Featured Article

Outcomes of cardiothoracic surgery in women with Turner syndrome

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Background: To describe short- and mid-term surgical outcomes of patients with Turner syndrome (TS) after cardiovascular interventions.

Methods: All individuals >12 years of age at the time of surgical repair for cardiovascular disease (valve or coarctation repairs, aortic disease, aortic dissection) from 2002 to 2022 were eligible. The primary endpoint was complications or death within 30 days of intervention. Secondary outcomes included late complications and reinterventions within six months. Combined data from the University of Texas Health Science Center at Houston and the Turner Syndrome Society of the United States were included in the analysis.

Results: We identified 22 patients who met the inclusion criterion. The median age was 46 years (range, 21–75 years), with 86% having estrogen replacement therapy. The most common medical condition was hypertension (77%), followed by hypothyroidism (59%). The most frequent indication for surgery was aortic root or ascending aortic aneurysms (68%), followed by symptomatic aortic stenosis in patients with bicuspid aortic valve (64%), coarctation of aorta (45%), and acute aortic dissection (18%). Respiratory complications were the most common (68%). Pleural effusions were the most frequent found sign on imaging studies (68%). Thoracentesis, or chest tube placement, was required in 33% (5/15). Respiratory failure requiring specific support with high flow oxygen and/or thoracentesis occurred in 36% (8/22).

Conclusions: Patients with TS may be at an increased risk for postoperative complications after aortic surgery. Bicuspid aortic valve (59%) and coarctation of the aorta (45%) were the most common congenital malformations among our study group. Our study showed that respiratory complications were the most common, with pleural effusions being the most common post-surgery complication.

Keywords: Aneurysm; aortic repair; Turner syndrome (TS); women

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Introduction

Cardiovascular disease, both congenital and acquired, is common in patients with Turner syndrome (TS), and has been identified as a primary cause of increased mortality in these patients (1-5). The most common congenital heart lesions are bicuspid aortic valve, coarctation of the aorta, aortic stenosis, patent ductus arteriosus, and persistent left superior vena cava (4-6). More than one-third of TS patients develop thoracic aortic aneurysms, and up to 25% require elective or emergent surgical repair of the aorta, due to enlarging aneurysms or acute aortic dissections (1-3). Since TS was first described in the 1930s, many studies have reported a unique constellation of comorbid conditions, including renal and genitourinary anomalies, autoimmune disease, and cardiovascular lesions (3,4,6-8).

Many patients with TS will require at least one cardiovascular procedure during their lifetime (9). Some display a great level of medical complexity, which may put them at increased risk for perioperative complications. Morbidity following aortic surgery in TS patients may include complications such as stroke, respiratory failure, arrhythmias, pericardial effusions, renal failure, infections, and pneumothorax. However, little is known about the exact prevalence and types of perioperative complications, and their effects on surgical outcomes (10).

It has been estimated that more than 100,000 patients with TS live in the United States (11). The high incidence of congenital problems makes this vulnerable population of special interest, with the potential for unique complications not seen in other patient groups with similar indications for surgery. Current international guidelines for aortic surgical indications, decision-making, and counseling for patients with TS do not factor in these potential risks. Careful analysis of perioperative outcomes may deliver new insights to close these knowledge gaps and improve the risk stratification of TS patients.

A high prevalence of aortic anomalies has also been found in patients with TS. Studies have found dilatation of the ascending and aortic arch, with a prevalence of transverse arch elongation of 49%, and aberrant right subclavian artery of 8%. Thoracic aneurysms and aortic dissections are also prevalent in TS patients. Venous anomalies may also be present in women with TS, including partial anomalous pulmonary venous return (13%) and persistent left superior vena cava (13%) (12).

