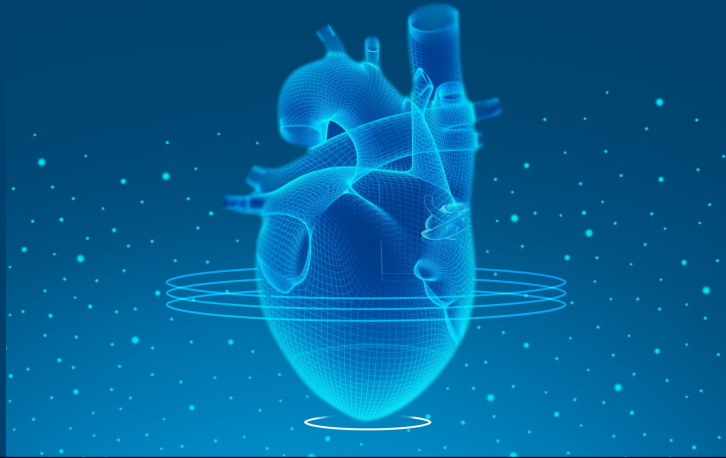




# GRAND ROUNDS



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## COMPLEX CONGENITAL CASES: THE NEW ERA

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## Growth of the patient population

- Swedish National Inpatient, Outpatient, and Cause of Death Registry
- Currently, 97% of children born with congenital heart disease are reaching adulthood
- At least 75% of patients with CHD alive at 18 years of age lived past middle age and became sexagenarians
- Mortality was 3.2 times higher (95% CI, 3.0–3.4;  $P < 0.001$ ) among patients with ACHD compared with matched controls
- Estimated 25% require reoperation as an adult

Adults With Congenital Heart Disease: Trends in Event-Free Survival Past Middle Age. Mikael Dellborg, MD, PhD, Kok Wai Giang, PhD, Peter Eriksson, MD, PhD, Hans Liden, MD, PhD, Maria Fedchenko, MD, PhD, Anders Ahnfelt, MD, Annika Rosengren, MD, PhD and Zacharias Mandelzakis, MD, PhD *Circulation* Volume 147, Number 12 <https://doi.org/10.1161/CIRCULATIONAHA.122.068834>

Surgery in Adults With Congenital Heart Disease. A.C. Zomer, MD, C.L. Verheugt, MD, PhD, I. Vaartjes, PhD, C.S.P.M. Uiterwaal, MD, PhD, M.M. Langemeijer, MSc, D.R. Koolbergen, MD, PhD, M.G. Hazekamp, MD, PhD, J.P. van Melle, MD, PhD, T.C. Konings, MD, L. Belleren, MD, D.E. Grobbee, MD, PhD, and B.J.M. Mulder, MD, PhD, *Circulation*. Volume 124, Number 28, <https://doi.org/10.1161/CIRCULATIONAHA.111.027763>

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## Complex Medical and Surgical Issues

- Decoupling of the right and left sided circulation
- Psychology of chronic illness
  - Adult body with child behavior
- Anatomy, Anatomy, Anatomy
  - So.. you are telling me there is no left ventricle and the right ventricle is connected to the aorta...??
  - Yes.
  - How the....

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## Different Language

### Non congenital anatomic description

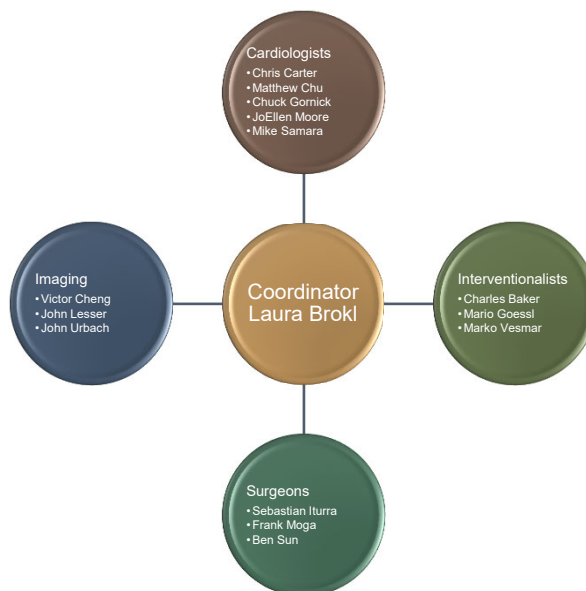
- Aortic valve replacement
- Mitral valve repair/replacement
- Tricuspid valve repair/replacement
- Coronary artery bypass times 1,2,3...
- Repair ascending aortic aneurysm
- Aortic root repair

