Cardiac Surgery in Patients With Trisomy 13 and 18: An Analysis of The Society of Thoracic Surgeons Congenital Heart Surgery Database

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Background—Congenital heart disease is common in patients with Trisomy 13 (T13) and Trisomy 18 (T18), but offering cardiac surgery to these patients has been controversial. We describe the landscape of surgical management across the United States, perioperative risk factors, and surgical outcomes in patients with T13 and T18.

Methods and Results—Patients in the Society of Thoracic Surgeons Congenital Heart Surgery Database with T13 and T18 who underwent cardiac surgery (2010–2017) were included. There were 343 operations (T13: n=73 and T18: n=270) performed on 304 patients. Among 125 hospitals, 87 (70%) performed at least 1 operation and 26 centers (30%) performed ≥5 T13/T18 operations. Operations spanned the full spectrum of complexity with 29% (98/343) being in the highest categories of estimated risk. The operative mortality rate was 15%, with a 56% complication rate. Preoperative mechanical ventilation was associated with an odds ratio of mortality >8 for both patients with T13 and T18 (both P<0.012) while presence of a gastrostomy tube (odds ratio, 0.3; P=0.03) or prior cardiac surgery (odds ratio, 0.2; P=0.02) was associated with better survival in patients with T18 but not patients with T13.

Conclusions—Data from this nationally representative sample indicate that most centers offer surgical intervention for both patients with T13 and T18, even in highly complex patients. However, the overall mortality rate was high in this select patient cohort. The association of preoperative mechanical ventilation with mortality suggests that this subset of patients with T13 and T18 should perhaps not be considered surgical candidates. This information is valuable to clinicians and families for counseling and deciding what interventions to offer. (J Am Heart Assoc. 2019;8:e012349. DOI: 10.1161/JAHA.119.012349.)

Key Words: cardiac surgery • congenital heart disease • outcomes • pediatrics • Trisomy 13 • Trisomy 18

Trisomy 13 (T13) and Trisomy 18 (T18) are the most commonly occurring types of aneuploidy after Trisomy 21 and are known to be associated with congenital heart defects in upwards of 85% of cases.1 Aside from congenital heart disease, these syndromes are associated with an array of extracardiac anomalies and neurodevelopmental abnormalities, which add to the challenge of caring for this population of patients.2,3 Traditionally, T13 and T18 have been considered “lethal” conditions, with the majority of patients dying within the first months of life and few living beyond 1 year.4,5 Common causes of death include central apnea and multiorgan system failure. Premature death is, surprisingly, less frequently attributed to cardiac defects.6,7

Over the past 20 years, improving surgical technique and knowledge about these syndromes has led to more aggressive interventions, including cardiac surgery. Prior studies from administrative databases have described outcomes in these patients; however, they have not been representative of the entire United States as a whole and lack the diagnostic and clinically granular data necessary to understand the care provided.8-11 Therefore, we still lack understanding of the current national landscape of care delivery and outcomes for this population, as well as stratification of outcomes by case complexity as a result of inadequate size and details of other...
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DOI: 10.1161/JAHA.119.012349

Journal of the American Heart Association

What Are the Clinical Implications?

- Surgical candidacy of patients with Trisomy 13 and 18 who require preoperative mechanical ventilation should be carefully considered.

What Is New?

- Analysis of the Society of Thoracic Surgeons Congenital Heart Surgery Database provides a national representation of surgery in this population and captures granular data regarding diagnoses and operations performed in patients with Trisomy 13 and Trisomy 18.
- Operative mortality when compared with patients without Trisomy 13 and 18 was high across all surgical complexity categories.
- Preoperative mechanical ventilation is associated with postoperative mortality.

Clinical Perspective

Patient Population

All cardiovascular operations performed from 2010 to 2017 in patients with T13 (Edwards syndrome) or T18 (Patau syndrome) and captured in the STS-CHSD were potentially eligible for inclusion. Analysis was based on the first cardiovascular surgical operation (index operation) of each episode of care (most often a single hospitalization, but includes transfer for extended care). One operation with missing operative mortality status at discharge was excluded. The final analytic cohort included 343 index cardiac operations from 304 patients performed at 87 centers.

