

# Operative and nonoperative outcomes in patients with trisomy 13 and 18 with congenital heart disease



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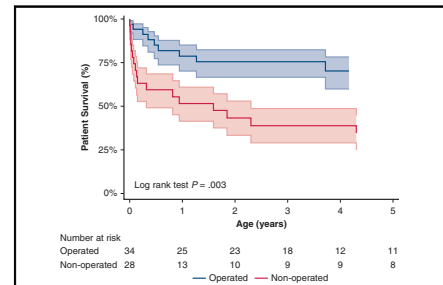
## ABSTRACT

**Objective:** To evaluate the short- and long-term outcomes of cardiac repair versus nonoperative management in patients with trisomy 13 and trisomy 18 with congenital heart disease.

**Methods:** An institutional review board-approved, retrospective review was undertaken to identify all patients admitted with trisomy 13/18 and congenital heart disease. Patients were divided into 2 cohorts (operated vs nonoperated) and compared.

**Results:** Between 1985 and 2023, 62 patients (34 operated and 28 nonoperated) with trisomy 13 (n = 9) and trisomy 18 (n = 53) were identified. The operated cohort was 74% girls, underwent mainly The Society of Thoracic Surgeons-European Association for Cardio-Thoracic Surgery mortality category 1 procedures (n = 24 [71%]) at a median age of 2.5 months (interquartile range [IQR], 1.3-4.5 months). This compares with the nonoperative cohort where 64% (n = 18) would have undergone The Society of Thoracic Surgeons-European Association for Cardio-Thoracic Surgery mortality category 1 procedures if surgery would have been elected. The most common diagnosis was ventricular septal defect. Postoperative median intensive care unit stay was 6.5 days (IQR, 3.7-15 days) with a total hospital length of stay of 15 days (IQR, 11-49 days). Thirty-day postoperative survival was 94%. There were 5 in-hospital deaths in the operated and 7 in the nonoperated cohort. Median follow-up was 15.4 months (IQR, 4.3-48.7 months) for the operated and 11.2 months (IQR, 1.2-48.3 months) for the nonoperated cohorts. One-year survival was 79% operated versus 51.5% nonoperated (P < .003). Nonoperative treatment had an increased risk of mortality (hazard ratio, 3.28; 95% CI, 1.46-7.4; P = .004).

**Conclusions:** Controversy exists regarding the role of primary cardiac repair in patients with trisomy 13/18 and congenital heart disease. Cardiac repair can be performed safely with low early mortality and operated patients had higher long-term survival compared with nonoperated in our cohort. (JTCVS Open 2024;20:123-31)



Survival in patients undergoing operation was significantly higher than those who did not.

## CENTRAL MESSAGE

Primary cardiac repair is safe and effective in patients with T13/T18 with CHD and can greatly improve survival. Despite comorbidities, most families are satisfied with their child's quality of life.

## PERSPECTIVE

Primary cardiac repair in patients with T13/T18 with CHD is controversial. Here we demonstrate it is safe and effective when standard operative criteria are followed and can greatly improve survival. Despite a high burden of morbidities, most families are satisfied with their child's quality of life. We advocate for early referral for primary cardiac repair in patients with T13/T18 with CHD.

Trisomy (T) 13 and T18 are the most common trisomy disorders following T21 with a prevalence of 1.68 and 4.08 per 10,000 live births, respectively.<sup>1,2</sup> Patients present with a variety of anomalies, including cleft palate, genitourinary

malformations, brain and spinal cord deformities, and congenital heart disease (CHD).<sup>3</sup> Almost 90% of patients with T13 and T18 have associated congenital heart defects.<sup>4</sup> Most commonly ventricular septal defects (VSDs), patent

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**Abbreviations and Acronyms**

ASD = atrial septal defect  
 CHD = congenital heart disease  
 T = trisomy  
 VSD = ventricular septal defect

ductus arteriosus, and atrial septal defects (ASDs).<sup>3-7</sup> Unlike T21, patients with T13 and T18 are often viewed to have such a dismal prognosis that they are not routinely referred for repair.<sup>7-14</sup> Instead, the majority are sent home to hospice without surgical consultation mirroring the experience of T21 patients 50 years ago.<sup>3,6,11,15,16</sup> Without intervention, the median mortality within the first week of life is reported to be 47% for T13 and 42% for T18 with the largest international registry reporting a 1-year mortality of 87% for T13 and 88% for T18.<sup>1</sup>

For the past decade, it has been the practice at our institution to offer primary cardiac repair to patients with T13/T18 and 2-ventricle heart disease with the rationale that it improves patient survival and quality of life for the patient and the family. Specifically, the diagnosis of T13/T18 is not a disqualifying metric for repair. These patients were evaluated against the same criteria that patients without a T diagnosis were measured against. More recently, with the advent of social media and online disease-specific forums, families are seeking out surgical programs willing to operate on their children when their local institutions refuse to offer cardiac repair highlighting the disparate views of this diagnosis.<sup>8,9,11</sup> Few studies compare primary cardiac repair to medical management, and conclusions are limited by small sample size.<sup>8,17</sup> We present our short and long-term outcomes of primary cardiac repair versus nonoperative management in T13 and T18 patients to aid treatment decision.

