Tracheal surgery for airway anomalies associated with increased mortality in pediatric patients undergoing heart surgery: Society of Thoracic Surgeons Database analysis

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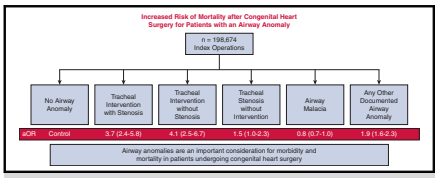
ABSTRACT

Objectives: Airway anomalies are common in children with cardiac disease but with an unquantified impact on outcomes. We sought to define the association between airway anomalies and tracheal surgery with cardiac surgery outcomes using the Society of Thoracic Surgery Congenital Heart Surgery Database.

Methods: Index cardiac operations in children aged less than 18 years (January 2010 to September 2018) were identified from the Society of Thoracic Surgery Congenital Heart Surgery Database. Patients were divided on the basis of reported diagnosis of an airway anomaly and subdivided on the basis of tracheal lesion and tracheal surgery. Multivariable analysis evaluated associations between airway disease and outcomes controlling for covariates from the Society of Thoracic Surgery Congenital Heart Surgery Database Mortality Risk Model.

Results: Of 198,674 index cardiovascular operations, 6861 (3.4%) were performed in patients with airway anomalies, including 428 patients (0.2%) who also underwent tracheal operations during the same hospitalization. Patients with airway anomalies underwent more complex cardiac operations (45% vs 36% Society of Thoracic Surgeons/European Association for Cardiothoracic Surgery Congenital Heart Surgery Mortality category ≥3 procedures) and had a higher prevalence of preoperative risk factors (73% vs 39%; both $P < .001$). In multivariable analysis, patients with airway anomalies had increased odds of major morbidity and tracheostomy ($P < .001$). Operative mortality was also increased in patients with airway anomalies, except those with malacia. Tracheal surgery within the same hospitalization increased the odds of operative mortality (adjusted odds ratio, 3.9; $P < .0001$), major morbidity (adjusted odds ratio, 3.7; $P < .0001$), and tracheostomy (adjusted odds ratio, 16.7; $P < .0001$).

Conclusions: Patients undergoing cardiac surgery and tracheal surgery are at significantly higher risk of morbidity and mortality than patients receiving cardiac surgery alone. Most of those with unoperated airway anomalies have higher morbidity and mortality, which makes it an important preoperative consideration. (J Thorac Cardiovasc Surg 2021;161:1112-21)



Airway anomalies and interventions are associated with increased morbidity and mortality.

CENTRAL MESSAGE

Cardiac surgery in pediatric patients and airway anomalies, with or without concomitant tracheal surgery, are associated with increased risk of morbidity and mortality.

PERSPECTIVE

Associations between outcomes from cardiac surgery in pediatric patients and coexisting airway anomalies, with or without concomitant tracheal surgery, are incompletely understood. This report based on analysis of a nationwide dataset demonstrates the elevated risk of adverse outcomes, including mortality. These findings are important when discussing expectations with families and providers.

See Commentaries on pages 1122, 1123, and 1124.

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Abbreviations and Acronyms

aOR = adjusted odds ratio
CHD = congenital heart disease
CI = confidence interval
STAT = Society of Thoracic/European Association for Congenital Heart Surgery Mortality
STS-CHSD = Society of Thoracic Surgeons Congenital Heart Surgery Database

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The most common extracardiac anomaly in children with congenital heart disease (CHD) is an associated airway anomaly, seen in an estimated 3% to 4% of patients.[1,2] However, this definition can be broad and often challenging to fully understand the specific airway anomaly and its impact on presentation and repair. Anomalies vary from tracheobronchial malacia to tracheal stenosis and tracheoesophageal fistulas, some of which may require surgical intervention.[2,3] It has also been reported that 50% to 70% of patients with congenital tracheal stenosis have a cardiac anomaly.[4,5]

Despite their frequent association, the outcomes of patients with cardiac disease who undergo repair of both cardiac anomalies and tracheal lesions during the same hospitalization are largely unknown. Furthermore, it is unknown to what extent airway pathology not requiring surgical intervention increases the risk for morbidity and mortality in patients undergoing cardiac surgery, including the need for a tracheostomy. Unfortunately, prior literature often focuses on comparing patients undergoing airway surgery, with or without cardiac defects.[6-8] Although several small, single-center studies have identified airway anomalies as a risk factor for morbidity and mortality in children with CHD and a nationwide study from Taiwan reported an adjusted mortality hazard ratio of 1.76,[1,9,10] these studies have all met challenges in defining and grouping the airway anomalies. Additionally, they included patients with CHD who did not undergo cardiac surgery, and the complexity of cardiac defect and repair was not stratified. Therefore, the current literature still leaves us asking how congenital airway anomalies affect survival in children undergoing surgery for various CHDs. We used the Society of Thoracic Surgeons Congenital Heart Surgery Database (STS-CHSD) to assess the association of airway anomalies with CHD requiring surgical repair and to quantify the morbidity and mortality for children with airway anomalies and tracheal surgeries who underwent cardiac surgery.

TABLE 1. Composition of airway group

Airway group	N = 6861 (100%)
Tracheal intervention for stenosis	283 (4.1%)
Tracheal intervention without stenosis	145 (2.1%)
Tracheal stenosis without intervention	516 (7.5%)
Airway malacia*	3853 (56.1%)
All other airway anomalies	2064 (30.0%)

*Includes tracheomalacia, laryngomalacia, or bronchomalacia not meeting criteria for the aforementioned groups.

MATERIALS AND METHODS

Data Source

The STS-CHSD includes data from more than 95% of all pediatric heart centers in the United States and Canada.[11] Perioperative, operative, and outcomes data are collected on all patients undergoing pediatric and congenital heart surgery at participating centers using standard definitions.[12] Evaluation of data quality includes intrinsic verification of data, along with formal data audits of 10% of participating sites each year, conducted by a panel of independent data-quality personnel and pediatric cardiac surgeons.[13-15] The Duke Clinical Research Institute serves as the primary analytic center for the STS databases. This study was approved by the Duke University Institutional Review Board with waiver of informed consent and by the STS-CHS Database Access and Publications Committee.

Patient Population

All first cardiac surgeries (index operation) of each episode of care performed in children aged less than 18 years from January 1, 2010, to September 30, 2018, were identified from the STS-CHSD (n = 198,674 operations from 128 centers). The patients were then grouped as patients with airway anomalies (airway group) or the control group. The airway group was then further classified into 5 mutually exclusive categories based on performance of a tracheal procedure and the type of airway anomaly (Table 1). Patients were grouped through a hierarchy so that any patient meeting criteria for subgroup higher on the table could not be in a lower group (ie, a patient having a tracheal intervention for tracheal stenosis who also had bronchial malacia documented would only be counted in the tracheal intervention group). A concomitant tracheal procedure was defined as any that occurred at the time of the index operation or later within the same hospitalization. Seventeen patients underwent tracheal intervention before cardiac surgery and were excluded from this analysis. Patients having a tracheal procedure were further divided into those with a diagnosis of tracheal stenosis versus another diagnosis (eg, tracheoesophageal fistula). Likewise, the patients who did not have a tracheal surgery were divided into those with tracheal stenosis, those with large airway malacia (laryngeal, tracheal, or bronchial), and those with any other airway anomaly.