Data Collection and Definitions

Collected data included demographics, cardiac diagnoses, preoperative risk factors, intraoperative details, surgical procedures, postoperative complications, length of stay, and operative mortality, defined as death before discharge from the hospital or within 30 days of the index operation if discharged. The STS/European Association for Cardiothoracic Surgery Congenital Heart Surgery Mortality (STAT) categories group patients on the basis of statistically estimated risk of mortality derived from empirical analysis of multiyear data pertaining to all patients in the databases, with the 5 STAT categories having been chosen to be optimal with respect to minimizing within-category variation and maximizing between-category variation. Thus, STAT category 1 includes the operations with the lowest risk of mortality, and STAT category 5 includes the operations with the highest overall risk of mortality. Apart from classification of operations based on this stratification of complexity or overall mortality risk, we selected apriori individual common preoperative risk factors that the investigative team felt likely to be of interest in this T13/T18 population, including prior cardiac surgery, gastrostomy tube, or mechanical ventilation. Postoperative complications were defined using standard STS data element definitions, whereas major complications for the purposes of this study were considered to be any \( \geq 1 \) of the following: postoperative cardiac arrest, arrhythmia requiring permanent pacemaker,
mechanical circulatory support, unplanned interventional cardiac procedure, renal failure, mediastinitis, neurologic deficit, or reoperation for any reason.¹⁷

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 number (percentage) for discrete variables. T13 and T18 operations were grouped by STS category and then compared with the overall national mortality rate for the respective STS categories using chi-square analysis and Fisher exact test when appropriate. Associations of preoperative risk factors with mortality were evaluated using 2-sided Fisher exact test. Statistical significance was established by a P<0.05. All analyses were conducted using SAS version 9.4 (SAS Institute).

Results
Case Distribution, Demographic, and Preoperative Characteristics
From 125 centers that reported data to the STS-CHSD during the study period, there were 304 patients identified as having T13 or T18, and these patients underwent 343 index operations. There were 73 operations in patients with T13 and 270 operations in patients with T18. The annual distribution of cases and mortality rate by aneuploidy diagnosis is presented in the Figure. Of the contributing centers to the STS-CHSD, over half (87/125, 70%) had at least 1 operation in a patient with T13 or T18 (Table 1). Of those centers, 26 (30%) reported performing ≥5 operations in patients with T13 or T18 during the study period (Table 1). The center with the largest number had 18 operations over the timespan of the study. A total of 49% and 39% of the T13 and T18 operations, respectively, came from the Southern region, although they have the overall largest volume (Table 2).

Trisomy 13
The 73 operations in patients with T13 had a slight female majority (54%), with a prematurity rate of 18% (Table 3). Whites represented approximately half of the patients undergoing surgery (47%), with an overall median age and weight of 4.5 months and 4.8 kg, respectively, at time of operation. Interestingly, only 37% (n=27) of the patients had a reported associated noncardiac anatomic abnormality, 21% (n=16) of the patients had undergone prior cardiothoracic surgery, and 22% (n=16) went to the operating room on mechanical ventilation. The primary diagnosis and procedure are reported in Table 3. STAT category 4 and 5 comprised 36% (n=26) of the patients with T13 who underwent operation. Overall, patients with T13 underwent a higher frequency of STAT 4/5 (highest complexity) index operations than the broader subset of patients included in the STS database (36% of T13 cases received STAT 4/5 index operations versus 24% of all STS participants, P=0.03). However, STAT 5 cases were rarely performed in patients with T13 (n=2 cases, 2.7%). Table S1 shows the distribution of STS cases.

Trisomy 18
Among the 270 operations in patients with T18, there was a 3:1 female to male ratio, and prematurity was present in 23.7% (n=64) of operations. Half of the patients were white (53.3%), with an overall median age of 3.7 months and a median weight of 3.5 kg. Noncardiac abnormalities were common (n=125; 46.3%). Prior cardiac surgery operations had been performed in 16.3% (n=44) of patients. Nearly one third (n=82) required mechanical ventilation before going to the operating room. The primary diagnosis and procedure are reported in Table 3. STAT category 4 comprised 26.7% (n=72) of the operations. Overall, patients with T18 underwent a similar frequency of STAT 4/5 (highest complexity) index operations when compared with the broader subset of patients included in the STS database (27% of patients with T18 received STAT 4/5 index operations versus 24% for all STS participants, P=0.28). Additionally, STAT 5 cases were rarely performed in patients with T18 (n=5 cases, 1.9%).

