Impact of trisomy 13 and 18 on airway anomalies and pulmonary complications after cardiac surgery

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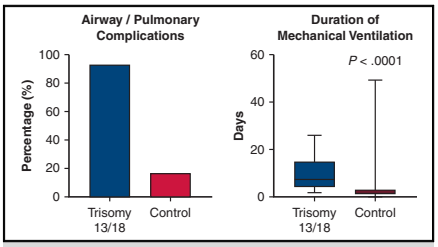
ABSTRACT

Objective: To determine the prevalence and influence of clinically significant airway and/or respiratory abnormalities in patients with trisomy 13 and 18 undergoing cardiac surgery.

Methods: We performed a retrospective, case-control cohort study of all patients with known trisomy 13 or 18 who underwent cardiac operations at our institution from 1994 to 2014. Cases were matched 3:1 by age, surgical date, and cardiac lesion with nontrisomy 13/18 patients. Baseline clinical characteristics and patient outcomes, including postoperative course and management were compared. Descriptive statistics and Wilcoxon rank-sum test or Fisher exact test as appropriate were used to determine significant differences.

Results: In the 14 trisomy 13/18 patients who underwent cardiac surgery, there was an increased incidence of postoperative complications. Specifically, 93% had airway or pulmonary complications, including prolonged mechanical ventilation (n = 8), prolonged noninvasive positive pressure ventilation (n = 6), reintubation (n = 7), tracheitis/pneumonia (n = 6), and tracheostomy (n = 2). The duration of intubation was longer (7.5 vs 2 days; $P < .0001$) as was the duration of noninvasive positive pressure ventilation (8 vs 2 days; $P < .04$) with longer hospital length of stay in the trisomy 13/18 cohort. There was 1 in-hospital mortality, with none in the control group.

Conclusions: Although most trisomy 13/18 patients survive cardiac surgery, these patients have an increased incidence of airway complications, requiring longer intensive respiratory support postoperatively that contributes to longer length of stay. Parental guidance before cardiac surgery should include a discussion about postoperative airway management. (J Thorac Cardiovasc Surg 2020;■:1-9)



Trisomy 13/18 patients have complicated postoperative courses with increased respiratory need.

CENTRAL MESSAGE

Whereas most patients with trisomy 13 and 18 survive cardiac surgery, higher prevalence of airway and respiratory issues leads to longer invasive and noninvasive ventilation and longer length of stay.

PERSPECTIVE

Our study suggests that despite low in-hospital mortality in trisomy 13 and 18 patients who undergo cardiac surgery, the postoperative course is significantly longer and is associated with more comorbidities compared with control patients. These are important considerations when counseling families about cardiac surgery and providing parental guidance regarding their postoperative course.

See Commentary on page XXXX.

CONG

Trisomy 13 and 18 (T13/18) are chromosomal abnormalities characterized by numerous congenital anomalies with a high percentage of these patients having congenital heart disease.[1,2] The most common heart defects in this population are ventricular septal defects (VSD), atrial septal defects, and a patent ductus arteriosus.[3] Population-based T13/18 studies have shown a median survival <2 weeks, with a small percentage (<5%-10%) surviving for 1 or more years.[4-8] Given the generally poor survival, in the past it was presumed that patients with congenital heart disease did not survive long enough for a heart defect to

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Scanning this QR code will take you to the article title page to access supplementary information.

Abbreviations and Acronym

CPAP = continuous positive airway pressure
HFNC = high-flow nasal cannula
ICU = intensive care unit
LOS = length of stay
T13/18 = trisomy 13 and 18
VSD = ventricular septal defect

be a significant cause of morbidity and therefore care focused mainly on nonintervention and palliative care approaches.[4,5] More recent reports have described increased long-term survival and subsequently care has shifted toward life-prolonging treatments, including surgical palliation or repair of cardiac defects.[9-15] Since the first reports of successful cardiac surgery in patients with T13/18 in 2004,[11] several groups have reported acceptable survival after cardiac surgery.[11-16] Although these reports establish that cardiac surgery is safe and feasible in T13/18 patients, data are lacking describing the patients' hospital course, including the current intensive care management strategies and complications following cardiac surgery. Among the major comorbidities in this group of patients, which can drastically influence their postoperative course, is the presence of airway and lung abnormalities. At the present time, although it has anecdotally been noted to be of significant concern in our own cardiac intensive care unit, there are no studies describing the prevalence or management of respiratory and airway anomalies and their influence on patient outcomes; particularly, intensity and duration of respiratory support. Therefore, we aimed to describe the prevalence of airway anomalies in patients with T13/18 who have undergone cardiac surgery and evaluate how these influence postoperative outcomes.

