Cardiac surgery in children with trisomy 13 or trisomy 18: How safe is it?

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ABSTRACT

Objective: Surgery for heart defects in children with trisomy 13 or 18 is controversial. We analyzed our 20-year experience.

Methods: Since 2002, we performed 21 operations in 19 children with trisomy 13 (n = 8) or trisomy 18 (n = 11). Age at operation was 4 days to 12 years (median, 154 days). Principal diagnosis was ventricular septal defect in 10 patients, tetralogy of Fallot in 7 patients, arch hypoplasia in 1 patient, and patent ductus arteriosus in 1 patient.

Results: The initial operation was ventricular septal defect closure in 9 patients, tetralogy of Fallot repair in 7 patients, pulmonary artery banding in 1 patient, patent ductus arteriosus ligation in 1 patient, and aortic arch/coarctation repair in 1 patient. There were no operative or hospital deaths. Median postoperative intensive care and hospital stays were 189 hours (interquartile range, 70-548) and 14 days (interquartile range, 8.0-37.0), respectively, compared with median hospital stays in our center for ventricular septal defect repair of 4.0 days and tetralogy of Fallot repair of 5.0 days. On median follow-up of 17.4 months (interquartile range, 6.0-68), 1 patient was lost to follow-up after 5 months. Two patients had reoperation without mortality. There have been 5 late deaths (4 with trisomy 18, 1 with trisomy 13) predominately due to respiratory failure from 4 months to 9.4 years postoperatively. Five-year survival was 66.6% compared with 24% in a group of unoperated patients with trisomy 13 or 18.

Conclusions: Cardiac operation with an emphasis on complete repair can be performed safely in carefully selected children with trisomy 13 or trisomy 18. Hospital resource use measured by postoperative intensive care and hospital stays is considerably greater compared with nontrisomy 13 and 18. (JTCVS Open 2022;12:364-71)

Trisomy 13 and trisomy 18 are uncommon genetic syndromes associated with multiple congenital defects, of which cardiac anomalies are the most common occurring





CENTRAL MESSAGE

Cardiac surgery can be performed safely in children with trisomy 13 or trisomy 18 with low mortality and morbidity and enhanced survival.

PERSPECTIVE

Cardiac surgery for children with trisomy 13 or trisomy 18 should be revisited to identify ideal candidates and long-term results.

in 57% to 91% of affected children.^{1,2} Historically, the natural history of children with trisomy 13 or trisomy 18 has been bleak with commonly reported 1-year survival of only 10% to 20%.³⁻⁶ The decision to offer cardiac surgical intervention for these patients has been controversial.⁷⁻¹² An early report from the Pediatric Cardiac Care Consortium cardiac registry suggested that cardiac surgery in children with trisomy 13 or trisomy 18 can be performed safely.¹³ We analyzed our 20-year single-institution experience with cardiac surgical intervention for children with trisomy 13 or trisomy 18 and compared our surgical outcomes with a similar population of children with trisomy 13 or trisomy 18 and cardiac lesions who were not offered surgical intervention.

PATIENTS AND METHODS

This retrospective study was approved by the Institutional Review Board of Children's Healthcare of Atlanta. The requirement for written

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Read at the 101st Annual Meeting of The American Association for Thoracic Surgery: A Virtual Learning Experience, April 30-May 2, 2021.

This retrospective study was approved by the Institutional Review Board of Children's Healthcare of Atlanta. The requirement for written informed consent was waived due to the retrospective nature of this study. IRB#04052022, approved May 18, 2022.

Received for publication May 15, 2021; revisions received Aug 4, 2022; accepted for publication Sept 21, 2022; available ahead of print Oct 28, 2022.

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Abbreviations and Acronyms

IQR = interquartile range

VSD = ventricular septal defect

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informed consent was waived due to the retrospective nature of this study (IRB s#04052022, approved May 18, 2022).

Data Collection

We searched for all patients with a diagnosis of trisomy 13 or trisomy 18 in our pediatric cardiac surgical database who underwent a cardiac operation from January 2002 to April 2021. Demographic, clinical,

TABLE 1. Surgical patients

echocardiographic, and operative details were extracted from the medical record. Survival was determined from the inpatient and outpatient medical records or from a phone call to the family within 3 months of termination of data collection (April 2021). Our inpatient echocardiographic database was also examined to identify children with a diagnosis of trisomy 13 or trisomy 18 from 2007 to 2021 to identify children with trisomy 13 or trisomy 18 with cardiac defects who did not undergo a cardiac operation during roughly the same time. Both database queries were designed to only identify patients admitted to Children's Healthcare of Atlanta.