Surgical Outcomes
Trisomy 13
Hospital mortality and postoperative length of stay are shown in Table 4. Overall, operative mortality was 11%, and varied greatly by STS category, but was only significantly higher in STAT category 2 than the corresponding STS overall mortality rate. Similarly, length of stay varied by STS category. The postoperative complication rate was high at 58% (n=42) for any complication compared with 36% (n=72 345) for all patients, P<0.001 (Table 5). There were no instances of permanent pacemaker required, 1 instance of postoperative mechanical circulatory support (1%), and 5 postoperative cardiac arrests (7%).

Trisomy 18
Hospital mortality and postoperative length of stay are shown in Table 4. Overall surgical mortality was 15.6%. In every STAT category, mortality was at least 3 times higher than the overall STS-CHSD mortality rate (all P<0.05), and in most cases it was 5 times higher. Median length of stay was
18 days and varied widely by STAT category. Postoperative complications (Table 5) were identified in over half of the patients (55.6%) compared with 36% (n=72,345) for all patients (P<0.001). There were 6 (2.3%) cases of arrhythmias necessitating a permanent pacemaker, 6 (2.3%) instances of postoperative mechanical circulatory support, and 18 (6.7%) postoperative cardiac arrests.

### Association Between Preoperative Factors and Postoperative Outcomes

Table 6 demonstrates the association between preoperative factors and in-hospital mortality. For patients with T13, preoperative mechanical ventilator support was the only factor associated with in-hospital mortality (P=0.011) with an unadjusted odds ratio of 8.2. Likewise, for patients with T18, preoperative mechanical ventilator support was associated with mortality (odds ratio, 8.5; P<0.001), but gastrostomy tube (odds ratio, 0.3; P=0.027) and prior cardiac surgery (odds ratio, 0.2; P=0.024) were associated with survival.

### Table 1. Number of Operations Per Center Over the 8-Year Study Period

| Total Cases, No. | Sites, No. (%) |
|------------------|---------------|
| 1                | 31 (35.6)     |
| 2                | 14 (16.1)     |
| 3                | 11 (12.6)     |
| 4                | 5 (5.7)       |
| 5                | 6 (6.9)       |
| 6                | 1 (1.1)       |
| 7                | 5 (6.9)       |
| 8                | 3 (3.4)       |
| 9                | 2 (2.3)       |
| 10+              | 9 (10.3)      |

Number of heart surgery cases being performed in patients with Trisomy 13 and 18 per site over the study period.

### Table 2. Regional Distribution of Operations

**Number of operations performed by the North American Society of Thoracic Surgeons geographic region and Trisomy diagnosis. T13 indicates Trisomy 13; T18, Trisomy 18.**
Table 3. Demographic and Preoperative Characteristics