**METHODS**

A retrospective review was performed to identify all patients admitted with a diagnosis of T13 or T18 and CHD between November 1985 and October 2023. The study was approved by our institutional review board (IRB-P00033023; September 3, 2019) and patient consent was waived. Patients with T13 or T18 with a cardiac defect were identified from the cardiac surgery and cardiology database. Mosaic T13/T18 patients were excluded because they are generally viewed to have a milder form of the syndrome.<sup>18-20</sup> Primary end point was survival, and secondary end points were cost and patient's health status.

The patients were subdivided into 2 groups: operated and nonoperated. Treatment choice was made by the family after interdisciplinary consultation. Surgical treatment was not offered for single-ventricle disease. Demographic, prenatal, postnatal, echocardiographic, catheterization, operative follow-up, and cost data were collected and analyzed. Follow-up for survival analysis was obtained in 100% of operated and 88% of nonoperated patients.

**Statistical Analysis**

Patient and operative characteristics are represented as number (%) for categorical variables and median and interquartile range (IQR) for continuous variables. Wilcoxon rank-sum test for continuous and Fisher exact test

for categorical data comparisons were used. Kaplan-Meier survival curves were compared between operated and nonoperated groups by log-rank test with numbers at risk presented for each year of age. Multivariable Cox proportional hazards regression analysis was used to determine the independent predictors of mortality. The multivariable model was created using stepwise backward elimination with criterion for removal of  $P > .1$  to fit a parsimonious model. All statistical analyses were performed using Stata version 17.1 (StataCorp LLC).

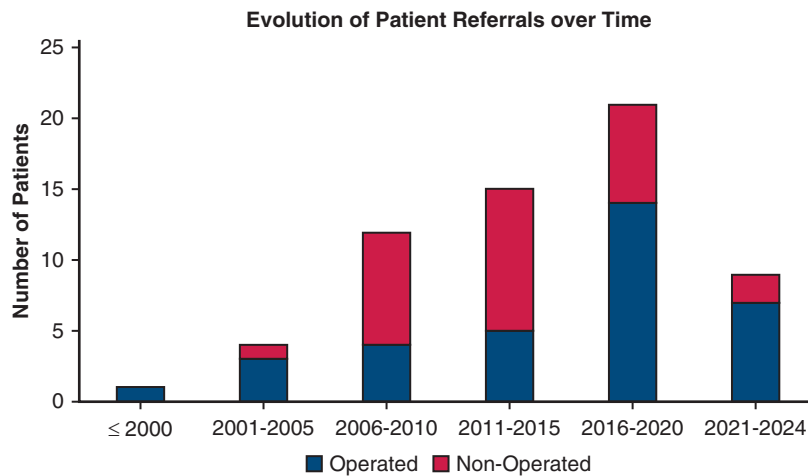
**RESULTS****Baseline Characteristics**

Between 1985 and May 2023, 62 patients with T13 (n = 9) or T18 (n = 53) and CHD were evaluated. **Figure 1** shows the evolution of patients treated at our center over time. Gestational age was 37 weeks (IQR, 36-39 weeks) and birth weight was 2.08 kg (IQR, 1.8-2.4 kg). Seventy-nine percent of patients were girls (n = 49). The most common CHDs were ASDs and VSDs (**Table 1**). Eleven patients (18%) had duct-dependent lesions and 7 patients (11%) had arch hypoplasia. Prenatal diagnosis was known in 35 patients (56%) and intrauterine growth retardation was present in 26 patients (42%). The most common concomitant lesions were structural brain deformities (n = 37 [59%]) and structural kidney deformities (n = 16 [26%]).

The study population was then divided into an operated (n = 34) and nonoperated (n = 28) cohort. Baseline characteristics were similar between the 2 groups on multivariable analysis (**Table 2**). There was no difference in the rates of preoperative mechanical ventilation or pulmonary hypertension. The only difference was 67% of the nonoperated cohort were initially sent home (n = 19), whereas only 32% of the operated cohort (n = 11) were. In the nonoperated cohort, 9 patients had bidirectional flow across the VSD (32%) and underwent catheterization. All 9 were found to have elevated pulmonary vascular resistance 14.7 Wood units (IQR, 14.1-25 Wood units). In the operated cohort, bidirectional flow across the VSD was seen in 15 patients (44%). Eight patients underwent catheterization with 6 diagnosed with elevated pulmonary vascular resistance.