Patient Population

Nineteen children were identified who had trisomy 13 or trisomy 18 and had a cardiac operation (Table 1). Eight children had trisomy 13 (3 mosaic), and 11 children had trisomy 18 (2 mosaic). All patients had biventricular anatomy and did not have any unrepaired major comorbidity. Only 2 patients underwent operation before 2012. The age at initial operation ranged from 4 days to 12.7 years (median 154 days, interquartile range [IQR], 77-465 days), and the weight ranged from 1.9 to 19.7 kg (median, 4.4 kg, IQR, 3.6-7.8 kg). Ten children had a ventricular septal defect (VSD), and 7

Patient no.	Chromosomal abnormality	Age at operation (d)	Weight (kg)	Primary diagnosis	Admission from home?	Preoperative respiratory support
1	Trisomy 18 (mosaic)	112	4.4	TOF-like DORV	Yes	None
2	Trisomy 18	4636	19.7	Sinus venosus ASD, VSD, aortic regurgitation	Yes	None
3	Trisomy 13 (mosaic)	4	3.6	Coarctation, hypoplastic aortic arch	Newborn	None
4	Trisomy 13	704	13.5	TOF	Admitted for 3 d preoperatively after at-home arrest	2 L/min NC
5	Trisomy 13	240	6.7	VSD, ASD	Admitted 6 d earlier with heart failure	1 L/min NC
6	Trisomy 18	58	1.9	VSD, ASD	Ex-premature 1.5 kg, in NICU	7 L/min HFNC
7	Trisomy 18	154	4.3	VSD, ASD	Ex-premature 32 wk 1.8 kg in NICU	Tracheostomy on ventilator
8	Trisomy 13 (mosaic)	39	4.6	TOF	Admitted 4 d preoperatively for TOF spells	None
9	Trisomy 18	28	1.7	VSD	Term 1.7 kg in NICU preoperatively	None
10	Trisomy 13 (mosaic)	568	10.9	VSD, ASD	Yes	None
11	Trisomy 13	487	13.4	VSD, ASD, PDA	Yes	Tracheostomy on ventilator
12	Trisomy 18	79	3.9	VSD	Admitted 5 wk preoperatively for respiratory failure	3 L/min HFNC
13	Trisomy 18 (mosaic)	288	6.3	VSD, ASD	Yes	None
14	Trisomy 18	127	4.2	TOF	Yes	None
15	Trisomy 13	465	7.8	TOF	Yes	None
16	Trisomy 13	77	3.1	PDA	Yes	2 L/min NC
17	Trisomy 18	308	4.7	TOF	Yes	None
18	Trisomy 18	104	3.1	VSD	Term 1.9 kg IUGR in NICU	4 L/min HFNC
19	Trisomy 18	159	4.1	TOF-like DORV, second VSD	Yes	1/16 L/min NC

TOF, Tetralogy of Fallot; DORV, double-outlet right ventricle; ASD, atrial septal defect; VSD, ventricular septal defect; NC, nasal cannula; HFNC, high-flow nasal cannula; PDA, patent ductus arteriosus; IUGR, intrauterine growth retardation; NICU, neonatal intensive care unit.

children had tetralogy of Fallot or double-outlet right ventricle of the tetralogy type. The other 2 patients had a patent ductus arteriosus or coarctation of the aorta with a hypoplastic aortic arch.

Thirteen of the 19 patients (68%) were admitted from home within 1 week of their operation. Nine of these 13 patients were admitted for an electively scheduled operation. Four patients were admitted to the hospital within 1 week before operation for heart failure in 3 patients or tetralogy spells in 1 patient. Five more patients had never been out of the hospital since birth before surgical repair. The final patient had been home briefly after birth but was readmitted for respiratory failure that was managed in the hospital for 5 weeks before operation.

In general, we excluded patients from consideration for operation who had never been weaned from mechanical ventilatory support and preferred that the patient had been under home care for at least a brief period during their lifetime. Ten patients required no respiratory support or supplemental oxygen at the time of operation. Seven patients had preoperative supplemental nasal cannula oxygen or high flow nasal cannula. Two of the patients had a prior tracheostomy and were mechanically ventilated, 1 of whom was admitted from home and the other having never left the hospital since birth.