### Congenital

- Unidirectional/bidirectional Glenn
- Mustard/Senning procedure
- Blalock-Taussig (BT) shunt
- Warden procedure
- LeCompte maneuver (is that legal in all 50 states?)
- Fontan (isn't that a cheese dip?)

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## The Team



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## Case 1

- 32 y.o healthy male doing triathlons and running on a daily basis
- Past 6 months he noticed DOE and mid sternal chest pain that for the past few days which has started radiating up his neck
- Having difficulty running 2 miles now.
- Stress EKG which showed ST depression the inferolateral leads

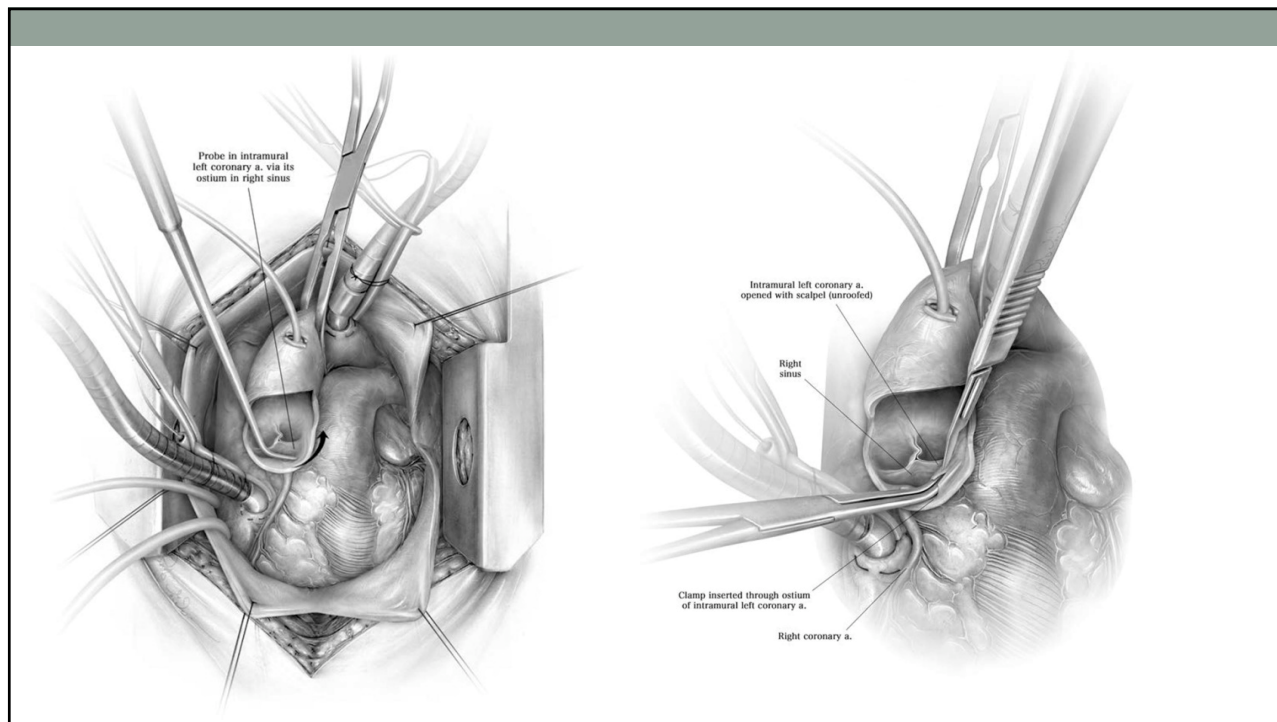


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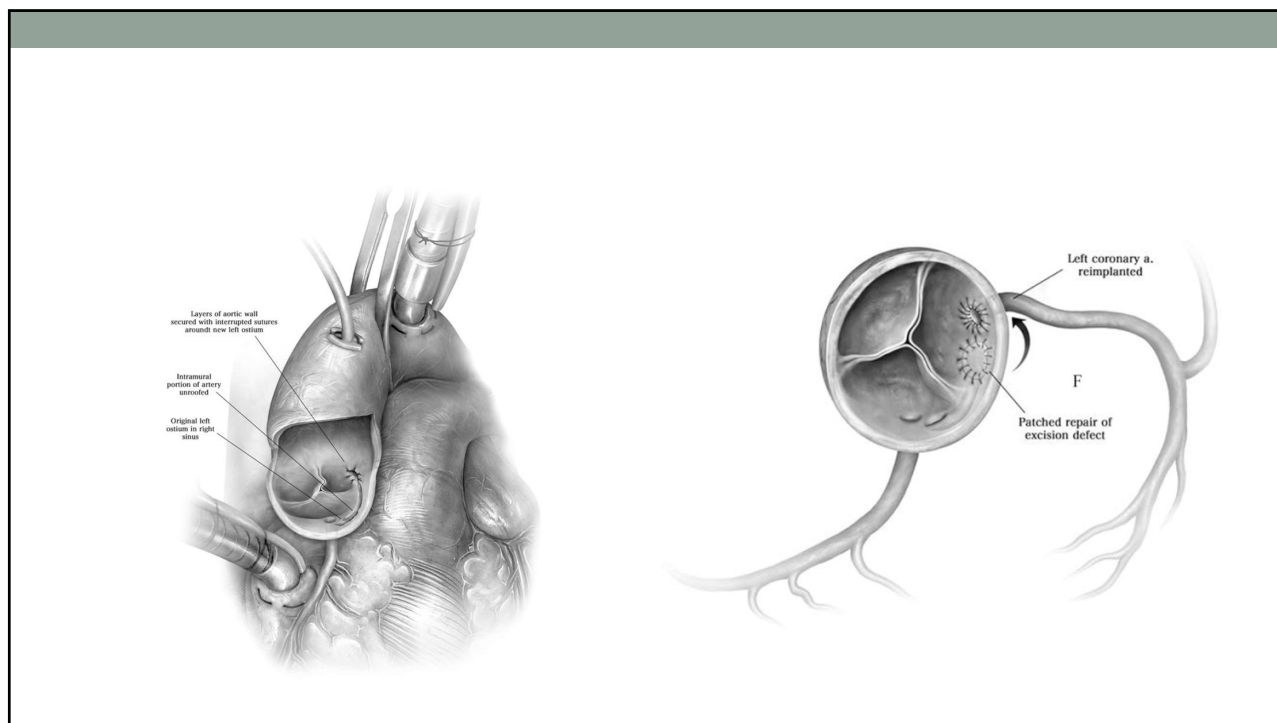
## Anomalous RCA



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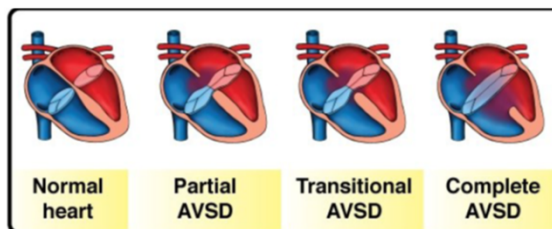
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## Case 2