Data Collection and Definitions

Data collection included demographics, cardiac diagnoses, preoperative risk factors, intraoperative details, surgical procedures, the Society of Thoracic/European Association for Congenital Heart Surgery Mortality (STAT) Category of the index cardiovascular surgical operation, postoperative complications, length of stay, and operative mortality. The STAT mortality categories are based on a statistical estimate of the risk of mortality for a given operation. STAT mortality category 1 represents operations

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TABLE 2. Preoperative demographics and risk factors

Variable	Overall, N = 198,674	Control group, N = 191,813	Airway anomaly group, N = 6861	*P* value
Birth weight (kg)	3.0 [2.5-3.4]	3.0 [2.5-3.4]	2.8 [2.3-3.2]	<.001
Premature	22.2% (41,615)	21.9% (39,433)	32.6% (2182)	<.001
Gender, female	45.1% (89,602)	45.1% (86,435)	46.2% (3167)	.182
Race				<.001
White	53.8% (99,071)	53.7% (95,394)	56.2% (3677)	
Black	16.3% (30,007)	16.2% (28,857)	17.6% (1150)	
Hispanic	18.9% (34,799)	19.0% (33,706)	16.7% (1093)	
Other	11.0% (20,339)	11.1% (19,712)	9.6% (627)	
STAT Category				<.001
Level 1-3	75.0% (147,197)	75.2% (142,392)	60.8% (4805)	
Level 4-5	25.0% (49,018)	24.8% (47,036)	29.2% (1982)	
Age at surgery (mo)	30.0 [6.1]	30.4 [6.1-221.9]	22.9 [6.0-83.9]	<.001
Single ventricle physiology	14.6% (28,976)	14.7% (28,132)	12.3% (844)	<.001
Any major noncardiac Anatomic anomaly	13.0% (25,007)	10.3% (19,110)	86.5% (5897)	<.001
Any preoperative risk factor	40.2% (77,941)	39.1% (73,025)	72.5% (4916)	<.001
Mechanical ventilation	13.5% (26,083)	12.9% (24,208)	27.7% (1875)	<.001
Tracheostomy	0.7% (1404)	0.5% (885)	7.7% (519)	<.001
Gastrostomy	5.6% (10,862)	4.9% (9229)	24.1% (1663)	<.001
Chromosomal Abnormality	17.6% (33,951)	17.0% (31,563)	35.1% (2388)	<.001

There is considerable difference in the preoperative demographics and risk factors between patients with and without airway anomalies. *STAT,* Society of Thoracic/European Association for Congenital Heart Surgery Mortality.

with the lowest overall predicted mortality risk, and STAT category 5 represents operations with the highest overall predicted mortality risk.[16] Single ventricle diagnosis was defined by the following fundamental diagnosis codes within the STS-CHSD reporting form: single ventricle, double inlet left ventricle; single ventricle, double inlet right ventricle; single ventricle, mitral atresia; single ventricle, tricuspid atresia; single ventricle, unbalanced atrioventricular canal; single ventricle, heterotaxia syndrome; single ventricle, other; single ventricle + total anomalous pulmonary venous connection; and hypoplastic left heart syndrome.

Primary outcomes of interest included operative mortality, defined as hospital mortality or outpatient mortality within 30 days of surgery, major morbidity, and postoperative tracheostomy. Major morbidity is defined as renal failure, neurologic deficit, arrhythmia necessitating pacemaker, postoperative mechanical circulatory support, bleeding requiring reoperation, unplanned cardiac reoperation, unplanned interventional cardiovascular catheterization, postoperative cardiac arrest, or phrenic nerve injury.

Statistical Analysis

Baseline demographic and clinical data as well as outcomes of interest were evaluated using standard descriptive statistics including median (interquartile range) for continuous variables and n (%) for discrete variables. Preoperative characteristics were compared using Kruskal–Wallis for continuous variables as well as chi-square analysis and Fisher exact when appropriate for discrete variables. Associations between airway disease and outcomes including operative mortality, composite major morbidity, and postoperative tracheostomy were assessed using multivariable hierarchical logistic regression analysis. Covariates used in the STS-CHSD mortality risk model were included in addition to single ventricle status (Table E1).[16,17] Clustering of patients within centers was handled by including a random intercept for each center. The same modeling approach was applied to characterize associations between risk factors and outcomes just in the airway disease cohort. Adjusted odds ratios (aORs) are presented with 95% confidence intervals (CIs). All analyses were performed using SAS 9.4 (SAS Institute Inc, Cary, NC).

RESULTS

Patient Population

Of the 198,674 index operations identified in the database, 6861 (3.4%) were performed in patients with airway anomalies, including 428 (0.2%) who underwent concomitant cardiac and tracheal operations. Of those, 283 (66.1%) had a diagnosis of tracheal stenosis, and the remainder (n = 145, 33.9%) had another indication for tracheal intervention. Of those, 34 had a diagnosis of tracheoesophageal fistula. For patients without tracheal intervention, 516 (8.0%) had stenosis without intervention, 3853 (59.9%) had large airway malacia, and 2064 (32.0%) had reported airway disease without further specification (Table 1).

Preoperative Characteristics

Table 2 shows the patient characteristics by presence of airway anomaly. Those with airway anomalies had lower birth weight, were more often premature, were younger and had a lower weight at time of cardiac surgery, were more likely to have other (nonairway, noncardiac) anatomic anomalies and chromosomal anomalies, and had

TABLE 3. Unadjusted intraoperative and postoperative outcomes

Variable	Overall, N = 198,674	Control group, N = 191,813	Airway anomaly group, N = 6861	*P* value
Cardiopulmonary bypass time (min)	99 [66-145]	98 [66-145]	115 [79-169]	<.001
Crossclamp time (min)	50 [24-82]	50 [24-82]	52 [22-88]	.099
Operative mortality	3.5% (6889)	3.4% (6454)	6.4% (435)	<.001
Major morbidity	11.9% (22,573)	11.6% (21,160)	21.4% (1413)	<.001
Postoperative mechanical circulatory support	2.8% (5290)	2.7% (4970)	4.8% (320)	<.001
Unplanned reoperation	7.7% (14,641)	7.5% (13,674)	14.6% (967)	<.001
Neurologic deficit	0.8% (1507)	0.8% (1401)	1.6% (106)	<.001
Prolonged mechanical ventilation	8.2% (15,611)	7.7% (14,143)	22.2% (1468)	<.001
Postoperative tracheostomy	1.0% (1914)	0.8% (1391)	7.9% (523)	<.001
Postoperative length of stay (d)	7 [4-17]	7 [4-16]	16 [6-51]	<.001

Nearly all reported surgical outcomes are different between the control group and the patients with airway anomalies except for crossclamp time.

preoperative risk factors including preoperative mechanical ventilation and tracheostomy. For patients undergoing tracheal intervention without a diagnosis of tracheal stenosis who had a preoperative tracheostomy (n = 21), most had malacia (n = 15, 71.4%) and 6 (28.6%) had tracheoesophageal fistula. They also underwent more complex operations by STAT category. Of note, we found that 416 (10.8%) of the patients with airway malacia had a vascular ring, and 115 patients (22.3%) with tracheal stenosis without intervention had a vascular ring. Baseline characteristics by subgroup are presented in Table E2.

Unadjusted Outcomes

Table 3 displays unadjusted major surgical outcomes comparing the airway group with the control group. Patients within the airway group demonstrated significantly higher overall mortality (6.4% vs 3.4%) and composite major morbidity (22.6% vs 12.1%), more frequently needed prolonged mechanical ventilation (22.2% vs 7.7%) and tracheostomy (7.9% vs 0.8%), and had longer postoperative length of stay (median 16 days vs 7 days), $P < .001$ for all. Primary postoperative outcomes by subgroup are displayed in Table 4, indicating increased risk for patients receiving a tracheal intervention. There were 523 patients with postoperative tracheostomy, of whom 447 (85.5%) had a complication of prolonged postoperative mechanical ventilation. Of the 320 patients with postoperative mechanical support, 8 (2.5%) had a procedure for ventricular assist device implantation. Tables E3 and E4 summarize data for the low complexity (STAT 1-3) and high complexity (STAT 4-5) groups with similar overall findings.

Of the 283 patients with tracheal stenosis undergoing tracheal intervention, 228 (80.6%) underwent tracheal intervention concomitant with their index cardiac operation. For those with tracheal stenosis who had a tracheal procedure after their index operation, the median time to intervention was 22 days (13-84 days). For those without tracheal stenosis undergoing tracheal intervention, 85 of 145 (58.6%) had the tracheal intervention performed concomitant with their index cardiac surgery. For the remainder, the median time to intervention was 49 days (23-89 days).

Adjusted Risk Analysis by Subgroup

Multivariable models for adjusted odds of mortality, major morbidity, and tracheostomy are presented in Table 5 with morbidity and mortality presented in Figure 1. Compared with the control group, patients undergoing a

TABLE 4. Major operative end points by subgroup

	Control group	Tracheal intervention for stenosis	Tracheal intervention without stenosis	Tracheal stenosis without intervention	Airway malacia	Any other airway anomaly	*P* value
Operative mortality	3.4% (6454)	14.4% (40)	18.1% (26)	5.9% (30)	4.5% (173)	8.1% (166)	<.001
Major morbidity	11.6% (21,160)	35.1% (85)	40.2% (55)	21.2% (102)	21.6% (812)	18.0% (359)	<.001
Postoperative tracheostomy	0.8% (1391)	13.6% (33)	31.4% (43)	5.6% (27)	8.8% (331)	4.5% (89)	<.001

There is considerable difference in the unadjusted outcomes among the different airway subgroups and the control group.