**Operative Details**

Overall, 34 patients underwent 35 operations. Patients weighed 2.95 kg (IQR, 2.2-4.3 kg) at the time of repair and were taken to the operating room at a median of 2.5 months (IQR, 1.35-4.5 months). Mechanical ventilation was required preoperatively in 44% (n = 15), and 26% (n = 9) had been treated for a previous infection with intravenous antibiotics. Nine patients (26%) had a gastrostomy tube in place at the time of operation. Cardiac diagnoses are listed in **Table 1**. Suprasystemic right ventricular pressures were present in 9 patients (26%). Eight patients (24%) underwent preoperative catheterization with only 1 patient receiving a patent ductus arteriosus stent.



**FIGURE 1.** Referrals to our center from 1985 until October 2023. Sixty-two patients were referred with 34 undergoing operative and 28 undergoing nonoperative treatment. There has been a steady increase in overall referrals and a majority of patients from the operated group have been treated during the past 5 years (21 out of 34 patients).

The most common procedures were ASD/VSD closures ( $n = 28$  [74%]). Distribution of Society of Thoracic Surgeons-European Association for Cardio-Thoracic Surgery mortality categories are presented in Figure 2. Cardiopulmonary bypass time was 107 minutes (IQR, 82-134 minutes), and crossclamp time was 73 minutes (IQR, 61-94 minutes). All patients successfully weaned off cardiopulmonary bypass. Postoperative echocardiogram showed good biventricular function in 94% ( $n = 32$ ). Patients were extubated 6 days postoperatively (IQR, 3-10 days). Eleven patients (32%) required postoperative tracheostomy with 1 patient known to be decannulated. Of these, 6 (54%) required mechanical ventilation preoperatively. Only 3 patients were able to establish oral feeds postoperatively. Thirteen were fed via nasogastric tube and 10 were fed via gastrostomy tube at discharge. Patients were transferred to the floor 6.5 days postoperatively (IQR, 3.75-15 days). Total length of stay was 15 days (IQR, 11.25-49 days).

### Operated Versus Nonoperated Survival

There was a 25% mortality rate ( $n = 7$ ) during the first 30 days of life in the nonoperated cohort versus 3% ( $n = 1$ ) in the operated group. Postoperative 30-day survival was 94% ( $n = 32$ ). Follow up of the operated and nonoperated group patients was 15 months (IQR, 4-49 months) and 11 months (IQR, 1.2-48 months), respectively. Survival at 1, 3, and 5 years for the operated cohort was 79%, 76%, and 70%, whereas survival for the nonoperated cohort was 52%, 39%, and 35% ( $P < .001$ ) (Figure 3).

Of those operated, 6 died of sepsis and respiratory failure, whereas 1 died of multiorgan failure. Of these, 4 (67%) were ventilated preoperatively. The most common cause of death in the nonoperated group was respiratory failure ( $n = 5$ ), followed by withdrawal of care ( $n = 4$ ) and infection ( $n = 2$ ). Two patients arrested while having outpatient

procedures (urologic repair and tonsillectomy). Nonoperative treatment had an increased risk of mortality, adjusted hazard ratio, 3.28 (95% CI, 1.46-7.4;  $P = .004$ ) adjusting for pulmonary hypertension and structural kidney disease.

Improved survival for the operated patients was observed throughout the course of the study and when T13 and T18 patients were evaluated individually (Figure 4). Nonoperated T13 patients had the worst survival overall. Currently, there are 7 long-term survivors with more than 5 years of

**TABLE 1. Congenital heart defects in the operated and nonoperated group patients**

Operated group (n = 34)	Nonoperated group (n = 28)
Aortic arch hypoplasia, CoA, multiple VSDs, PDA, PFO (n = 7)	AVC
ASD, PDA	DORV, VSD, PA, dysplastic AV, PDA, PFO
CoA	DORV, VSD, PDA (n = 2)
Multiple muscular VSDs, ASD, PDA	Mitral dysplasia
Multiple VSDs, s/p PAB	TOF/AVSD
TOF, ASD, PDA (n = 2)	TOF/PA/PDA (n = 3)
Tracheoinnominate fistula, vascular ring	VSD (n = 5)
VSD (n = 3)	VSD, ASD (n = 9)
VSD, ASD (n = 5)	VSD, ASD, PDA (n = 2)
VSD, ASD, PDA (n = 5)	VSD, MR, TR, AI
VSD, PFO, PDA (n = 6)	VSD, PDA (n = 2)
VSD, PFO, PDA, severe AI	