- 5/20/1990
- 9/1/1990 Atrioventricular canal repair dropping AV valve to top of septal ridge, which was small; mitral valvuloplasty & annuloplasty; pericardial patch closure, atrial primum defect portion of AV canal - Dr. Helseth,
- 9/06/1990 re-repair of mitral valvuloplasty due to severe mitral regurg - Dr. Helseth,
- 11/15/1995 mitral valve replacement 23 mm St. Jude prosthesis - Dr. Helseth
- 7/27/2009 mitral valve replacement 25 mm St. Jude Epic porcine prosthesis - Dr. Moga,
- 2/08/2012 mitral valve replacement 29 mm Mosaic porcine valve - Dr. Moga,
- 11/27/2019 mitral valve replacement using a 33 mm Epic tissue valve - Dr. Sun & Moga



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## Case 2

- William Kessler pectus 1008403145
- Michelle King pectus 1007551260

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## Pectus Excavatum

- Most common deformity of the thoracic wall
- Marfan's Syndrome
- Congenital cardiac disease
- Patients with pectus excavatum have slightly higher incidence of valvular issues

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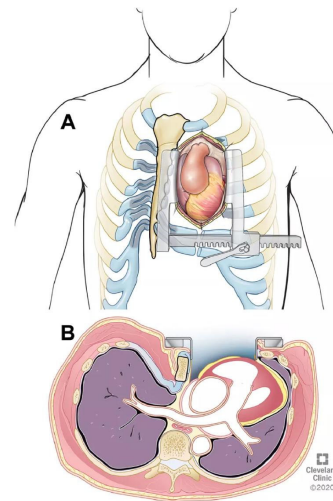
## How to access?



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## Surgical Approach

- Remove cartilage on the left side preserving the IMA
- Remove the cartilage on the right side preserving the IMA
- Exposure to all cardiac structures



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## How to access?



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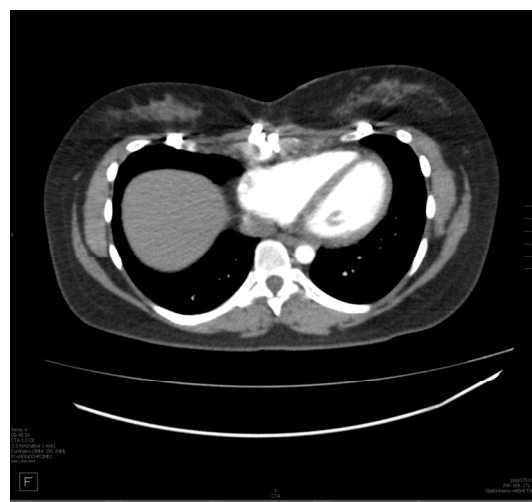
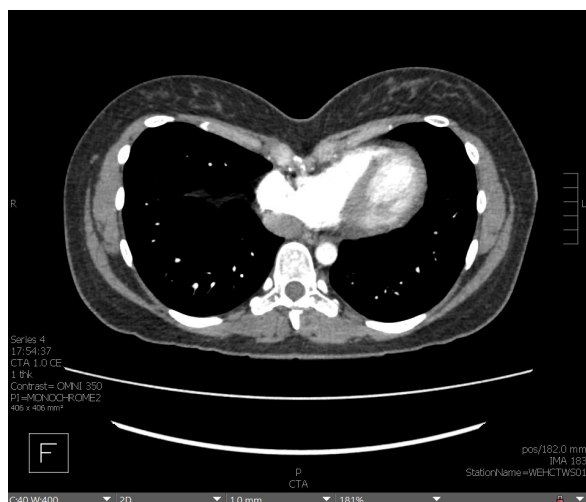


## Modified Ravitch

- Remove costal cartilage on both sides of the lower sternum
- Reshape the sternum
- Rigid fixation of sternum
- Rigid fixation of boney ribs to the sternum



