



# Adult Congenital Heart Disease for the General Cardiologist

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@drwaynefranklin



# Disclosures

- *I have no disclosures*

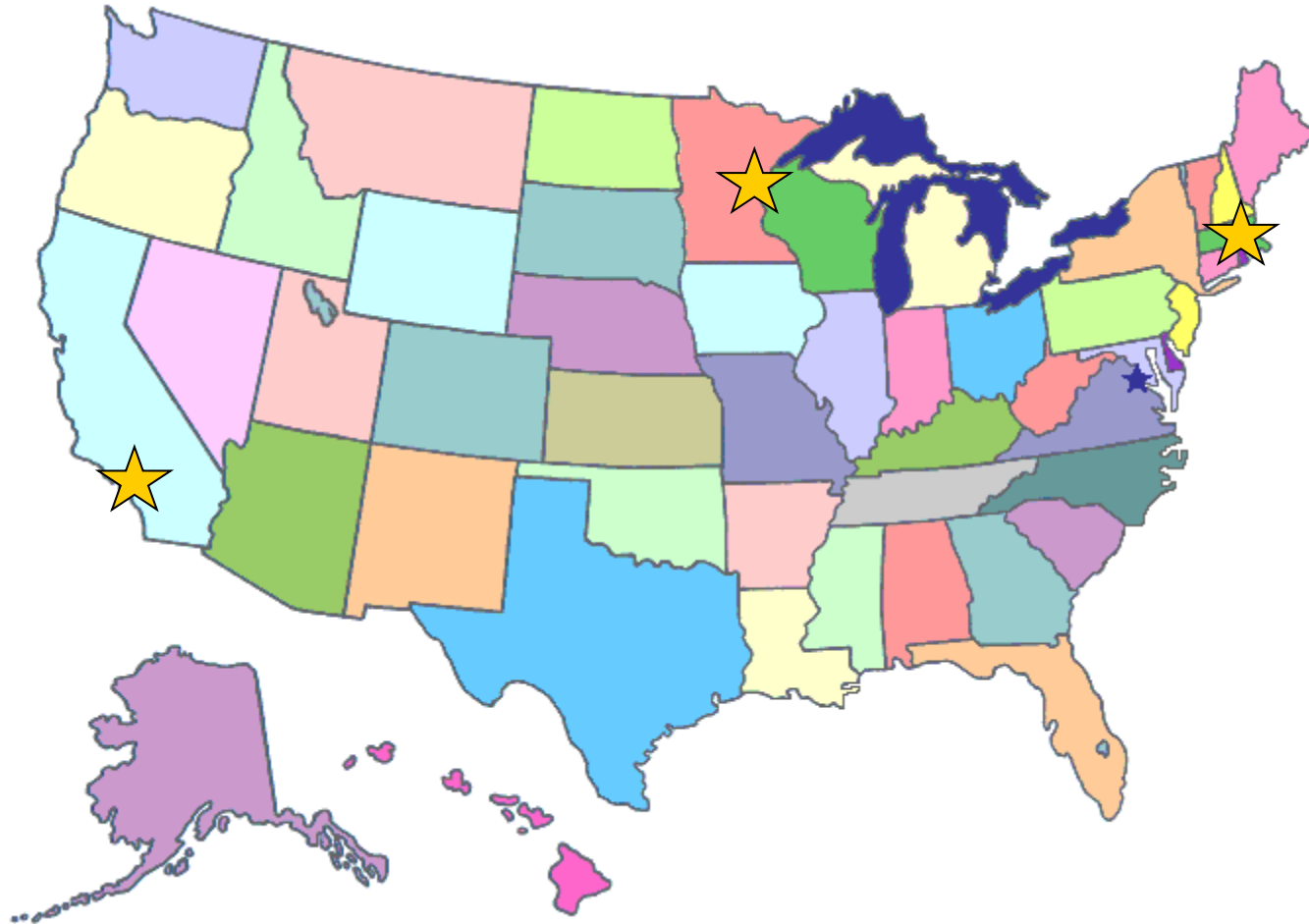
# Name Change!

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# Center for Heart Care Update

PHOENIX CHILDREN'S HOSPITAL

# ACHD Centers in the U.S. -- 1990s

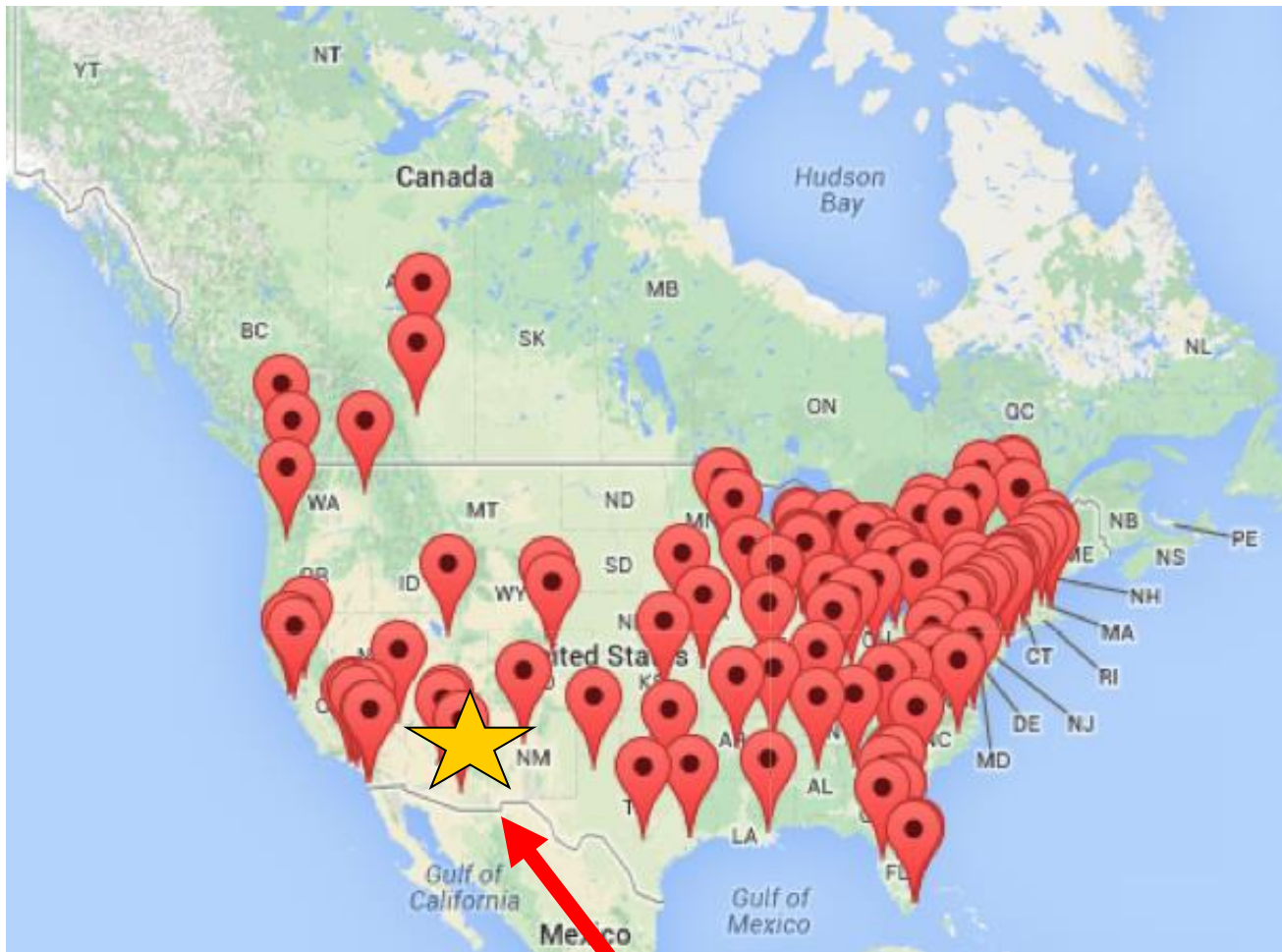


# Overview

- Congenital heart disease - Epidemiology
- Adult Congenital Heart Association
  - Accreditation = Quality
- Top 10 Things You Need to Know about ACHD
  - Case examples
  - Rationale
  - CHD History
  - Evidence → 2018 ACHD Guidelines from ACC/AHA



# ACHD Centers in the U.S. -- 2021





# Adult Congenital Heart Association





# Adult Congenital Heart Association



Accredited - 2020



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Children's**



**Dignity Health**<sup>TM</sup>  
St. Joseph's Medical Center

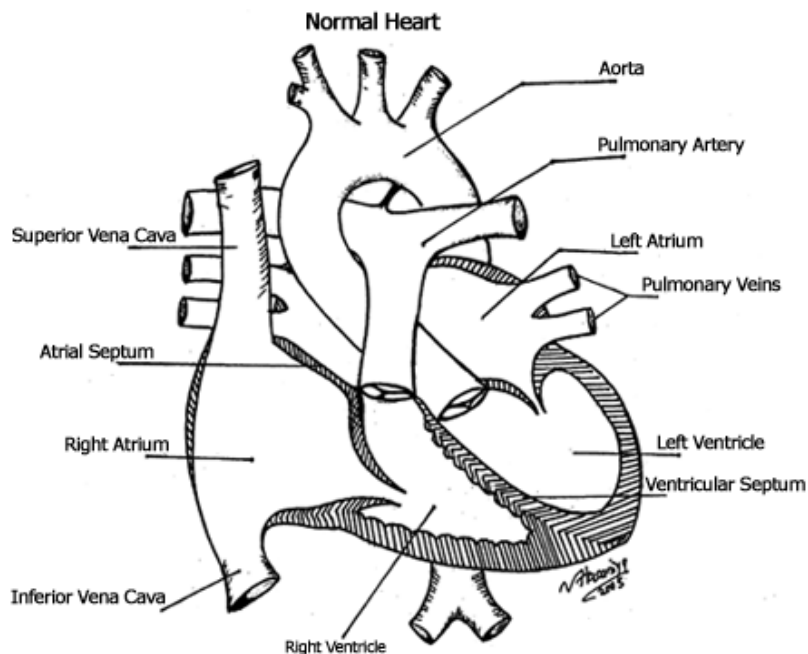


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# What is *Congenital Heart Disease*?

## More than a simple “birth defect” ...



Cardiac Catheterization Laboratories

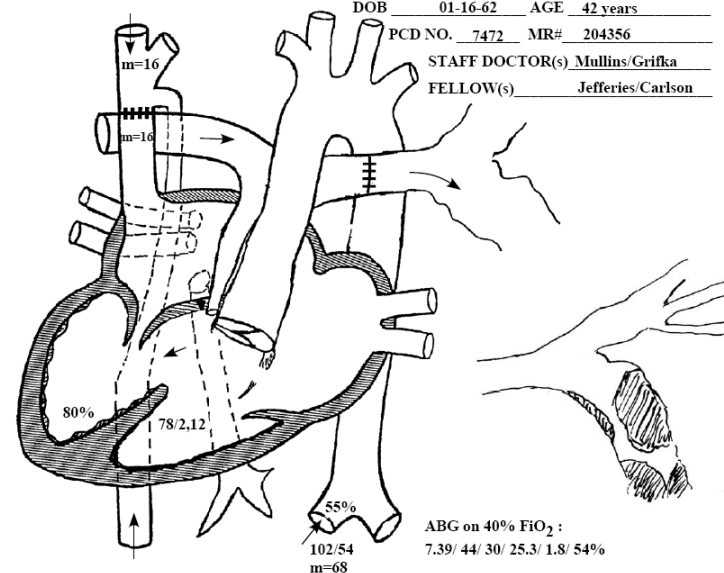
CATH NO. C040989 DATE 12-29-04

DOB 01-16-62 AGE 42 years

PCD NO. 7472 MR# 204356

STAFF DOCTOR(s) Mullins/Grifka

FELLOW(s) Jefferies/Carlson



Ht 179 cm Wt 59.1 kg BSA 1.75 m<sup>2</sup> Hgb 10.5 Hct      %

### DIAGNOSIS:

1. Dextrocardia with situs solitus of the viscera and the atria.
2. Ventricular inversion with transposition of the great arteries.
3. Absence of suprarenal inferior vena cava with azygous continuation to the superior vena cava.
4. Partial anomalous venous return with pulmonary veins to the right atrium.
5. Hepatic veins to right atrium.
6. Atrial septal defect.
7. Status post takedown of Pott's shunt.
8. Status post takedown of central aortopulmonary shunt.
9. Status post atrial septectomy.
10. Status post left pulmonary artery repair.
11. Status post right cavopulmonary anastomosis - Kawashima connection.
12. Status post amputation of left atrial appendage.
13. Extensive thrombus in distal left pulmonary artery segments.

# Congenital Heart Disease: Not JUST children!



# ACHD professional athletes...



**Shaun White**



**Erik Compton**



# Patient, Erik Compton in 2008: 3 months after 2<sup>nd</sup> heart transplant





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# Famous People with CHD



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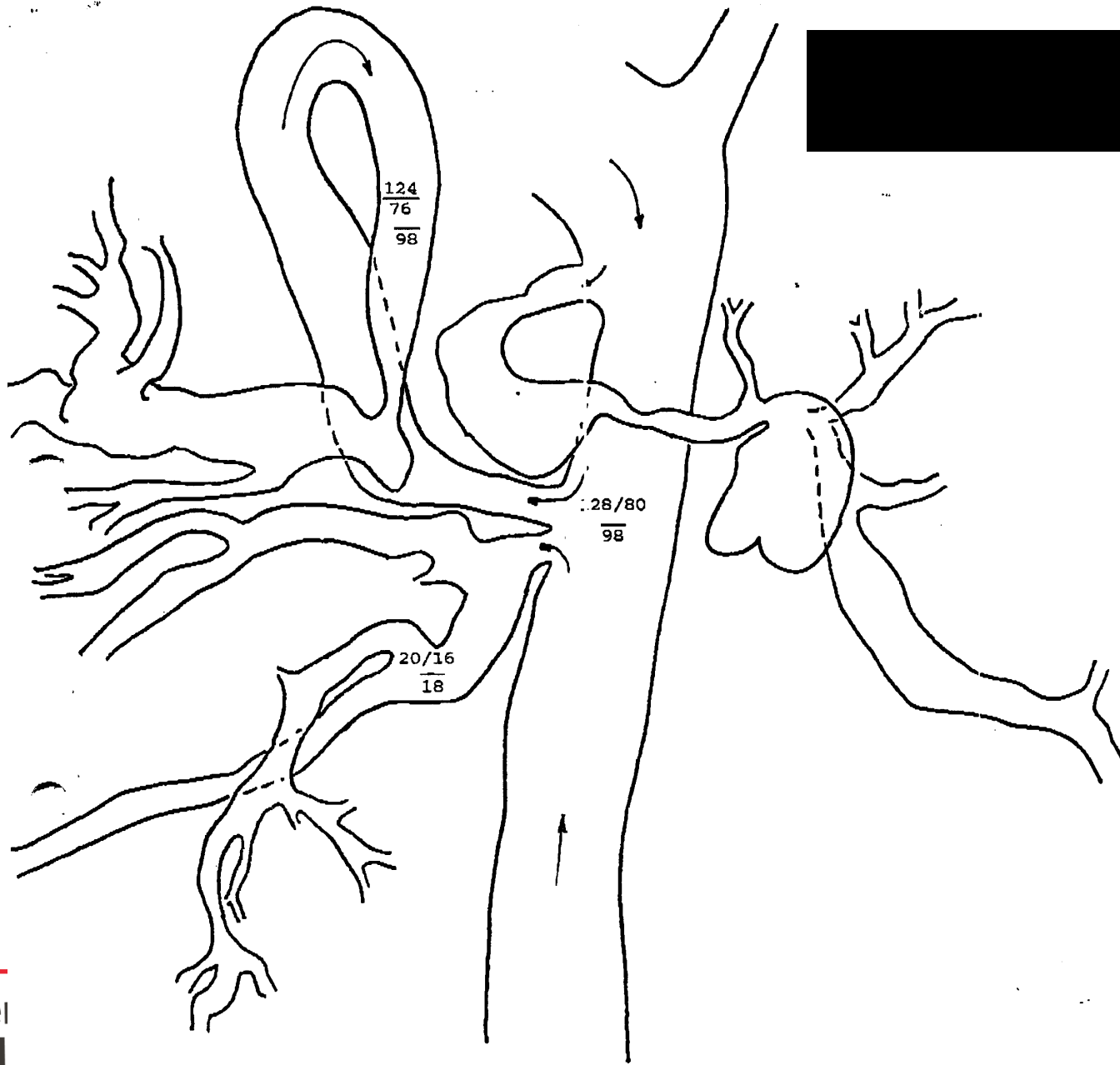
# Famous People with CHD