Overall, aortic disease in TS patients is very common, with left-sided congenital heart lesions, bicuspid aortic valves, thoracic aneurysms, and acute aortic dissections being 30–50 times more common in TS patients than in the general population (2). The goal of this study is to describe the surgical outcomes of patients with TS after cardiovascular interventions. We hypothesize that patients with TS are at increased risk for postoperative complications after aortic surgery due to their small body size and frequent congenital thoracic malformations. Understanding the risks predisposing to postoperative events after aortic procedures in TS patients may enable us to predict, avoid or mitigate these complications.

Methods

This retrospective case series includes patients with TS who underwent any cardiovascular procedure between 2002–2023 and were at least 12 years of age at the time of the procedure. Any open, hybrid, or endovascular procedure was eligible for inclusion. Eligible participants were identified from four sources: the Inspiring New Science in Guiding Healthcare in Turner Syndrome Registry (INSIGHTS), the Turner Syndrome Research Registry, the UTHealth Houston TS Registry, and inpatient records at Memorial Hermann Hospital-Texas Medical Center. Study participants were also prospectively recruited from the Turner Syndrome Society of the United States membership using social media and newsletter posts. The University of Texas Health (UTH) Institutional Review Board approved the study protocol.

Variables on perioperative outcomes were abstracted from medical records. In-hospital, postoperative complications and mortality were the primary endpoints. Mid-term morbidity and mortality rates were secondary outcomes. In-hospital respiratory complications included pleural effusions, pneumonia, pneumothorax, chylothorax, reintubation and prolonged ventilation. Respiratory failure was defined as those patients requiring prolonged ventilation or reintubation. Prolonged mechanical ventilation in this study was defined as greater than 48 hours. Perioperative risks were estimated using the EuroSCORE II calculator (13).

Statistical analysis

Descriptive statistics were used to characterize the variability of measurements. We report categorical variables as percentages and continuous variables as mean ± standard deviation (SD) or median for skewed data. Data was compared using standard parametric and nonparametric tests, depending on the distribution. The statistical analysis was performed by SPSS-V21.0 software (IBM, Armonk, New York, USA)

Results

We identified 22 patients who met the inclusion criteria. The final demographic characteristics of the patient cohort are listed in *Table 1*. Karyotypes were available for 20 patients (90%), most of which were 45X (70%) or mosaic (45X/46XX or 45X/47XXX, 20%). The median age at the time of the first procedure was 46 years (range, 21–75 years), and only two were older than 70 years of age. A high percentage took estrogen replacement therapy (86%). The most common medical condition was hypertension, found in 77% of patients, followed by hypothyroidism (59%). More

Table 1 Baseline patient demographic features and medications (total n=22)

Characteristics	Values
Age, years	46 [21–75]
45X Karyotype	14 [64]
Height (cm)	147±14
Weight (kg)	55±23
BMI (kg/m^2)	25±7
Other comorbidities	
Hypertension	17 [77]
Hypothyroidism	13 [59]
CAD	6 [27]
Autoimmune disease	4 [18]
Pulmonary disease	3 [14]
Arrythmias	2 [9]
Lymphedema	2 [9]
Diabetes	1 [5]
Cardiovascular medications	
Warfarin	9 [41]
Aspirin	16 [73]
Beta blocker	19 [86]
Statin	16 [73]
Other medication	
Levothyroxine	13 [59]
Estrogens	19 [86]

Data are presented as mean ± SD or median [IQR] or n [%], computed using SPSS-V21.0 software (IBM, Armonk, New York, USA). BMI, body mass index; CAD, coronary artery disease; SD, standard deviation; IQR, interquartile range.

Table 2 Patient types of congenital cardiovascular anomalies (total n=22)

Cardiovascular anomalies	Number [%]
Aortic aneurysm	14 [64]
Bicuspid aortic valve	13 [59]
Coarctation of aorta	10 [45]
Aortic dissection	4 [18]
Partial anomalous pulmonary venous return	4 [18]
ASD	1 [5]
VSD	1 [5]
PDA	0 [0]

ASD, atrial septal defect; VSD, ventricular septal defect; PDA, patent ductus arteriosus.

than one-quarter of patients were diagnosed with coronary artery disease but only 5% were diagnosed with diabetes.