MATERIALS AND METHODS

Study Design

We performed a retrospective case-control study of all patients with T13/18 who underwent cardiac operations at our institution between 1994 and 2014. Institutional review board approval was obtained with waiver of written informed consent. All patients with a diagnosis of T13/18 undergoing cardiac surgery at our institution were included. Patients were identified using electronic medical record review and local surgical database search to ensure complete case capture. A 3:1 diagnosis-matched control group was identified by searching our local surgical database. These individuals were further matched by age (±6 months) at surgery and then date of surgery (±1 year). In 5 patients, the range of date of cardiac surgery necessitated extension of the date of surgery to ±3 years resulting in adequate control selection.

Study Measurements

Baseline clinical characteristics and postoperative clinical course data up to time of discharge or in-hospital death were obtained by electronic medical record review, which includes medical records from outside our institution. These data included demographic characteristics, clinical data before cardiac surgery (birth history, genetic testing, and known airway anomalies), cardiac operative data (age, weight and height at time of surgery, type of surgical repair, duration of cardiopulmonary bypass, aortic crossclamp time, and circulatory arrest), and intraoperative complications. Additionally, details from the postoperative course included intensive care unit (ICU) and hospital length of stay (LOS), maximal vasoactive-inotropic support score in the first 24 hours postoperatively,[17,18] airway management course and details, postoperative complications, and in-hospital death. Any airway evaluation (otolaryngology or pulmonology) consult notes were reviewed. Postoperative complications were reviewed and categorized by system. Airway/pulmonary complications were classified as prolonged need for mechanical ventilation (≥7 days), prolonged noninvasive positive pressure ventilator support with either continuous positive airway pressure (CPAP) or high flow nasal cannula (HFNC) (≥7 days), repeated episodes of apnea requiring escalation of respiratory support, reintubation within 72 hours of extubation, tracheostomy, and/or tracheitis (positive respiratory culture) or pneumonia (positive respiratory culture with radiographic evidence of pneumonia or worsening respiratory status).

Statistical Analysis

Descriptive statistics for patient characteristics and outcomes were summarized as median with interquartile ranges (IQRs) for continuous variables and count with percentage for categorical variables when appropriate. Intergroup comparisons were made using Wilcoxon rank-sum test, as appropriate, for continuous variables, and Fisher exact test, as appropriate for each categorical outcome using GraphPad Prism version 8.4.2 for Windows (GraphPad Software Inc, San Diego, Calif).

RESULTS

Patient Characteristics

Fourteen patients with T13/18 (4 men and 10 women) underwent cardiac surgery at a median age of 93 days during the study period. Baseline characteristics of the cases and controls are summarized in Table 1. Most patients had trisomy 18 (86%). There was 1 mosaic trisomy 13 and 1 mosaic trisomy 18 patient. The patient population, matching selection and cardiac disease is graphically depicted along key postoperative findings in Figure 1. Cardiac diseases included VSD (n = 9 [64%]), VSD with coarctation of the aorta (n = 2 [14%]), tetralogy of Fallot (n = 2 [14%]), and atrioventricular septal defect (n = 1 [7%]). Forty-two control patients were matched to this cohort. Nine (64%) T13/18 patients compared with 13 (31%) control patients were admitted before surgery, which included all patients with VSD and coarctation of the aorta requiring neonatal repair. If the patient was admitted before cardiac surgery, the length of hospitalization before surgery was similar for both groups (median, 5 days).

Preoperative Airway or Respiratory Status

Preoperatively, 3 T13/18 patients had an airway evaluation by specialists (pulmonary and/or otolaryngology). These patients were diagnosed with micrognathia/retrognathia and mild laryngomalacia, micrognathia with tracheomalacia resulting in upper airway obstruction and severe sleep apnea, and chronic respiratory insufficiency. The patient with micrognathia,

TABLE 1. Baseline characteristics

Characteristic	T13/18 (n = 14)	Control (n = 42)	*P* value
Male	4 (29)	28 (67)	.03
Premature infant	2 (14)	3 (7)	.59
Admitted before surgery	9 (64)	13 (31)	.55
If admitted, LOS before surgery (d)	5 (2-9)	5 (4-7)	.88
Respiratory support at time of cardiac repair	10 (71)	6 (14)	.0001
Intubated/mechanical ventilation	4 (28)	3 (7)	.06
Other support, including CPAP, HFNC, or NC	6 (43)	3 (7)	.005
Preoperative airway evaluation	3 (21)	0 (0)	.01

Values are presented as total n (%) or median (interquartile range). *T13/18*, Trisomy 13 and 18; *LOS*, length of stay; *CPAP*, continuous positive airway pressure; *HFNC*, high-flow nasal cannula; *NC*, nasal cannula.

tracheomalacia, severe sleep apnea with upper airway obstruction underwent testing, including bronchoscopy and a sleep study before cardiac surgery. One additional patient was noted to have apnea before surgery but did not have a preoperative airway evaluation (pulmonology was consulted postoperatively for continued apnea spells). In the control group, there were no preoperative airway evaluations.