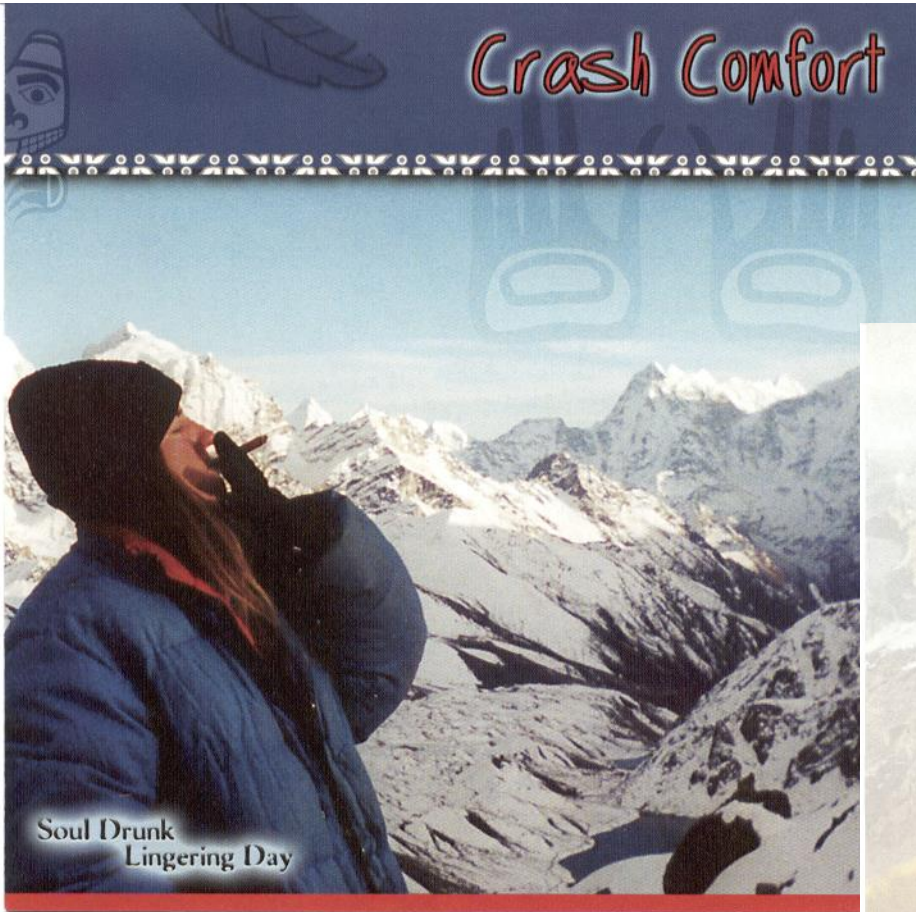


# Famous People with CHD









Works at a bank part-time and...

Soul Drunk

**THANKS FOR KEEPING ME GOING! TAKE CARE! '02**

You move too fast  
thoughts can't compete  
hardly time for  
lying in the shade

I'm into dark  
feeling out of reach  
one last try  
a passing proclamation with you

Feeling that I'm simply not real  
feeling all your 100 hesitations girl

It gets too dark  
feeling out of reach  
simple as a single line of truth from you

Surrendered so long  
before a child  
I'm watching all of your wheels  
move before mine

I can not slow  
(yes, love goes)  
I wish you'd show  
(yes, love goes)

I'm waiting now you feel  
So I can redo and repeat  
Soul Drunk Lingering Day

Keep a love of faith  
or just move on  
hardly time for

wasting younger days

Can I get that high  
Rising above the flame  
on  
in passing and the gifts so rare

Feeling that I'm living every day for  
Feeling all your 100 hesitations girl

It gets too dark  
feeling out of reach  
simple as a single line of truth from you

Old man sittin'  
talking about love  
About the ones  
that made him high  
So high have he'd reach back down  
and kick up to the ground --

Drifting so long in you I have found  
hope to watch you spin  
Kick up the clouds

I can not slow  
(yes, love goes)  
I wish you'd show  
(yes, love goes)

I'm waiting now you feel  
So I can redo and repeat  
Soul Drunk Lingering Day

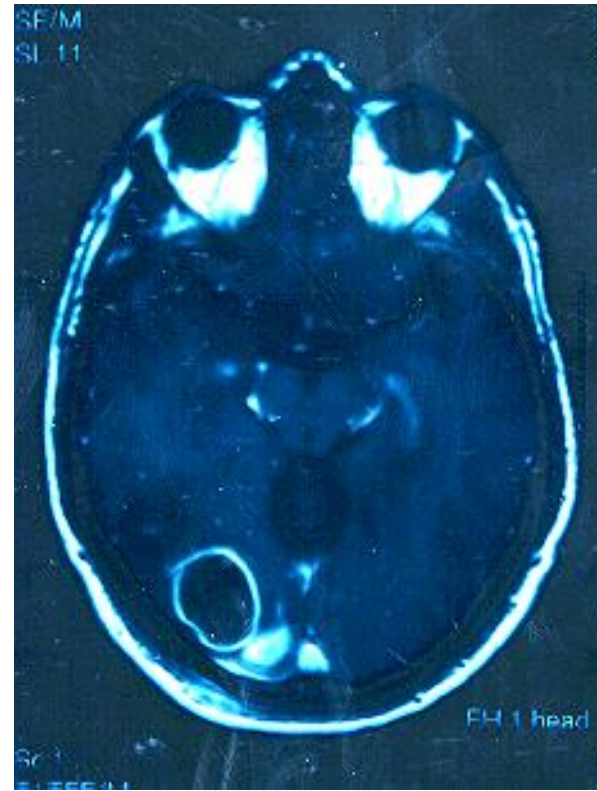
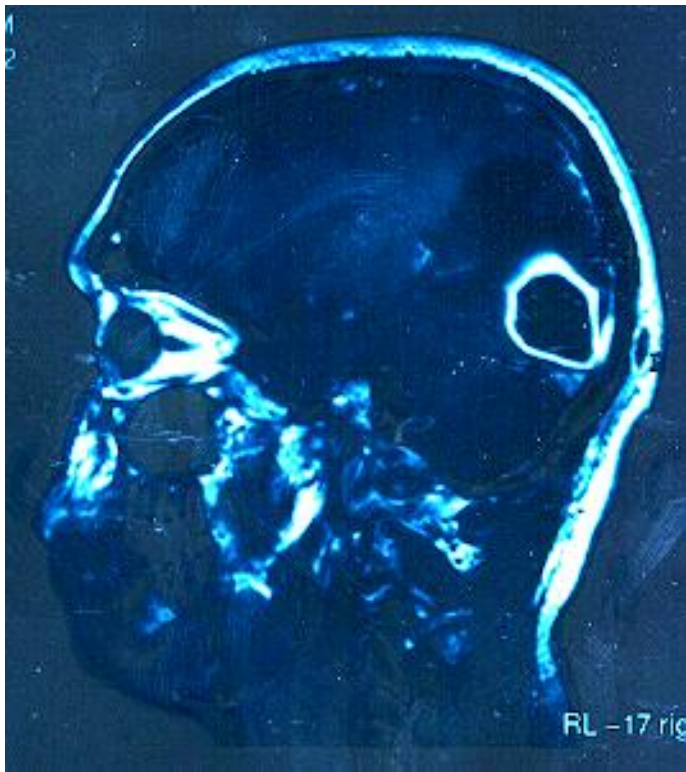
(All Lyrics can be found at [crashcomfort.com](http://crashcomfort.com))

Montrose Records, 2001

# Case: 34 y/o male

- Arose in AM and “forgot” how to blow nose.
- Expressive aphasia.
- Exam: RA Sat 82%
  - CV: clubbing, active precordium, single S2, III/VI HSM at LLSB
  - Neuro: unremarkable
- ***Labs: HCT 67%***
- Head MRI was ordered...

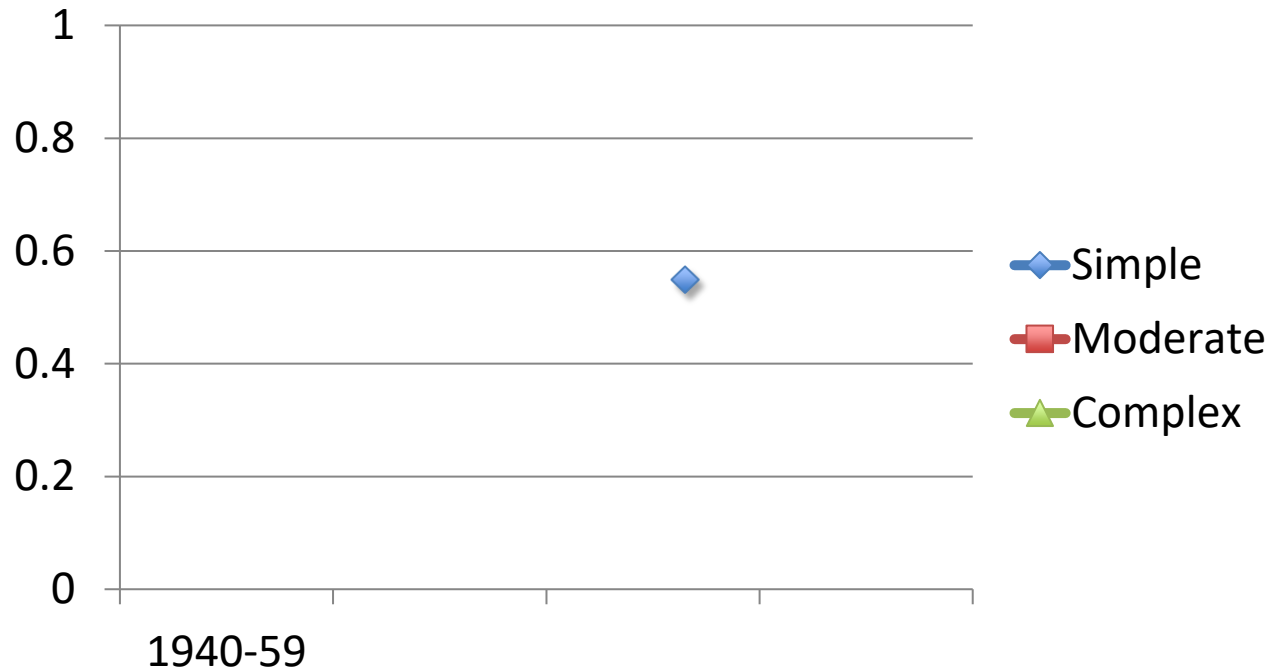
# Case: 34 y/o male with cyanotic CHD, headache, neuro sx



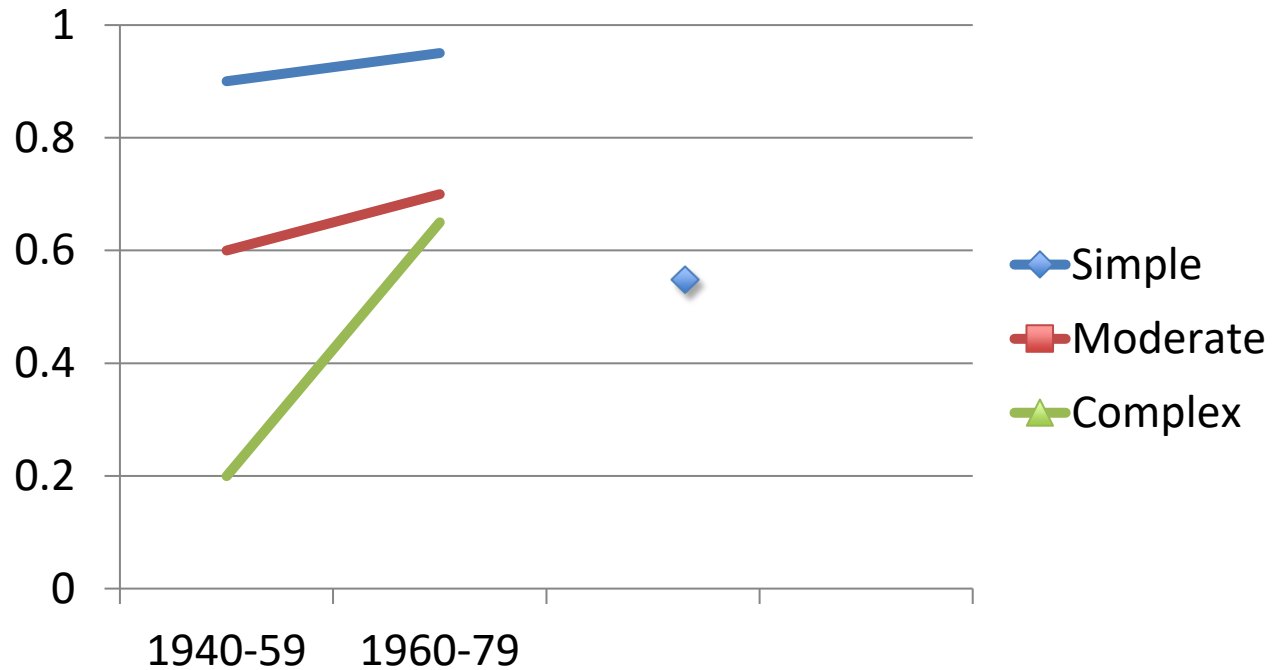
***#1:*** Congenital heart disease (CHD) patients survive until adulthood and more Adult CHD pts are coming.



