

# TURNER SYNDROME AND AORTIC SURGERY

**Presenter: Lucas Ribé MD,**

**Yuki Ikeno, Rana O Afifi, Akiko Tanaka, Ferial D Shihadeh, Anthony L Estrera, Siddarth Prakash.**

April 25<sup>th</sup>, 2024. Aortic Symposium. AATS- The American Association for Thoracic Surgery 104<sup>th</sup> Annual Meeting.

The University of Texas Health Science Center at Houston.

Department of  
Cardiothoracic &  
Vascular Surgery



# INTRODUCTION

- **Up to 50%** of patients with Turner Syndrome will have a congenital heart disease, or may develop throughout their life-time some type of congenital heart disease.
- Congenital heart anomalies are currently the most frequent **cause of early mortality** in these patients.

Silberbach M, Roos-Hesselink JW, Andersen NH, Braverman AC, Brown N, Collins T, et al.  
Cardiovascular Health in Turner Syndrome: A Scientific Statement From the American Heart Association.  
Circ. Genom. Precis. Med. 2018;11:e000048.

# INTRODUCTION

- Some of these patients display a great level of medical complexity, which may put them at increased risk for perioperative complications.
- However, **little is known** about the prevalence and types of perioperative complications and their effects on surgical outcomes.

Fuchs MM, Attenhofer Jost CH, Said SM, Hagler DJ, Connolly HM, Dearani JA, Egbe AC.  
Cardiovascular surgery in Turner syndrome- early outcome and long-term follow-up.  
World J Cardiol. 2020 Mar 26;12(3):97-106.

# AIM

Describe the surgical and medical outcomes of patients with Turner syndrome (TS) after aortic and cardiovascular interventions

# METHODS

- **INCLUSION:**

- **DESIGN:**

- Retrospective observational. Non-randomized, non-interventional.

- **Eligible participants** were identified from four sources:

1. Inspiring New Science in Guiding Healthcare in Turner Syndrome Registry (INSIGHTS).
2. Turner Syndrome Research Registry (TSRR).
3. UTHealth Houston TS Registry.
4. Inpatient records at Memorial Hermann Hospital-Texas Medical Center.

- Variables on perioperative outcomes were abstracted from medical records.

	Variables PreOP	Cardiovascular malformations PreOP
1	Age	Bicuspid aortic valve
2	Karyotype	Coarctation
3	Weight	Aortic aneurysm
4	Height	Aortic dissection
5	Smoking (ever/never/former)	Septal defects
6	CV medication	Patent ductus arteriosus
7	Hypertension	Anomalous pulmonary veins
8	Diabetes mellitus	ASD
9	Hypothyroidism	VSD
10	Lymphedema	
11	Estrogen supplementation	
12	Autoimmune disease	
13	Pulmonary disease	
14	Coronary artery disease	
15	Arrhythmia	
16	Operation type (Emergent, urgent or elective)	
17	Indication for operation	
18	Prior cardiac/aortic interventions	

	Variables POST- OP	Variables POST- OP Long- term
1	Alive/dead	Survival at 1 year
2	Re-operation	All-cause re-hospitalization within 6 months
3	Stroke	Additional cardiovascular procedures
4	Respiratory failure	
5	Renal failure. Dialysis (Y/ N).	
6	Time to extubation ( <24 hrs, >24 hrs).	
7	Time to discharge	
8	Pneumothorax (CT placement Y/S)	
9	Pleural effusion (CT placement Y/S)	
10	Pericardial effusion	
11	Pacemaker	
12	Arrhythmia	
13	Hemoglobin	
14	Infection	

# RESULTS

CHARACTERISTICS	TYPE (Numerical- Percentage)
Age	46 (21- 75)
Karyotype	45 XO (64%)
Height (cm)	147 +/- 14
Weight (kg)	55 +/- 23
BMI (kg/m <sup>2</sup> )	25 +/- 7
<b><i>Other comorbidities</i></b>	
Hypertension	17/ 22 (77%)
Hypothyroidism	13/ 22 (59%)
CAD	6/ 22 (27%)
Autoimmune disease	4/ 22 (18%)
Pulmonary disease	3/ 22 (13.6%)
Arrhythmias	2/ 22 (9%)
Lymphedema	2/ 22 (9%)
Diabetes	1/ 22 (4.5%)



# RESULTS

## CARDIOVASCULAR ANOMALIES

## TYPES (Numerical/ Percentage)

1. Aortic aneurysm	14/ 22 (64%)
2. Bicuspid aortic valve	13/ 22 (59%)
3. Coarctation of aorta	10/ 22 (45%)
4. Aortic dissection	4/ 22 (18%)
5. Partial anomalous pulmonary venous return	4/ 22 (18%)
6. Atrial septal defect (ASD)	1/ 22 (4.5%)
7. Ventricular septal defect (VSD)	1/ 22 (4.5%)
8. Patent ductus arteriosus (PDA)	0/ 22 (0%)

# RESULTS

<b>OUTCOMES</b>	<b>Types (Numerical/ Percent)</b>
Survival	<b>95 % (21/22)</b>
Open surgery	<b>18/ 22 (82%)</b>
Endovascular surgery	<b>2/ 22 (9%)</b>
Hybrid surgery	<b>1/ 22 (4.5%)</b>
Re-interventions	<b>6/ 22 (27%)</b>
<b><i>Complications</i></b>	
Pleural effusion	<b>15/ 22 (68%)</b>
Bleeding (transfusion)	<b>9/ 22 (41%)</b>
Respiratory failure	<b>8/ 22 (36%)</b>
Arrhythmias	<b>9/ 22 (41%)</b>
Infection	<b>5/ 22 (23%)</b>
Pericarditis	<b>4/ 22 (18%)</b>
Pacemaker	<b>2/ 22 (9%)</b>
Renal failure	<b>2/ 22 (9%)</b>
Stroke	<b>2/ 22 (9%)</b>
Pneumothorax	<b>1/ 22 (4.5%)</b>
Dialysis	<b>0/ 22 (0%)</b>

# RESULTS

- **Respiratory complications** were the most common: 68% (15/22). Pleural effusions were the most frequently found sign on imaging studies, 68% (15/22).
- 5/15 patients required thoracentesis/ chest tube placement **(33%)**.
- 9 patients: RBC **transfusion (41%)**. POD 1 in 7/ 9 (77%).

# DISCUSSION

- Our small cohort suggested that **postoperative bleeding** requiring transfusion may be higher than expected (40-50%).
- Acute **renal failure** after surgery is uncommon.
- **33% of TS** patients with pleural effusions required thoracentesis. This rate is higher than those of other published reports.

Labidi M, Baillet R, Dionne B, Lacasse Y, Maltais F, Boulet L-P.  
Pleural effusions following cardiac surgery: prevalence, risk factors, and clinical features.  
Chest. 2009;136:1604-11.

# DISCUSSION

- **This is the 1<sup>st</sup> study** to focus exclusively on cardiovascular procedures in TS cohort & specific complications that are highly relevant to patients over a 20-year period.
- There is still little data in the literature as regards the indications for aortic surgery in TS patients, & mid and long- term outcomes.