39)	38 (37–39)	0.06	38 (36–39)	38 (36–39)	0.54	39 (37–40)	37 (36–39)	0.19
Prematurity (<37 wk), % (*n*)	12.7% (3030)	65.2% (43)	0.03	22.5% (3839)	22.7% (15)	1.00	13.3% (1727)	26% (6)	0.11
Any syndrome/genetic abnormality, % (*n*)	21.7% (5157)	43.9% (29)	<0.001	46.7% (7955)	57.6% (38)	0.08	27.1% (3518)	47.8% (11)	0.03
Preoperative mechanical venitilation, % (*n*)	30.1% (7155)	65.2% (43)	<0.001	12.5% (2133)	31.8% (21)	<0.001	4.3% (565)	8.7% (2)	0.27
Preoperative mechanical circulatory support, % (*n*)	0.7% (177)	12.1% (8)	<0.001	0.99% (168)	1.5% (1)	0.49	3.0% (395)	0	1.00
Single lung, % (*n*)	0.05% (12)	9.1% (6)	<0.001	0.11% (19)	6.1% (4)	<0.001	0.1% (13)	4.3% (1)	0.03

CHD, congenital heart disease; IQR, interquartile range; SD, standard deviation.
*Days.
†Years.

Table 4. Operative Mortality, Postoperative Length of Stay, and Prevalence of Major Postoperative Complications Stratified by Cardiac Diagnosis and Primary Cardiac Procedure Based on the Presence of Congenital Diaphragmatic Hernia

	Operative Mortality, % (*n*)			Postoperative Length of Stay, Median (IQR)			Major Postoperative Complications, % (*n*)		
	No CDH	CDH	*P* Value	No CDH	CDH	*P* Value	No CDH	CDH	*P* Value
Cardiac diagnosis									
Coarctation	2.1% (140)	15.4% (4)	0.002	8.0 (5–16)	55 (21–99)	<0.001	10.6% (696)	42.3% (11)	<0.001
Coarctation with VSD	4.2% (127)	20% (2)	0.066	15 (8–31)	59 (57–86)	<0.001	20.9% (608)	33.3% (3)	0.407
Tetralogy of Fallot	1.9% (165)	14.6% (6)	<0.001	7 (5–12)	25 (8–57)	<0.001	10.1% (859)	23.7% (9)	0.012
Complete AVSD	4.5% (267)	21.1% (4)	0.001	9 (6–20)	40 (9–97)	<0.001	15.1% (868)	36.8% (7)	0.017
Single ventricle	6.3% (349)	27.3% (3)	0.029	11 (6–27)	37 (9–126)	0.061	23.3% (1226)	27.3% (3)	0.725
Hypoplastic left heart syndrome	13.3% (1021)	25% (2)	0.289	21 (9–43)	84 (6–140)	0.1498	32.4% (2396)	44.4% (4)	0.484
Transposition of the great arteries	4.3% (283)	0	1.00	12 (8–21)	28.5 (10–46)	0.0845	20.4% (1297)	16.7% (1)	1.00
Primary cardiac procedure									
Aortic arch repair	4.1% (137)	21.4% (3)	0.018	13 (8–26)	56.5 (30–107)	<0.001	18.8% (613)	40% (6)	0.047
Complete AVSD repair	2.9% (141)	15.4% (2)	0.054	8 (6–16)	16 (9–44)	0.007	12.6% (597)	23.1% (3)	0.219
Coarctation repair	1.4% (74)	10.5% (2)	0.029	6 (4–12)	28 (6–83)	0.003	7.3% (378)	33.3% (6)	0.001
Tetralogy of Fallot repair	2% (170)	13.3% (4)	0.003	7 (5–12)	24.5 (8–62)	<0.001	10.4% (846)	25.9% (7)	0.018
Norwood	15.9% (766)	41.7% (5)	0.03	29 (18–51)	85.5 (20.5–132.5)	0.049	42.6% (1984)	63.6% (7)	0.223
Glenn	2.3% (146)	0	1.00	7 (5–12)	9 (7–14)	0.2519	14.3% (880)	11.1% (1)	1.00
Hybrid	27.6% (236)	28.6% (2)	1.00	36 (18–83)	119.5 (59–145)	0.013	38.1% (317)	62.5% (5)	0.27
Pulmonary artery banding	9.8% (309)	41.2% (7)	<0.001	19 (8–44)	55 (19–140)	0.014	23.2% (710)	23.5% (4)	1.00
PDA ligation	6.3% (609)	15.6% (5)	0.049	56 (6–99)	46.5 (11–119.5)	0.402	8% (773)	25% (8)	0.003
VSD repair	0.9% (80)	8.2% (4)	<0.001	5 (4–8)	20.5 (7–52.5)	<0.001	5.2% (481)	8.9% (4)	0.302
Systemic to pulmonary shunt	8% (409)	17.6% (3)	0.153	17 (9–33)	48 (20–103)	<0.001	27.7% (1351)	43.8% (7)	0.165