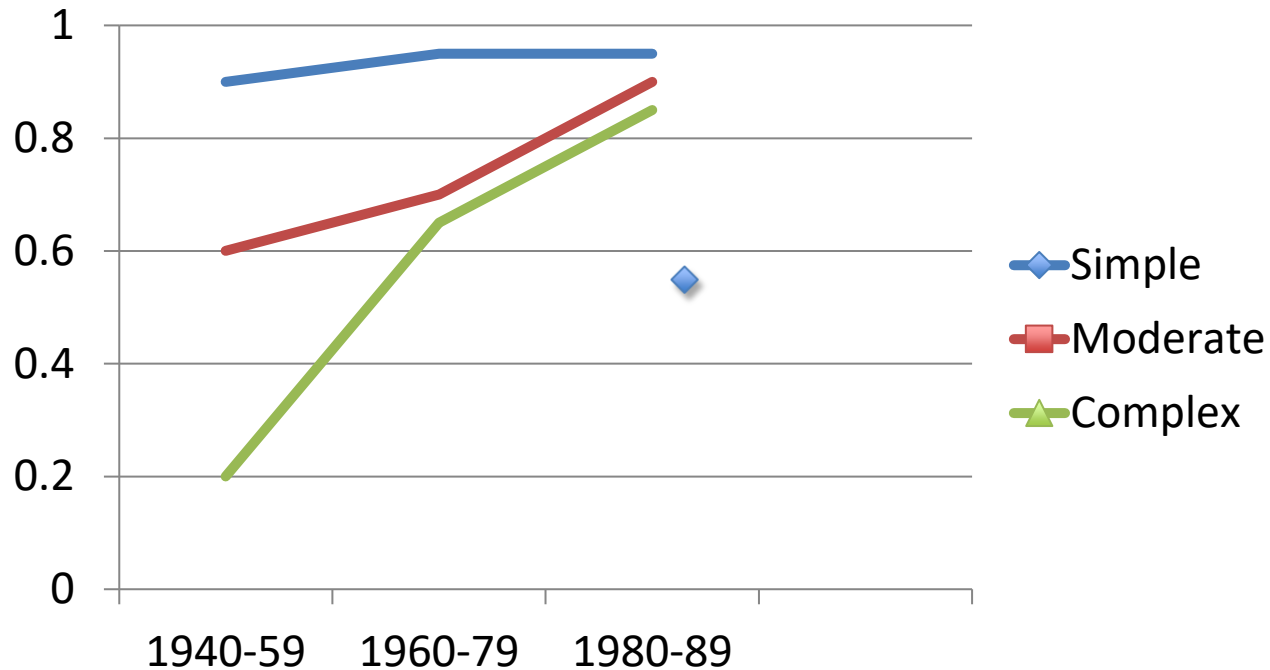
# Survival to 1 Year of Life



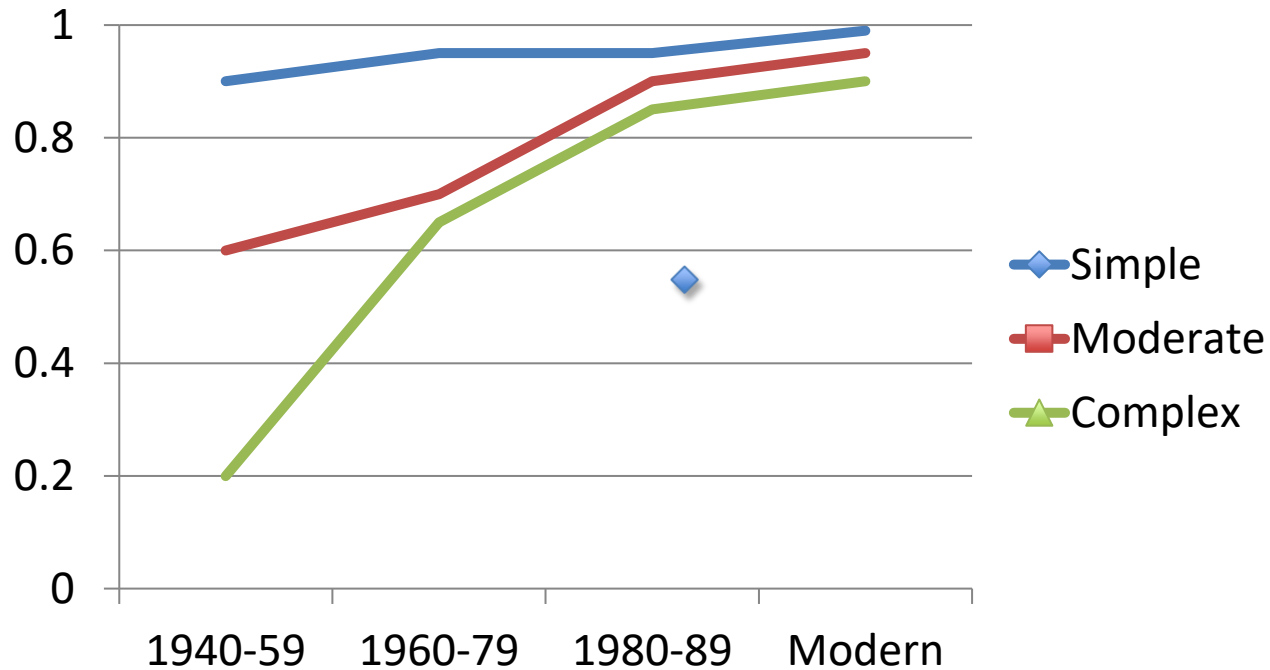
# Survival to 1 Year of Life



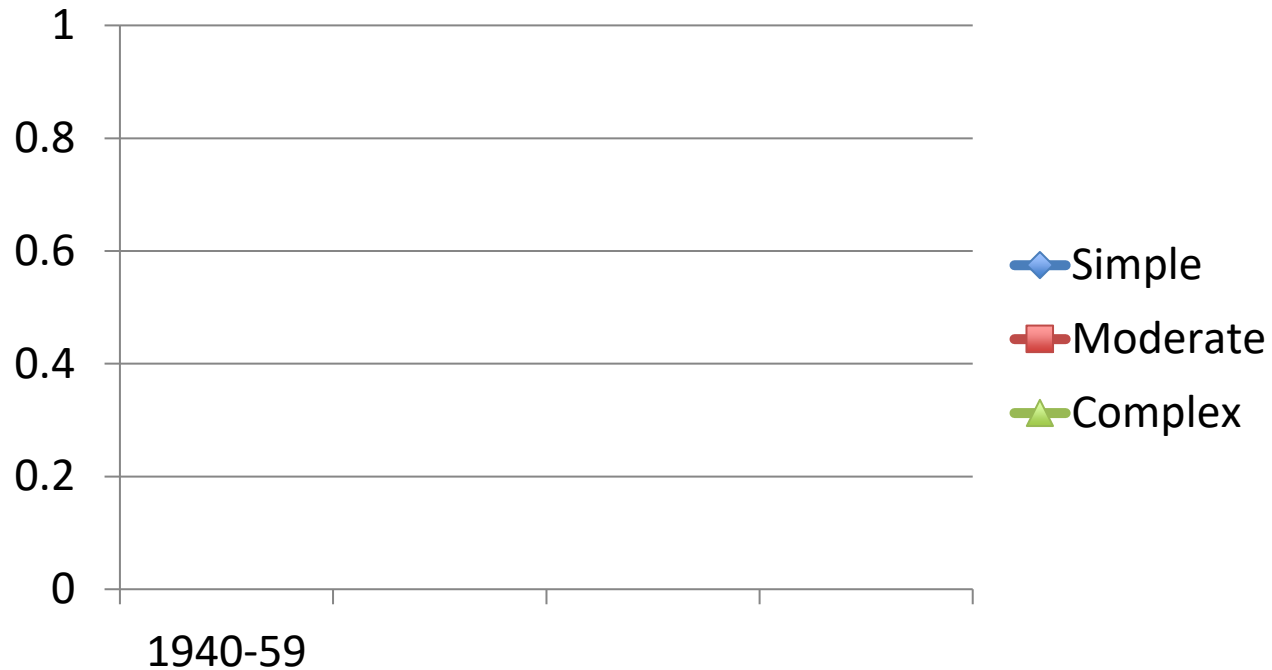
# Survival to 1 Year of Life



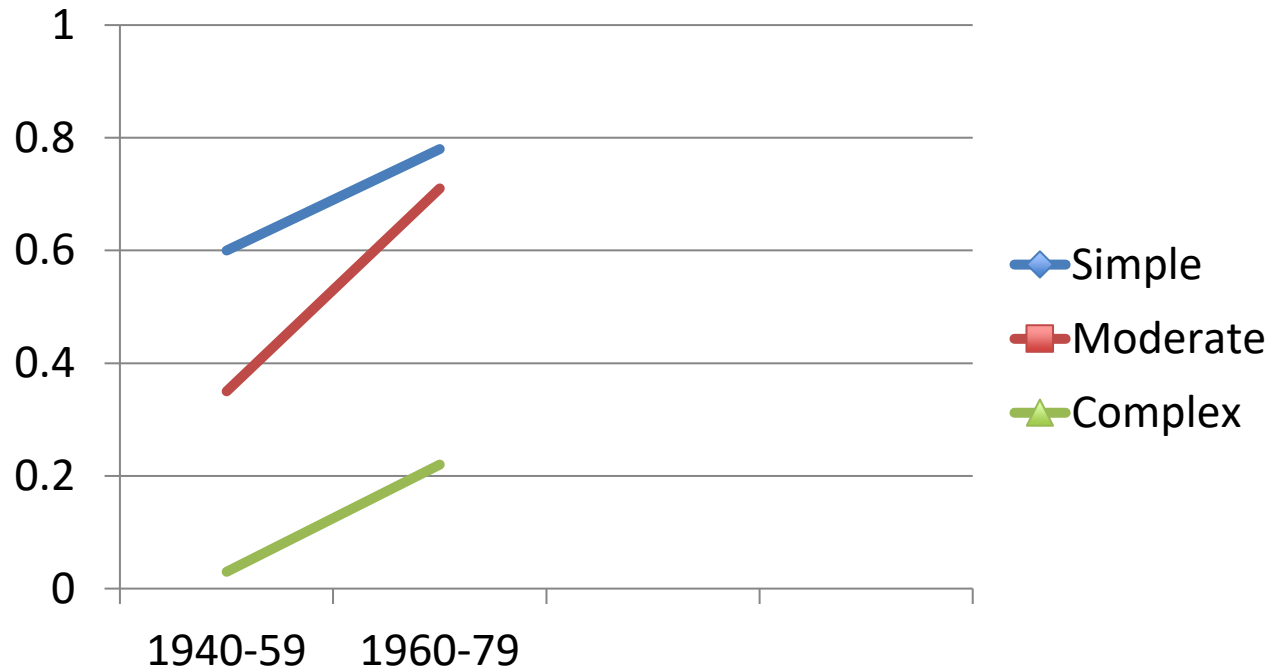
# Survival to 1 Year of Life



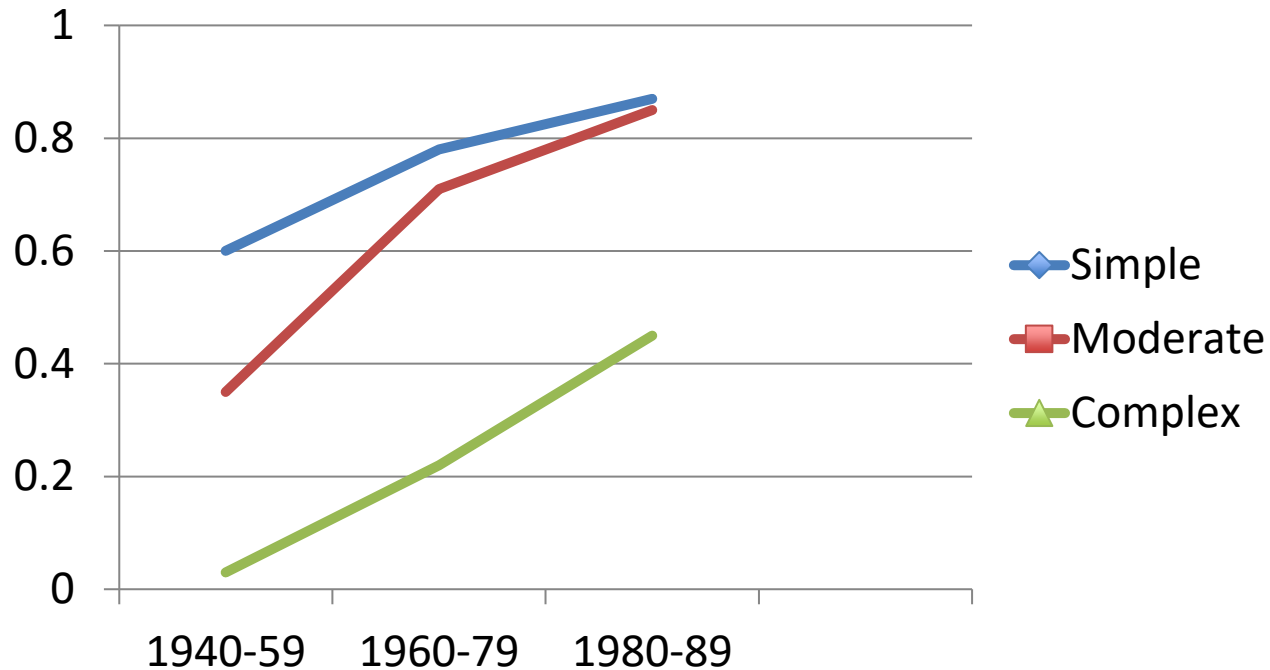
# Survival to Adulthood



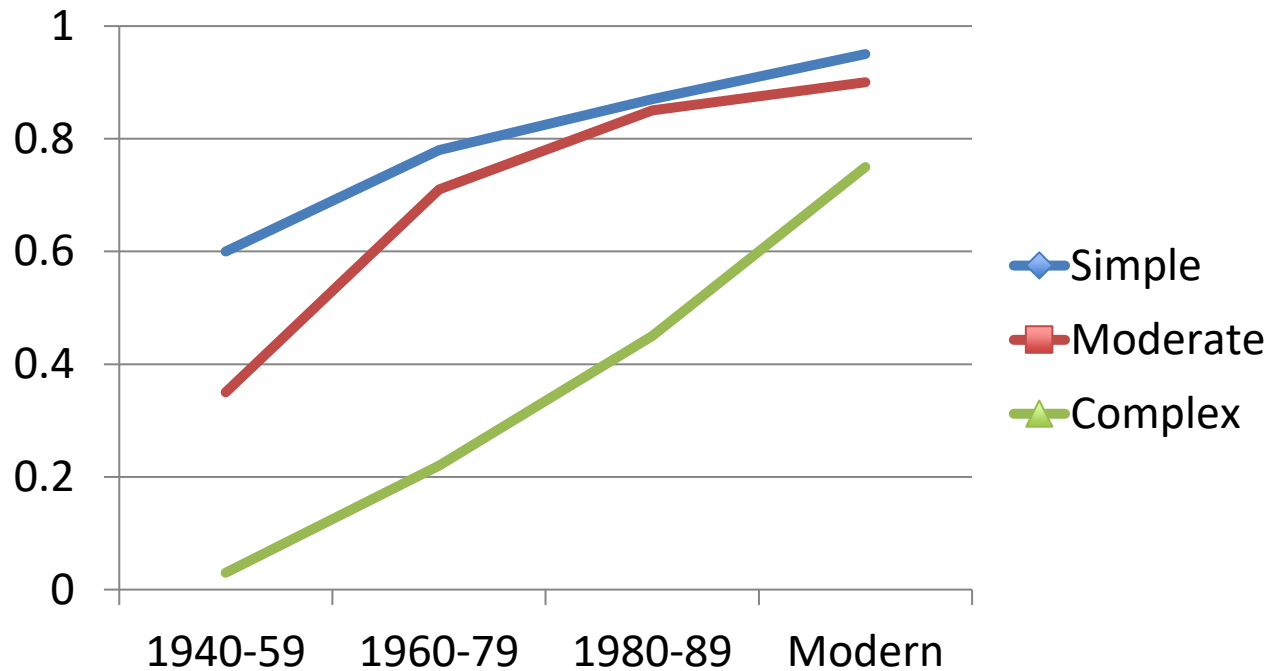
# Survival to Adulthood



# Survival to Adulthood



# Survival to Adulthood



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## **STATE-OF-THE-ART PAPER**

# The Adult With Congenital Heart Disease

Born to Be Bad?