TABLE 5. Adjusted odds for morbidity and mortality

Airway group	Adjusted odds of mortality	*P* value	Adjusted odds of major morbidity	*P* value	Adjusted odds of tracheostomy	*P* value
Nonairway patients	Reference	—	Reference	—	Reference	—
Tracheal intervention for tracheal stenosis	3.71 (2.4-5.75)	<.0001	3.57 (2.59-4.91)	<.0001	8.79 (5.41-14.29)	<.0001
Tracheal intervention without tracheal stenosis	4.07 (2.46-6.73)	<.0001	3.74 (2.51-5.57)	<.0001	33.21 (21.12-52.21)	<.0001
Tracheal stenosis without intervention	1.52 (1-2.3)	.0504	1.89 (1.46-2.44)	<.0001	4.85 (3.06-7.67)	<.0001
Airway malacia	0.80 (0.68-0.95)	.0117	1.44 (1.32-1.57)	<.0001	7.38 (6.37-8.55)	<.0001
All other airway anomalies	1.94 (1.62-2.33)	<.0001	1.55 (1.37-1.75)	<.0001	3.59 (2.82-4.57)	<.0001

There is an increased adjusted risk for morbidity, mortality, and tracheostomy for most airway anomaly subgroups. aOR (95% CI).

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tracheal intervention had increased odds of mortality (aOR, 3.9; 95% CI, 2.8-5.4), major morbidity (aOR, 3.7; 95% CI, 2.9-4.8), and tracheostomy (aOR, 8.8; 95% CI, 5.4-14.3), all $P < .001$. Patients with airway malacia without intervention had increased odds of major morbidity (aOR, 1.4; 95% CI, 1.3-1.6) and tracheostomy (aOR, 7.4; 95% CI, 6.4-8.6) (both $P < .001$). Patients with unclassified airway anomalies had increased odds of mortality (aOR, 1.9; 95% CI, 1.6-2.3), major morbidity (aOR, 1.6; 95% CI, 1.4-1.8), and tracheostomy (aOR, 3.6; 95% CI, 2.8-4.6) (all $P < .001$).

Adjusted Risk Analysis Within Airway Group

Multivariable analysis identified several risk factors for poor outcome among patients with airway anomalies (Table 6). The greatest risk factors for mortality were higher STAT category (aOR, 5.3; 95% CI, 3.0-9.4; $P < .001$ for STAT 5 vs STAT 1); omphalocele, gastroschisis, or congenital diaphragmatic hernia (aOR, 3.8; 95% CI, 2.2-6.6; $P < .001$); and tracheal intervention (aOR, 3.0; 95% CI, 2.1-4.2; $P < .001$). Additionally, patients undergoing a tracheal intervention had a 2.2-fold (95% CI, 1.8-2.8) increased odds of major morbidity and a 2.7-fold (95% CI, 2.0-3.7) increased odds of tracheostomy (both $P < .001$) (Figure 2).

DISCUSSION

This nationwide analysis of the STS-CHSD demonstrates on a broad scale that the presence of an airway anomaly in pediatric patients, particularly one who requires surgery concomitant with or after a cardiovascular surgical operation but during the same hospitalization, is a risk factor for major morbidity and mortality compared with similarly complex operations in children without airway disease. By stratifying analyses across different levels of case complexity, we have shown that airway anomalies affect outcomes for all children. The study found that more than 3% of children undergoing cardiac surgery have an associated airway anomaly, with laryngeal, tracheal, and bronchial malacia being the most commonly associated airway anomalies. Although concurrent airway intervention was less common, it increased the risk within the airway group by 3-fold, and this was most often an intervention for tracheal stenosis.

There are only a few studies that have measured the impact of airway anomalies and interventions on the outcomes of children undergoing cardiac surgery.[1,4,9,10] However, they do suggest an elevated risk. Although being born with an airway anomaly is not modifiable, perhaps certain steps can be taken to better support these children and improve their outcomes. Additionally, further consideration could be given to whether intervening on the tracheal or cardiac anomaly first would change the outcome; however, certain operations such as tracheal stenosis with a vascular ring are probably best addressed by 1 surgical intervention.[6] Our study excluded 17 patients who had their tracheal operation before their index cardiac operation, but timing of cardiac versus tracheal intervention and time between the 2 are clinically important questions that have proven difficult to answer. Furthermore, the breadth of cardiac defects and airway anomalies make a case-by-case analysis impossible, but the overall sentiment is to approach these cases with a heightened sense of preparation and caution. This highlights the utility of the STS-CHSD definitions, which were developed with input from experts from each related field to harmonize with existing definitions from other societies, in stratifying the complexity of the cardiac cases to best elucidate the impact of the airway anomalies.

Patients with airway anomalies were younger at the time of their cardiac surgery, which may be due to their more complex cardiac anomalies or an inability to compensate for their cardiac defects longer because of their airway disease. Additionally, the greater presence of other preoperative risk factors may have been an impetus for more urgent

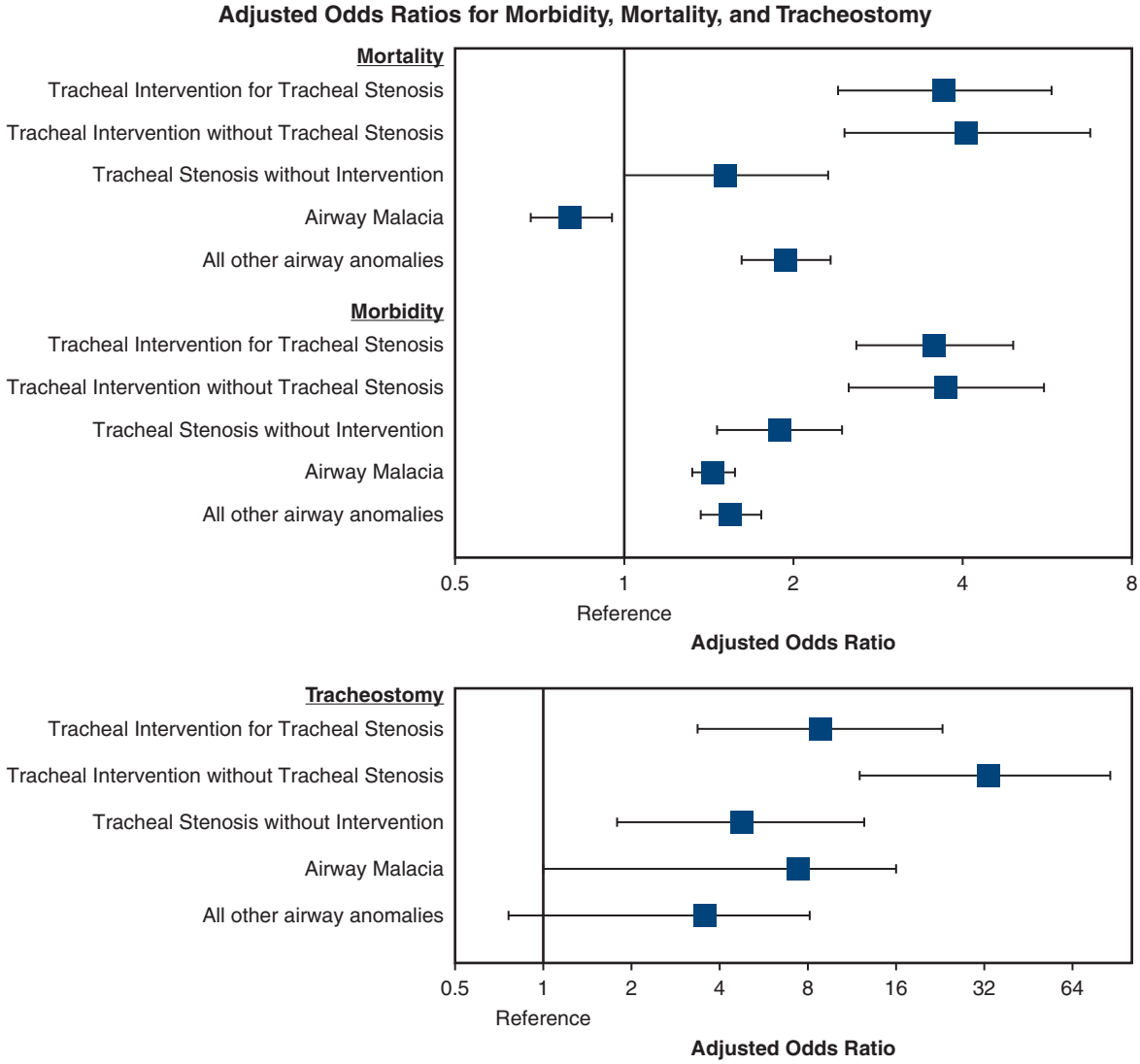

FIGURE 1. aORs for morbidity, mortality, and tracheostomy for each of the airway anomaly subgroups. The reference group is the control group of patients who did not have any airway anomalies or interventions with a null value of 1.0. Nearly every subgroup has an increased risk of morbidity, mortality, and tracheostomy.