Nonoperated Patients

Review of our inpatient echocardiographic database revealed 60 additional children with trisomy 13 or trisomy 18 diagnosed during the same time period. Of these patients, 45 (11 trisomy 13 and 34 trisomy 18, Table 2) had significant cardiac lesions that would have required repair but were not offered surgery for a variety of reasons, and 15 had echocardiography studies that demonstrated structurally normal heart or hemodynamically insignificant cardiac lesions (ie, trivial patent ductus arteriosus or patent foramen ovale). The most common diagnoses were VSD in 29 patients or tetralogy of Fallot or variants of tetralogy of Fallot in 9 patients. Interestingly, only 1 patient had single ventricle anatomy.

Statistics

Percentages of total were reported as indicated. Significant differences were calculated using chi-square or Kruskal–Wallis testing as indicated in individual tables. Survival estimates were calculated using the

TABLE 2. Overall cohort statistics

Kaplan–Meier curves with 95% confidence intervals. Significant difference was calculated with a Wilcoxon test, and Cox models were used to provide hazard ratios. Data analysis was performed using SAS/STAT.

RESULTS

Early Surgical Outcomes

Operative details are delineated in Table 3. Of the 19 patients, the initial operation was VSD closure in 9, including 1 patient who also required aortic valvuloplasty, and repair of tetralogy of Fallot or double-outlet right ventricle with tetralogy physiology in 7 patients. One patient was palliated with a pulmonary artery band for a large inlet VSD as a permanent management strategy. One patient had ligation of a large ductus arteriosus that was not amenable to device closure, and the final patient had repair of an aortic coarctation with hypoplastic aortic arch on day of life 4 before a genetic diagnosis. There was no operative mortality defined as death before hospital discharge or within 30 days of operation, whichever came later. One 7-week 1.9-kg baby with trisomy 18 undergoing VSD repair had delayed sternal closure.

There were 3 early reoperations. One patient with trisomy 18 and preoperatively unrecognized severe pulmonary artery hypertension required extracorporeal membrane oxygenation support instituted 18 hours postoperatively for 6 days after repair of tetralogy-like doubleoutlet right ventricle and multiple VSDs. Another patient with trisomy 18 who had repair of tetralogy of Fallot was returned to the operating room on the first postoperative day for relief of residual right ventricular outflow tract obstruction. A final patient with trisomy 13 who had ligation of a large patent ductus arteriosus required removal

		Underwent surgery	Did not undergo	
	Total (N = 64)	for CHD $(N = 19)$	surgery (N = 45)	P value*
Chromosomal abnormality, n (%)				.0158†
Trisomy 13	15 (23.4%)	5 (26.3%)	10 (22.2%)	
Trisomy 13 (mosaic)	4 (6.3%)	3 (15.8%)	1 (2.2%)	
Trisomy 18	43 (67.2%)	9 (47.4%)	34 (75.6%)	
Trisomy 18 (mosaic)	2 (3.1%)	2 (10.5%)	0 (0.0%)	
Sex, n (%)				.8755†
F	38 (59.4%)	11 (57.9%)	27 (60.0%)	
М	26 (40.6%)	8 (42.1%)	18 (40.0%)	
Age at surgery or age referral, d				<.0001
Median (IQR)	28.0 (1.0-108.0)	154.0 (77.0-465.0)	5.0 (1.0-43.0)	
Weight at operation or referral, kg				
Median (IQR)	-	4.4 (3.6-7.8)	-	-
Days of follow-up				.0002‡
Median (IQR)	137.0 (56.5-506.5)	529.0 (181.0-2067.0)	91.0 (21.0-185.0)	
Mortality, n (%)				<.0001†
No	22 (34.4%)	14 (73.7%)	8 (17.8%)	
Yes	42 (65.6%)	5 (26.3%)	37 (82.2%)	

CHD, Congenital heart disease; *IQR*, interquartile range. **P* value comparing the group that underwent surgery versus those with CHD who did not. †Chi-square *P* value. ‡Kruskal–Wallis *P* value.