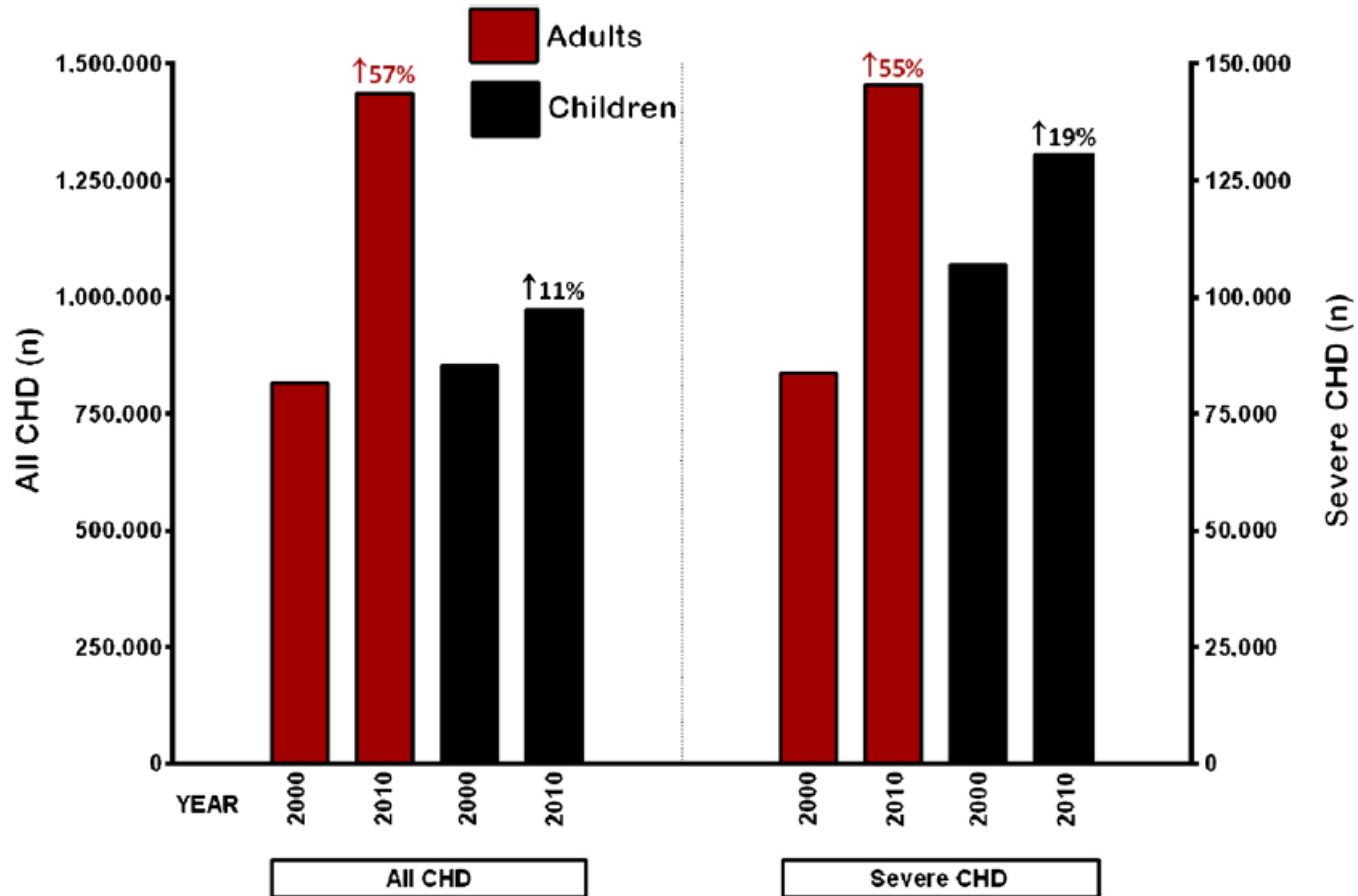
Carole A. Warnes, MD, MRCP, FACC

*Rochester, Minnesota*

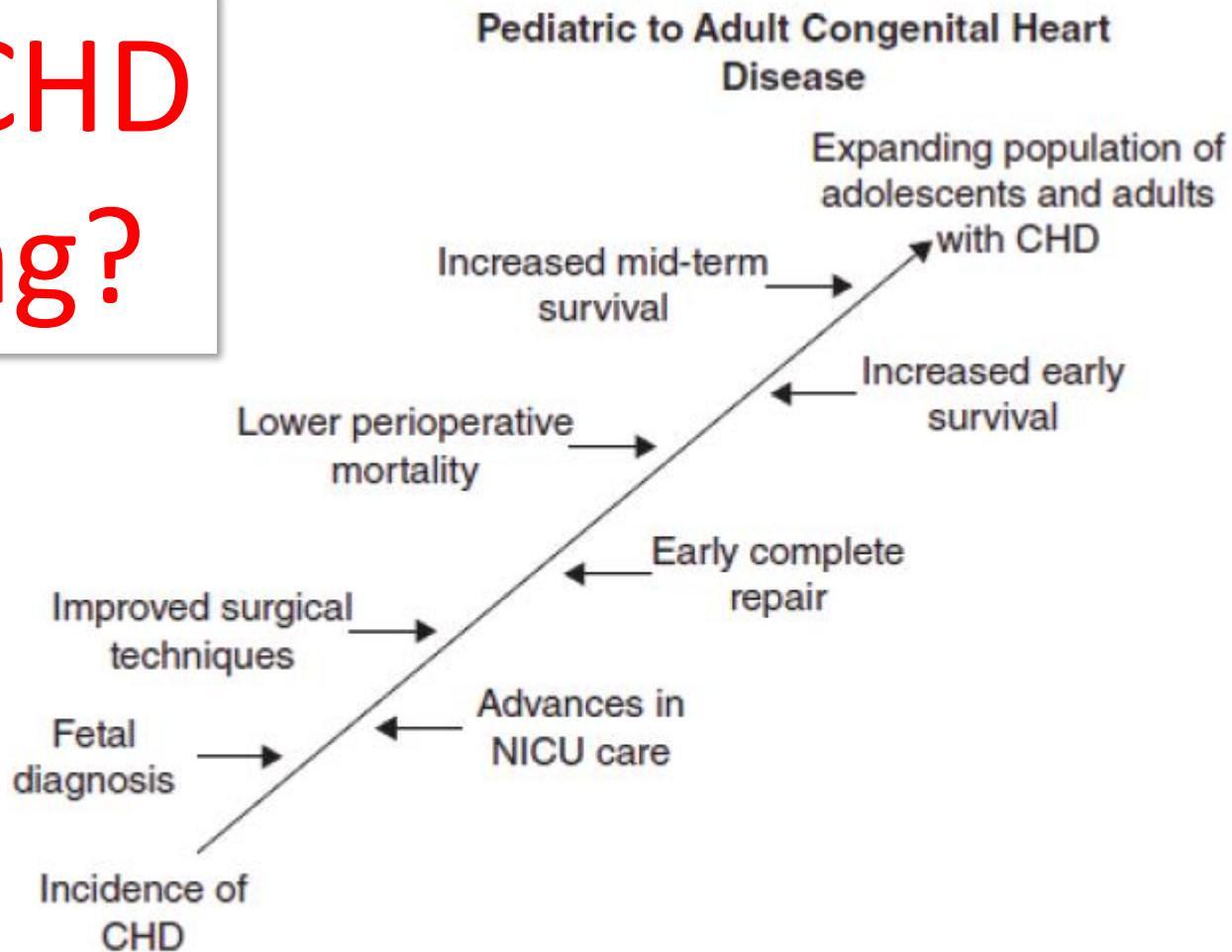


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# Adults v Children with CHD



# Why is ACHD Increasing?



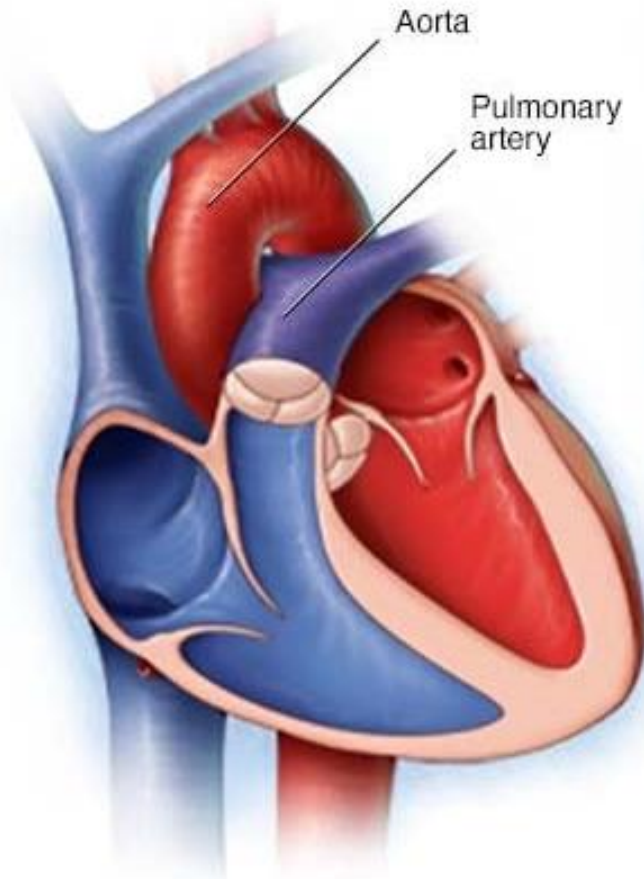
Source: Curt J. Daniels, Ali N. Zaidi  
Color Atlas and Synopsis of Adult Congenital Heart Disease  
[www.cardiology.mhmedical.com](http://www.cardiology.mhmedical.com)  
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# #2: Arrhythmias are common!

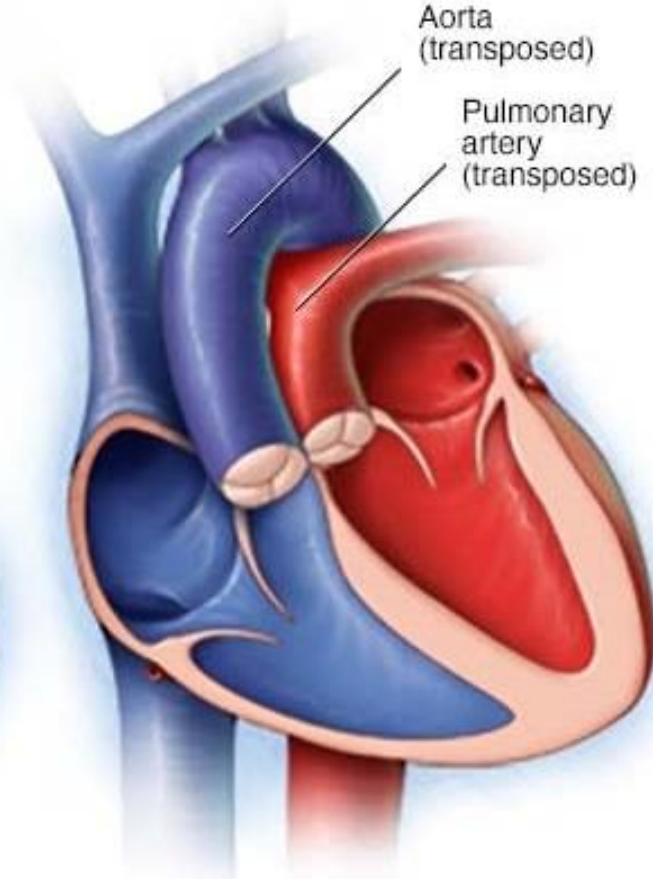
Arrhythmias	Associated Defects
<b>Tachycardias</b>	
Accessory pathways	Ebstein's anomaly; L-TGA
Twin AV nodes	Heterotaxy syndrome
Intra-atrial reentrant tachycardia (atrial flutter)	Postoperative Mustard; postoperative Senning; postoperative Fontan; others
Atrial fibrillation	Mitral valve disease; aortic stenosis; unrepaired single ventricle
Ventricular tachycardia	Tetralogy of Fallot; congenital aortic stenosis; others
<b>Bradycardias</b>	
Congenital sinus node dysfunction	Heterotaxy syndrome
Acquired sinus node dysfunction	Postoperative Mustard; postoperative Senning; postoperative Fontan; postoperative Glenn; others
Congenital AV block	Endocardial cushion defects; L-TGA
Acquired AV block	VSD closure; subaortic stenosis relief; AV valve replacement