cardiac repair. This was associated with greater mortality, and therefore patients being less likely to undergo further palliative or corrective procedures at a later age. Although a greater incidence of prolonged mechanical ventilation and greater prevalence of tracheostomy in the airway group seems intuitive, these are both associated with high mortality after cardiac surgery.[18-20] Overall complication rates varied widely by complication with a relatively low incidence of neurologic defects in both groups, 0.8% versus 1.6% compared with unplanned reoperation and 7.5% in controls versus 14.6% in patients with airway malacia. Nonetheless, our multivariable analysis confirmed that major morbidity was significantly higher in all airway anomaly subgroups ranging from an aOR of 1.44 (95% CI, 1.32-1.57) in patients with airway malacia to 3.74 (95% CI, 2.51-5.57) in those receiving tracheal intervention without tracheal stenosis.

Longer cardiopulmonary bypass times with similar crossclamp times make us wonder if bypass may have been useful during repair of the airway defects. In some cases, it may even be that the tracheal lesion was the primary reason to operate on these patients, and the cardiac lesions necessitated repair for the patient to recover from a prolonged bypass run. Particularly in the intervention for tracheal stenosis group, which is known to be associated with vascular rings and pulmonary slings, this stands out because they had a median bypass time of 167 minutes, but a median crossclamp time of 6 minutes. Conversely,

TABLE 6. Adjusted odds of mortality within the airway group

Variable	aOR for mortality	*P* value	aOR for morbidity	*P* value	aOR for tracheostomy	*P* value
Omphalocele, gastroschisis, or CDH	**3.79 (2.17-6.63)**	**<.0001**	**2.12 (1.36-3.31)**	**.0010**	**2.34 (1.27-4.29)**	**.0062**
Tracheal intervention	**3.01 (2.18-4.17)**	**<.0001**	**2.21 (1.75-2.79)**	**<.0001**	**2.74 (2.02-3.73)**	**<.0001**
Preoperative mechanical circulatory support	**2.67 (1.42-5.02)**	**.0023**	**2.34 (1.37-4.01)**	**.0020**	1.03 (0.47-2.24)	.9445
Preoperative mechanical ventilator support	**1.91 (1.51-2.42)**	**<.0001**	**1.57 (1.36-1.82)**	**<.0001**	**3.02 (2.43-3.75)**	**<.0001**
Preoperative shock, persistent at time of surgery	**1.79 (1.00-3.21)**	**.0488**	1.54 (0.97-2.45)	.0691	1.14 (0.61-2.14)	.6757
Any other preoperative factor	**1.69 (1.30-2.20)**	**.0001**	**1.36 (1.15-1.61)**	**.0003**	0.92 (0.72-1.18)	.5203
Single ventricle	**1.51 (1.09-2.10)**	**.0133**	**1.74 (1.43-2.12)**	**<.0001**	**1.45 (1.04-2.01)**	**.0277**
Prematurity among neonates and infants	**1.42 (1.09-1.85)**	**.0095**	**1.19 (1.01-1.41)**	**.0430**	1.14 (0.89-1.45)	.2952
Prior cardiothoracic operation	1.12 (0.80-1.58)	.5109	**1.64 (1.36-1.99)**	**<.0001**	1.08 (0.78-1.50)	.6376
Age in years for children	**1.08 (1.01-1.15)**	**.0162**	0.98 (0.95-1.02)	.3481	1.03 (0.94-1.12)	.5084
STAT Category 5 vs 1	**5.33 (3.04-9.36)**	**<.0001**	**5.09 (3.62-7.16)**	**<.0001**	**2.18 (1.31-3.61)**	**.0026**
STAT Category 4 vs 1	**3.87 (2.59-5.78)**	**<.0001**	**4.05 (3.26-5.02)**	**<.0001**	**1.85 (1.34-2.57)**	**.0002**
STAT Category 3 vs 1	**2.10 (1.33-3.33)**	**.0015**	**2.37 (1.86-3.01)**	**<.0001**	**1.56 (1.08-2.25)**	**.0179**
STAT Category 2 vs 1	**1.71 (1.13-2.61)**	**.0118**	**1.62 (1.30-2.02)**	**<.0001**	1.17 (0.84-1.64)	.3518
Infants	**0.70 (0.52-0.94)**	**.0160**	**0.61 (0.51-0.73)**	**<.0001**	0.89 (0.68-1.17)	.4104

The risk factors for adverse outcomes restricted to patients within the airway group are shown. Notably, a tracheal intervention increases the risk of mortality by 3-fold. Bold indicates statistically significant predictors of morbidity, mortality, and tracheostomy. *aOR*, Adjusted odds ratio; *CDH*, congenital diaphragmatic hernia; *STAT*, Society of Thoracic/European Association for Congenital Heart Surgery Mortality.

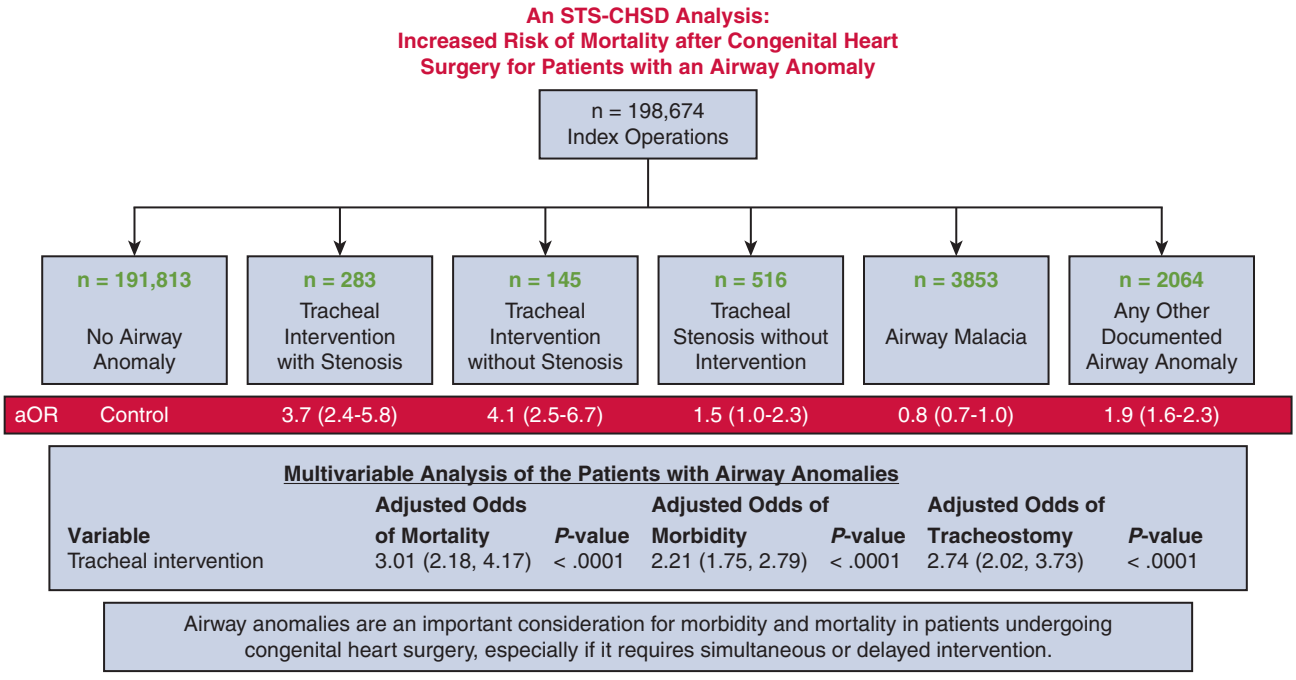

FIGURE 2. This chart depicts the breakdown of all patients undergoing cardiac surgery included in the study. The vast majority had no airway anomaly. Most categories of airway anomalies and interventions had an increased adjusted odds ratio of mortality highlighted by the red bar. The multivariable results of having a tracheal intervention for the group of patients with airway anomalies is displayed below. *STS-CHSD*, Society of Thoracic Surgeons Congenital Heart Surgery Database; *aOR*, adjusted odds ratio.