AVSD, atrioventricular septal defect; PDA, patent ductus arteriosus; VSD, ventricular septal defect.

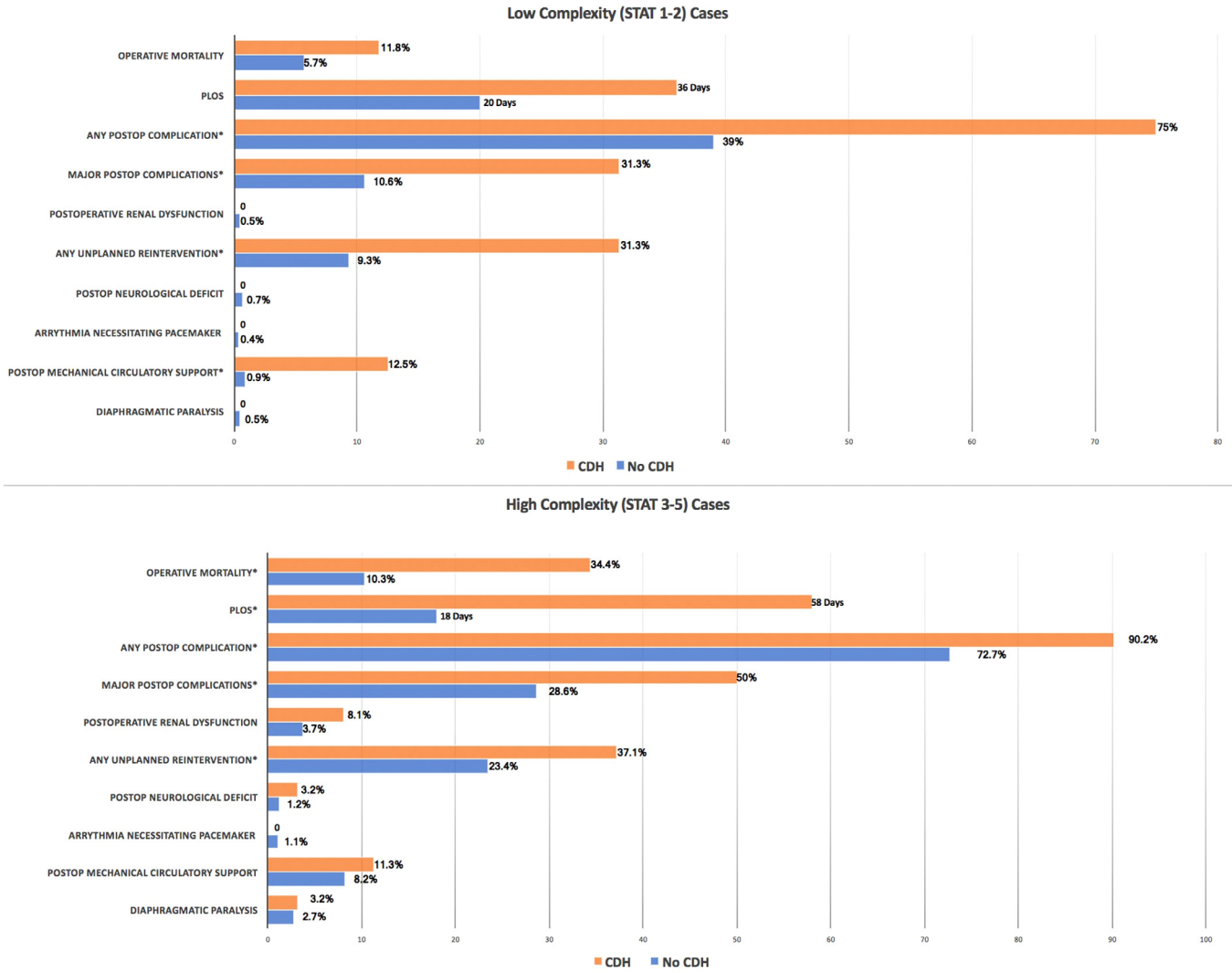

Figure 1. Outcomes following index operation in neonates with and without CDH based on procedural complexity. PLOS, postoperative length of stay. * Statistically significant difference ($P < 0.05$).

Table E2). Infants with CDH also suffered higher rates of major postoperative complications regardless of case complexity category. Neonates and infants with CDH undergoing cardiac surgery also experienced longer PLOS when compared to those without CDH (Figs. 1 and 2). However, in children older than 1 year, mortality did not differ significantly between those with and without CDH undergoing low complexity (1.1% vs 0.5%, $P = 0.40$) or high complexity (4.3% vs 2.5% $P = 0.45$) cardiac procedures (Fig. 3 and Table E3). No differences in PLOS or postoperative morbidity were observed in children undergoing high complexity cases. In children undergoing low complexity cases, those with CDH experienced significantly longer PLOS, higher incidence of major postoperative complications (12% vs 5.2%, $P = 0.008$), and higher rates of unplanned reintervention (9.8% vs 3.8%, $P = 0.008$; Fig. 3 and Table E3).

DISCUSSION

In this study, the presence of CDH in pediatric patients undergoing surgery for congenital heart disease was rare. However, as expected, a concomitant diagnosis of CDH did portend worse outcomes in children undergoing cardiac surgery, particularly in neonates and infants. Awareness of the impact of a concomitant CDH in pediatric patients with congenital heart disease can have important implications for risk stratification, medical and surgical decision making, prognosis, and family counseling. Previous studies have investigated the impact of cardiac anomalies on children undergoing repair of CDH.[4,10,11] Ours is the first large multicenter study to look at the problem from the converse perspective, ie, investigating the impact of CDH on patients undergoing surgery to palliate or repair cardiac anomalies. This analysis was undertaken to provide empirically derived data based on multi-institutional