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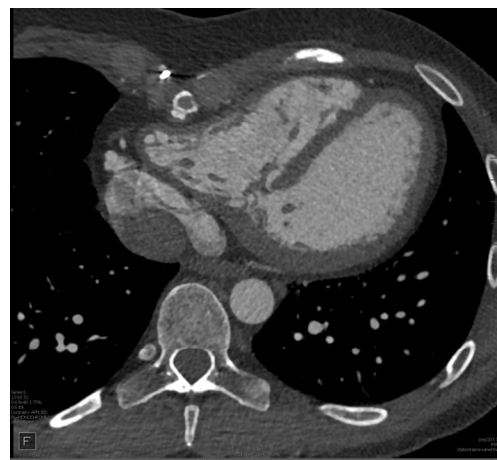
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## Case

- 32 y. o. male with history of -transposition of the great arteries who underwent arterial switch procedure in March 1992 with a LeCompte maneuver
- Pulmonary valvectomy/pericardial patch over PAs October 1992
- 24 mm pulmonary homograft implanted in August 2006
- Now with pulmonary valve stenosis mild RV dysfunction
- Evaluated for transcatheter options and found not to be a good candidate for TPVR

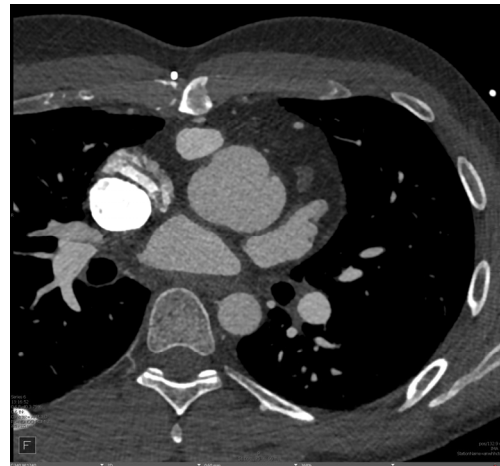
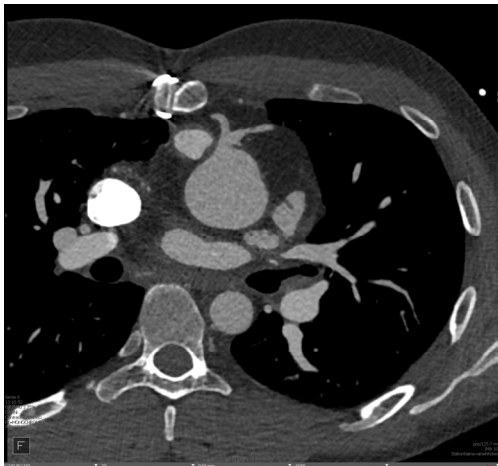
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## LeCompte and pectus with partial sternal non-union



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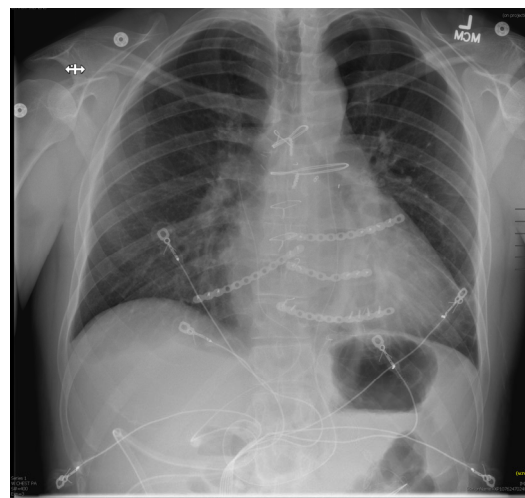
## Right Coronary going to be problematic