|                          | T13     | T18     | T13+T18  |
|--------------------------|---------|---------|----------|
|                          | n=73    | n=270   | n=343    |
| Age at surgery, mo       | 4.5 (1.4–18.6) | 3.7 (1.5–9.4) | 3.8 (1.5–11.0) |
| Male sex                 | 33 (45.2) | 64 (23.7) | 97 (28.3) |
| Race                     |         |         |          |
| White                    | 34 (46.6) | 144 (53.3) | 178 (51.9) |
| Black                    | 12 (16.4) | 45 (16.7) | 57 (16.6) |
| Hispanic                 | 18 (24.7) | 40 (14.8) | 58 (16.9) |
| Premature birth          | 13 (17.8) | 69 (25.6) | 82 (23.9) |
| Birth weight, kg         | 2.7 (2.4–3.1) | 2.0 (1.8–2.4) | 2.2 (1.8–2.6) |
| Weight at surgery, kg    | 4.8 (3.5–9.4) | 3.5 (2.5–5.6) | 3.8 (2.6–6.1) |
| Noncardiac abnormalities | 27 (37.0) | 125 (46.3) | 152 (44.3) |
| Previous cardiothoracic operation(s) | 16 (21.9) | 44 (16.3) | 60 (17.5) |
| Mechanical ventilatory support | 16 (21.9) | 82 (30.4) | 98 (28.6) |
| Cardiopulmonary resuscitation | 1 (1.4) | 3 (1.1) | 4 (1.2) |
| Gastrostomy present      | 18 (24.7) | 61 (22.6) | 79 (23.0) |
| STAT complexity level     |         |         |          |
| STAT 1                   | 25 (34.2) | 113 (41.9) | 138 (40.2) |
| STAT 2                   | 14 (19.2) | 69 (25.6) | 83 (24.2) |
| STAT 3                   | 8 (11.0) | 12 (4.4) | 20 (5.8) |
| STAT 4                   | 24 (32.9) | 67 (24.8) | 91 (26.5) |
| STAT 5                   | 2 (2.7) | 5 (1.9) | 7 (2.0) |
| STAT unknown             | 0 (0.0) | 4 (1.5) | 4 (1.2) |
| Primary diagnosis         |         |         |          |
| Ventricular septal defect | 21 (28.8) | 146 (54.1) | 167 (48.7) |
| TOF                      | 14 (19.2) | 16 (5.9) | 30 (8.7) |
| Patent ductus arteriosus | 1 (1.4) | 20 (7.4) | 21 (6.1) |
| VSD-CoA/aortic arch hypoplasia | 2 (2.7) | 15 (5.6) | 17 (5.0) |
| Atrial septal defect      | 5 (6.8) | 9 (3.3) | 14 (4.1) |
| Coarctation of the aorta | 5 (6.8) | 8 (3.0) | 13 (3.8) |
| Single ventricle          | 2 (2.7) | 7 (2.6) | 9 (2.6) |
| AVSD, complete            | 2 (2.7) | 5 (1.9) | 7 (2.0) |
| TOF with pulmonary atresia | 4 (5.5) | 3 (1.1) | 7 (2.0) |
| Other                    | 17 (23.3) | 41 (15.2) | 58 (16.9) |
| Primary procedure         |         |         |          |
| VSD repair                | 13 (17.8) | 90 (33.3) | 103 (30.0) |
| Pulmonary artery banding  | 7 (9.6) | 50 (18.5) | 57 (16.6) |
| PV/RVOT reconstruction    | 7 (9.6) | 26 (9.6) | 33 (9.6) |
| Repair of TOF            | 10 (13.7) | 14 (5.2) | 24 (7.0) |
| PDA closure               | 1 (1.4) | 20 (7.4) | 21 (6.1) |
| Blalock-Taussig shunt     | 6 (8.2) | 5 (1.9) | 11 (3.2) |
| CoA repair                | 3 (4.1) | 8 (3.0) | 11 (3.2) |

Continued
Discussion

Our study is the largest description of outcomes among patients with T13 or T18 undergoing cardiac surgery that is further classified by operative complexity and representative of the practice and outcomes across the entire United States and Canada. In this highly selective population, we found that complex surgeries (STAT 4 and 5) are being performed in this complicated patient population, and for patients with T18 there is significantly increased mortality regardless of surgical complexity. Furthermore, the incidence of complications in both patients with T13 and T18 are high. Lastly, preoperative mechanical ventilation is highly associated with mortality and should be considered in the decision to offer surgery to these patients. While physiology may play a part in this, it is often in cardiac defects that usually do not require mechanical ventilation as part of their preoperative management. Therefore, when unable to extubate these patients, caution is advisable.

There are few single-center studies, and even fewer multicenter studies, that describe surgical outcomes in patients with T13 and T18. A recent study found an overall 5-year survival rate of 9.7% and 12.3% for patients with T13 and T18, respectively, but it did not distinguish between patients who did and did not undergo cardiac surgery. Combined with other recent additions to the literature, there appears to be a trend toward more frequently providing cardiac surgery to these patients.