Ten of the 14 T13/18 patients (71%) required some type of respiratory support before cardiac surgery. Four were intubated and mechanically ventilated, 1 required HFNC, and 5 required a nasal cannula. This was a significantly larger percentage compared with the control group ($P = .0001$) who by comparison had only 14% of patients who required respiratory support immediately before cardiac surgery (3 patients [7%] were intubated/mechanical ventilation, 1 patient [2%] required HFNC, and 2 patients [5%] required nasal cannula). No patients in either group had a tracheostomy before surgery.

Cardiac Surgery

Cardiopulmonary bypass time, crossclamp time, and need for circulatory arrest were not significantly different between the 2 groups (Table 2). In the T13/18 cohort, 50% of patients had a "difficult" or "critical airway designation" assigned by anesthesia based on visual assessment of the airway during intubation in the operating room, compared with 5% in the control group.

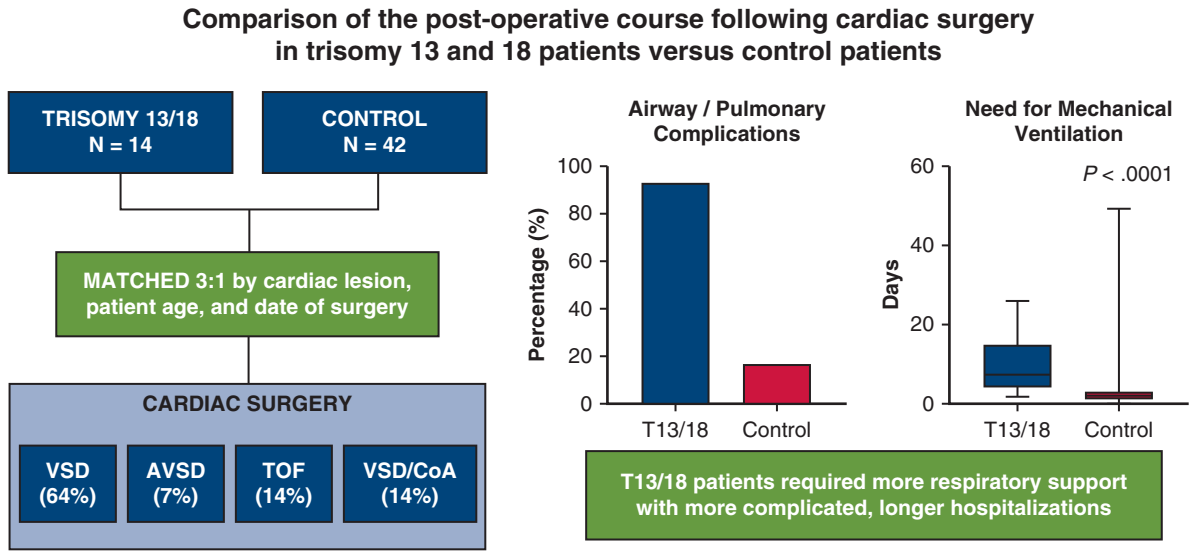


FIGURE 1. Trisomy 13 and 18 (T13/18) patients and control patients were matched 3:1 by cardiac lesion, patient age, and date of surgery. There were 14 T13/18 patients who underwent cardiac surgery during our study time period. Patients underwent cardiac surgery for ventricular septal defect (*VSD*) (64%), atrioventricular septal defects (*AVSD*) (7%), tetralogy of Fallot (*TOF*) (14%), and VSD with coarctation of the aorta (*CoA*) (14%). T13/18 patients had more complicated postoperative courses requiring more respiratory support with longer hospitalizations. Ninety-three percent of T13/18 patients (compared with 17% in the control group) had airway/pulmonary complications, including prolonged mechanical ventilation, prolonged noninvasive positive pressure ventilation, failed extubation attempts, apnea requiring escalation of support, new diagnosis of tracheitis/pneumonia, and need for tracheostomy. Additionally, the T13/18 cohort required longer duration of intubation with mechanical ventilation compared with the control group ($P < .0001$).