CoA, Coarctation of the aorta; VSD, ventricular septal defect; PDA, persistent ductus arteriosus; PFO, persistent foramen ovale; AVC, atrioventricular septal defect; ASD, atrial septal defect; AV, aortic valve; DORV, double outlet right ventricle; PA, pulmonary artery; s/p PAB, pulmonary artery banding; TOF, tetralogy of Fallot; AVSD, atrioventricular septal defect; VSD, ventricular septal defect; MR, mitral regurgitation; TR, tricuspid regurgitation; AI, aortic insufficiency.

**TABLE 2. Comparison of baseline factors between operated and nonoperated patients**

Variable	Operated group (n = 34)	Nonoperated group (n = 28)	P value
Sex			.35
Female	25 (73.5)	24 (85.7)	
Male	9 (26.5)	4 (14.3)	
Trisomy			.999
13	5 (14.7)	4 (14.3)	
18	29 (85.3)	24 (85.7)	
Gestational age (wk)	37 (35-41)	38 (36-39)	.453
Birth weight (kg)	2.2 (1.8-2.5)	2 (1.8-2.2)	.329
Prenatal diagnosis	23/32 (71.9)	15/25 (60)	.404
IUGR	16 (47.1)	13/26 (50)	.821
Initially sent home	11 (32.4)	19 (67.9)	.01*
Cleft palate	5 (14.7)	6 (21.4)	.523
PHTN	7/33 (21.4)	12 (42.9)	.097
TEF	3 (8.8)	1/27 (3.7)	.623
Structural brain anomalies	21 (61.8)	16/25 (64)	.999
Structural kidney	11 (32.4)	8 (28.6)	.788
Ventilator support	17 (50)	8 (28.6)	.12
Duct dependent	5/33 (15.2)	5 (17.9)	.999
ASD	23/31 (74.2)	22 (78.6)	.766
VSD	29/32 (90.6)	26 (92.9)	.999
Arch hypoplasia	6/32 (18.8)	2 (7.1)	.264
Pulmonary HTN	6/8 (75)	9/9 (100)	.206

Values are presented as median (interquartile range) for continuous data and n (%) for categorical data. P values were calculated using the Wilcoxon rank-sum test and Fisher exact test. IUGR, Intrauterine growth restriction; PHTN, pulmonary hypertension; TEF, tracheoesophageal fistula; ASD, atrial septal defect; VSD, ventricular septal defect; HTN, hypertension. \*Statistically significant.

follow-up in the operated group versus 3 in the nonoperated group. All patients have T18.

**Patient’s Health Status**

Of patients alive, 63% (15 out of 24) of operated versus 71% (5 out of 7) of nonoperated patients were described by their parents as doing well in their last clinic note. In the operated cohort, 3 patients (13%) have seizures that significantly reduce their quality of life and 3 are ventilator-dependent (13%). Two operated group patients were noted to be attending school, whereas 1 patient, age 39 years, lives independently. In the nonoperated cohort, 4 patients experienced congestive heart failure and pulmonary hypertension (66%).

**Number of Admissions and Cost**

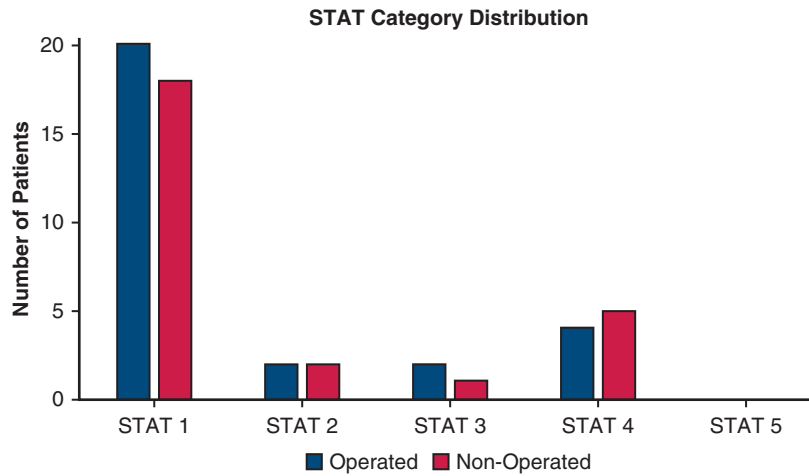
Admission and cost data were available for 47 patients. The number of admissions for alive patients in the operated

cohort was 1 (IQR, 1-3.75) over 54 months (IQR, 9.6-99 months) versus 2 (IQR, 1.5-3.5) over 47 months (IQR, 37-60 months) in the nonoperated cohort (P = .67). Operated patients were in the hospital longer than nonoperated patients (34 vs 12 days; P = .034) with greater total hospital charges (\$210,754 vs \$53,050; P = .22) (Table 3). On average, they received more echocardiograms (6 vs 3; P = .016) and a similar number of catheterizations. Total cost per year of life was more in the nonoperated group than the operated group, although this was not statistically significant (\$70,743 vs \$25,487).