Table 3. Continued

| Procedure                              | T13 n=73 | T18 n=270 | T13+T18 n=343 |
|----------------------------------------|----------|-----------|---------------|
| Atrial septal defect repair            | 4 (5.5)  | 6 (2.2)   | 10 (2.9)      |
| VSD with CoA/aortic arch repair        | 1 (1.4)  | 8 (3.0)   | 9 (2.6)       |
| Other                                  | 22 (30.1)| 45 (16.7) | 67 (19.5)     |

Values are expressed as number (percentage). Preoperative data for patients with Trisomy 13 (T13) and Trisomy 18 (T18). AVSD indicates atrioventricular septal defect; CoA, coarctation of the aorta; PDA, patent ductus arteriosus; PV, pulmonary valve; RVOT, right ventricular outflow tract; STAT, Society of Thoracic Surgeons/European Association for Cardiothoracic Surgery Congenital Heart Surgery Mortality; TOF, Tetralogy of Fallot; VSD, ventricular septal defect.

Table 4. Surgical Outcomes

| Procedure            | T13 P Value to Overall | T18 P Value to Overall | T13 or T18 Overall STS Data |
|----------------------|------------------------|------------------------|----------------------------|
| In-hospital mortality| 8/73 (11)              | 42/270 (16)            | 50/343 (15)                |
| STAT 1               | 0/25 (0)               | 0.747                  | <0.001                     |
| STAT 2               | 4/14 (29)              | <0.001                 | 11/138 (8)                 |
| STAT 3               | 0/8 (0)                | 0.655                  | 12/69 (17)                 |
| STAT 4               | 4/24 (17)              | 0.024                  | 2/12 (17)                  |
| STAT 5               | 0/2 (0)                | 0.123                  | 1/14 (8)                   |
| Unknown STAT level   | 0/0 (N/A)              | 0.545                  | 0.033                      |
| Postoperative LOS, d | 13 (6–46)              | 18 (8–46)              | 16 (7–46)                  |
|                      | 8 (5–14)               | 14 (6–34)              | 13 (6–31)                  |
|                      | 15 (6–46)              | 12 (6–28)              | 13 (6–32)                  |
|                      | 14 (8–38)              | 35 (19–77)             | 28 (8–66)                  |
|                      | 27 (9–65)              | 31 (14–65)             | 31 (13–65)                 |
|                      | 50 (45–55)             | 51 (36–61)             | 51 (36–61)                 |

Values are expressed as number or number (percentage). Surgical outcomes divided by Trisomy diagnosis and compared with overall Society of Thoracic Surgeons (STS) data. LOS indicates length of stay; N/A, not available; STAT, Society of Thoracic Surgeons/European Association for Cardiothoracic Surgery Congenital Heart Surgery Mortality; T13, Trisomy 13; T18, Trisomy 18.
Historically, given the poor long-term prognosis in patients with T13 or T18, surgical therapy has not been frequently offered. Despite this common perception among healthcare providers, we found that 70% of STS centers performed surgery in these patients and 30% of those centers performed ≥5 operations over the 8-year period. While our data describe the patients undergoing heart surgery and outcomes, it unfortunately does not shed light on the number of patients offered surgery who declined or other circumstances that influenced whether surgery was performed. A large study by Peterson et al8 reported that 54% of the patients with T13 and T18 underwent an operation, whereas Maeda et al9 found that 26% of 161 patients surveyed with T13 or T18 had undergone cardiac surgery. The findings of these 2 large studies may have been very focused though, because more recently Kosiv et al10 reported that from 2004 to 2015 using the Pediatric Health Information System database, only 7% of 1668 patients diagnosed with T13 or T18 underwent congenital heart surgery. These observations suggest that our study results are representative of a highly selected group of patients with trisomy who ultimately undergo cardiac surgery.

Although we found a difference in the number of cases performed in these patients across regions, with the south representing 41.4% of all operations in patients with T13 and T18 and the northeast only 14.3%, these numbers paralleled the case distribution from around the country for the period of the study and demonstrate consistency in practice across the country. Although we cannot comment on the reasons or percentage of patients undergoing cardiac surgery, patients with T13 and T18 are known to frequently have extracardiac anomalies such as central nervous system anomalies, urogenital malformations, facial dysmorphia, and spina bifida, which may influence the offering of cardiac surgery.1,5 Like other studies, we found a high incidence (44.3%) of associated extracardiac anomalies.6,9 While this may be an underestimate and somewhat limited by the STS-CHSD data collection platform classification of an extracardiac anatomic anomaly, we speculate that it could indicate that the presence of other anomalies may only partially influence the offering of surgery from the care team as it is a known predictor of poor outcomes.6,12