TABLE 2. Operative data

Variable	T13/18 (n = 14)	Control (n = 42)	*P* value
Age at time of cardiac repair (d)	92 (40-144)	88 (48-144)	.94
Weight at time of cardiac repair (kg)	3.7 (2.7-4.2)	4.6 (3.2-5.6)	.02
Surgical cardiac repair			
Cardiopulmonary bypass time (min)	69 (53-107)	65 (49-81)	.45
Crossclamp time (min)	36 (26-51)	40 (26-52)	.93
Required circulatory arrest	4/14 (29)	8/42 (19)	.47
Arrest time if needed (min)	38 (33-56)	48 (26-53)	>.99

Values are presented as total n (%) or median (interquartile range). *T13/18*, Trisomy 13 and 18.

Postoperative Course

The ICU LOS following cardiac surgery was longer in the T13/18 cohort compared with the control group (14 days; IQR, 6-27 days vs 4 days; IQR, 2-5 days; $P < .0001$), as was the hospital LOS (31 days; IQR, 19-42 days vs 8 days; IQR, 6-15 days; $P < .0001$) (Figure 2). All T13/18 patients (100%) required vasoactive support in the first 24 hours following cardiac surgery compared with 74% in the control group. The median vasoactive-inotropic support score was 13 (IQR, 4.5-20.5) in the T13/18 group, compared with 5 (IQR, 0-10) in the control group ($P = .01$). All (100%) of the T13/18 patients arrived intubated to the ICU following cardiac surgery compared with 93% in the control group.

Postoperative Complications

Postoperative complications are summarized in Table 3. In the T13/18 cohort, 13 of 14 patients experienced at least 1 postoperative complication with all 13 having an airway/pulmonary complication as described in more detail below and in Table 4. Additionally, 8 T13/18 patients (58%) had nonpulmonary complications. In comparison, 12 control patients (29%) had some type of postoperative complication. There was 1 in hospital death in the T13/18 group in a trisomy 13 patient following tetralogy of Fallot repair whose postoperative course was complicated with necrotizing enterocolitis with intestinal pneumatosis, sepsis, and aspiration pneumonia. At that time, the family redirected goals of care (do not intubate/do not resuscitate) and the patient developed worsening respiratory failure and died postoperative day 27 in the hospital. There were no deaths in the control cohort.

Airway Course and Complications

T13/18 patients had a significantly higher percentage of airway/pulmonary complications postoperatively (Tables 4 and 5, Figure 1). Thirteen of 14 (93%) T13/18 patients had at least 1 of the following airway/pulmonary complications postoperatively: prolonged mechanical ventilation ≥7 days (n = 7), prolonged CPAP and/or HFNC ≥7 days (n = 2), reintubation within 72 hours (n = 5), need for tracheostomy (n = 2), new diagnosis of tracheitis/pneumonia (n = 6), or apnea requiring escalation of support (n = 1). In comparison, in the control cohort, 7 of 42 patients (17%) had postoperative airway complications specifically prolonged mechanical ventilation (n = 4) and reintubation within 72 hours (n = 3).

In the T13/18 cohort, the median duration of intubation with mechanical ventilation was significantly longer compared with the control group (7.5 vs 2 days; $P < .0001$) (Figure 3, *A*). Fifty percent (50%) of T13/18 patients required prolonged mechanical ventilation ≥7 days compared with 7% of the control group ($P = .0012$). A significantly higher percentage ($P = .01$)

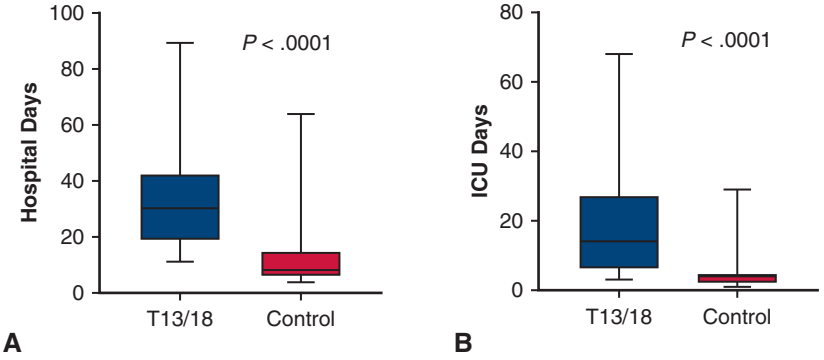


FIGURE 2. Box plots (median, interquartile range, range) demonstrate a significantly longer postoperative (A) total hospital length of stay ($P < .0001$) and (B) intensive care unit (*ICU*) length of stay ($P < .0001$) in patients with trisomy 13/18 compared with the control group.