**DISCUSSION**

This study evaluates the short- and long-term outcomes of primary cardiac repair versus nonoperative management in T13 and T18 patients with CHD at a large tertiary referral pediatric hospital. Over a 39-year period, 62 patients with T13/T18 and CHD were admitted to our hospital with steadily increasing numbers over the past decade. More than half of the patients (n = 34 [55%]) underwent cardiac repair, which is higher than previously reported.<sup>6,8,14,16,19-21</sup> This results from an institutional approach to offer definitive surgical repair rather than palliation to families when possible. Of note, 1 patient with multiple muscular VSDs underwent pulmonary artery band placement before definitive repair. It also likely reflects the trend that families seeking treatment for their children get increasingly referred to our center. The decision to pursue surgery is made after extensive multidisciplinary counseling. The majority had simple cardiac lesions, mainly VSDs, that are amenable to durable repair and associated with low mortality (100% 30-day survival) in the general population. More complex lesions that require multiple interventions like pulmonary atresia or double outlet right ventricle were exclusively treated nonoperatively. However, the operative cohort certainly also includes complex procedures such as aortic arch repair. There was no difference seen in Society of Thoracic Surgeons-European Association for Cardio-Thoracic Surgery category distribution between groups.

Aside from the families’ wishes the most important determinant of operative candidacy in 2-ventricle T13/T18 patients with CHD is the presence of pulmonary hypertension. Patients with evidence of pulmonary hypertension on echocardiogram (ie, bidirectional flow across the VSD) were referred for catheterization. Those found to have elevated pulmonary vascular resistance were managed nonoperatively because the window to benefit from operative closure of the VSD had passed. These same operative criteria apply to non-T patients. Age at the time of catheterization in the nonoperative group was 289 days (IQR, 248-621 days) versus 81 days (IQR, 51-161 days) in the operative group, although due to small sample size this did not achieve statistical significance



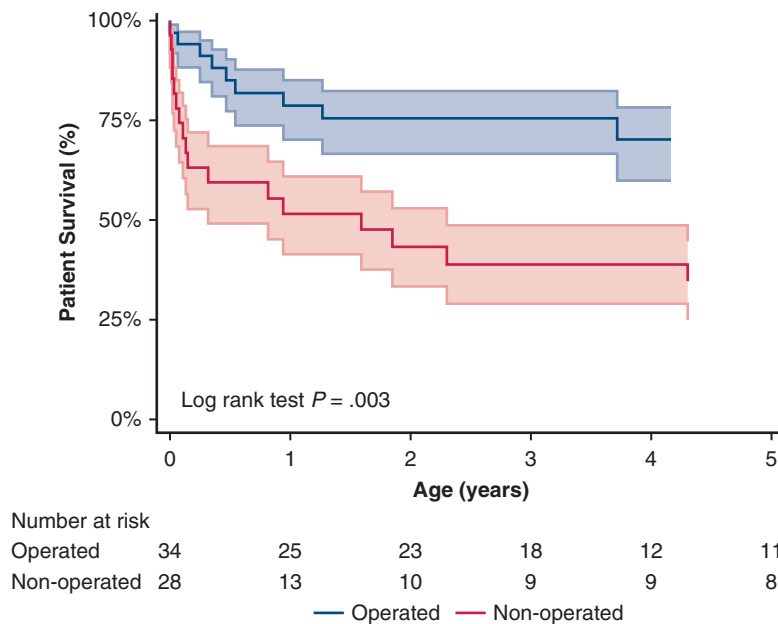
**FIGURE 2.** There was no difference in Society of Thoracic Surgeons-European Association for Cardio-Thoracic Surgery (STAT) category distribution between the 34 operated and 28 nonoperated patients ( $P = .63$ ).

( $P = .15$ ). For this reason, we advocate for early operative evaluation, so families still have the option for operative repair if they so choose.