The most common indication for elective repair was aortic root or ascending aortic aneurysms (68%), followed by symptomatic aortic stenosis in patients with bicuspid aortic valve (64%), coarctation (45%), acute aortic dissection (18%), and heart failure symptoms in patients with partial anomalous pulmonary venous return (PAPVR, 18%; *Table 2*).

Ten patients (45%) had both bicuspid aortic valves and an aortic aneurysm of the aortic root or ascending aorta. Eight (36%) were found to have both an aortic aneurysm of the thoracic aorta and coarctation of the aorta, with four presenting with an acute aortic dissection (18%), all of whom had dissection and aortic aneurysm greater than 50 mm in diameter. Among those four, one had an acute type A dissection and a bicuspid aortic valve and two had prior surgery for coarctation of the aorta, both of whom were treated with open aortic repair.

The most frequent surgical procedure was ascending aorta with hemiarch repair, performed in nine patients, six of whom received both a hemiarch repair and aortic valve replacement. The David procedure (valve-sparing aortic root replacement surgery) was performed on one patient, and the Bentall-de Bono procedure was performed on two patients. One open repair of a proximal descending thoracic aortic (DTA) aneurysm was performed in one patient at the site of a previous coarctation using a 24-mm Hemashield woven Dacron tube graft. Other operations included one mitral valve repair with atrial septal defect closure and one repair of left partial anomalous pulmonary venous return using the Warden procedure with direct anastomosis to the left atrial appendage. All aortic tissue sampling from the surgeries were assessed intraoperatively and sent for pathology inspection and evaluation.

Complications and mortality

Complications after aortic surgery are summarized in

Table 3 Patient outcomes and results after aortic surgery (total n=22)

Outcomes	Values
Survival, n [%]	21 [95]
Open surgery	18 [82]
Endovascular surgery	2 [9]
Hybrid surgery	1 [5]
Re-interventions	6 [27]
Complications, n [%]	
Pleural effusion	14 [64]
Bleeding (transfusion)	9 [41]
Respiratory failure	8 [36]
Arrythmias	8 [36]
Infection	5 [23]
Pericarditis	4 [18]
Pacemaker	2 [9]
Renal failure	2 [9]
Stroke	2 [9]
Pneumothorax	1 [5]
Dialysis	0 [0]
Other data, median (range)	
Length of stay, days	8 (4 to 25)

Table 3. The most frequent complications were respiratory events in 68% of the study cohort (15/22). Pleural effusions were the most frequently found abnormality on imaging studies [68% (15/22)]. Five of 15 patients with effusions required thoracentesis or chest tube placement (33%). Respiratory failure requiring specific support and high-flow oxygen or thoracentesis occurred in 8/22 (36%). Three patients required prolonged ventilation (>48 hours) due to pneumonia or multiorgan failure. Another was extubated in postoperative day (POD) 2 but required home oxygen on discharge on POD 5. A third patient was reintubated on POD 6, after acquiring ventilator-associated pneumonia on POD 5, requiring a tracheostomy on POD 10, and discharged on POD 16. Nine patients required red blood cells transfusion (41%). Transfusion was performed on POD 1 in seven of nine patients (77%). Arrhythmias occurred in 41% (9/22) of the study cohort. Five patients developed postoperative atrial fibrillation or atrial flutter, one developed junctional tachycardia, and one developed third-degree AV nodal block, which required a dual chamber pacemaker on POD 7. One patient underwent radiofrequency ablation of a supraventricular tachycardia due to focal atrial tachycardia from the right anteroseptum ten months after surgery. One patient presented with premature supraventricular complexes and ventricular complexes, with no ventricular tachycardia, and was discharged home on POD 4 on atenolol (25 mg twice daily). Notably, seven of the nine patients who developed arrhythmias (77%) also had postoperative pleural effusions. Only two patients who presented with arrhythmias required postoperative transfusion. Less-common complications included infection and pericarditis. Three patients were diagnosed with pericarditis (14%) and four presented with infections (18%). Two developed pneumonias on POD 2 and 5, respectively. One had a sternal wound infection and dehiscence one month after surgery and required readmission, antibiotics, and wound care with a vacuum-assisted closure device. One patient presented with superficial sternal wound infection on POD 7 and was treated with closed incision negative pressure therapy (ciNPT)-Prevena™ device.