TABLE 3. Postoperative complications

Complication	T13/18 (n = 14)	Control (n = 42)	*P* value
Arrhythmia	6 (43)	9 (21)	.16
Tracheitis/pneumonia	**6 (43)**	**0 (0)**	**<.0001**
Sepsis	**3 (21)**	**0 (0)**	**.01**
Sternal wound infection	0 (0)	0 (0)	.99
Acute kidney injury	1 (7)	1 (2)	.44
Hemodialysis	0 (0)	0 (0)	.99
Stroke/seizure	0 (0)	0 (0)	.99
Vascular*	1 (7)	1 (2)	.44
Cardiac arrest	3 (21)	3 (7)	.16
ECMO	0 (0)	0 (0)	.99
Delayed sternal closure	1 (7)	2 (5)	.99
Unplanned postoperative operation			
Cardiac	2 (14)	1 (2)	.15
Noncardiac			
Tracheostomy	2 (14)	0 (0)	.06
Gastrostomy tube placement	**3 (21)**	**0 (0)**	**.01**
In-hospital mortality/death	1 (7)	0 (0)	.25

Values are presented as total n (%) or median (interquartile range). Items in bold represent statistically significant associations where *P* value is < .05. *T13/18*, Trisomy 13 and 18; *ECMO*, extracorporeal membrane oxygenation. *Vessel injury or occlusion.

of the T13/18 cohort had difficulty with extubation with 7 of 14 (50%) requiring re-intubation during the hospitalization (5 requiring re-intubation within 72 hours). Three of 7 (29%) failed twice. In comparison, only 12% of the control group failed extubation. Reasons for failure included carbon dioxide retention and hypoventilation, poor effort, apnea, or respiratory distress leading to bradycardia and/or cardiac arrest. Two T13/18 patients required tracheostomy (14%). These patients had clinical suspicion for airway disease with failed extubation attempts. Both underwent an airway evaluation, including bronchoscopy that demonstrated significant disease (bronchomalacia or tracheomalacia). The time between cardiac surgery and tracheostomy placement was 18 and 29 days for the 2 patients. No control patients required tracheostomy. At time of extubation, T13/18 patients were transitioned to nasal cannula only 43% of the time compared with 85% of the control group. A significantly higher percentage of the T13/18 cohort required CPAP and/or HFNC (71% vs 24%; $P < .003$) at some point during the postoperative period, and when utilized, the duration of CPAP and/or HFNC was significantly longer in the T13/18 cohort compared with the control group (Figure 3, *B*) (8 vs 2 days; $P < .04$). Additionally, 2 T13/18 patients who did not require prolonged mechanical ventilation required prolonged CPAP and/or HFNC support lasting >7 days (compared with no control patients) in the postoperative period.

During the hospitalization, 7 T13/18 patients (50%) underwent airway evaluation by specialists, resulting in the diagnosis of a significant airway abnormality or respiratory disease in all 7 patients. Comparatively, only 1 control patient (2%) who had a prolonged course of mechanical ventilation postoperatively underwent a similar evaluation and diagnosis ($P = .0001$). The diagnoses resulting from the above evaluations included tracheomalacia, bronchomalacia, micrognathia/retrognathia, apnea, and chronic lung disease. Bronchoscopy, which was performed in 4 T13/18 patients and 1 control patient, were utilized to diagnose broncho- or laryngomalacia.

TABLE 4. Morbidity and mortality in trisomy 13 and 18 patients

Variable	1	2	3	4	5	6	7	8	9	10	11	12	13	14	Total (N = 14)
Preoperative															
MV at time of surgery		✓		✓	✓		✓								4 (29)
Airway evaluation										✓		✓		✓	3 (21)
Postoperative airway complications															
Prolonged MV		✓			✓		✓	✓	✓		✓	✓		✓	8 (57)
Prolonged CPAP/HFNC							✓		✓	✓	✓	✓		✓	6 (43)
Re-intubation		✓					✓	✓			✓	✓	✓	✓	7 (50)
Tracheitis/pneumonia				✓		✓		✓			✓	✓		✓	6 (43)
Postoperative airway evaluation	✓						✓	✓			✓	✓	✓	✓	7 (50)
Tracheostomy								✓			✓				2 (14)
Other postoperative complications															
Arrhythmia				✓		✓	✓		✓		✓		✓		6 (43)
Cardiac arrest						✓			✓			✓			3 (21)
Sepsis				✓		✓						✓			3 (21)
Cardiac reoperation												✓		✓	2 (14)
Death				✓											1 (7)

Values are listed as total n (%). Individual trisomy 13 and 18 patients are listed from 1 to 14. Checkmarks indicate the presence of the complication/evaluation for that individual. *MV*, Mechanical ventilation; *CPAP*, continuous positive airway pressure; *HFNC*, high-flow nasal cannula.