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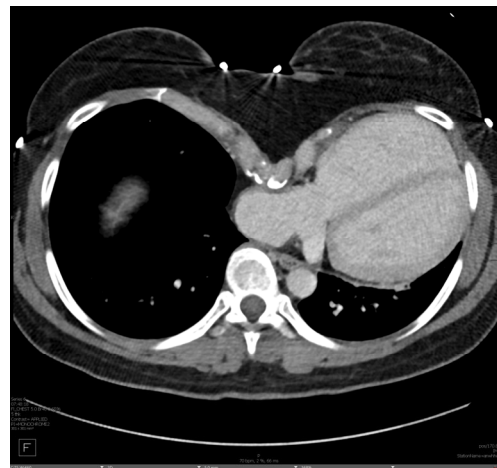
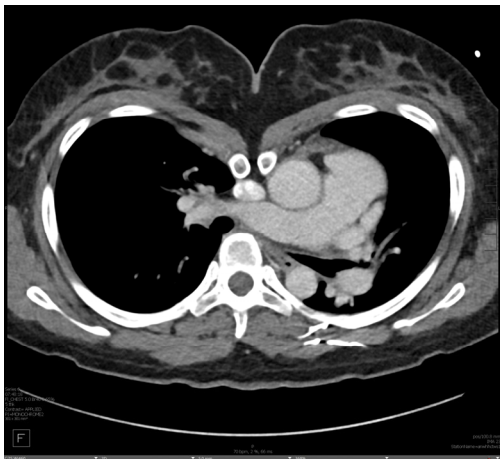
## Procedure

- Modified Ravitch
- 25 mm Mosaic tissue valve
- Pericardial patch augmentation



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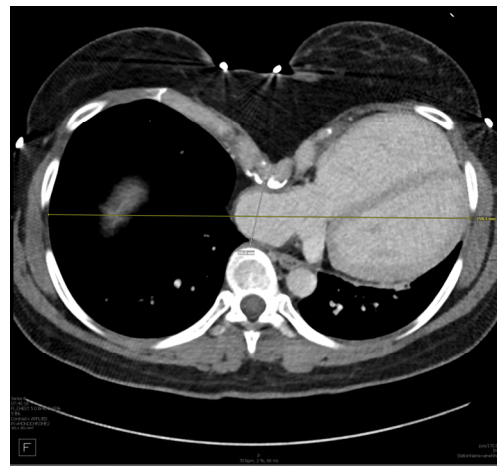
## Pulmonary Valve Stenosis/Insufficiency



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## Haller Index=sternum to spine/chest diameter

- Normal < 2.0
- Mild pectus = 2.0-3.2
- Moderate pectus = 3.2-3.5
- Severe pectus > 3.5
- Surgical correction recommended > 3.25
- Haller Index 8.1



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## Case 3

- Cole Young 1007887595
- Morgan Bartlett 1008736473

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## Case

- 37 y.o. with history of tricuspid atresia and L-transposition of the great vessels
- 6/23/1984 Right Blalock-Taussig shunt
- 12/12/1988 Fontan procedure w/ connection of RA appendage to main & confluent large PA w/ bovine pericardial gusset enlargement; pericardial closure of common atrium to keep single large AV valve to the left; ligation of BT shunt; plus connection of anomalous left superior pulmonary vein to the left atrial appendage
- 1/02/1989 removal of BT shunt; R innominate to RPA Gore-Tex shunt
- 8/11/1999 Lateral tunnel revision of the Fontan, to include incorporation of the SVC directly into the RPA; atrial septectomy; placement of permanent & temporary AV sequential pacing leads; done with 25mm St. Jude aortic valve replacement
- He also has a history of non-compliance and mechanical aortic valve thrombosis requiring lytic therapy in 2010.
- Presented in atrial flutter due to noncompliance with amiodarone as well as subtherapeutic INR
- PEA arrest after IV amiodarone and placed on ECMO
- Imaging showed mechanical aortic valve fixed leaflet