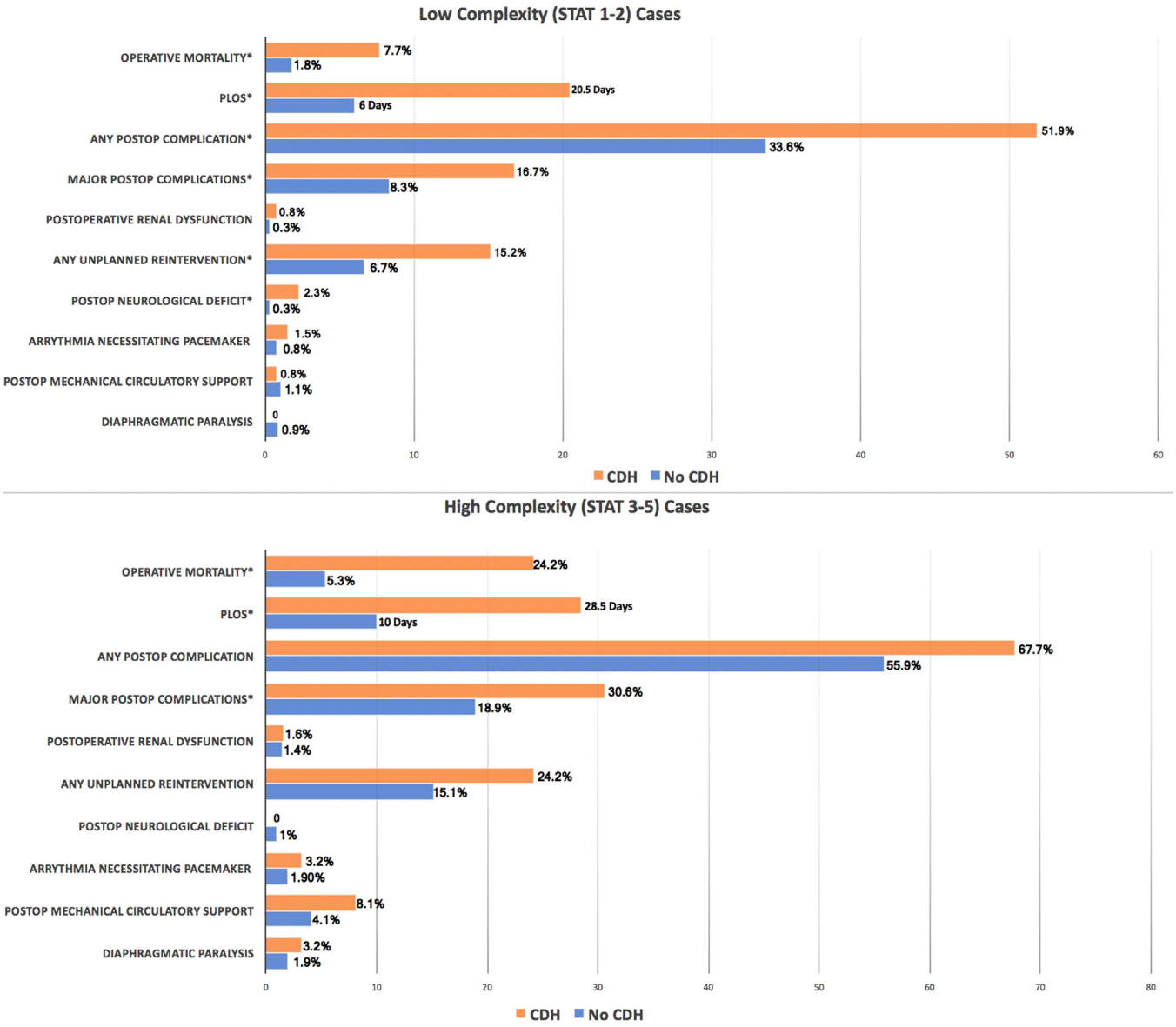

Figure 2. Outcomes following index operation in infants with and without CDH based on procedural complexity. PLOS, postoperative length of stay. * Statistically significant difference ($P < 0.05$).

experiences in the management of this high-risk subset of pediatric patients requiring cardiac surgery.

Previous studies have shown that mortality in children with CDH is dependent on the presence of a number of associated risk factors, of which congenital heart disease is the most impactful.[12] Further, survival has been shown to be significantly worse in infants with CDH with major cardiac anomalies compared with minor cardiac anomalies, with some studies suggesting that only one third of CDH patients with major cardiac anomalies will survive to discharge.[13] However, comparisons of our findings to previous studies in CDH patients are difficult as these studies focus on outcomes in patients undergoing repair of CDH to investigate the impact of congenital heart disease. Our study investigates the impact of a concomitant diagnosis of CDH on children undergoing surgical repair of cardiac anomalies, and as such, it may be imprudent to extrapolate outcomes from the aforementioned studies which focus solely on children with CDH. Other studies and case series have investigated the impact of noncardiac anomalies on patients with congenital heart disease, showing that these concomitant anomalies are particularly impactful in neonates with major cardiac anomalies including hypoplastic left heart syndrome and TGA.[14–17] The presence of genetic and noncardiac abnormalities was highly variable across diagnostic groups but was frequently associated with increased morbidity and mortality in patients with congenital heart disease.