10% of patients with airway malacia had an associated vascular ring, but still had a median crossclamp time of 54 minutes, likely due to their other cardiac defects being more complex.

Multivariable analysis models identified greater than 3-fold increased risk of major morbidity, mortality, and tracheostomy for any tracheal intervention. This risk has not been appreciated in prior analyses, especially those that considered cardiac surgery a risk factor for tracheal interventions.[6-8] Although the risk likely applies to both operations as a culmination of the patients' overall state of health, it is important to analyze the impact from both points of view. For the patients with airway malacia not requiring tracheal intervention, an increased risk remained in all categories except mortality for tracheal stenosis without intervention and airway malacia. Surprisingly, airway malacia was found to be protective against mortality in this analysis, which is counterintuitive and incongruent with the other results of this article. This may be due to an association with prematurity as well as the definition of malacia reported to the STS that does not require bronchoscopic confirmation.

Study Limitations

Limitations primarily relate to the nature of the STS-CHSD and those associated with a retrospective review. Comparison of the effects of tracheal intervention is not based on randomized assignment, and associations could be due to other concomitant factors not fully captured in the risk adjustment. The study is limited by the variables collected by the STS-CHSD, whereas other factors may be important to decision making and the overall respiratory status. Although most patients have a specific airway diagnosis, many reported "unknown" airway anomaly and bronchoscopy are not required for reporting the diagnosis of airway malacia. However, the strength is the multicenter patient population for studying a rare combination of diseases that make the findings generalizable. Additionally, patients with cardiac and airway disease who died before a cardiac operation was performed would not be captured by this database. Last, this study only captures in-hospital complications and hospital mortality or mortality up to 30 days after discharge.

CONCLUSIONS

Children undergoing congenital heart surgery who have airway anomalies, whether or not requiring an operation, are at increased risk of morbidity, mortality, and tracheostomy. When children do undergo concurrent tracheal surgery, they incur more morbidity and higher mortality than patients only undergoing cardiac surgeries of similar STAT categories. This sheds light on an often overlooked, but intuitively understood, risk factor in congenital cardiac surgery. Moving forward, it is imperative that any airway anomaly in a pediatric cardiac surgery candidate be identified preoperatively. This should help guide discussions of postoperative expectations with family and care teams, as well as heighten our attention in caring for these patients.

Webcast

You can watch a Webcast of this AATS meeting presentation by going to: https://aats.blob.core.windows.net/media/20AM/Presentations/Tracheal%20Surgeries%20and%20Anomalies%20Inc.mp4.

Conflict of Interest Statement

The authors reported no conflicts of interest.

The *Journal* policy requires editors and reviewers to disclose conflicts of interest and to decline handling or reviewing manuscripts for which they may have a conflict of interest. The editors and reviewers of this article have no conflicts of interest.

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Key Words: bronchi, congenital cardiac, trachea

CONG

Discussion

Presenter: Dr Kyle W. Riggs

Dr Charles B. Huddleston *(St Louis, Mo)*. Registry and database studies provide a powerful tool for clinical investigation just because of the sheer numbers that are available for analysis. In this case, there were approximately 200,000 patients total, of whom 7000 had airway anomalies identified. But there are some inherent weaknesses to these sorts of studies. These include the accuracy of the data entry, the lack of consistency in a diagnosis or treatment, and the skills of each center in making these particular diagnoses and establishing these treatments.

On top of that, in studies like this where noncardiac diagnoses are kind of front and center, the STS database offers only a drop-down menu with limited options to choose in terms of the type of airway disease that the patients have. To that end, I'd like to focus on 2 of those diagnoses: tracheomalacia and unspecified airway disease, because these 2 diagnoses (or categories, if you will) comprise approximately 90% of the 7000 patients you study.

The diagnosis of tracheomalacia can range, at least in my experience, from the clinical impression of a neonatologist to a fairly well-documented diagnosis provided by an otolaryngologist using bronchoscopy. Do you know how often this diagnosis was actually confirmed with bronchoscopy in this group of 7000 patients?

Dr Kyle W. Riggs *(Manhasset, NY)*. I think that's an important question because that makes up a great number of our patients, as you pointed out. Unfortunately, for the STS reports, which we all submit, it doesn't require that bronchoscopy was performed; it simply requires checking of the box of trachea or laryngeal or bronchomalacia. That's unfortunately as specific we can be and how the center's diagnosed it.

Dr Huddleston. But I thought that any procedure performed on a patient during their hospitalization such as bronchoscopy would be entered into the database; is that not correct?

Dr Riggs. I believe it is entered, but it may be on a different form and may be a different submission within that same hospitalization. We didn't look for all bronchoscopies performed in the same hospitalization; perhaps we could go back, and that could give us more insight as to if that was performed in all patients with malacia. However, it could have been performed in prior outpatient studies as well. So that definitely is a hang-up in our data analysis.

Dr Huddleston. Now, for this other category of "other airway anomalies," do you have any idea what is included in that? Tracheoesophageal fistula is one of those potentially—but what else?

Dr Riggs. Yes, I think that could be one of them. This is a newer diagnostic category for them, and it's at the surgeon's discretion. Unfortunately, there's nothing more specific. It could be someone with tracheomalacia, and they didn't specify what the airway disease was and just checked the box for "airway anomaly." So that is a big category, but it is up to the surgeon to determine if there was a significant airway anomaly.

Dr Huddleston. The tracheal interventions included repair of tracheal stenosis and repair of tracheal esophageal fistula, but yet there were other tracheal interventions—another "other" category. I don't suppose you have any idea what those other interventions were?

Dr Riggs. Again, similar to my prior responses and as you pointed out, the database doesn't specify further, so we're left to speculate but I do think a significant amount of them were tracheoesophageal fistula repairs, possibly some tracheopexy, but we can't comment further with the data that we have.

Dr Huddleston. I presume that this study included patients with the more common vascular rings such as double arch or right aortic arch, anomalous left subclavian artery, and so forth. Virtually all those patients have tracheomalacia. Were those patients included in your study? Do you have any idea how many of the 7000 came from that diagnosis?

Dr Riggs. They were included in our inclusion criteria. We didn't ask the Duke data center to specifically separate them out. But again, before presenting the manuscript perhaps that's something we could ask them—who had tracheal surgery for a ring and did they have associated malacia—and that might be an important cohort to capture.

Dr Huddleston. About 4 years ago, a publication appeared in the *Annals of Thoracic Surgery* regarding tracheostomies after congenital heart surgery. This was also an STS Database–driven study. The factors associated with the need for tracheostomy in that study (and again, a fairly robust study) included injuries to the recurrent laryngeal nerve, phrenic nerve injuries, neurologic deficit (presumably hypoxic ischemic encephalopathy), delayed sternal closure, major mechanical assistance, and on and on; interestingly, there was no mention of airway anomalies in that analysis. Do you have a comment about that?

Dr Riggs. That is a good point. I reviewed that article and even discussed it with our statisticians. They said that in that study (from ∼6 years ago now), airway anomaly was not a category within the STS report form. So our study is somewhat of an update on that, I believe. Additionally, we found similar postoperative complications with increased incidence of phrenic nerve injury and everything you said, so I think it's just something that wasn't fully captured within their study, but was probably true back at that time.

Dr Huddleston. I found it interesting that you didn't include that in the references of your article.

Dr Riggs. I agree, we could have included it, but I think it was a bit different than what we started out looking for.

Dr Huddleston. My final comment and question is that many registry studies that look at these large groups of patients tend to confirm, in an objective way, what a lot of us have already been suspicious of from the more subjective sort of overview of something like this. I don't think anyone would be particularly surprised that patients with the concomitant airway problem would have worse outcomes with congenital heart surgery than those who did not. And this study I think confirms my and perhaps others' suspicions about that. Was there anything in this study that surprised you at all or was there something that you found that was unexpected?

Dr Riggs. It's a good point. I agree with what you're saying, that many of our findings may seem obvious. But there really wasn't a whole lot in the literature confirming them, which is why we investigated this in the STS database. I think part of the value is quantifying the risk; it's increased, but how much increased within different categories is what we elucidated. One of the strangest findings to us and the statisticians was the protective effect of malacia on the airway groups.

But further analysis, as I pointed out, shows that's probably a confounding finding, and I wouldn't necessarily say that those patients have a protective effect. But overall, I think our findings were intuitive, and we were happy to confirm what we were seeing clinically.

Dr Huddleston. Great. Very nice job.