CONG

TABLE 5. Postoperative airway management

Variable	T13/18 (n = 14)	Control (n = 42)	*P* value
Critical airway designation	7 (50)	2 (5)	.0004
Duration of mechanical ventilation (d)	7.5 (4-15)	2 (1-3)	<.0001
Prolonged mechanical ventilation	7 (50)	3 (7)	.0012
Successful extubation on first attempt	7 (50)	37 (88)	.01
Failed/required reintubation ×1	4 (29)	6 (12)	–
Failed/required reintubation ×2	3 (21)	0 (0)	–
Postextubation CPAP/HFNC required (%)	71	24	.003
Time on CPAP/HFNC if required (d)	8 (2-11)	2 (1-3)	.04
Airway evaluation	7 (50)	1 (2)	.0001
Bronchoscopy	4 (28)	1 (2)	.01
Tracheostomy at time of hospital discharge	2 (14)	0 (0)	.06

Values are presented as total n (%) or median (interquartile range) except where otherwise noted. *T13/18*, Trisomy 13 and 18; *CPAP*, continuous positive airway pressure; *HFNC*, high-flow nasal cannula.

At time of discharge, 6 (46%) of 13 patients with T13/18 required no respiratory support. Five (38%) required a nasal cannula. Two T13/18 patients were discharged home with a tracheostomy and mechanical ventilation. In comparison, in the control group, 40 patients (95%) were discharged home on room air without any respiratory support. Two (5%) required a nasal cannula.

The 4 T13/18 patients receiving mechanical ventilation at the time of cardiac surgery all experienced at least 1 airway complication (Table 4). Three required prolonged mechanical ventilation during the postoperative period. Two of these patients were neonates with coarctation of the aorta and had postoperative courses complicated by prolonged mechanical ventilation and failed extubation attempts. Of note, the 1 patient who died during the postoperative period also received mechanical ventilation before surgical repair. However, the 2 patients who required tracheostomy were not preoperatively mechanically ventilated.

Cardiac Complications

In the T13/18 cohort, 6 patients had an abnormal rhythm noted during the immediate postoperative period, including junctional rhythm (5 patients) or sinus bradycardia with inadequate perfusion (1 patient). For junctional rhythms, no patients required long-term medication management. The patient with sinus bradycardia required temporary postoperative pacing and had cardiac arrest secondary to bradycardia after his pacemaker malfunctioned with electrolyte imbalances. In comparison, arrhythmia was seen in 9 control patients (21%) and included sick sinus syndrome with bradycardia or transient atrioventricular block, supraventricular tachycardia, and junctional rhythms. This was not statistically significant between groups ($P = .16$). The frequency of postoperative cardiac arrest (T13/18, 21% and control, 7%) was also not statistically significant between groups ($P = .16$). Two T13/18 patients underwent unplanned repeat surgical cardiac intervention (partial closure of a patent foramen

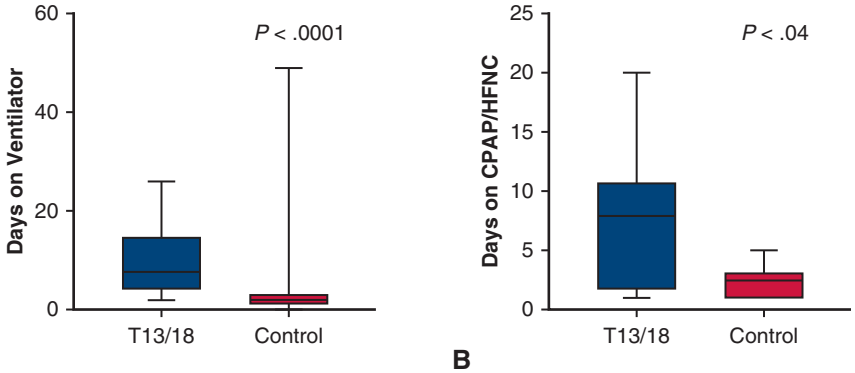


FIGURE 3. Box plots (median, interquartile range [IQR], range) demonstrate days of mechanical ventilation (A) and if required, days of continuous positive airway pressure (*CPAP*) or high flow nasal cannula (*HFNC*) (B) during the postoperative period for trisomy 13/18 (T13/18) and control groups. The T13/18 cohort required longer duration of mechanical ventilation compared with the control group ($P < .0001$). A significantly higher percentage of T13/18 patients (71% vs 24%) required CPAP and/or HFNC ($P = .003$), and if required, T13/18 individuals required it for a significantly longer duration ($P < .04$).

ovale given hypoxia, and ascending aortopexy) compared with 1 unplanned cardiac intervention (repair of an unstable sternum) in the control group ($P = .15$). No patients required extracorporeal membrane oxygenation.