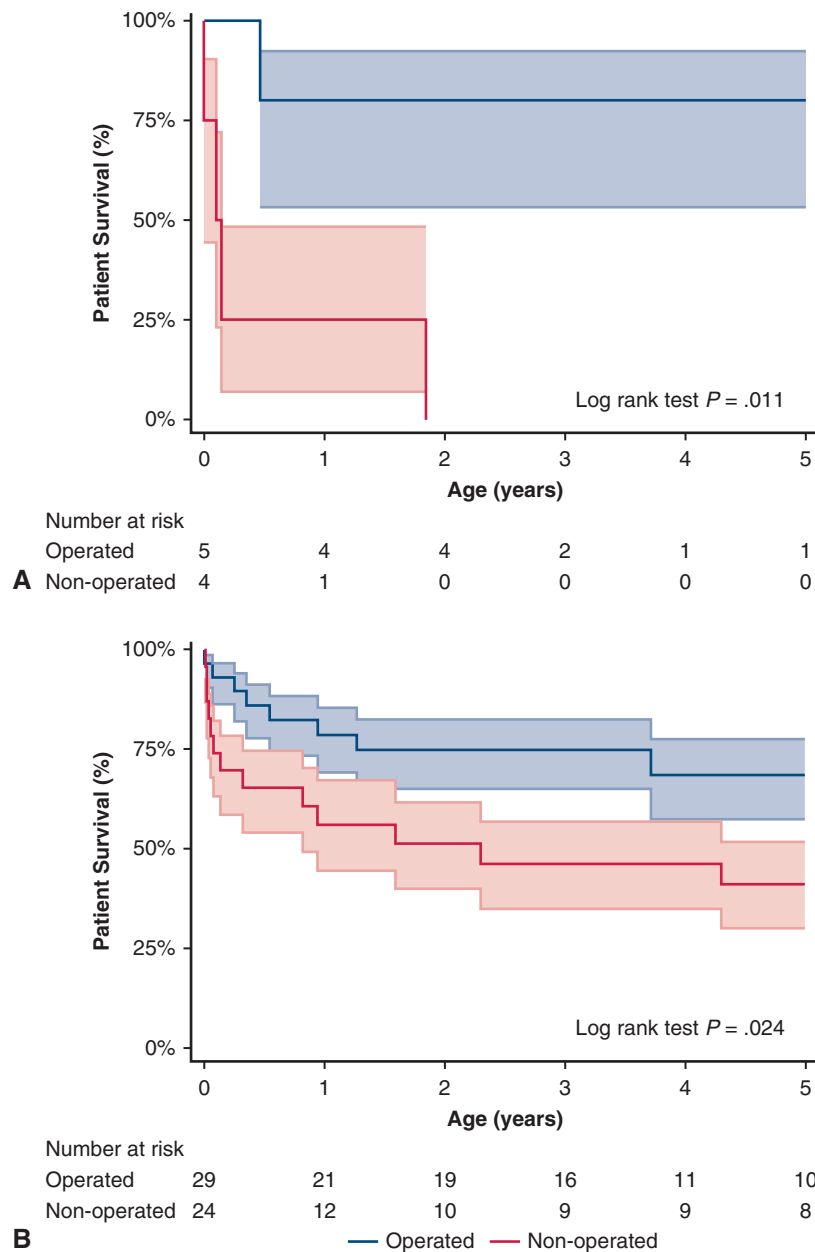
Our nonoperated cohort had a 30-day mortality of 25% and 5-year mortality of 65.4%. This compares with data from the largest international registry with reported median mortality within the first week of life of 47% for T13 and 42% for T18 and 1-year mortality of 87% for T13 and 88% for T18.<sup>1</sup> In contrast, the operated cohort had a post-operative 30-day mortality of 6% and 5-year mortality of 30%, highlighting that surgical repair can be safely performed in T13/T18 patients and a considerable number might survive long-term if treated. Because this study

does not include all patients born with T13/T18 and CHD and there was no prospective allocation to a treatment group, there is certainly some selection bias, and it is unclear how many patients died before referral.

Nevertheless, the long-held belief that T13/T18 and CHD is universally fatal in infancy cannot be supported by our data and seems outdated and incorrect (Figure 5).<sup>8,11-13,22</sup> Most of our patients were born after 2010 and therefore represent a much more contemporary cohort compared with previously published series. Additionally, it should be noted that whereas the 30-day survival was 75% without intervention, it decreased to 28% by 6-months, which suggests a window for possible intervention passing. These



**FIGURE 3.** Survival for operated patients was significantly higher than nonoperated patients throughout the study period ( $P < .001$ ). Shading indicates 70% CI.