TABLE E1. Covariates in the STS-CHSD risk model

Primary procedure (within age group) (modeled as random effects)
Prematurity among neonates and infants
Prior cardiothoracic operation
Any other preoperative factor
Preoperative mechanical circulatory support
Preoperative shock, persistent at time of surgery
Preoperative renal dysfunction or dialysis
Preoperative mechanical ventilator support
Preoperative neurologic deficit
Weight among neonates
Weight among infants
Infants (vs neonates)
Children (vs neonates)
Adults (vs neonates)
Age in days for neonates
Age in days for infants
Age in years for children
Age in years for adults
STAT Category 2 (vs 1)
STAT Category 3 (vs 1)
STAT Category 4 (vs 1)
STAT Category 5 (vs 1)
Congenital diaphragmatic hernia
Omphalocele
Intestinal malrotation
Atresia of large intestine
Gastroschisis
Hirschsprung's disease (congenital aganglionic megacolon)
Highest risk group 5: syndrome and CA (vs no syndrome and CA)
Risk group 4: syndrome and CA (vs no syndrome and CA)
Risk group 3: syndrome and CA (vs no syndrome and CA)
Risk group 2: syndrome and CA (vs no syndrome and CA)
Lowest risk group 1: syndrome and CA (vs no syndrome and CA)

Description of the hierarchical multivariable logistic regression model to estimate odds ratios comparing factors of primary interest and adjusted for the variables included in the STS congenital mortality risk model. Covariates include in the risk model are listed and include patient-level demographic factors such as age, weight and prematurity, procedural complexity (STAT category), and baseline risk factors such as other chromosomal anomalies and noncardiac anatomic abnormalities. We also included as a covariate, "any single ventricle diagnosis," because this was considered an important additional risk factor in this patient population.[16] *STAT,* Society of Thoracic/European Association for Congenital Heart Surgery Mortality; *CA*, chromosomal abnormality.

TABLE E2. Preoperative patient demographics by airway subgroups

Variable	Level	Overall (N = 198,674)		Control group (N = 191,813)		Tracheal intervention for tracheal stenosis (N = 283)		Tracheal intervention without tracheal stenosis (N = 145)		Tracheal stenosis without intervention (N = 516)		Airway malacia (N = 3853)		All other airway anomalies (N = 2064)		*P* value*
Hospitalization																
Birth weight (kg)†	Median	96,332	3.00	91,394	3.01	150	2.77	61	2.87	309	2.83	3379	2.83	1039	2.67	<.0001
	25th		2.50		2.51		2.30		2.02		2.32		2.31		2.13	
	75th		3.40		3.41		3.12		3.26		3.21		3.29		3.12	
	Mean		2.87		2.88		2.65		2.63		2.76		2.74		2.59	
	STD		0.85		0.85		0.68		0.88		0.74		0.83		0.79	
	Min		0.10		0.10		0.72		0.51		0.49		0.38		0.40	
	Max		9.97		9.97		3.90		4.43		4.54		5.45		4.77	
Demographics																
Prematurity (<37 wk completed gestation)	Yes	41,615	22.24	39,433	21.85	75	27.99	44	31.43	151	30.08	1186	31.26	726	36.45	<.0001
	No	145,518	77.76	141,004	78.15	193	72.01	96	68.57	351	69.92	2608	68.74	1266	63.55	
Gender	Ambiguous	26	0.01	25	0.01	1	0.35	0	0.00	0	0.00	0	0.00	0	0.00	.0004
	Female	89,602	45.14	86,435	45.10	117	41.49	67	46.21	243	47.09	1776	46.12	964	46.86	
	Male	108,871	54.85	105,188	54.89	164	58.16	78	53.79	273	52.91	2075	53.88	1093	53.14	
Race	Other	10,864	5.90	10,502	5.91	12	4.78	12	9.76	17	3.39	191	5.12	130	6.69	<.0001
	Native American	1430	0.78	1380	0.78	2	0.80	1	0.81	4	0.80	30	0.80	13	0.67	
	Asian	8045	4.37	7830	4.41	20	7.97	2	1.63	16	3.19	111	2.98	66	3.40	
	Hispanic	34,799	18.89	33,706	18.97	40	15.94	20	16.26	67	13.35	613	16.44	353	18.17	
	Black	30,007	16.29	28,857	16.24	41	16.33	22	17.89	99	19.72	712	19.10	276	14.20	
	White	99,071	53.78	95,394	53.69	136	54.18	66	53.66	299	59.56	2071	55.55	1105	56.87	
Preoperative factors																
STS-EACTS complexity level†	5	8570	4.37	8261	4.36	2	0.71	5	3.55	18	3.54	224	5.87	60	2.94	<.0001
	4	40,448	20.61	38,775	20.47	44	15.71	44	31.21	110	21.61	982	25.73	493	24.17	
	3	22,129	11.28	21,088	11.13	122	43.57	23	16.31	81	15.91	539	14.12	276	13.53	
	2	69,723	35.53	67,653	35.71	59	21.07	46	32.62	184	36.15	1081	28.32	700	34.31	
	1	55,345	28.21	53,651	28.32	53	18.93	23	16.31	116	22.79	991	25.96	511	25.05	
Any NCAA	Yes	25,007	12.99	19,110	10.29	170	61.82	82	56.94	318	62.85	3837	99.58	1490	73.08	<.0001
	No	167,453	87.01	166,533	89.71	105	38.18	62	43.06	188	37.15	16	0.42	549	26.92	
Noncardiac congenital anatomic abnormalities																
Major abnormality of head	Yes	3125	1.62	2607	1.40	6	2.18	5	3.47	21	4.15	343	8.90	143	7.01	<.0001
	No	189,335	98.38	183,036	98.60	269	97.82	139	96.53	485	95.85	3510	91.10	1896	92.99	
Major abnormality of brain	Yes	3588	1.86	3054	1.65	14	5.09	7	4.86	29	5.73	376	9.76	108	5.30	<.0001
	No	188,872	98.14	182,589	98.35	261	94.91	137	95.14	477	94.27	3477	90.24	1931	94.70	

(Continued)

TABLE E2. Continued

Variable	Level	Overall (N = 198,674)		Control group (N = 191,813)		Tracheal intervention for tracheal stenosis (N = 283)		Tracheal intervention without tracheal stenosis (N = 145)		Tracheal stenosis without intervention (N = 516)		Airway malacia (N = 3853)		All other airway anomalies (N = 2064)		*P* value*
Major abnormality of spinal cord	Yes	546	0.28	433	0.23	7	2.55	1	0.69	15	2.96	63	1.64	27	1.32	<.0001
	No	191,914	99.72	185,210	99.77	268	97.45	143	99.31	491	97.04	3790	98.36	2012	98.68	
Major abnormality of spine	Yes	2006	1.04	1680	0.90	12	4.36	3	2.08	25	4.94	164	4.26	122	5.98	<.0001
	No	190,454	98.96	183,963	99.10	263	95.64	141	97.92	481	95.06	3689	95.74	1917	94.02	
Major abnormality of larynx	Yes	5712	2.97	0	0.00	135	49.09	64	44.44	291	57.51	3833	99.48	1389	68.12	<.0001
	No	186,748	97.03	185,643	100.00	140	50.91	80	55.56	215	42.49	20	0.52	650	31.88	
Major abnormality of lung	Yes	1190	0.62	985	0.53	25	9.09	3	2.08	10	1.98	127	3.30	40	1.96	<.0001
	No	191,270	99.38	184,658	99.47	250	90.91	141	97.92	496	98.02	3726	96.70	1999	98.04	
Major abnormality of abdominal wall	Yes	1086	0.56	977	0.53	6	2.18	3	2.08	8	1.58	57	1.48	35	1.72	<.0001
	No	191,374	99.44	184,666	99.47	269	97.82	141	97.92	498	98.42	3796	98.52	2004	98.28	
Major abnormality of gastrointestinal system	Yes	948	0.49	754	0.41	6	2.18	4	2.78	8	1.58	105	2.73	71	3.48	<.0001
	No	191,512	99.51	184,889	99.59	269	97.82	140	97.22	498	98.42	3748	97.27	1968	96.52	
Major abnormality of kidney, ureter, or bladder	Yes	3643	1.89	3128	1.68	17	6.18	9	6.25	40	7.91	304	7.89	145	7.11	<.0001
	No	188,817	98.11	182,515	98.32	258	93.82	135	93.75	466	92.09	3549	92.11	1894	92.89	
Previous interventions																
No. of prior cardiothoracic operations†	Median	194,460	0.00	187,646	0.00	277	0.00	141	0.00	505	0.00	3847	0.00	2044	0.00	<.0001
	25th		0.00		0.00		0.00		0.00		0.00		0.00		0.00	
	75th		1.00		1.00		0.00		1.00		1.00		1.00		1.00	
	Mean		0.62		0.62		0.29		0.66		0.81		0.70		0.72	
	STD		1.29		1.28		0.83		1.17		1.75		1.49		1.53	
	Min		0.00		0.00		0.00		0.00		0.00		0.00		0.00	
	Max		159.00		159.00		6.00		7.00		22.00		36.00		29.00	
No. of prior CPB cardiothoracic operations†	Median	190,345	0.00	183,628	0.00	269	0.00	137	0.00	498	0.00	3788	0.00	2025	0.00	<.0001
	25th		0.00		0.00		0.00		0.00		0.00		0.00		0.00	
	75th		1.00		1.00		0.00		0.00		1.00		1.00		1.00	
	Mean		0.38		0.38		0.17		0.34		0.39		0.39		0.40	
	STD		0.78		0.78		0.60		0.78		0.84		0.80		0.80	
	Min		0.00		0.00		0.00		0.00		0.00		0.00		0.00	
	Max		50.00		50.00		4.00		5.00		6.00		12.00		8.00	