For patients who underwent aortic procedures, the mean time to extubation was 0.6 days, with all patients extubated by POD 4. The length of stay was 8 days (range, 4–25 days). Four patients (18%) had at least one reoperation during their index admission, one due to bleeding, two had pericardial tamponade, requiring a pericardial window. One of those patients required a pericardial window on POD 4 after presenting with tamponade. This patient underwent a second surgery with evacuation of chylothorax and pericardial hematoma (with a redo sternotomy) due to tamponade on POD 20, with concomitant ligation of a tributary of the thoracic duct along the anterior surface of the mid-transverse arch. Two patients were non-mosaic.

The 30-day mortality rate was 5%. One patient died on POD 14 due to multiorgan failure after presenting with a complicated right pneumonia on POD 2, sepsis requiring inotropic support on POD 3, respiratory failure requiring mechanical ventilation, right MCA embolic stroke on POD 7, and left pneumothorax on POD 8. The overall intervention-free survival rate (*Figure 1*) was 95% (21/22) at a mean follow-up interval of 61 months (range, 2–92).

Patients with body mass index (BMI) ≤20 kg/m^2 developed more frequent pleural effusions (100%) than the overall TS group (68%). Only one patient (20%) required thoracentesis. For patients with BMI >30 kg/m^2,

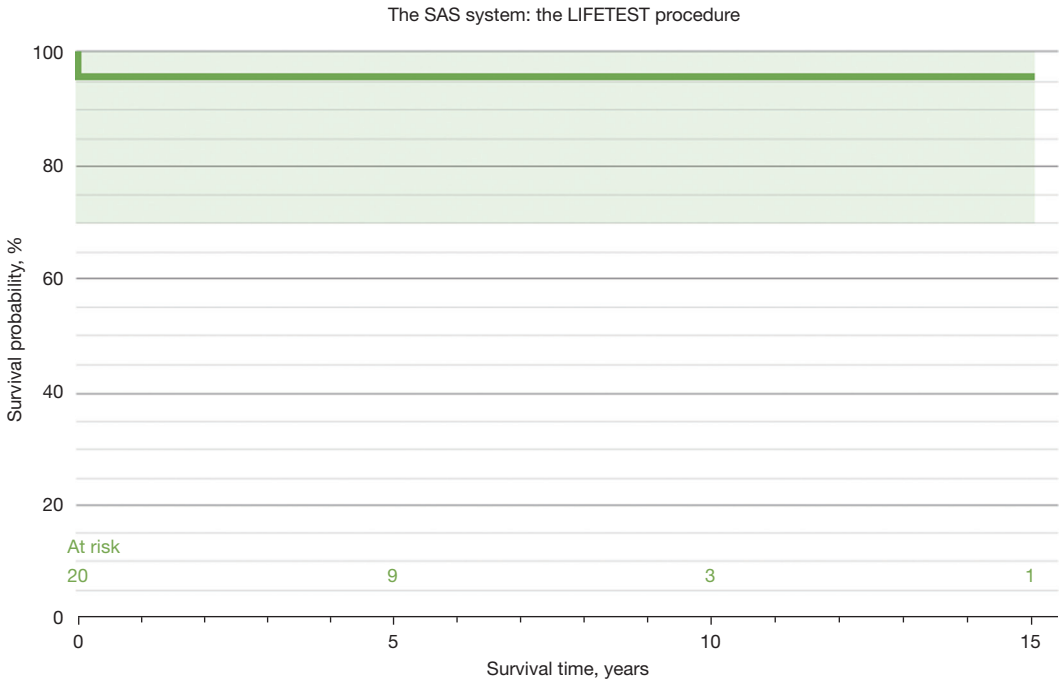

Figure 1 Kaplan-Meier estimates for survival in our TS group. TS, Turner syndrome.

postoperative anemia requiring transfusion was more common (80% *vs.* 40%). There were no significant differences in the overall rates of complications in patients with pleural effusions or hypertension, which was the most frequent medical diagnosis.