**FIGURE 4.** A, Survival of operated trisomy (T) 13 and 18 patients versus nonoperated. Shading indicates 70% CI. B, Survival for operated T18 patients was significantly higher than nonoperated T18 patients ( $P = .006$ ). Patients with T13 had worse survival compared with patients with T18. Survival for patients with T13 undergoing operation was better than nonoperated patients with T13 but was limited by small sample size and was not statistically significant ( $P = .06$ ).

survival rates are in sharp contrast to previous reports by Rasmussen and colleagues<sup>23</sup> of 1-month survival of 30% and 6-month survival of 10% to 15% in a cohort from 1968 to 1999.

Apart from potential selection bias, important advances in critical care have occurred over the past decades, which might also improve our patients' overall survival. Central apnea is a known comorbidity of T13/T18 patients and an important determinant of survival.<sup>10</sup> It has become our

practice to use chronic continuous positive airway pressure therapy and or perform tracheostomies for patients exhibiting life-threatening central apnea. Thus, 32% of our repaired patients have tracheostomies and are ventilator dependent, which is reflective of the underlying condition rather than a postoperative complication. It should be noted that half of the patients who did not survive in the operative cohort were vent dependent at the time of operation (5 out of 10). Additionally, almost all patients will require access for

**TABLE 3. Comparison of accrued costs and procedures by operated status**

Variable	Operated group (n = 20)	Nonoperated group (n = 27)	P value
Years alive	6.5 (0.9-11)	1.8 (0.05-5)	.027*
Days in hospital	34 (16-146)	12 (4-59)	.034*
Total hospital charges† (\$) <ul style="list-style-type: none"> <li>Total hospital charges per year alive</li> <li>Total hospital charges per day in hospital</li> </ul>	210,754 (21,995-1,126,312)	53,050 (15,767-307,089)	.227
	49,553 (1326-379,194)	139,165 (3662-940,312)	.518
	10,736 (302-12,498)	8508 (7306-10,610)	.896
Total hospital cost† (\$) <ul style="list-style-type: none"> <li>Total hospital cost per year alive</li> <li>Total hospital cost per day in hospital</li> </ul>	100,526 (13,563-556,457)	33,470 (9398-198,464)	.253
	25,487 (499-217,540)	70,743 (1639-571,389)	.504
	5150 (75-6331)	4471 (3826-6113)	.991
No. of echocardiograms	6 (3-9)	3 (1-6)	.016*

Values are presented as median (interquartile range) for continuous data and n (%) for categorical data. P values were calculated using the Wilcoxon rank-sum test and Fisher exact test. \*Statistically significant. †Cost and charges data were available on n = 47 patients.

enteral nutrition because only 3 patients in our cohort were able to achieve oral feeds. The likelihood of requiring postoperative mechanical ventilation and feeding tubes should be discussed with families preoperatively so the family's goals of care can be aligned with the expected outcome. It is important to note that 19% of patients in the nonoperative cohort died after withdrawal of care.

Notwithstanding the rate of postoperative mechanical ventilation, 63% of families with living children describe their child as happy and doing well. Despite ventilator dependence, seizure disorder limited quality of life in 18%. Although quality of life could not be assessed with a validated questionnaire, data from chart review reflects family satisfaction. Our findings are consistent with a recent study from the University of Nebraska where parents described their child's quality of life as "high."<sup>9</sup> These data in concert with operative outcomes are incredibly important to families trying to make a well-informed decision about the best course of action for their child and what to expect long-term.

Our cost analysis was inconclusive in showing a significant financial difference between operative and nonoperative management of patients with T13/T18 and CHD. Patients in the nonoperated group accumulated significant cost over a brief period of time and then died, whereas patients in the operated group accumulated significant cost and survived.

Although the cohort size limited our ability to make direct comparisons between those patients who survived and those who did not in the operated cohort, patients who did well exhibited the following trends: age at operation of 2.1 months (IQR, 1.45-4.79 months), weight >3.09 kg (IQR, 2.47-4.58 kg), and gestational age >37 weeks' gestation. Nonsurvivors tended to be older at operation (2.8 months; IQR, 1.38-3.47 months), weigh less (2.5 kg, IQR, 2.15-3.2 kg), be preoperatively ventilated

(5 out of 10) and have more structural brain abnormalities (7 out of 10) than survivors.

### Limitations

This is a retrospective study of a rare disease affecting a small cohort of patients. Although we did include all patients admitted to our hospital, our cohort is not representative of all T13/T18 patients because many parents seeking treatment rather than palliation presented to our center, devoted to the care of their children, which might have positively influenced overall outcome. There was no structured quality of life assessment and data are limited to chart review. Rigorous quality of life metrics are the focus of a forthcoming separate publication. Our cost analysis only covers the admission and charges accrued at our center. Some patients received additional care at their local centers, which was not captured.