# D-Transposition of the Great Arteries

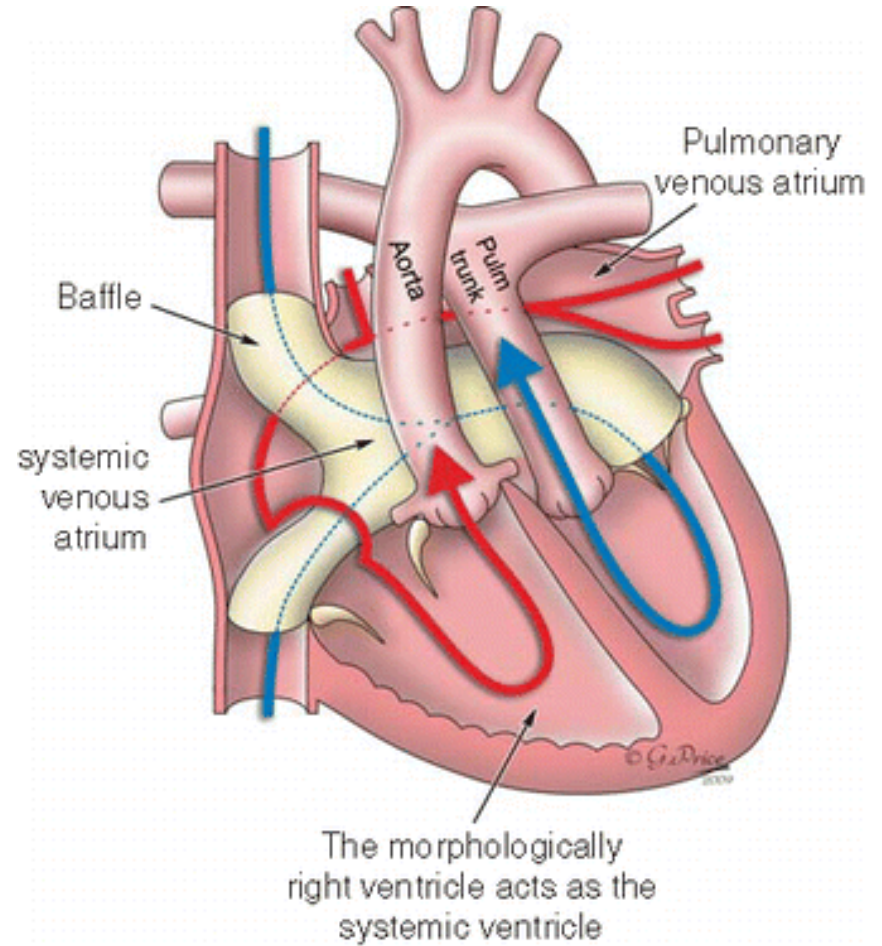
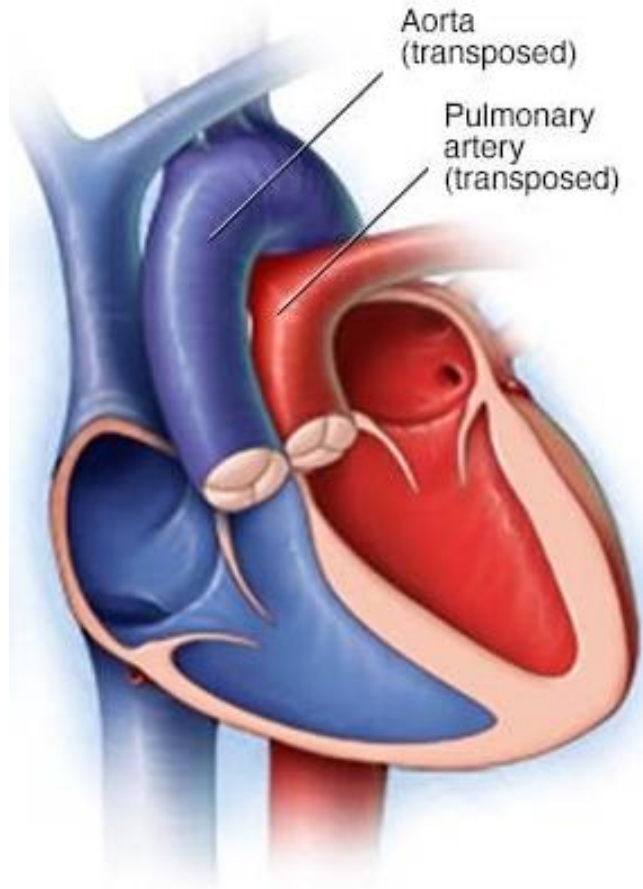
Typical heart



Heart with transposition of the great arteries



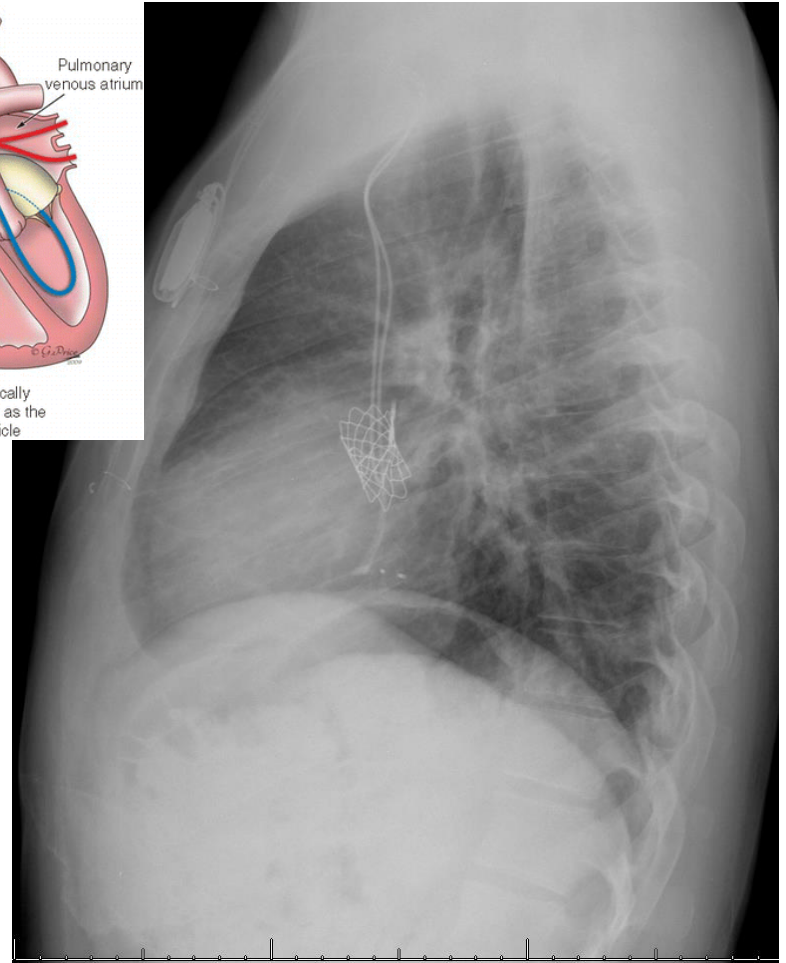
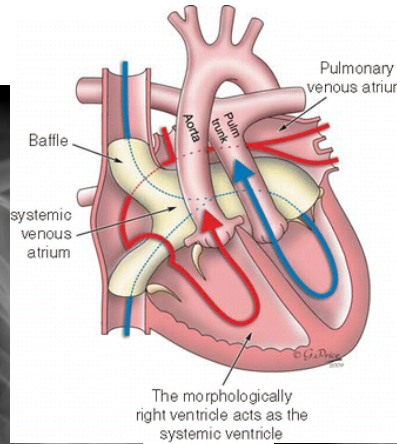
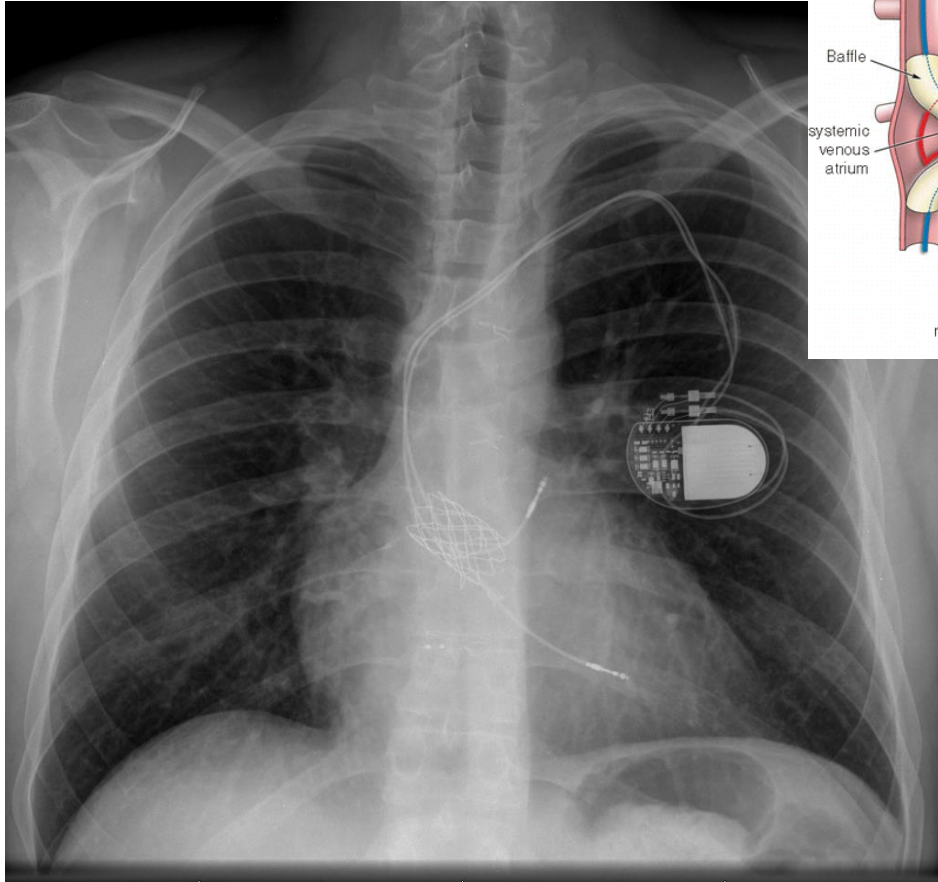
# Atrial Switch Operation for D-TGA (Mustard or Senning)



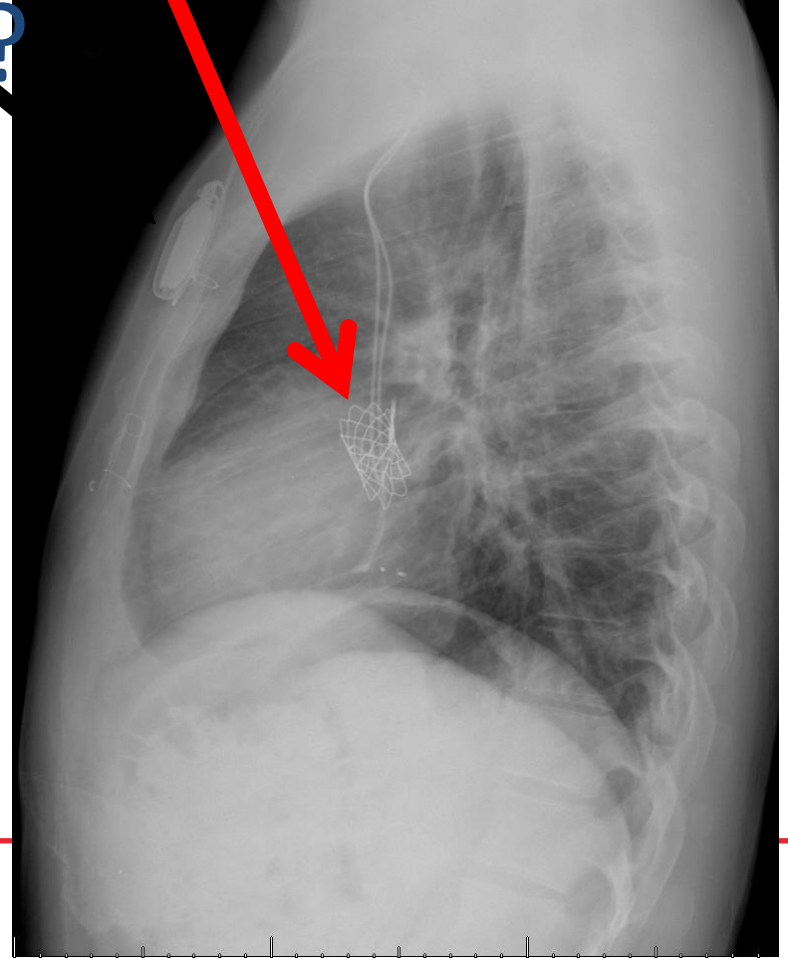
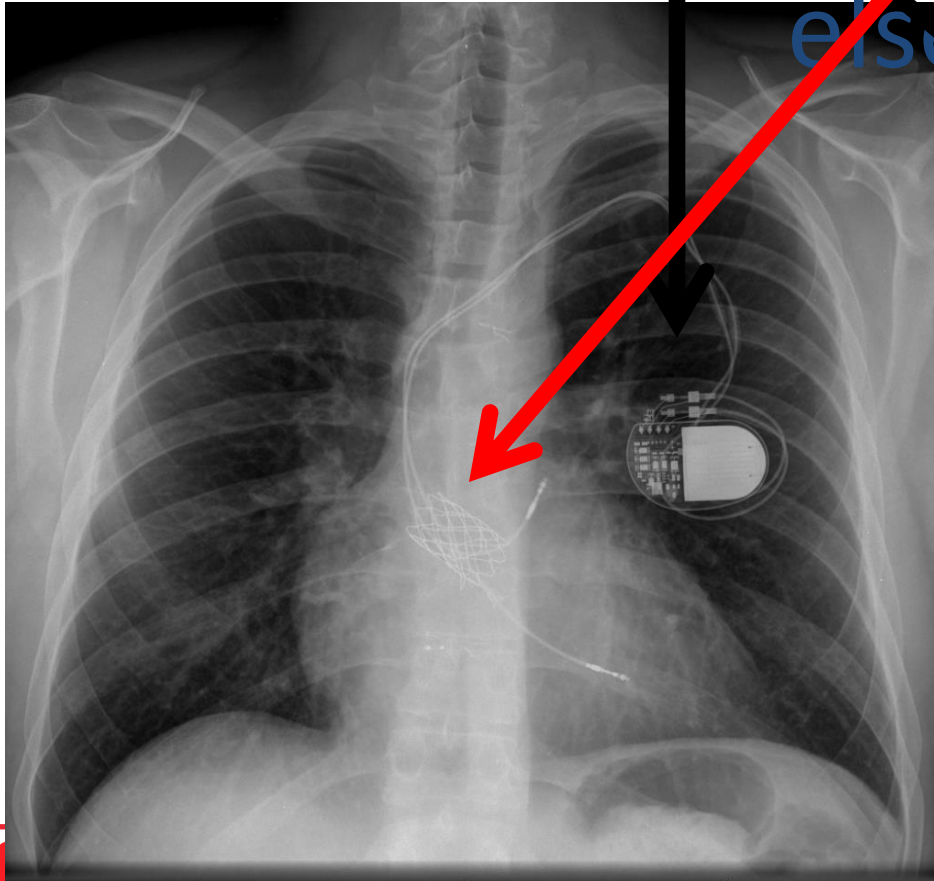
# Special Considerations for PM or ICD in ACHD patients

- Congenital venous anomalies
- Acquired venous occlusion
- Inability to access chamber
- Surgical obstacles (patches, conduits, baffles)
- Intracardiac shunts
- Ventricular geometry and position