In total, 16 patients (73%) presented with aortic aneurysms or aortic dissections, and the mean maximum aortic diameter was 45 mm (range, 36–82 mm). The average Z-score was 6.2 (range, 4–17). The highest Z-score (score =17) was found in a 41-year-old TS patient who presented with a ruptured DTA aneurysm and successfully underwent emergent thoracic endovascular aortic repair (TEVAR). There was insufficient data to evaluate correlations between congenital heart lesions and perioperative events.

Discussion

Summary of key findings

Our findings highlight the significance of respiratory and cardiac complications after cardiovascular procedures in TS. More than two-thirds of patients in this series developed significant pleural effusions, arrhythmias or pneumothorax after surgical or hybrid procedures. In our TS study group, out of 14 patients with pleural effusions, four (29%) required interventions. One patient needed a left-sided chest tube insertion due to pneumothorax on POD 8 and one required bilateral thoracentesis (1.2 and 1 L drained on initial evacuation, respectively) due to large pleural effusions on POD 18. One patient required thoracentesis for a right pleural effusion on POD 4 (draining 600 mL). A fourth patient had a large, left-pleural effusion requiring thoracentesis on POD 3 after a thoracic branched endoprosthesis for a distal arch aneurysm. The most common aortic intervention for TS patients was ascending aortic and hemiarch repair 40% (9/22), with five patients receiving both a hemiarch repair and aortic valve replacement.

The most frequent chromosomal abnormality that causes TS is known as monosomy X (45X), characterized by the complete absence of one sex chromosome (12). TS is a fairly common chromosomal disorder that affects around 1 in 2,000 newborn females (9,14). The most common physical characteristics of individuals with TS are short stature and gonadal dysgenesis (12). Most patients with TS develop menstrual abnormalities, and many are diagnosed with TS during workups for infertility. Other common clinical findings include skeletal anomalies, such as scoliosis or kyphosis and cubitus valgus (15-18).

Cardiovascular complications and surgical outcomes: insights from the literature

The most serious clinical complication of TS is congenital heart disease, which may manifest as many different cardiac lesions, including bicuspid aortic valve, coarctation of the aorta, or septal defects. Up to 50% of patients with TS have at least one congenital cardiovascular lesion (9). Also, up to 50% of patients with TS are born with congenital heart disease or develop complications related to congenital cardiovascular lesions during their lifetimes (9). Congenital heart lesions, primarily left heart obstructive lesions, are the most frequent cause of early mortality in TS (3,19). Bicuspid aortic value is the most common congenital lesion in TS (15–30%) while aortic coarctation is the second-most common lesion (7–18%) (3,19-21). Acute aortic dissections are up to 100 times more frequent in young individuals with TS compared to population controls and are associated with very high morbidity and mortality (2,22-26). TS patients also acquire adult-onset cardiovascular diseases, including coronary artery disease and valvular heart disease, at higher-than-expected rates. Factors that increase risk for acquired cardiovascular diseases include dyslipidemia, insulin resistance, hypertension, and uncorrected congenital heart lesions (27-29).

Alam *et al.* evaluated the long-term mortality after congenital heart surgeries in TS patients who survived to hospital discharge and found they had excellent survival rates, comparable to the population mean (10). The series featured pediatric cases (47%) who underwent very different surgical procedures to correct severe congenital malformations, such as hypoplastic left heart, and they did not collect data on immediate perioperative complications, other than mortality. A recent study from the Mayo Clinic presented long-term outcomes of 51 pediatric and adult patients with TS who underwent a wider variety of surgical procedures. In-hospital mortality (8%) was comparable to our series, and their long-term survival was also excellent (89% at 20 years) (4).