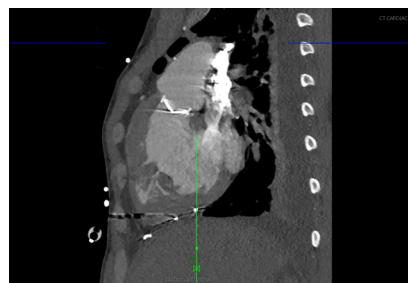
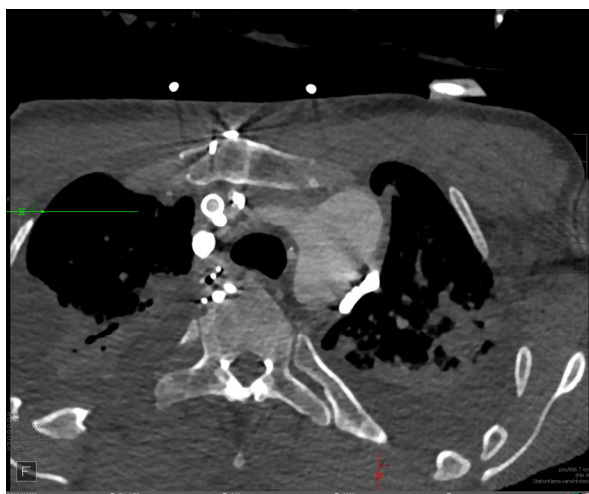
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### Case

Tricuspid atresia and L-  
transposition of the great vessels  
Fontan  
St Jude mechanical aortic valve  
with one of the leaflets stuck  
On ECMO  
Two attempts at lytics were  
attempted without change



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## Surgery

- Left anterior thoracotomy 2<sup>nd</sup> interspace
- Identified anomalous venous drainage to left atrium
- Converted ECMO to CPB
- Cross clamped aorta
- Hockey stick aortotomy and excised mechanical valve
  - One leaflet had pannus and was fused
- 23 mm Magna Ease tissue valve
- Decannulated from ECMO 48 hrs later

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## Case

- 31 year-old female with double outlet right ventricle
- Transposition of the great vessels
- Pulmonic stenosis
- Right ventricular outflow tract stenosis
- Pulmonary insufficiency
- LVOT subaortic membrane with stenosis
- Single coronary artery s/p intervention
- Surgical repair in infancy including pulmonary artery banding
- redo sternotomy in December 2020 with subaortic membrane resection, pulmonary valve replacement with a 25 mm bioprosthetic Mosaic valve, patch augmentation of the RVOT
- Complicated by postoperative VT and RV dysfunction.
- She required VA ECMO and an open chest
- Angiogram at that time showed patent coronary arteries. She eventually underwent subcutaneous ICD implantation on 6.28.2023.



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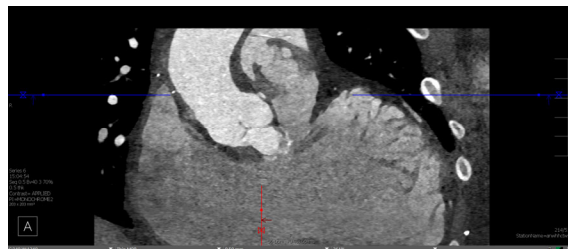
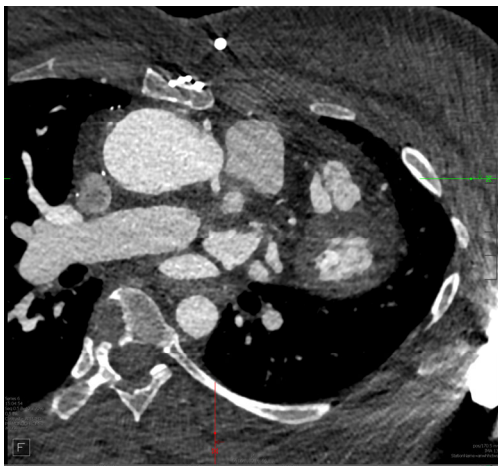


- Presented with polymorphic VT and terminated with ICD shock
- Left heart cath by Dr Goessl