The primary diagnosis resulting in surgical therapy was diverse among the patients with T13 and T18, with ventricular septal defects being extremely common in patients with T18, representing over half of the primary diagnoses and accounting for 29% of the defects among patients with T13. This finding is consistent with previous studies where ventricular septal defect was the most common diagnosis in both groups9,21; however, not all studies distinguish between the 2 trisomies. Although there were only 7 patients in the highest complexity level group (STAT category 5), there were 91 in STAT category 4, which together represent 28.5% of all operations and show a willingness to operate on patients with complex heart disease. It was surprising to find 13 operations for patients with single ventricle disease including 2 Norwood

| Table 5. Complications Following Cardiac Surgery |
|-------------------------------------------------|
| T13, No. (%) | T18, No. (%) | T13 and T18, No. (%) | Overall STS Data, % |
|--------------|--------------|---------------------|-------------------|
| n=73         | n=270        | n=343               | N=198 185         |
| Any postoperative complication 42 (57.5) | 150 (55.6) | 192 (56.0) | 36.4 |
| Cardiac arrest 5 (6.8) | 18 (6.7) | 23 (6.7) | 2.5 |
| Arrhythmia requiring pacemaker 0 (0.0) | 6 (2.2) | 6 (1.7) | 1.2 |
| Mechanical circulatory support 1 (1.4) | 6 (2.2) | 7 (2.0) | 2.3 |
| Unplanned interventional cardiovascular procedure 2 (2.7) | 4 (1.5) | 6 (1.7) | 2.1 |

Table 6. Preoperative Factors Associated With Operative Mortality

| T13 (n=73) | T18 (n=270) |
|------------|-------------|
| Survivor (n=65) | Mortality (n=8) | P Value | Unadjusted OR | Survivor (n=228) | Mortality (n=42) | P Value | Unadjusted OR |
| Mechanical ventilation 11 (17) | 5 (63) | 0.01 | 8.2 | 52 (23) | 30 (71) | <0.0001 | 8.5 |
| Gastrostomy tube 16 (25) | 2 (25) | 1 | 1.0 | 57 (25) | 4 (10) | 0.027 | 0.3 |
| Prior cardiothoracic surgery 15 (23) | 1 (13) | 0.68 | 0.5 | 42 (18) | 2 (5) | 0.024 | 0.2 |

Values are expressed as number (percentage) unless otherwise indicated. Association between selected preoperative factors and mortality by Fisher exact test analysis also showing unadjusted odds ratios (ORs). T13 indicates Trisomy 13; T18, Trisomy 18.
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This study can only comment on variables collected that were predetermined by the STS-CHSD and is not specific to these trisomies. However, a strength of this study lies in the multicenter patient population, which makes the findings highly generalizable. Importantly, this study only captures patients who underwent a surgical intervention and cannot comment on the decision making as to why certain patients were selected for surgery, nor does it allow for comparison with those not offered surgery, which is an important question. As such, preoperative deaths and cardiac catheter-ization interventions were not captured. In addition, postop-erative complications occurring after transfer to another institution from the center where the cardiac operation was performed were not recorded in the STS-CHSD, and such transfers may be more common in patients with chronic medical conditions, such as those with T13 or T18. Last, this study assessed only in-hospital complications and mortality and did not reveal intermediate or long-term survival or overall quality of life.

Conclusions

Cardiac surgery in patients with T13 or T18 is associated with high morbidity and mortality in most STAT categories across the United States. Ventricular septal defect repair is the most common repair in patients with T13 and T18, but the spectrum of surgical procedures covers all aspects of congenital cardiac surgery and all levels of complexity, with the majority of patients surviving to discharge, even in highly complex cardiac operations. Although surgical experience at any one center remains limited, a surprising number of operations are being performed in these patients overall. Preoperative mechanical ventilation was associated with risk of postoperative mortality and may be considered a relative contraindication to surgery. With regard to patients with T18, perhaps the use of gastrostomy tubes to optimize nutrition and a staged approach to surgical therapy may result in greater survival. More long-term follow-up is required in these patients, including quality-of-life assessment, to determine whether surgical intervention truly alters their natural history.