Another recent study by Fuchs *et al.*, which included a cohort of 317 patients with TS, described long-term mortality at 14% and a mean age at death of 53 years. In that series, coarctation was the most frequent aortic repair (28%), followed by aortic replacement in 14% and aortic valve replacement in 12% (4). The principal limitations were that it was a single-center, retrospective study and aortic images were not available for all patients. They found that cardiac-related events were the most common cause of death (22%) (25). Natural history studies continue to show that older patients with TS may experience increased cardiovascular events related to ischemic heart disease, hypertension, and diabetes (3,4,19).

A large study incorporating data from The Society of Thoracic Surgeons Congenital Heart Surgery Database (STS-CHSD) included 780 patients with TS from 95 different institutions (5). That study focused primarily on early congenital surgical procedures in a neonatal and pediatric TS cohort, including Norwood, superior cavopulmonary anastomosis (Glenn), and Fontan operations. Moreover, the study population included non-syndromic patients, as well as TS patients, and did not evaluate long-term outcomes. While coarctation interventions were the most common procedure in the STS data (35%), open aortic aneurysm repair was the most frequent intervention in our series (14/22, 64%).

There is controversy in the literature regarding the mid- and long-term outcomes of TS patients who undergo cardiovascular interventions. Although many series have published results showing an overall decrease in long-term survival in patients with TS (3,4,25), other publications have presented data indicating increased morbidity for select operations without an associated increase in operative mortality for TS patients (5,10).

How our study differs

Several case series have concluded that TS patients remain at risk for late cardiovascular events after procedures. However, significant gaps remain in data about in-hospital and 30-day complications. Our study comprehensively assesses these aspects of perioperative management. This is the first study to focus exclusively on cardiovascular procedures in a TS cohort and specific complications that are highly relevant to TS over a 20-year period.

Our study also provides new detailed information on procedural outcomes in patients with TS. We found that respiratory complications are prevalent among TS patients after aortic surgery, with more than 50% experiencing pleural effusions and less than 10% developing pneumothorax or respiratory failure. Acute renal failure following surgery is uncommon, and temporary dialysis is rarely required. However, our small cohort suggested that postoperative bleeding requiring transfusion may be higher than expected, up to 40–50%. We found that 33% of TS patients with pleural effusions required thoracentesis

or chest tube placement. This rate is higher than those of other non-TS patients after aortic interventions in our region, and compared to other published reports (30).

The higher-than-expected perioperative event rate highlights the importance of shared decision making about the timing of elective procedures and may justify a more extensive preoperative workup for some patients. Awareness about unique perioperative risks in TS may also impact counseling about operative risks, particularly for patients with prior cardiovascular procedures, multiple cardiovascular risk factors, or small body sizes.

Limitations

This retrospective study may be subject to inherent biases that are associated with the study design. There are also inherent limitations related to data abstraction from electronic health records. We recognize that the small number of cases precludes extensive comparisons and limits our ability to discover associations between clinical features and outcomes. Images were not available for all patients to confirm preoperative cardiovascular anatomy.

Conclusions

Patients with TS may be at an increased risk for postoperative complications after aortic surgery. Our study showed that cardiac and respiratory complications are frequent, with the latter being the most common, and pleural effusions being the most common overall complication after surgery. Bicuspid aortic valve (64%) and coarctation of the aorta (45%) were the most common congenital malformations among our study group, and might be related to the postoperative outcomes. In more than 40–50% of patients, postoperative bleeding requiring transfusion may be necessary. These findings should be validated in larger cohorts with longer follow-up periods before this data is used to inform new guidelines for the perioperative management of TS patients.

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Footnote

Conflicts of Interest: ROA is a consultant for Medtronic and EndoRon Ltd. ALE is a consultant for WL Gore, CryoLife, Edwards Lifesciences, and Terumo Aortic. SKP is supported by a gift from the Turner Syndrome Society of the United States. The other authors have no conflicts of interest to declare.

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