Infectious Complications

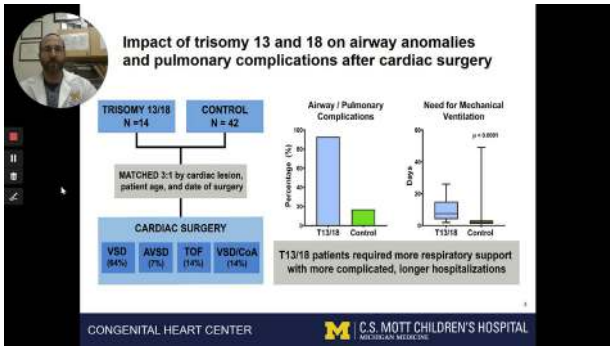
In the T13/18 cohort, there was a significantly higher rate of postoperative infections ($P < .0001$). Six of 14 T13/18 patients had a postoperative course complicated by infection. Most commonly this was tracheitis or pneumonia with or without sepsis (3 of 6 had sepsis with findings of tracheitis/pneumonia), but 1 patient became acutely ill with *Enterobacter bacteremia*, uremia, and tracheitis following cystoscopy and given the close timing with cardiac repair with intracardiac Gore-Tex patch (W. L. Gore and Associates, Newark, Del), was treated for presumed endocarditis. In comparison, in the control cohort, no postoperative infections were present.

Long-Term Outcomes

Of 13 T13/18 patients who left the hospital alive (93% survival rate; 92% if mosaic T13/18 patients are excluded), follow-up data are available on 10 patients because several patients were followed outside our institution. Of these 10 patients, 3 died: 1 patient died shortly after discharge home at age 2 months after the family redirected care, 1 patient died at age 6 months (5 months after cardiac surgery) after out-of-hospital cardiac arrest, and 1 patient died at an unknown age. The other 7 patients who left the hospital alive are still alive at final follow-up within this study time frame (2016) with a median survival of 3.4 years (3.0 years when excluding mosaic patients; range, 2.1-10.5 years). One-year survival was 70% (67% when excluding mosaic patients).

DISCUSSION

This study is the first to evaluate the postoperative course in detail, including a specific focus on airway and respiratory issues, in patients with T13/18 who have undergone cardiac surgery and highlights differences in their postoperative course compared with a control cohort (Video 1). A better understanding of the postoperative course in this subgroup of patients is important for cardiothoracic surgeons, intensivists, and cardiologists caring for these patients before and after cardiac surgery. As other studies have shown, cardiac surgery can be performed in T13/18 patients with good short-term survival during the perioperative period.[19] In our cohort, 93% of T13/18 patients who underwent cardiac surgery survived to hospital discharge, which is similar to what has been reported in the most contemporary study of survival in T13/18 patients.[9] However, our study suggests that despite low in-hospital mortality with cardiac surgery, the postoperative course, especially the ICU length of stay, of patients with T13/18 undergoing cardiac surgery is significantly longer and is associated with more comorbidities compared with control patients. Thus, these are important considerations when counseling families about cardiac surgery in T13/18 patients.



VIDEO 1. Dr. Jeffrey Zampi reviews the key findings and significance of this research study. Video available at: https://www.jtcvs.org/article/S0022-5223(20)32473-9/fulltext.

Although determinants of ICU LOS are multifactorial in all patients, in this cohort much of the LOS appears related to pulmonary complications. Specifically, prolonged mechanical ventilation, a higher incidence of failed extubation attempts, and a higher frequency and duration of noninvasive respiratory support, including CPAP and/or HFNC following cardiac surgery are key drivers of ICU LOS in our patient population. Additionally, 2 patients with T13/18 required tracheostomy (vs none in the control group). Correspondingly, the ICU LOS for non-T13/18 patients was much shorter because these patients had far fewer respiratory complications. In turn, the longer ICU LOS likely directly influenced the longer overall hospital LOS in the T13/18 cohort.