(Continued)

TABLE E2. Continued

Variable	Level	Overall (N = 198,674)		Control group (N = 191,813)		Tracheal intervention for tracheal stenosis (N = 283)		Tracheal intervention without tracheal stenosis (N = 145)		Tracheal stenosis without intervention (N = 516)		Airway malacia (N = 3853)		All other airway anomalies (N = 2064)		*P* value*
Demographics																
Age at surgery (d) among those aged ≤1 y†	Median	114,705	68.00	109,994	67.00	229	78.00	109	83.00	315	106.00	2766	83.00	1292	76.50	<.0001
	25th		11.00		11.00		30.00		17.00		27.00		18.00		21.00	
	75th		159.00		159.00		156.00		155.00		203.00		164.00		173.00	
	Mean		95.80		95.35		104.44		105.85		128.47		103.91		106.35	
	STD		93.35		93.21		91.53		100.61		106.63		94.08		96.55	
	Min		0.00		0.00		1.00		0.00		1.00		0.00		0.00	
	Max		365.00		365.00		364.00		360.00		362.00		365.00		365.00	
Age at surgery (y)†	Median	198,674	0.57	191,813	0.58	283	0.30	145	0.33	516	0.68	3853	0.39	2064	0.52	<.0001
	25th		0.12		0.12		0.12		0.08		0.19		0.10		0.12	
	75th		4.16		4.25		0.77		0.99		2.56		1.23		2.96	
	Mean		3.12		3.17		1.14		1.87		2.35		1.45		2.46	
	STD		4.68		4.72		2.71		3.87		3.82		2.70		4.03	
	Min		0.00		0.00		0.00		0.00		0.00		0.00		0.00	
	Max		18.00		18.00		17.34		17.58		17.11		17.66		17.94	
Age at surgery (mo)†	Median	198,674	30.00	191,813	30.43	283	15.43	145	17.43	516	35.36	3853	20.43	2064	27.07	<.0001
	25th		6.14		6.14		6.14		4.14		10.07		5.43		6.43	
	75th		217.29		221.86		40.00		51.43		133.57		64.29		154.21	
	Mean		162.85		165.28		59.58		97.62		122.69		75.68		128.41	
	STD		244.38		246.22		141.22		202.18		199.40		140.77		210.20	
	Min		0.00		0.00		0.14		0.00		0.14		0.00		0.00	
	Max		939.14		939.14		904.57		917.43		892.86		921.29		936.00	
Hospitalization																
Weight at surgery (kg)†	Median	197,514	6.80	190,667	6.82	281	4.90	144	4.90	515	7.33	3849	5.50	2058	6.07	<.0001
	25th		3.71		3.73		3.34		3.45		3.90		3.48		3.40	
	75th		15.60		15.90		7.38		8.24		12.50		9.30		12.90	
	Mean		14.19		14.37		7.12		9.04		11.65		8.16		11.01	
	STD		18.15		18.33		8.25		11.63		13.72		9.14		13.20	
	Min		0.00		0.00		2.00		1.35		0.80		0.41		0.00	
	Max		193.50		193.50		63.50		70.00		89.50		120.00		133.00	
Preoperative factors																
Any preoperative risk factors	Yes	77,941	40.22	73,025	39.05	199	70.82	110	77.46	312	62.53	2877	75.29	1418	69.75	<.0001
	No	115,857	59.78	113,997	60.95	82	29.18	32	22.54	187	37.47	944	24.71	615	30.25	
Preoperative/ preprocedural mechanical circulatory support (IABP, VAD, ECMO, or CPS)	Yes	1614	0.83	1538	0.82	16	5.69	2	1.41	7	1.40	32	0.84	19	0.93	<.0001
	No	192,184	99.17	185,484	99.18	265	94.31	140	98.59	492	98.60	3789	99.16	2014	99.07	

(Continued)

TABLE E2. Continued

Variable	Level	Overall (N = 198,674)		Control group (N = 191,813)		Tracheal intervention for tracheal stenosis (N = 283)		Tracheal intervention without tracheal stenosis (N = 145)		Tracheal stenosis without intervention (N = 516)		Airway malacia (N = 3853)		All other airway anomalies (N = 2064)		*P* value*
Persistent shock	Yes	2278	1.18	2189	1.17	3	1.07	3	2.11	5	1.00	59	1.54	19	0.93	.2394
	No	191,520	98.82	184,833	98.83	278	98.93	139	97.89	494	99.00	3762	98.46	2014	99.07	
Mechanical ventilatory support	Yes	26,083	13.46	24,208	12.94	114	40.57	68	47.89	116	23.25	1059	27.72	518	25.48	<.0001
	No	167,715	86.54	162,814	87.06	167	59.43	74	52.11	383	76.75	2762	72.28	1515	74.52	
Failure requiring dialysis	Yes	296	0.15	279	0.15	3	1.07	0	0.00	2	0.40	6	0.16	6	0.30	.0010
	No	193,502	99.85	186,743	99.85	278	98.93	142	100.00	497	99.60	3815	99.84	2027	99.70	
Neurologic deficit	Yes	4790	2.47	4316	2.31	15	5.34	10	7.04	39	7.82	297	7.77	113	5.56	<.0001
	No	189,008	97.53	182,706	97.69	266	94.66	132	92.96	460	92.18	3524	92.23	1920	94.44	
Cardiopulmonary resuscitation	Yes	1614	0.83	1516	0.81	9	3.20	3	2.11	4	0.80	47	1.23	35	1.72	<.0001
	No	192,184	99.17	185,506	99.19	272	96.80	139	97.89	495	99.20	3774	98.77	1998	98.28	
Preoperative complete AV block	Yes	3087	1.59	2992	1.60	1	0.36	1	0.70	9	1.80	60	1.57	24	1.18	.3186
	No	190,711	98.41	184,030	98.40	280	99.64	141	99.30	490	98.20	3761	98.43	2009	98.82	
Steroids for adrenal insufficiency	Yes	1237	0.64	1147	0.61	4	1.42	3	2.11	6	1.20	60	1.57	17	0.84	<.0001
	No	192,561	99.36	185,875	99.39	277	98.58	139	97.89	493	98.80	3761	98.43	2016	99.16	
Steroids for other reason	Yes	2056	1.06	1844	0.99	13	4.63	3	2.11	18	3.61	127	3.32	51	2.51	<.0001
	No	191,742	98.94	185,178	99.01	268	95.37	139	97.89	481	96.39	3694	96.68	1982	97.49	
Colostomy	Yes	1000	0.52	855	0.46	12	4.27	4	2.82	8	1.60	51	1.33	70	3.44	<.0001
	No	192,798	99.48	186,167	99.54	269	95.73	138	97.18	491	98.40	3770	98.67	1963	96.56	
Esophagostomy	Yes	48	0.02	34	0.02	0	0.00	3	2.11	0	0.00	1	0.03	10	0.49	<.0001
	No	193,750	99.98	186,988	99.98	281	100.00	139	97.89	499	100.00	3820	99.97	2023	99.51	
Gastrostomy	Yes	10,862	5.60	9229	4.93	31	11.03	42	29.58	88	17.64	916	23.97	556	27.35	<.0001
	No	182,936	94.40	177,793	95.07	250	88.97	100	70.42	411	82.36	2905	76.03	1477	72.65	
Hepatic dysfunction	Yes	1149	0.59	1083	0.58	4	1.42	1	0.70	4	0.80	34	0.89	23	1.13	.0012
	No	192,649	99.41	185,939	99.42	277	98.58	141	99.30	495	99.20	3787	99.11	2010	98.87	
Necrotizing enterocolitis treated medically	Yes	1527	0.79	1398	0.75	3	1.07	1	0.70	4	0.80	84	2.20	37	1.82	<.0001
	No	192,271	99.21	185,624	99.25	278	98.93	141	99.30	495	99.20	3737	97.80	1996	98.18	
Necrotizing enterocolitis treated surgically	Yes	962	0.50	909	0.49	0	0.00	1	0.70	3	0.60	32	0.84	17	0.84	.0075
	No	192,836	99.50	186,113	99.51	281	100.00	141	99.30	496	99.40	3789	99.16	2016	99.16	
Hypercoagulable state	Yes	1134	0.59	1069	0.57	1	0.36	0	0.00	4	0.80	43	1.13	17	0.84	.0003