# D-TGA s/p Mustard: Hardware?



# PM ,SVC Stent in a Mustard patient. Anything else?

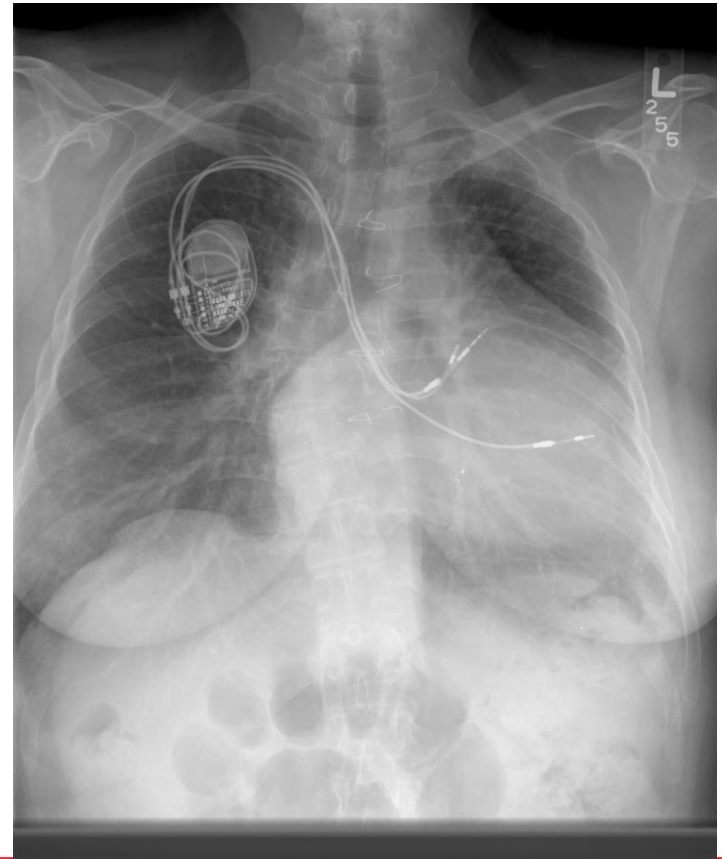
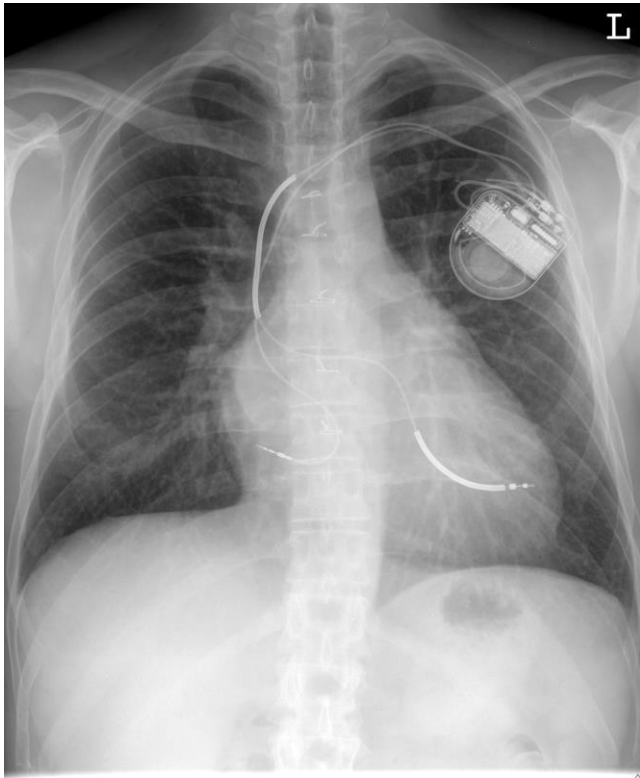


Amplatzer ASD occluder  
for baffle leak



# D-TGA s/p Mustard: Pacer leads

Incorrect                      Correct



# D-TGA s/p Mustard: Pacer leads

Incorrect                      Correct



# After lead removal & placement of epicardial leads

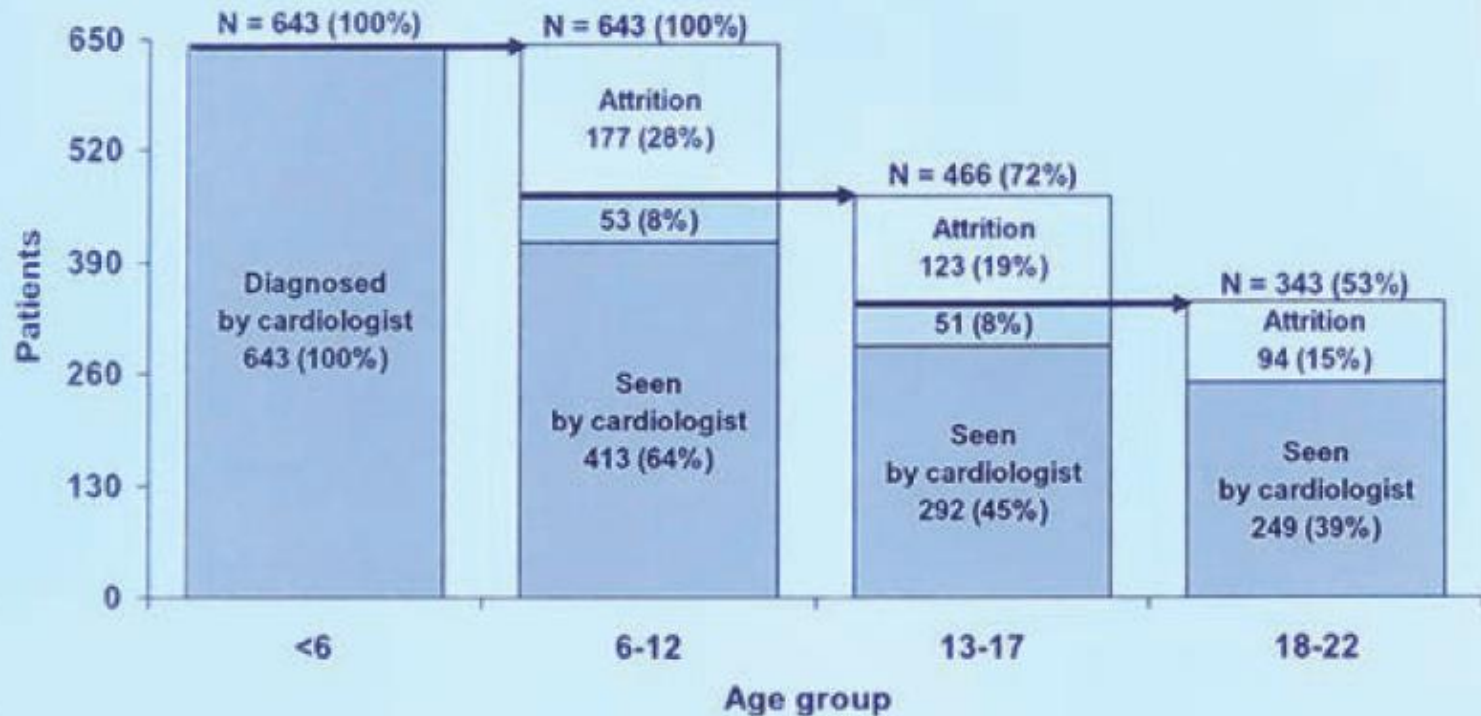


# #3: ACHD pts Get “Lost”

- Competent adult care not available
- Many patients were simply lost to follow-up
- Patients and families not well educated about their conditions, so fell out of care or into unskilled hands
- Caregivers don't know what to expect, or understand their responsibilities



# Chronic Disease Adolescents and Transition to Adult Care



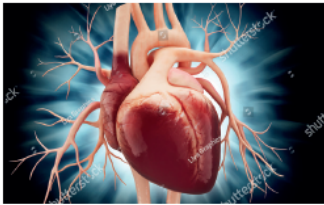
# How Were They Lost?

- Inadequate numbers of ACHD providers
- Inadequate ACHD training opportunities
- Inadequate institutional interest
- Inadequate financial incentives
- Inadequate political and advocacy profile

SECTION

IX

# Unique Issues Regarding the Young Adult with Congenital Heart Disease

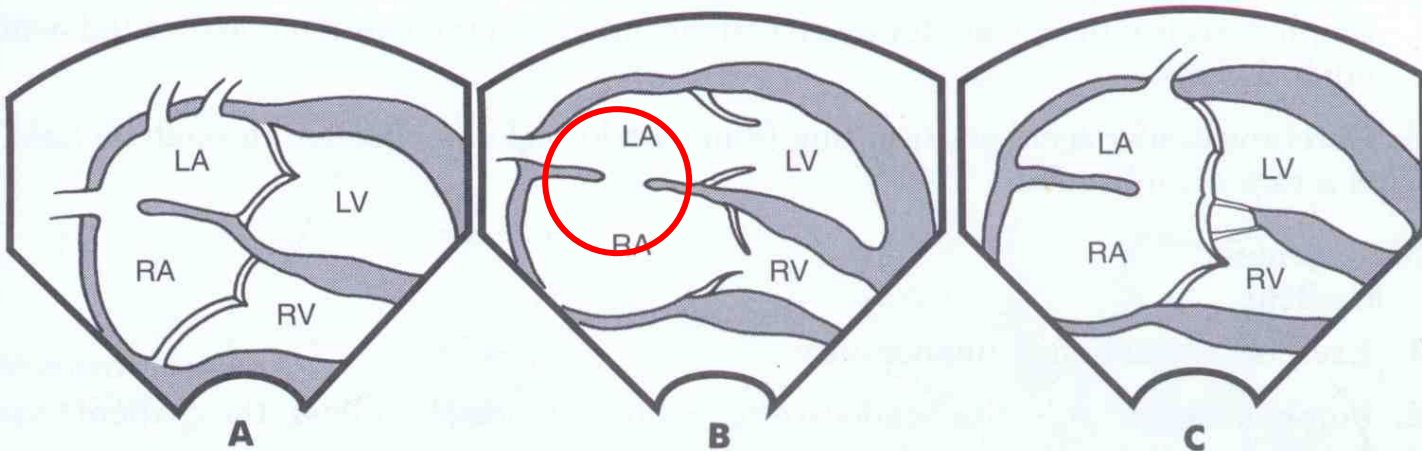
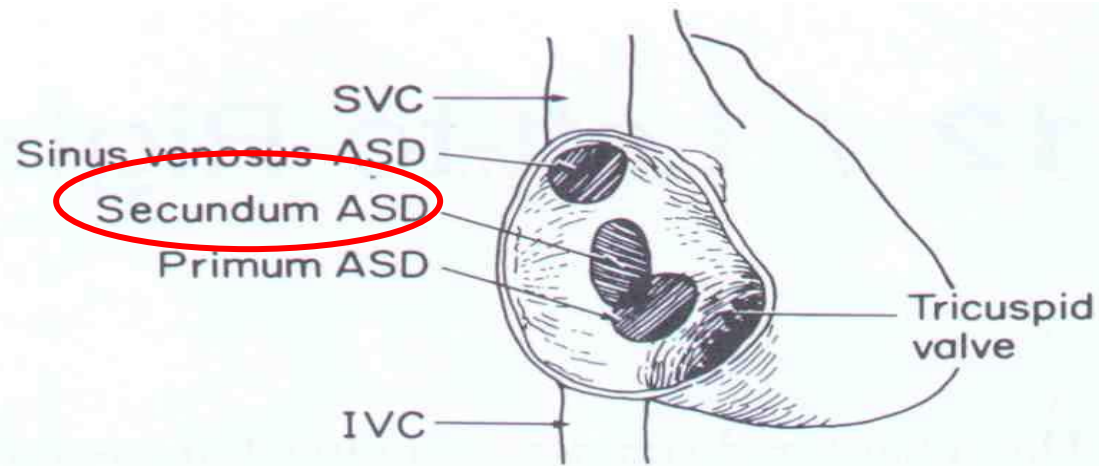


67

## Transition to Adult Care

Jordan D. Awerbach ■ Wayne J. Franklin ■ Peter R. Ermis

# #4: Some of these defects can be closed in the cath lab



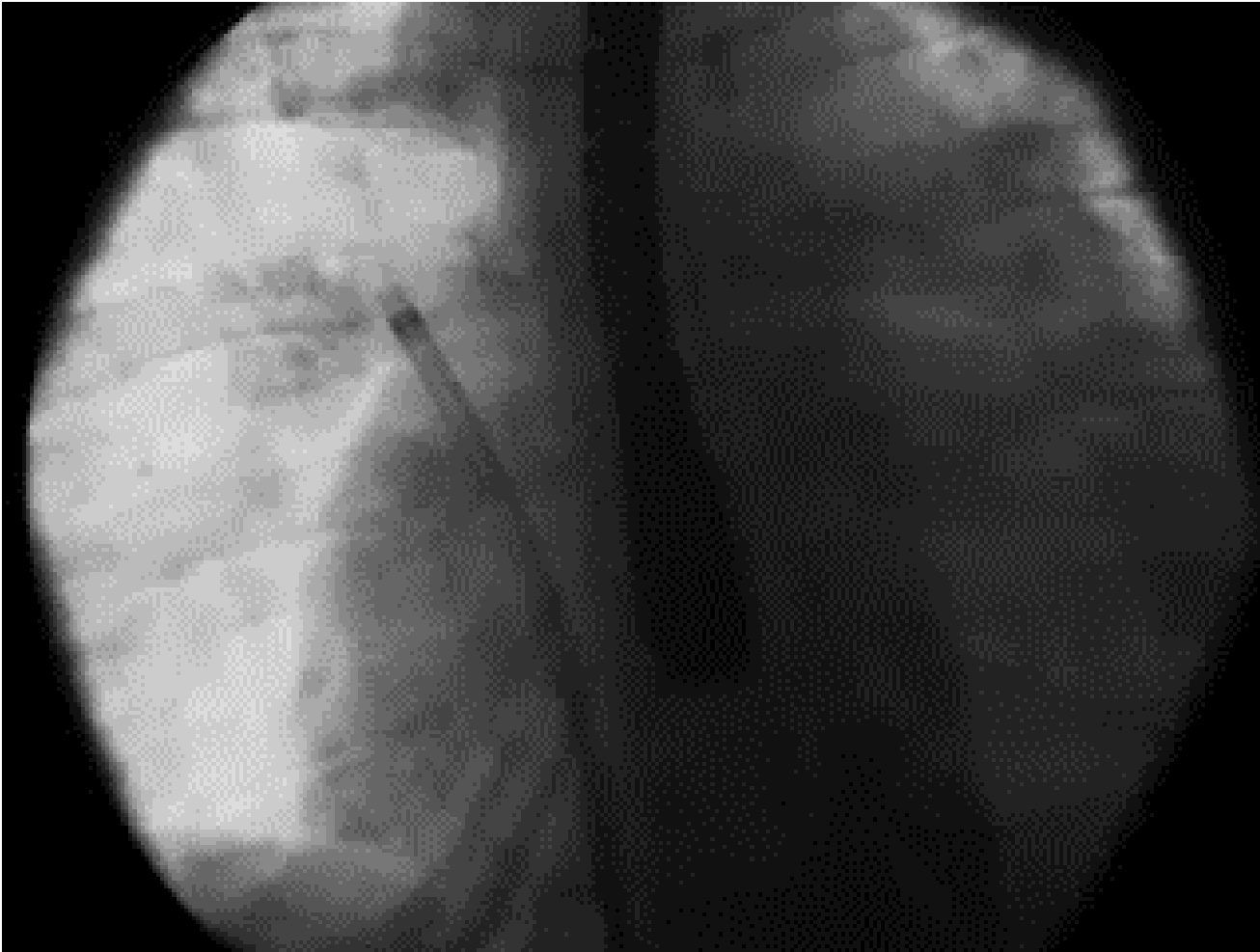
# ASD Closure in the Cath Lab



Phoenix  
Children's

*C. Machado*  
M.D.  
© IBN

# ASD Device Closure



**#5:** These pts are not “fixed” as children

# Types of Surgical Intervention

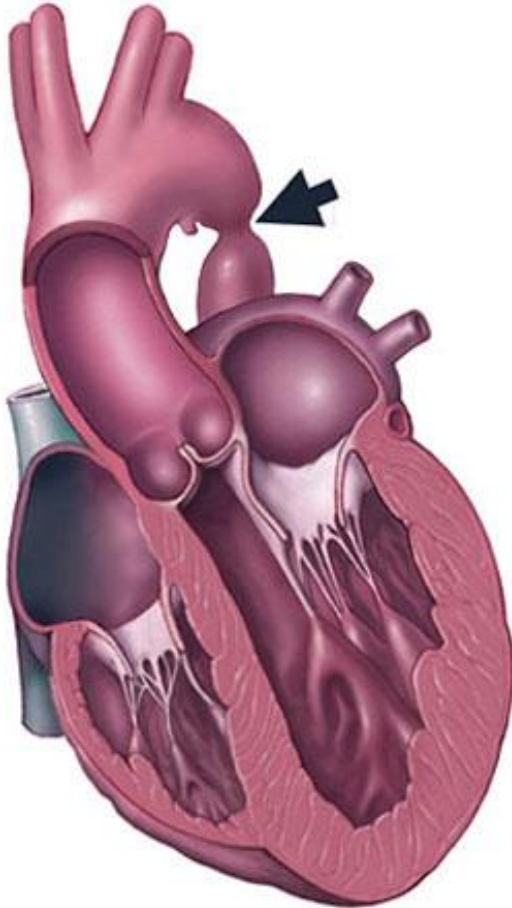
- Palliative
  - Shunt operations that provide intermediate bridge
- Corrective
  - Anatomic: with and without residua
  - Physiologic: to restore proper blood flow
- Curative
  - No residua or sequelae