Whereas only a few patients with T13/18 had a preoperative airway evaluation, larger percentages were mechanically ventilated before cardiac surgery. In a recently published study of the Society of Thoracic Surgeons database on a large cohort of T13/18 patients, preoperative mechanical ventilation was associated with an 8-fold greater risk of postoperative mortality.[9] Our 1 in-hospital death in the T13/18 cohort did require preoperative mechanical ventilation. After surgery, many T13/18 patients ultimately underwent airway evaluation because of prolonged mechanical ventilation or unsuccessful extubation attempts yielding a diagnosis of airway anomalies. This is similar to other reports in patients with T13/18 with many patients having airway abnormalities and a smaller percentage ultimately requiring tracheostomy.[20-23] This has important implications for these patients both during the immediate postoperative period and also long-term.

There were some other notable postoperative complications more commonly seen in the T13/18 group.

First, infectious complications (tracheitis and sepsis) were more common. The higher incidence of tracheitis could be explained by the longer duration of mechanical ventilation and higher re-intubation rate in the T13/18 patients. We were not able to differentiate sepsis based on the underlying etiology (ie, underlying tracheitis vs a catheter-associated bloodstream infection) due to the retrospective nature of this study. However, it is not surprising that the incidence of sepsis was higher in the T13/18 patients because this is commonly associated with longer duration of mechanical ventilation and longer ICU LOS. Second, gastrostomy tube placement was more common in the T13/18 patients. This matches a study by Josephson and colleagues[24] that demonstrated procedures, including gastrointestinal procedures and mechanical ventilation in addition to cardiac surgery in T13/18 patients during their first year of life were common. In our center, gastrostomy tube placement is typically performed after patients have been transferred out of the cardiac ICU, so this likely is not a key driver of longer ICU LOS. Third, the 24-hour maximal vasoactive inotropic support score was significantly higher in patients with T13/18 with all patients requiring some inotropic support versus only 26% requiring inotropes in the control group. This difference supports that the trisomy patients were overall sicker after cardiac surgery, which is also evident by the trend toward a higher incidence of arrhythmia and cardiac arrest in the T13/18 patients.

Overall, our data show that there is a significant prevalence of respiratory/airway abnormalities in T13/18 patients and these have the potential to influence the postoperative course after cardiac surgery. There is also a higher incidence of other nonpulmonary complications, like postoperative infections and the need for gastrostomy tube placement, which can affect postsurgical LOS, short- and long-term morbidity, and potentially quality of life. Therefore, parental guidance before cardiac surgery should include a discussion about postoperative management and possible complications with specific emphasis on how airway anomalies and pulmonary complications can influence the postoperative course. Additionally, given the high incidence of airway and pulmonary complications during the postoperative period, we now advocate for a multidisciplinary preoperative evaluation for all T13/18 patients being considered for cardiac surgery. In T13/18 patients who underwent preoperative airway evaluation, if there were specific concerns, additional testing was completed prior or in conjunction with surgery. Given the wide spectrum of comorbidities and cardiac disease in T13/18 patients, a preoperative evaluation is extremely helpful to identify medical problems that may influence postoperative care. In our own practice, a preoperative multidisciplinary assessment of known and potential medical comorbidities, including airway evaluation and palliative care consultation, is now routine. As the care of T13/18 evolves and more patients are considered for cardiac surgical interventions, palliative care team involvement is valuable, especially with complicated medical decision making with at times ethical complexity[25] in patients with complex cardiac disease and/or multiorgan disease. In our experience, we have not found, nor do we recommend, that noncardiac comorbidities preclude performing cardiac operations on T13/18 children. Rather, this information allows the medical care team to provide families with more accurate expectations of clinical course and thus allows families to make more informed decisions about their child's care.

The main limitation of this study is the retrospective design, which does not allow detailed understanding of the multitude of clinical decisions that lead to patient outcomes. Because this was a surgical cohort, we were unable to determine the number of T13/18 patients who were not referred or did not undergo cardiac surgery. Additionally, because this study focused on the postoperative in-hospital course and many patients received their medical care outside our institution, detailed long-term outcomes are limited.

CONCLUSIONS

Although survival is good, ICU and hospital LOS are longer compared with nontrisomy patients undergoing similar operations. Airway anomalies and respiratory complications are likely key drivers of these postoperative outcomes, but there are several other important postoperative complications that are common in patients with T13/18. These findings should be considered during preoperative parental counseling and for postoperative planning and expectations in T13/18 patients.

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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000 Impact of trisomy 13 and 18 on airway anomalies and pulmonary complications after cardiac surgery

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Whereas most patients with trisomy 13 and 18 survive cardiac surgery, higher prevalence of airway and respiratory issues leads to longer invasive and noninvasive ventilation and longer length of stay.