TABLE 3. Surgical patient outcomes

	Total (N = 19)
Age at surgery, d	
Median (IQR)	154.0 (77.0-465.0)
Weight, kg	
Median (IQR)	4.4 (3.6-7.8)
Procedure name, n (%)	
VSD repair	8 (42.1%)
TOF repair	7 (36.8%)
PDA closure, surgical	1 (5.3%)
Sinus venosus ASD, VSD,	1 (5.3%)
valvuloplasty, aortic	
COA repair	1 (5.3%)
PA banding	1 (5.3%)
ICU h	
Median (IQR)	189.0 (70.0-548.0)
LOS (d) from surgery to discharge	
Median (IQR)	14.0 (8.0-37.0)
Total LOS (d)	
Median (IQR)	19.0 (8.0-45.0)
STAT Mortality Category, n (%)	
1	10 (52.6%)
2	6 (31.6%)
4	3 (15.8%)
STS major complications, n (%)	
No	15 (78.9%)
Delayed sternal closure POD #2	1 (5.3%)
Yes postoperative ECMO ~18 h	1 (5.3%)
postoperative x6 d	
Yes RVOT revision POD 13	1 (5.3%)
Yes removal laryngeal granulation tissue	1 (5.3%)

IQR, Interquartile range; *VSD*, ventricular septal defect; *TOF*, tetralogy of Fallot; *PDA*, patent ductus arteriosus; *ASD*, atrial septal defect; *COA*, coarctation; *PA*, pulmonary artery; *ICU*, intensive care unit; *LOS*, length of stay; *STAT*, *The Society of Thoracic Surgeons-European Association for Cardio-Thoracic Surgery*; *STS*, Society of Thoracic Surgeons; *POD*, postoperative day; *ECMO*, extracorporeal membrane oxygenation; *RVOT*, right ventricular outflow tract.

of laryngeal granulation tissue by the otorhinolaryngology service due to failure to remain extubated postoperatively despite requiring only nasal cannula supplemental oxygen preoperatively.

Mean postoperative intensive care unit stay was 11.4 days (median, 7.9 days, IQR, 2.9-22.8 days). Median postoperative total hospital stay was 14 days (IQR, 8.0-37.0 days). This compares with a center-specific median length of stay at our institution from the Society of Thoracic Surgeons Congenital Heart Surgery database of 4.0 days after VSD closure and 5.0 days after repair of tetralogy of Fallot.

Nonoperative Patient Outcomes

Forty-five patients presented to our center during this timeframe diagnosed with trisomy 13 or trisomy 18 and cardiac defects on echocardiography but were not offered

 TABLE 4. Main diagnosis (nonsurgery group)

Diagnosis	Frequency (%)
VSD	23 (51.1)
PA/VSD	4 (8.9)
VSD/PS	4 (8.9)
ToF	3 (6.7)
ASD	2 (4.4)
AP window	1 (2.2)
DORV	1 (2.2)
DORV/arch hypoplasia	1 (2.2)
PDA	1 (2.2)
Single ventricle	1 (2.2)
ToF/CAVC	1 (2.2)
ToF/PA	1 (2.2)
VSD/MS	1 (2.2)
VSD/TR	1 (2.2)
Total	45 (100)

VSD, Ventricular septal defect; *PA*, pulmonary atresia; *PS*, pulmonary stenosis; *TOF*, tetralogy of Fallot; *ASD*, atrial septal defect; *AP*, aortopulmonary; *DORV*, double-outlet right ventricle; *PDA*, patent ductus arteriosus; *CAVC*, complete atrioventricular canal; *MS*, mitral stenosis; *TR*, tricuspid regurgitation.

surgical intervention. Median age at the time of initial referral for surgical evaluation was 5.0 days (IQR, 1.0-43.0 days, Table 2). Similar to the operative cohort, most patients had a VSD or tetralogy of Fallot variant as the predominant cardiac lesion (Table 4). Although likely multifactorial in most cases, the primary reasons for not offering surgical intervention were the presence of multiple other severe medical comorbidities and significant cardiac or respiratory failure (Table 5). Four patients (8.9%) had complex cardiac lesions that would have required multistage palliation or repair. Six patients died before meeting indications for surgical repair or before surgical repair could be undertaken.

Midterm Follow-up and Survival

Median follow-up for the entire cohort was 137 days (IQR, 56.5-506.5 days): 529 days in the surgical group and 91 days in the nonoperative group. Two patients have had late reoperation without mortality: 1 child with mosaic trisomy 13 who had undergone repair of his aortic coarctation and hypoplastic aortic arch at 4 days of age required closure of a large atrial septal defect due to ongoing heart failure at 3 months of age, and 1 patient with mosaic trisomy 18 had pulmonary valve replacement for severe right ventricular enlargement due to pulmonary valve insufficiency at 13 years of age after repair of tetralogy of Fallot at 3 months of age using a transannular right ventricular outflow tract patch.