# Weren't These Patients Fixed as Children?

- Most were left with abnormal circulations
- Abnormal native valves
- Conduits and artificial valves
- Cardiac scars
- Depressed cardiac function

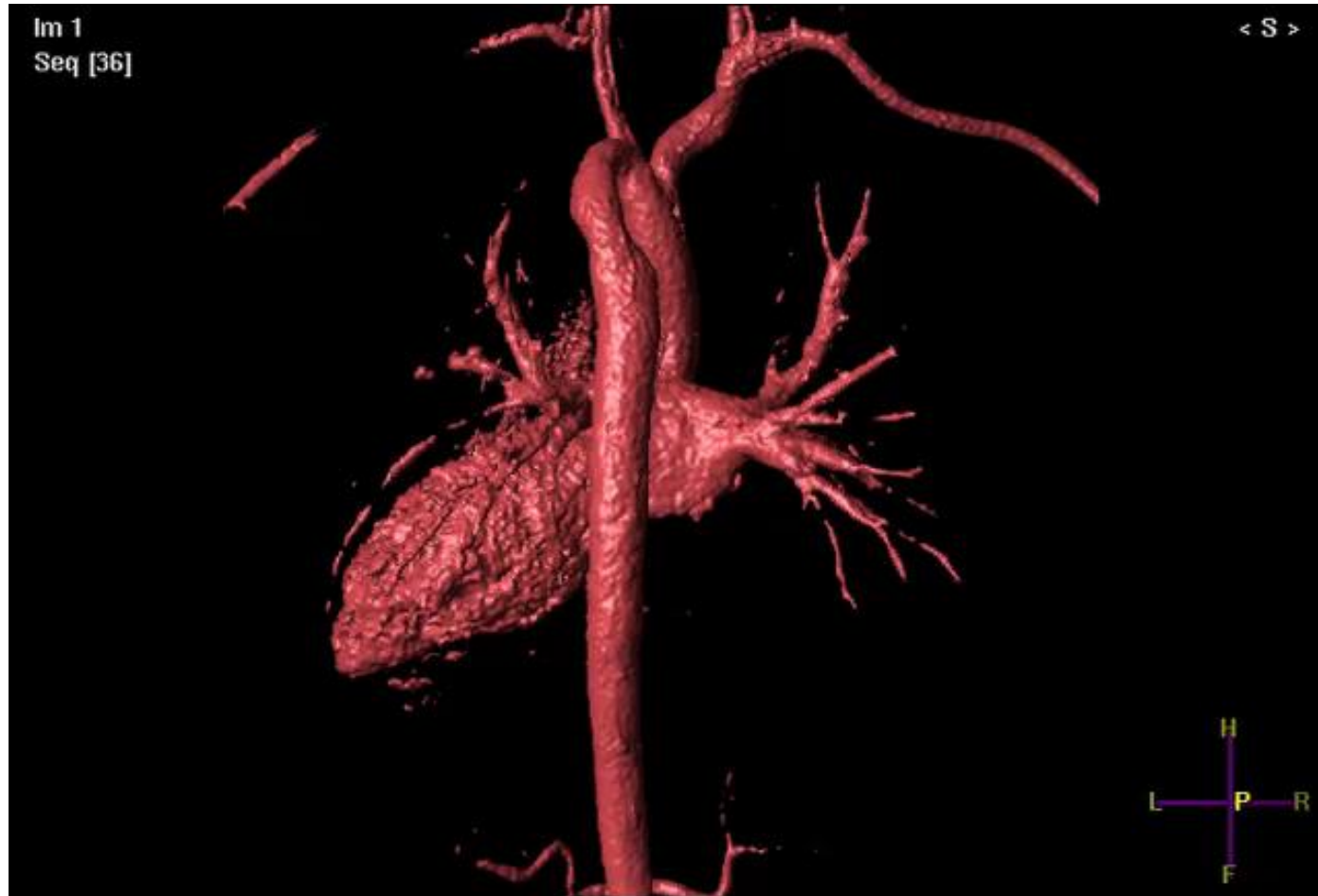


# Coarctation of the Aorta: A Fix is not often “Fixed”



- Definition: simple vs. complex, significant has >20 mmHg gradient

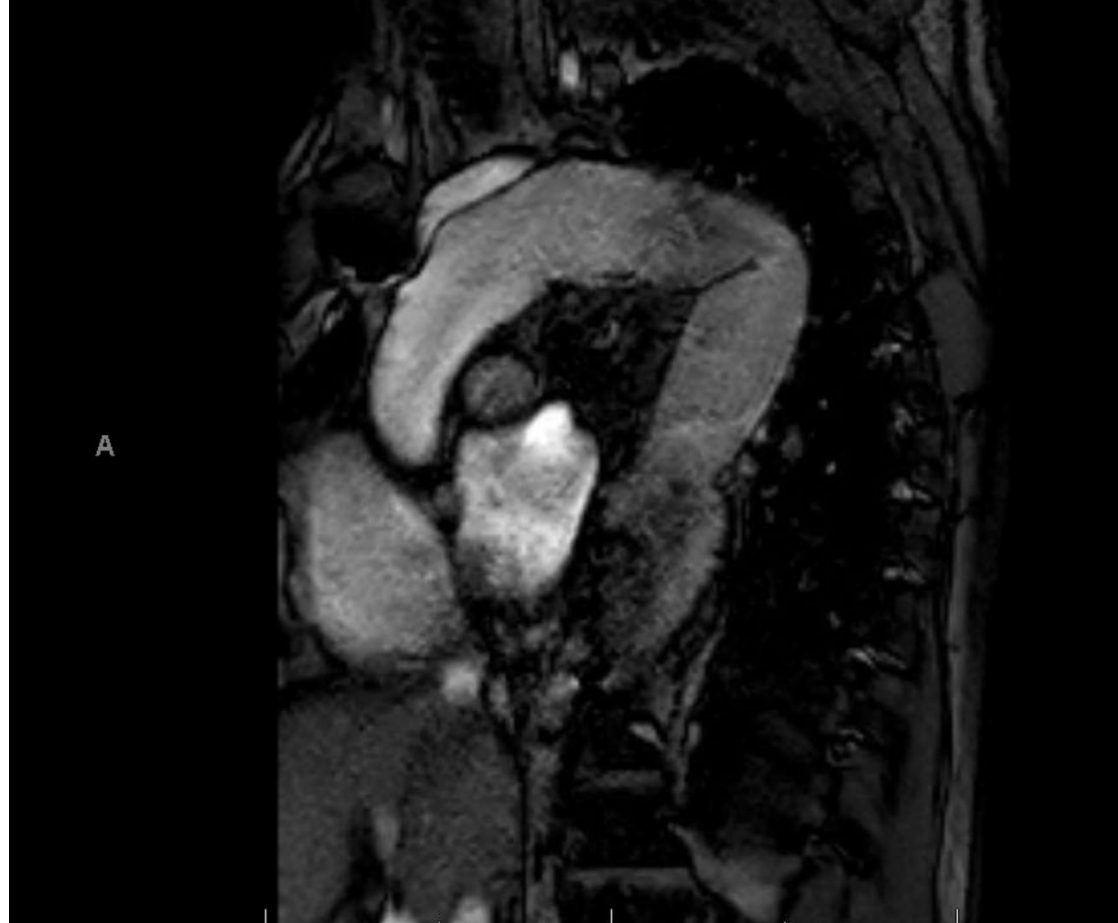
# Coarctation: MRI is best



# Post-op Coarct Repair--Aneurysm

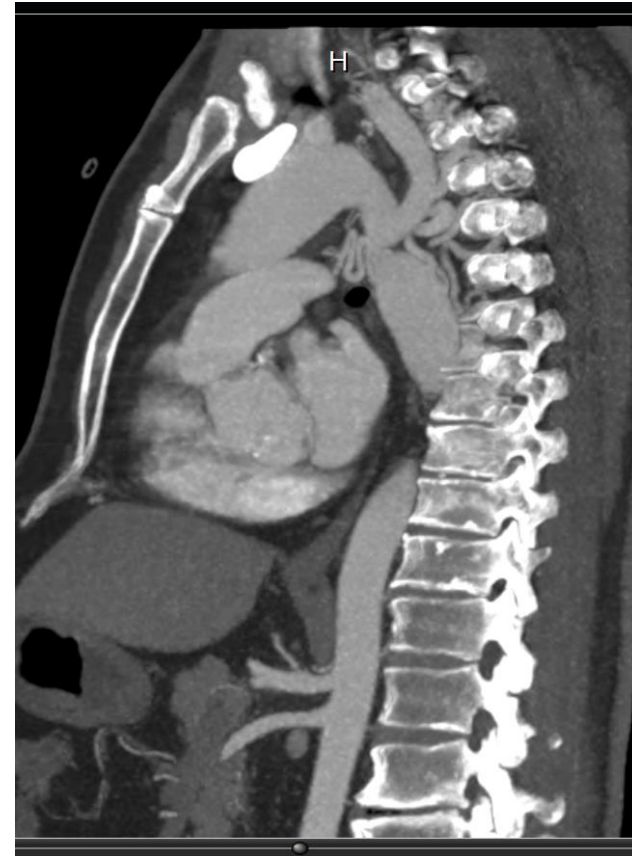


# Post-op Coarct Repair—Redo Repair

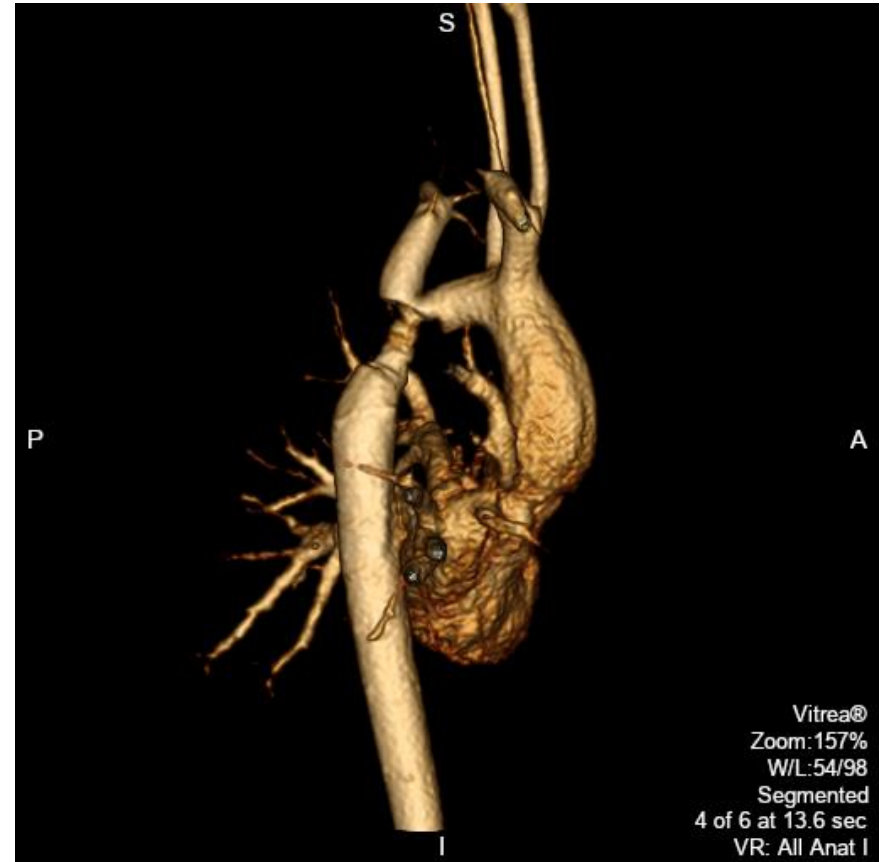
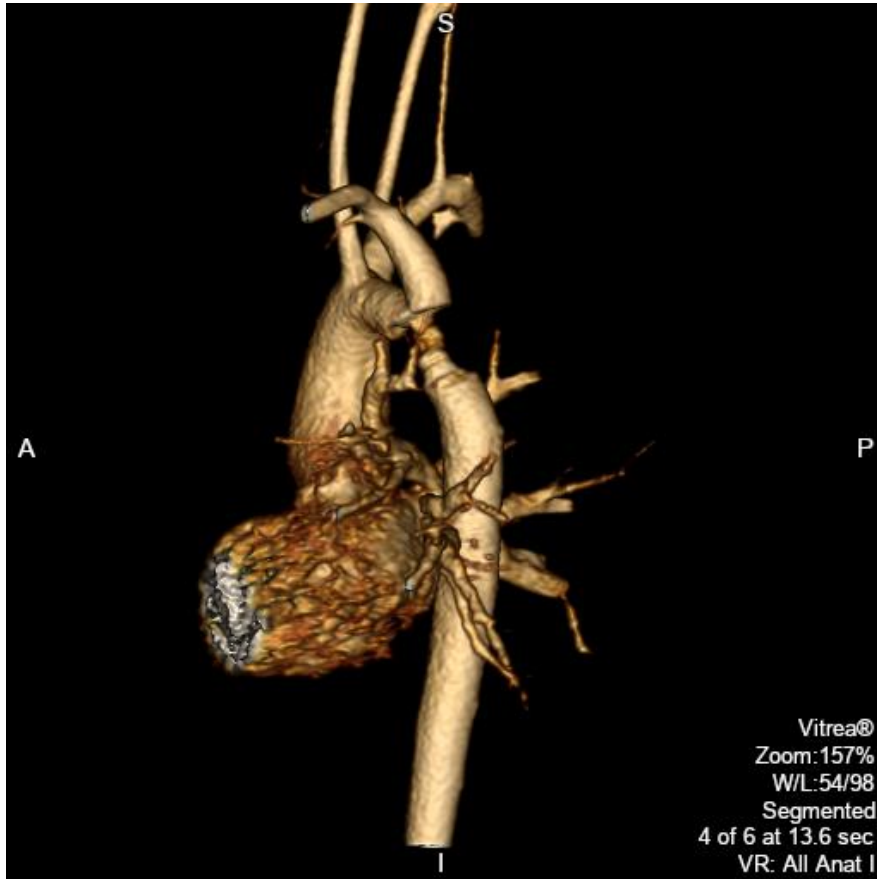