When analyzing the entire population based on the most common fundamental cardiac diagnoses and primary cardiac procedures, the presence of CDH was associated with worse operative mortality, postoperative length of stay, and major

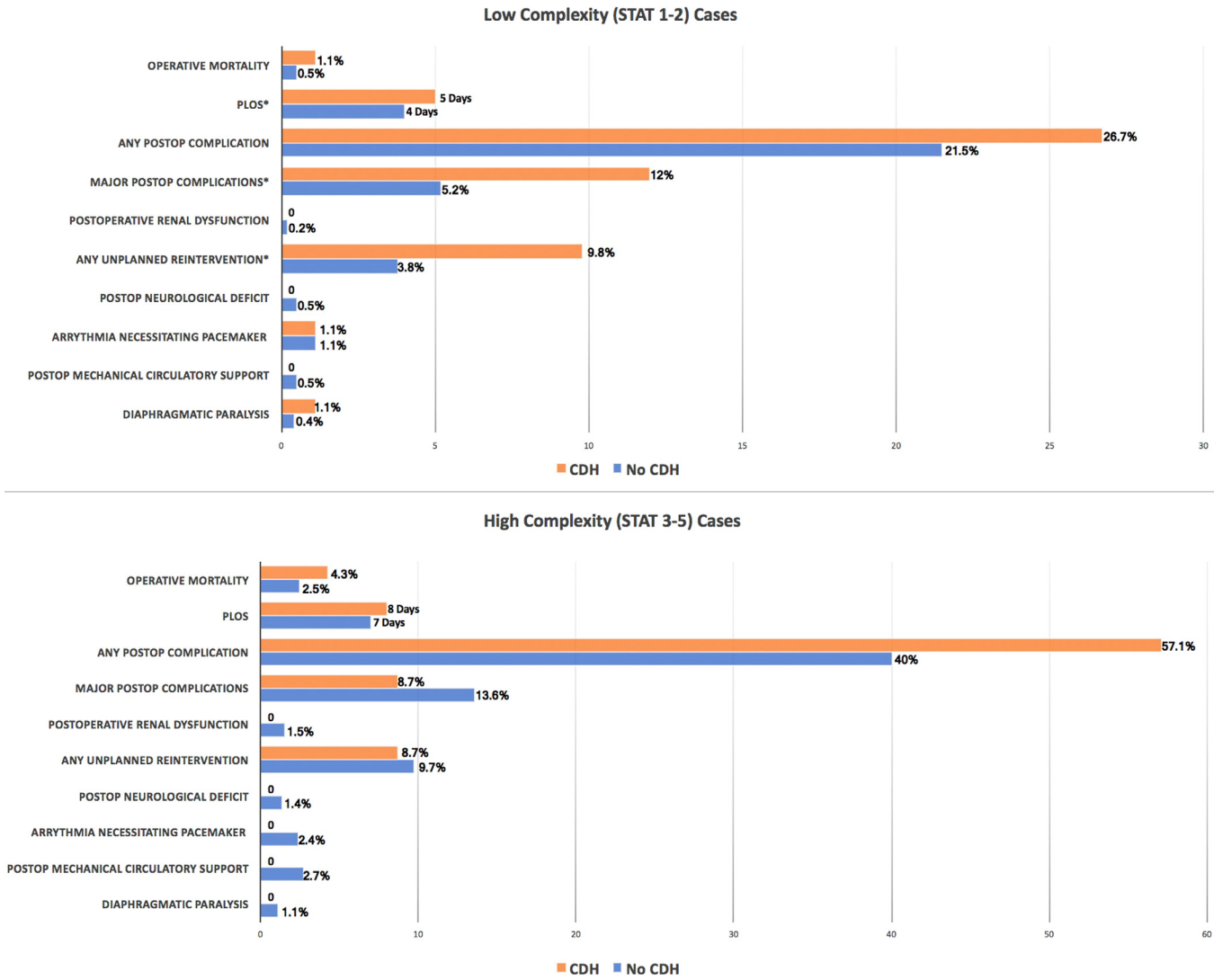

Figure 3. Outcomes following index operation in children with and without CDH based on procedural complexity. PLOS, postoperative length of stay. * Statistically significant difference ($P < 0.05$).

complications in the majority of the most common diagnostic and procedural groups. When the population is stratified by age groups, neonates, and infants requiring cardiac surgery suffer worse outcomes when a concomitant diagnosis of CDH is present regardless of cardiac case complexity, although these differences appear to be more pronounced in higher complexity cases. Children older than 1 year of age experienced less disparate postoperative outcomes based on presence of a concomitant diagnosis of CDH. This may be a reflection of survival bias as children surviving past 1 year of life with a diagnosis of CDH likely have either tolerated prior intervention for their CDH or may have a clinically insignificant diaphragmatic defect. Nonetheless, older children do not appear to suffer the negative impact of a diaphragmatic hernia that is observed in younger patients undergoing cardiac surgery.

The current series represents the largest study in the literature to evaluate outcomes in this high-risk subset of children undergoing cardiac surgery. We believe that these data will be helpful in providing surgeons, cardiologists, pediatric surgeons, neonatologists, and maternal-fetal medicine specialists with realistic prognostic information to be used when counseling families. The majority of cases of CDH and major congenital cardiac anomalies are diagnosed prenatally.[1,18] As such, these data can be used as a resource in prenatal counseling for families when faced with challenging concomitant diagnoses. In addition, although outcomes in most diagnostic and procedural categories are worse for patients with concomitant CDH, variation in the rates of morbidity and mortality exist across procedural groups.