There have been 5 late deaths (4 patients with trisomy 18 and 1 patient with trisomy 13) from 4 months to 9.7 years

TABLE 5. Reasons for not undergoing surgery

Reason for not undergoing surgery	Frequency (%)
Cardiorespiratory failure	10 (22.2)
CHF, central apnea	8 (17.8)
CNS lesions	5 (11.1)
Complex CHD	4 (8.9)
Did not meet surgical indications	4 (8.9)
Family wishes	3 (6.7)
Hospital death before cardiac surgery	2 (4.4)
Large omphalocele	2 (4.4)
Medical NEC	2 (4.4)
Multiple anomalies	1 (2.2)
Prematurity	1 (2.2)
Pulmonary HTN	1 (2.2)
Respiratory failure	1 (2.2)
Respiratory failure, complex CHD	1 (2.2)
Total	45 (100)

CHF, Congestive heart failure; CNS, central nervous system; CHD, congenital heart disease; NEC, necrotizing enterocolitis; HTN, hypertension.

postoperatively, mostly due to respiratory failure. Three of the 5 deaths were within 1 year of the initial operation. Both patients with mosaic trisomy 18 and all 3 patients with mosaic trisomy 13 survived on follow-up, although 1 of the patients with mosaic trisomy 13 was lost to followup after moving out of state. Neither of the patients who had late reoperation died.

Kaplan–Meier survival estimates for both groups of patients are shown in Figure 1, with survival estimates and hazard ratios shown in Table 6. In the surgical cohort, 1-year and 5-year survival estimates were 79.9% and 66.6%, respectively. In the nonoperative group, the most mortality occurred in the first 6 months after referral (survival 36.6%) with 5-year survival estimate of 24%.

DISCUSSION

The natural history of children with trisomy 13 or trisomy 18 is bleak with a 1-year mortality described as high as 91% to 100%^{6,14} with many of the deaths in the first week of life. Data from more recent population-based registries still report 1-year survival less than 20% in both syndromes.³⁻⁵ Cardiac disease is common in these patients, occurring in 80% to 91% of children with trisomy 18 and 57% to 86% of children with trisomy 13.^{1,2,9} Because of the poor prognosis without intervention, the advisability of offering cardiac surgery to these patients has been controversial.^{1,7-12} However, an early article by Graham and colleagues¹³ representing the Pediatric Cardiac Care Consortium reported a 91% hospital survival in 35 infants and children with trisomy 13 or trisomy 18 who received

cardiac surgery. Since that report, several individual institution articles describe survival of 71% to 100%.¹⁵⁻¹⁸ Likewise, several registry reports or multi-institutional reports show improved survival in patients with trisomy 13 or trisomy 18 who undergo cardiac surgery.^{2,19-25}

In this article, we report our experience with 21 cardiac operations in 19 children with trisomy 13 or trisomy 18 since 2002. It is difficult to ascertain the true denominator of patients with trisomy 13 or trisomy 18 during this study, although review of our inpatient echocardiographic database during a comparable time period identified 45 children with trisomy 13 or trisomy 18 and cardiac lesions who did not undergo surgical repair. Certainly, this under-represents the number of patients with trisomy 13 or trisomy 18 who did not have a cardiac operation because it would not capture patients seen at outside hospitals and nurseries who were not referred to our local institution nor would it identify those children without significant cardiac disease or just pulmonary artery hypertension who were managed in the outpatient setting (Figure 2).

In the earlier era of this study, we did not have formalized criteria for selecting patients as candidates for cardiac operation. In our series, all candidates for operation had cardiac disease as the dominant clinical problem with no unrepaired other major problems. We preferred that these patients had demonstrated a "will to live" and a "will to breathe" as demonstrated by spending some time at home and not requiring mechanical ventilation as has been previously recommended.^{19,26} Despite these preferences, 5 of the 19 patients (Table 1) had never been out of the hospital since birth and 2 patients had a tracheostomy with mechanical ventilation (1 of these patients came in from home for elective repair). Over time, we relaxed our criteria for surgical intervention; only 2 of the 19 initial operations were performed before 2012. Our current institutional philosophy and guidelines are that we will offer surgical repair of congenital cardiac lesions to patients with trisomy 13 or 18 if they have been home at some point. We do not offer surgery for lesions that may require multiple or lifelong surgical interventions (eg, pulmonary atresia with VSD and major aortopulmonary collaterals), and we do not offer single-ventricle palliation.