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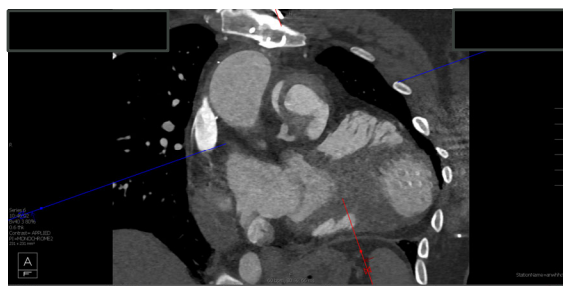
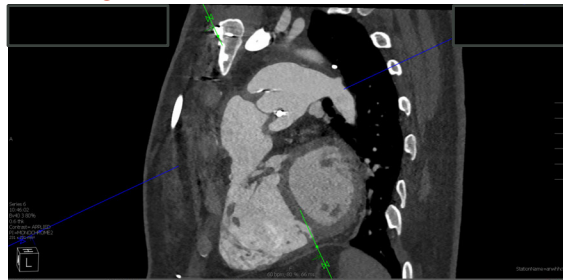
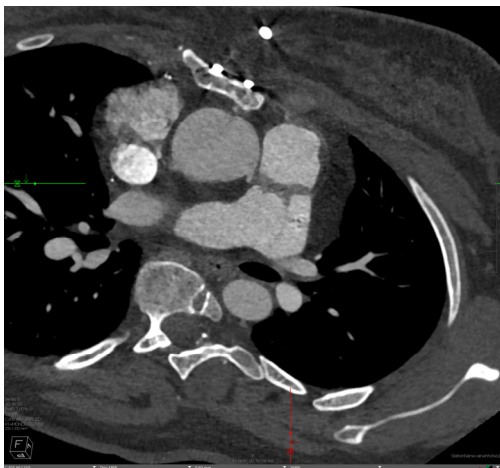
## Tissue Valve Kinking Coronary



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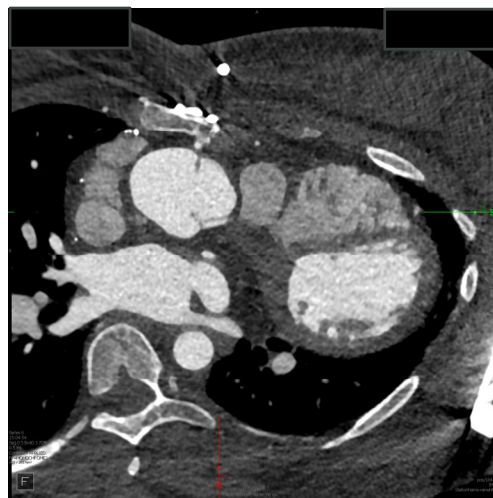
## Tissue Valve Kinking Coronary



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## Other Hazard

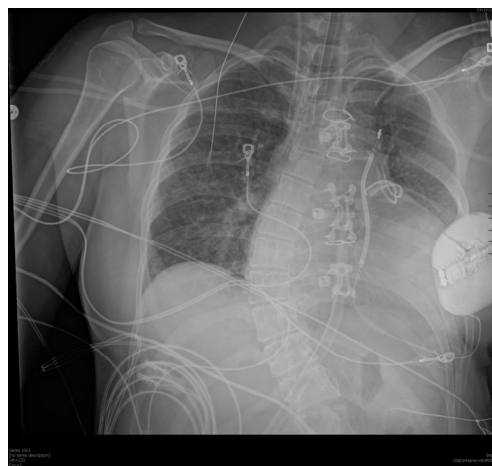
- RCA touches the back of the sternum



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## Operation 6/10/2024

- Femoral cannulation for bypass
- Limited left anterior thoracotomy
- Removal of stented valve
- 25 mm pulmonary homograft
- Bypass time 59 min
- Discharged home POD 3  
6/13/2024



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## Final Thoughts

- Adult Congenital Heart Disease is a rapidly growing field
- Complex anatomy
- Complex physiology
- All reoperative surgery
- Dedicated and focused multidisciplinary specialists are critical for a successful program
- We are truly unique in this space.

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