This study has several limitations. Although the STS-CHSD collects data pertaining to a robust set of variables, unmeasured confounding factors may have influenced the associations observed in our results. Furthermore, the STS-CHSD does not collect detailed clinical data related to the noncardiac diagnosis of congenital diaphragmatic hernia. The database does not collect data regarding size or location of the diaphragmatic hernia,

management of CDH, or the timing of surgical repair of CDH in relation to cardiac intervention. For any individual patient we are unable to ascertain whether CDH repair had been performed, or the timing of CDH repair. Therefore, we are only able to analyze these data with CDH as a binary diagnostic variable and are unable to investigate the impact of prior CDH management or defect morphology on outcomes in pediatric patients undergoing cardiac surgery. Additional limitations inherent to the retrospective study design, and the voluntary nature of the database may also impact our findings. Finally, for many of our diagnostic, procedural and age subcategories, the numbers of patients with CDH were quite limited, often resulting in substantially under-powered analyses. We have elected to report *P* values to aid readers in interpreting truly significant findings. However, we recommend caution in interpreting nonsignificant *P* values, as the lack of significance may often simply reflect underpowering.

CONCLUSION

CDH is a rare malformation and occurs in a small percentage of patients undergoing surgical repair of congenital heart disease. Concomitant diagnosis of diaphragmatic hernia portends increased morbidity and mortality in infants and neonates undergoing surgery for congenital heart disease. For older children undergoing cardiac surgery beyond infancy, a diagnosis of CDH is not consistently associated with worse outcomes.

SUPPLEMENTARY MATERIAL

Supplementary material associated with this article can be found in the online version at https://doi.org/10.1053/j.semtcvs.2018.09.014.

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