### CONCLUSIONS

Although primary cardiac repair in patients with T13/T18 and CHD is controversial,<sup>24</sup> we demonstrate that primary cardiac repair is safe and effective in patients with T13/T18 and CHD who meet standard operative criteria and can greatly improve survival. Despite a high burden of comorbidities, most families are satisfied with their child's quality of life. Therefore, we advocate for early referral and consideration of primary cardiac repair in children with T13/T18. Detailed family counseling is needed regarding long-term comorbidities. Treatment decisions remain center, as does individual choice.

### Conflict of Interest Statement

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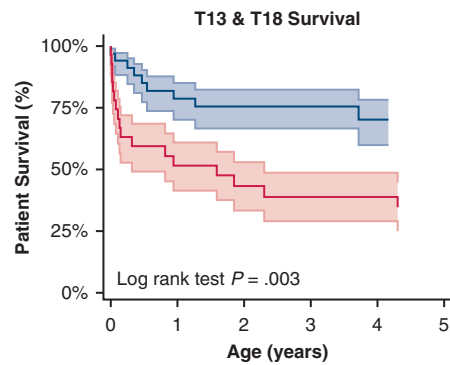
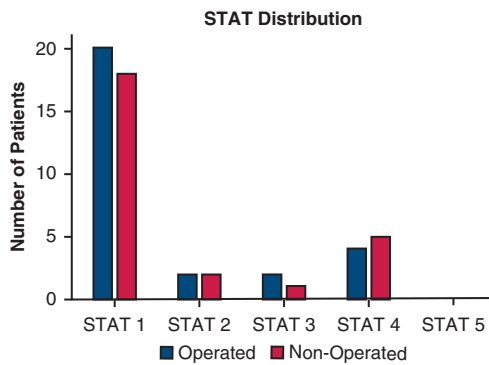
Operative and Non-Operative Outcomes in Trisomy 13 and 18 Patients with Congenital Heart Disease

Methods

Retrospective chart review of all Trisomy 13 (T13) and Trisomy 18 (T18) patients undergoing primary cardiac repair vs non-operative management at Boston Children's Hospital from 1985 to 2023. • 62 patients (34 operated, 28 non-operated) • T13 (n = 9), T18 (n = 53) • 74% female • 71% STAT1 cases, most commonly VSD • Median age at OR 2.5 months (IQR 1.3-4.5)

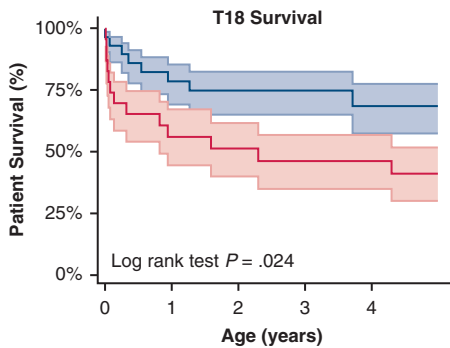
Results

Post-Op median ICU stay 6.5 days (IQR 3.7-15) • Total Hospital LOS 15 days (IQR 11-49) • 30-day Post Op Survival 94% • Median FU 15.4 months (IQR 4.3-48.7) Operated vs 11.2 months (IQR 1.2-48.3) Non-Operated • One-year Operated Survival 79% vs 51.5% Non-Operated  $P < .003$   
 • Non-Op Treatment increased risk of mortality (HR = 3.28, 95% CI: 1.46, 7.4,  $P = .004$ )



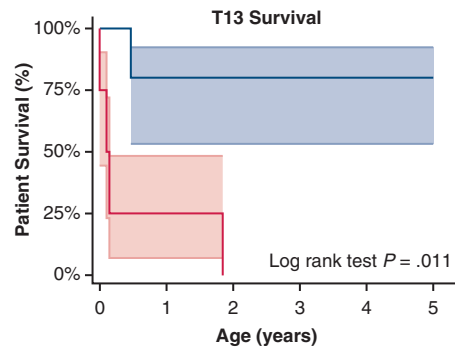
Number at risk

	0	1	2	3	4	5
Operated	34	25	23	18	12	11
Non-operated	28	13	10	9	9	8



Number at risk

	0	1	2	3	4
Operated	29	21	19	16	11
Non-operated	24	12	10	9	9



Number at risk

	0	1	2	3	4	5
Operated	5	4	4	2	1	1
Non-operated	4	1	0	0	0	0

Implications

Cardiac repair can be performed safely with low early mortality and operated patients had higher long-term survival compared to non-operated in our cohort.

**FIGURE 5.** Patients with trisomy 13 and trisomy 18 undergoing operation with congenital heart disease had improved survival compared with nonoperated patients.

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:** trisomy 13, trisomy 18, congenital heart disease