(Continued)

TABLE E2. Continued

Variable	Level	Overall (N = 198,674)		Control group (N = 191,813)		Tracheal intervention for tracheal stenosis (N = 283)		Tracheal intervention without tracheal stenosis (N = 145)		Tracheal stenosis without intervention (N = 516)		Airway malacia (N = 3853)		All other airway anomalies (N = 2064)		*P* value*
	No	192,664	99.41	185,953	99.43	280	99.64	142	100.00	495	99.20	3778	98.87	2016	99.16	
Hypocoagulable state not secondary to medication	Yes	1833	0.95	1714	0.92	5	1.78	1	0.70	2	0.40	70	1.83	41	2.02	<.0001
	No	191,965	99.05	185,308	99.08	276	98.22	141	99.30	497	99.60	3751	98.17	1992	97.98	
Endocarditis	Yes	903	0.47	881	0.47	0	0.00	0	0.00	1	0.20	14	0.37	7	0.34	.5058
	No	192,895	99.53	186,141	99.53	281	100.00	142	100.00	498	99.80	3807	99.63	2026	99.66	
Sepsis	Yes	1375	0.71	1290	0.69	2	0.71	1	0.70	4	0.80	39	1.02	39	1.92	<.0001
	No	192,423	99.29	185,732	99.31	279	99.29	141	99.30	495	99.20	3782	98.98	1994	98.08	
Seizure within 48 h before surgery	Yes	197	0.10	187	0.10	0	0.00	0	0.00	2	0.40	6	0.16	2	0.10	.3028
	No	193,601	99.90	186,835	99.90	281	100.00	142	100.00	497	99.60	3815	99.84	2031	99.90	
Tracheostomy	Yes	1404	0.72	885	0.47	16	5.69	21	14.79	36	7.21	316	8.27	130	6.39	<.0001
	No	192,394	99.28	186,137	99.53	265	94.31	121	85.21	463	92.79	3505	91.73	1903	93.61	
Pacemaker present (data versions 3.22, 3.3)	Yes	2271	1.17	2181	1.17	1	0.36	1	0.70	4	0.80	62	1.62	22	1.08	.0951
	No	191,527	98.83	184,841	98.83	280	99.64	141	99.30	495	99.20	3759	98.38	2011	98.92	
Other risk factor	Yes	33,629	17.35	31,190	16.68	96	34.16	50	35.21	155	31.06	1486	38.89	652	32.07	<.0001
	No	160,169	82.65	155,832	83.32	185	65.84	92	64.79	344	68.94	2335	61.11	1381	67.93	
Any chromosomal abnormality	Yes	33,951	17.62	31,563	16.98	66	23.66	43	30.07	160	31.56	1573	41.01	546	26.82	<.0001
	No	158,743	82.38	154,330	83.02	213	76.34	100	69.93	347	68.44	2263	58.99	1490	73.18	
Chromosomal abnormalities																
22q11 deletion DiGeorge Syndrome	Yes	5781	2.99	5193	2.79	11	3.93	15	10.42	44	8.66	393	10.21	125	6.12	<.0001
	No	187,315	97.01	181,079	97.21	269	96.07	129	89.58	464	91.34	3455	89.79	1919	93.88	
Trisomy 21	Yes	16,309	8.45	15,508	8.33	31	11.07	10	6.94	40	7.87	561	14.58	159	7.78	<.0001
	No	176,787	91.55	170,764	91.67	249	88.93	134	93.06	468	92.13	3287	85.42	1885	92.22	
Trisomy 18	Yes	307	0.16	251	0.13	0	0.00	1	0.69	3	0.59	33	0.86	19	0.93	<.0001
	No	192,789	99.84	186,021	99.87	280	100.00	143	99.31	505	99.41	3815	99.14	2025	99.07	
Trisomy 13	Yes	82	0.04	74	0.04	0	0.00	0	0.00	0	0.00	8	0.21	0	0.00	<.0001
	No	193,014	99.96	186,198	99.96	280	100.00	144	100.00	508	100.00	3840	99.79	2044	100.00	
Preoperative factors																
Any syndromes	Yes	43,278	22.43	40,041	21.51	93	33.21	67	46.53	202	39.76	1893	49.19	982	48.07	<.0001
	No	149,674	77.57	146,088	78.49	187	66.79	77	53.47	306	60.24	1955	50.81	1061	51.93	

AV, Aortic valve; *CPB,* cardiopulmonary bypass; *CPS,* cardiopulmonary support; *ECMO,* extracorporeal membrane oxygenation; *IABP,* intra-aortic balloon pump; *NCAA,* noncardiac congenital anatomic abnormality; *STD,* standard deviation; *VAD,* ventricular assist device. All tests treat the column variable as nominal. **P* values are based on Pearson chi-square tests for all categorical row variables. †*P* values are based on chi-square rank based group means score statistics for all continuous/ordinal row variables (designated by †). ††This is equivalent to Kruskal–Wallis tests.

TABLE E3. Intraoperative and postoperative outcomes for Society of Thoracic/European Association for Congenital Heart Surgery Mortality category 1 to 3

Variable	Overall, N = 147,197	Control group, N = 142,392	Airway anomaly group, N = 4805	*P* value
Cardiopulmonary bypass time (min)	88 [61-128]	88 [60-127]	103 [72-149]	<.001
Crossclamp time (min)	45 [21-75]	45 [21-75]	47 [16-79]	.590
Operative mortality	1.8% (2568)	1.7 (2380)	3.9 (188)	<.001
Major morbidity	7.2 (10,149)	7.0 (9498)	14.0 (651)	<.001
Postoperative mechanical circulatory support	1.1% (1558)	1.1% (1451)	2.3% (107)	<.001
Unplanned reoperation	4.6% (6439)	4.4% (6020)	9.0% (419)	<.001
Neurologic deficit	0.5% (705)	0.5% (662)	0.9% (43)	<.001
Prolonged mechanical ventilation	5.2% (7351)	4.8% (6602)	16.1% (749)	<.001
Postoperative tracheostomy	0.7% (939)	0.5% (670)	5.8% (269)	<.001
Postoperative length of stay (d)	6 [4-11]	6 [4-11]	10 [5-35]	<.001

TABLE E4. Intraoperative and postoperative outcomes for Society of Thoracic/European Association for Congenital Heart Surgery Mortality category 4 and 5

Variable	Overall	Control group, N = 47,036	Airway anomaly group, N = 1986	*P* value
Cardiopulmonary bypass time (min)	137 [95-190]	137 [95-190]	145 [100-205]	<.001
Crossclamp time (min)	65 [37-106]	65 [37-106]	66 [33-107]	.267
Operative mortality	8.5% (4150)	8.4% (3910)	12.2% (240)	<.001
Major morbidity	25.7% (11,996)	25.2% (11,250)	39.2% (746)	<.001
Postoperative mechanical circulatory support	7.8% (3620)	7.6% (3414)	10.8% (206)	<.001
Unplanned reoperation	17.0% (7903)	16.5% (7366)	28.2% (537)	<.001
Neurologic deficit	1.6% (759)	1.6% (698)	3.2% (61)	<.001
Prolonged mechanical ventilation	17.3% (8056)	16.5% (7351)	37.1% (705)	<.001
Postoperative tracheostomy	2.0% (939)	1.6% (693)	12.9% (246)	<.001
Postoperative length of stay (d)	16 [8-35]	15 [8-33]	37 [16-82]	<.001