For the neonate who presents with unrelenting congestive heart failure due to a large VSD, for example, we have prolonged discussions with family members and a multidisciplinary group within our heart center. If the likelihood of complete surgical repair is good and the family is committed to the possibility of needing tracheostomy and home mechanical ventilation in the postoperative recovery period, we are generally agreeable to offering surgery. We have found that this commitment and education up-front facilitates discussions postoperatively for patients who ultimately cannot safely separate from mechanical ventilation.



FIGURE 1. Kaplan–Meier survival estimates with 95% confidence intervals (*shaded*) for patients with trisomy 13 or 18 and cardiac disease who underwent surgical repair (*blue line*) or were unrepaired (*red line*). *HR*, Hazard ratio; *CHD*, congenital heart disease.

There was no operative or hospital mortality in this series. Nonetheless, these patients required considerable hospital resource use postoperatively as evidenced by increased length of stay in the intensive care unit and in the hospital as has been reported by others.^{18,20,22} Unplanned early reoperation was needed in 3 of the 19 patients. Two patients had elective reoperation 3 months and 13 years after the initial operation without incident.

On average follow-up of 3.4 years, there have been 5 late deaths with 3 within the first year after operation. Although

the scope of this study does not permit direct comparison of survival with children who had trisomy 13 or trisomy 18 and cardiac disease who did not undergo operation, examination of the natural history of these patients without an operation would suggest that their survival is considerably enhanced.^{21,26} Actuarial survival analysis of our patients demonstrates improved midterm survival with an estimated median survival between 9 and 10 years (Figure 1). Interestingly, all patients in our series who had mosaic trisomy 18 survived long-term even though one

TABLE 6.	Midterm s	survival for	operative and	nonoperative	patients v	with trisomy	13 and 18
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		Hazard ratio (95% CI)	Survival estimates (95% CI)	
	Event/total	Cox model	Kaplan–Meier method	P value
Survival from surgery/initial C	HD referral			
Group				<.0001*
No surgery	33/45	5.68 (2.00-16.09)	6 mo: 36.6 (24.8%-54.1%)	
			1 y: 34.2 (22.6%-51.6%)	
			2 y: 24.0 (13.9%-41.2%)	
			3 y: 24.0 (13.9%-41.2%)	
			4 y: 24.0 (13.9%-41.2%)	
			5 y: 24.0 (13.9%-41.2%)	
Underwent surgery	4/19	Reference	6 mo: 94.4 (84.4%-100.0%)	
			1 y: 79.9 (61.8%-100.0%)	
			2 y: 79.9 (61.8%-100.0%)	
			3 y: 79.9 (61.8%-100.0%)	
			4 y: 66.6 (42.9%-100.0%)	
			5 y: 66.6 (42.9%-100.0%)	

CI, Confidence interval; CHD, congenital heart disease. *Wilcoxon test.



FIGURE 2. Midterm results of patients with trisomy 13 or 18 undergoing cardiac surgery at a single institution as compared with similar, unoperated patients. *VSD*, Ventricular septal defect; *TOF*, Tetralogy of Fallot; *ECHO*, echocardiography.

was lost to follow-up at 5 months postoperatively. Although our numbers are small, perhaps this suggests that those patients with mosaicism can be expected to have better longterm outcomes.

Study Limitations

Quality of life and family satisfaction were not measured in this study. Anecdotally, however, our experience is that for these highly selected patients, the parents are grateful and happy with their decision. Enhanced parental satisfaction with cardiac surgical intervention has been reported by the group from Pennsylvania State University.¹⁶

CONCLUSIONS

This series shows that carefully selected patients with trisomy 13 or trisomy 18 can undergo cardiac operation with excellent early survival, acceptable early morbidity despite prolonged length of stay postoperatively, and reasonable midterm survival. An intriguing recent analysis from the Pediatric Cardiac Care Consortium indicated that patients with trisomy 13 or trisomy 18 who survive hospitalization after cardiac surgery have a greatly prolonged survival (median survival of 14.8 years for trisomy 13 and 16.2 years for trisomy 18).²⁵ In view of improved early surgical results demonstrated in this study and others previously cited, it is important to study the long-term fate of these operated children with respect to quality of life and family satisfaction and to directly compare those outcomes with children with trisomy 13 or trisomy 18 and cardiac lesions that are left unrepaired. Perhaps with proper planning, family education, and appropriate patient selection, we should be operating on more patients with trisomy 13 or trisomy 18 who have cardiac disease.

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Conflict of Interest Statement

The authors reported no conflicts of interest.

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Key Words: congenital cardiac surgery, trisomy 13, trisomy 18