Author Contributions

Drs Cooper, Zafar, and Bryant conceptualized and designed the study, drafted the initial article, and reviewed and revised the article. Dr Riggs drafted the initial article and reviewed and revised the article. Drs Jacobs, Pasquali, Hill, Gelehrter, Swanson, Morales, and Jacobs conceptualized and designed the study, helped analyze the data, and reviewed and revised the article. Dr Wallace performed the initial analyses and reviewed and revised the article. All authors approved the final

Study Limitations and Strengths

Limitations of our study are primarily related to the nature of the STS-CHSD and those typical of a retrospective review.

operations, a bidirectional Glenn procedure, and 3 Fontan operations. When deciding on surgical intervention it is often debated whether to offer palliative or corrective treatment. This is strongly influenced by the goals of treatment and the wishes of the family. In 2004, Graham et al\(^2\) describe a multicenter report of 21 complete repairs in 35 patients with T13 and T18 with 90% survival to discharge. Of the other 14 patients (40%), 93% survived to discharge, which raised the possibility that complete repair may be comparable to palliative surgeries when it is an option.\(^2\) Costello et al\(^19\) went so far as to advocate that nearly all patients with cardiovascular indications for operative intervention should be offered complete surgical repair over palliative approaches for moderately complex congenital cardiac anomalies. This is in contrast to our findings where 21% of patients with T13 and 16.3% of patients with T18 had received previous cardiothoracic operations. The patients with T18 appeared to have benefited by having prior cardiac surgery, while there was no difference in patients with T13. However, despite the higher complete repair rate in our study, we found a higher mortality rate that was independent of surgical complexity. This is likely reflective of national practice versus case selection at an individual institution.

Because of the high risk of surgery in patients with T13 and T18, we explored risk factors associated with mortality. We chose common clinical associations in these trisomies that relate to organ system dysfunction (need for gastrostomy and mechanical ventilation) or treatment strategy (prior cardiothoracic procedure). We identified that preoperative mechanical ventilation was associated with significantly increased risk of postoperative mortality. Likewise, Graham et al\(^2\) identified mechanical ventilation as a risk factor for mortality. The strong association of mechanical ventilation and mortality suggests that preoperative mechanical ventilator support may be a clinically useful indicator of whether to offer cardiac surgery or influence the strategy of treatment (palliation versus complete repair). Interestingly, we found that having a gastrostomy tube present preoperatively was associated with improved survival in patients with T18. This could represent the importance of optimizing preoperative nutrition in these patients, as a 2017 study from Ross et al\(^22\) demonstrated the negative impact of malnutrition on 30-day survival in 0- to 5-year-old patients undergoing cardiac surgery. As previously mentioned, prior cardiothoracic surgery was also protective against mortality in patients with T18 and may reflect the use of palliation before complete repair in those with complex congenital heart disease or as a purposeful strategy to optimize the patient before complete repair.

DOI: 10.1161/JAHA.119.012349

Journal of the American Heart Association

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Disclosures

None.

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SUPPLEMENTAL MATERIAL
Table S1. STS STAT Complexity Level Case Distribution.

| STAT Complexity Level | Trisomy 13 (n=73) | Trisomy 18 (n=270) | Overall STS (n=198,185) |
|-----------------------|--------------------|--------------------|------------------------|
| STAT 1                | 25 (34.2%)         | 113 (41.9%)        | 56,155 (28%)           |
| STAT 2                | 14 (19.2%)         | 69 (25.6%)         | 70,268 (35%)           |
| STAT 3                | 8 (11.0%)          | 12 (4.4%)          | 21,403 (11%)           |
| STAT 4                | 24 (32.9%)         | 67 (24.8%)         | 38,893 (20%)           |
| STAT 5                | 2 (2.7%)           | 5 (1.9%)           | 7,865 (4%)             |
| Unknown               | 0 (0.0%)           | 4 (1.5%)           | 3,601 (2%)             |

This table displays the distribution of STS cases in relation to the Trisomy 13 and Trisomy 18 patients. STAT, The Society of Thoracic Surgeons/European Association for Cardiothoracic Surgery Congenital Heart Surgery Mortality; STS; Society of Thoracic Surgeons.