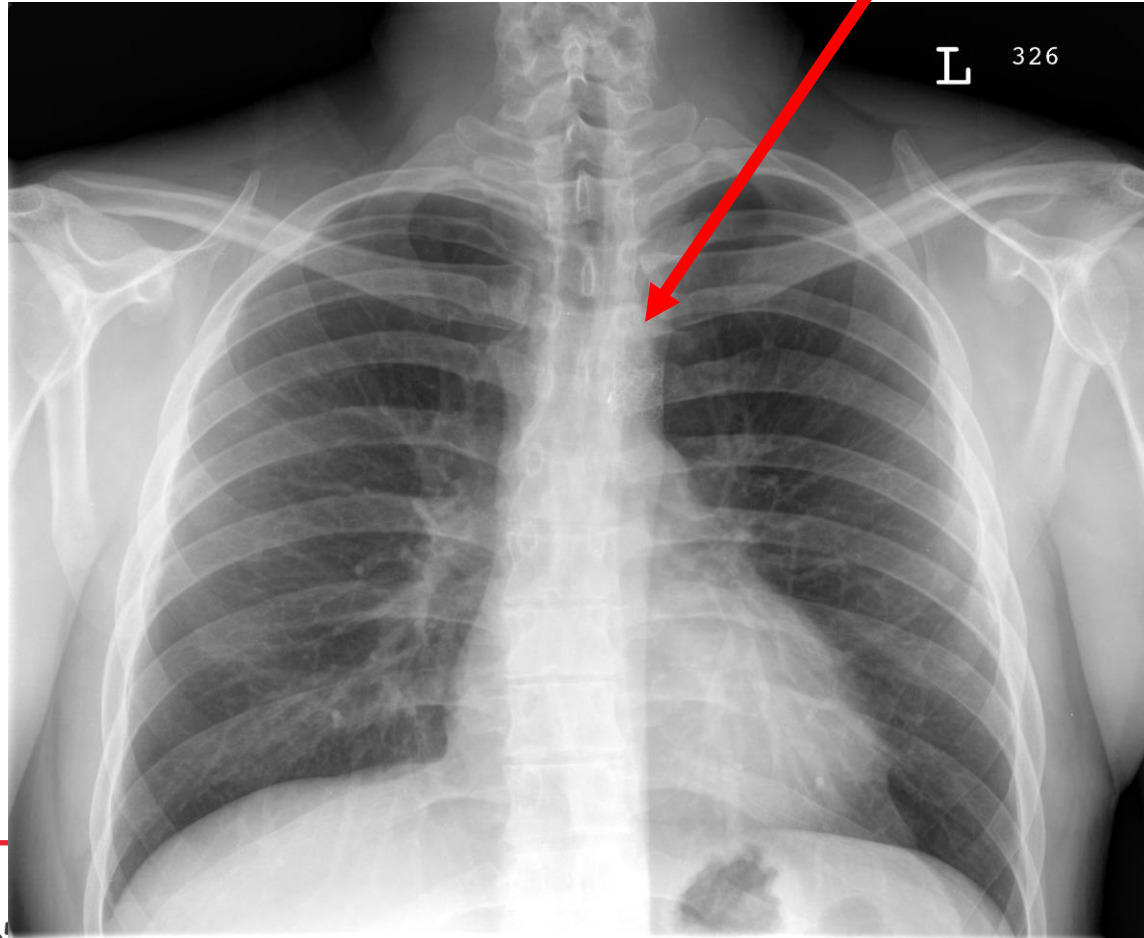
# Coarctation: Unrepaired in adult



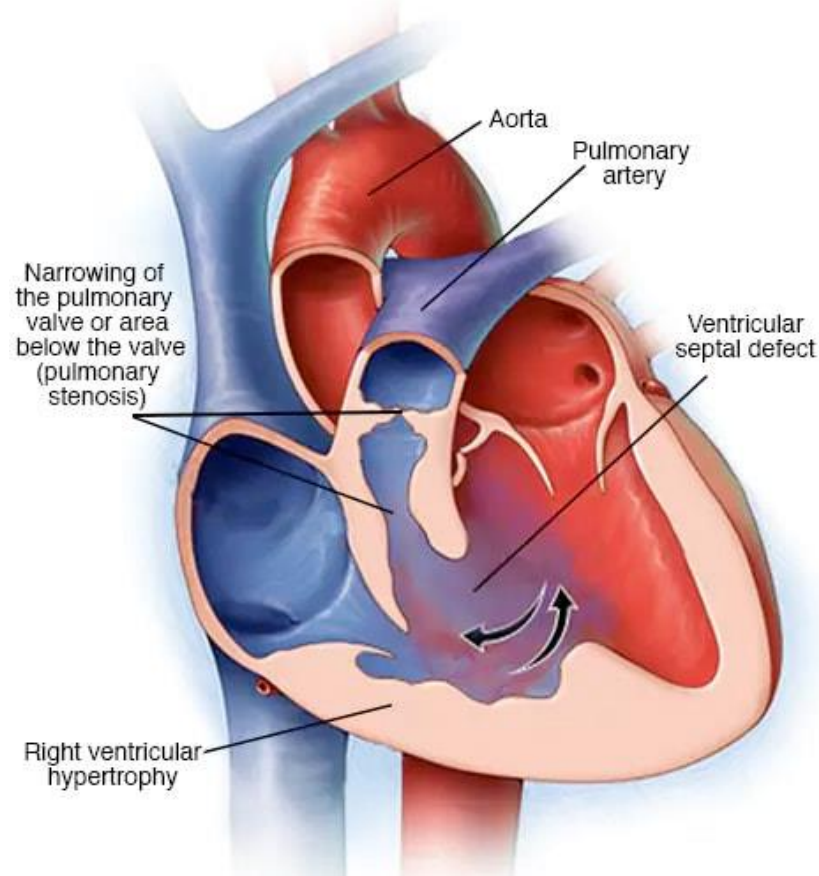
# Beware of Artifact on MRI!



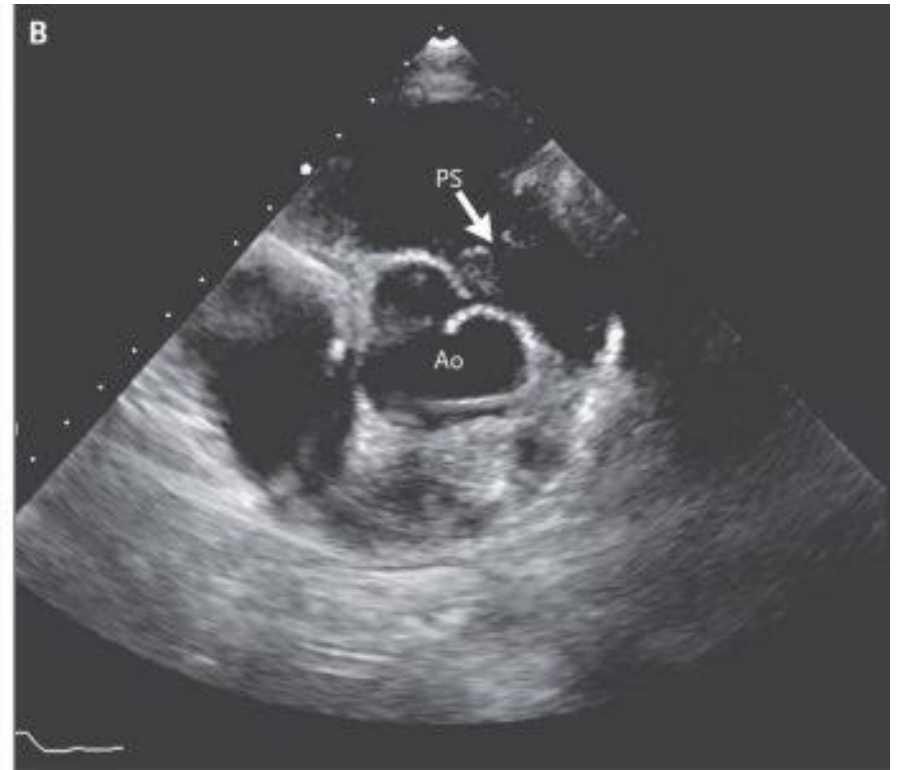
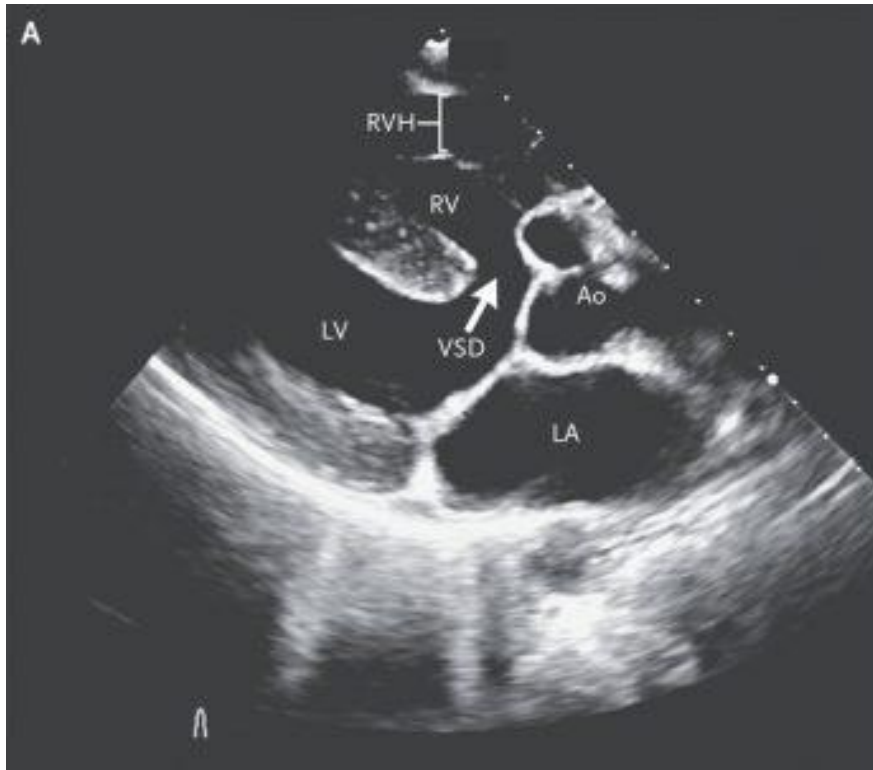
# Coarctation, s/p metallic stent



# #6: Tetralogy of Fallot is the most common complex ACHD lesion



# ToF on Echo (unrepaired)

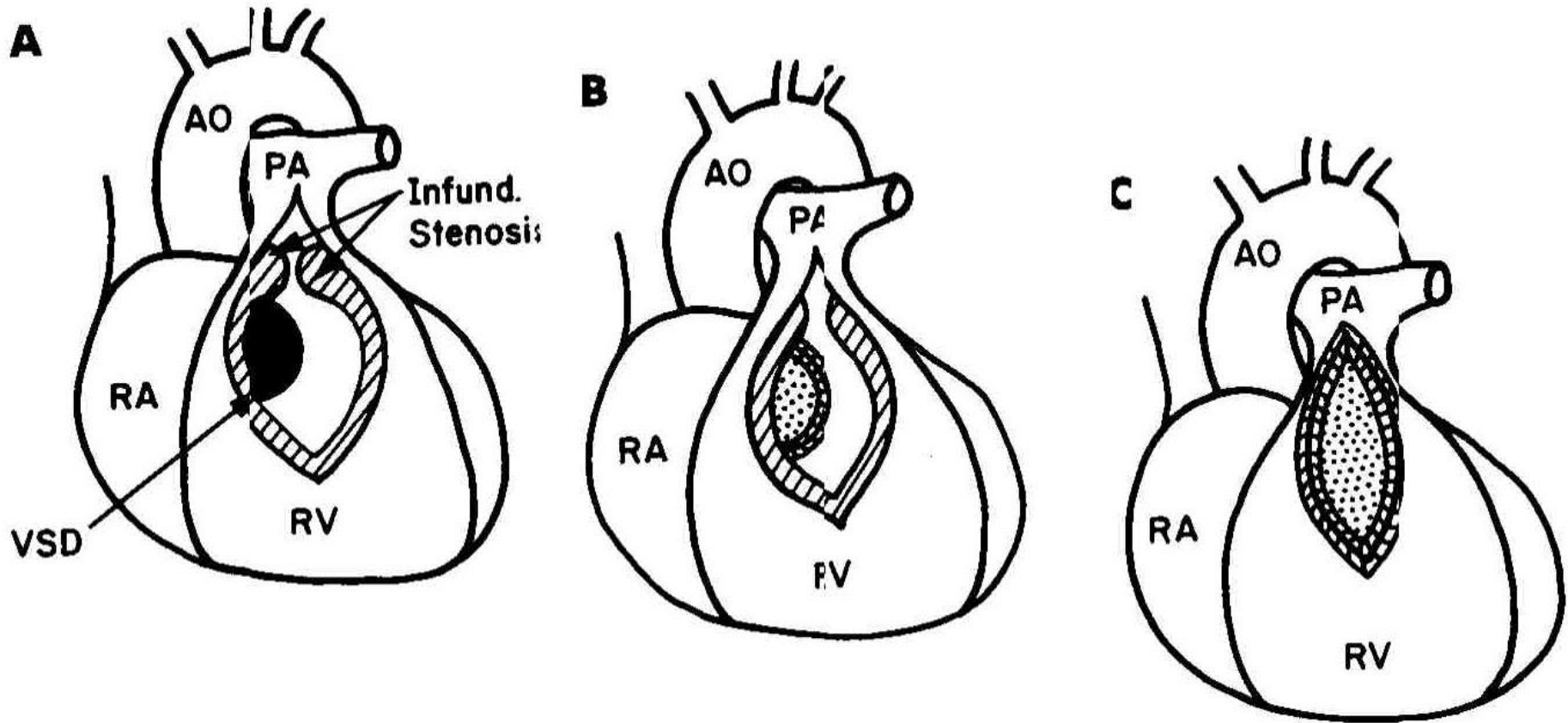


# Frequency of CHD Lesions

- VSD 20%
- ASD 8%
- PDA 8%
- Tetralogy 5%
- P. stenosis 5%
- Ao. Stenosis 5%
- d-Transposition 3%
- Hypoplastic LV 1%
- Hypoplastic RV 1%
- Truncus Art 1%
- TAPVR 1%
- Tric Atresia 1%
- Single ventricle 1%
- DORV 1%
- Others 5-10%



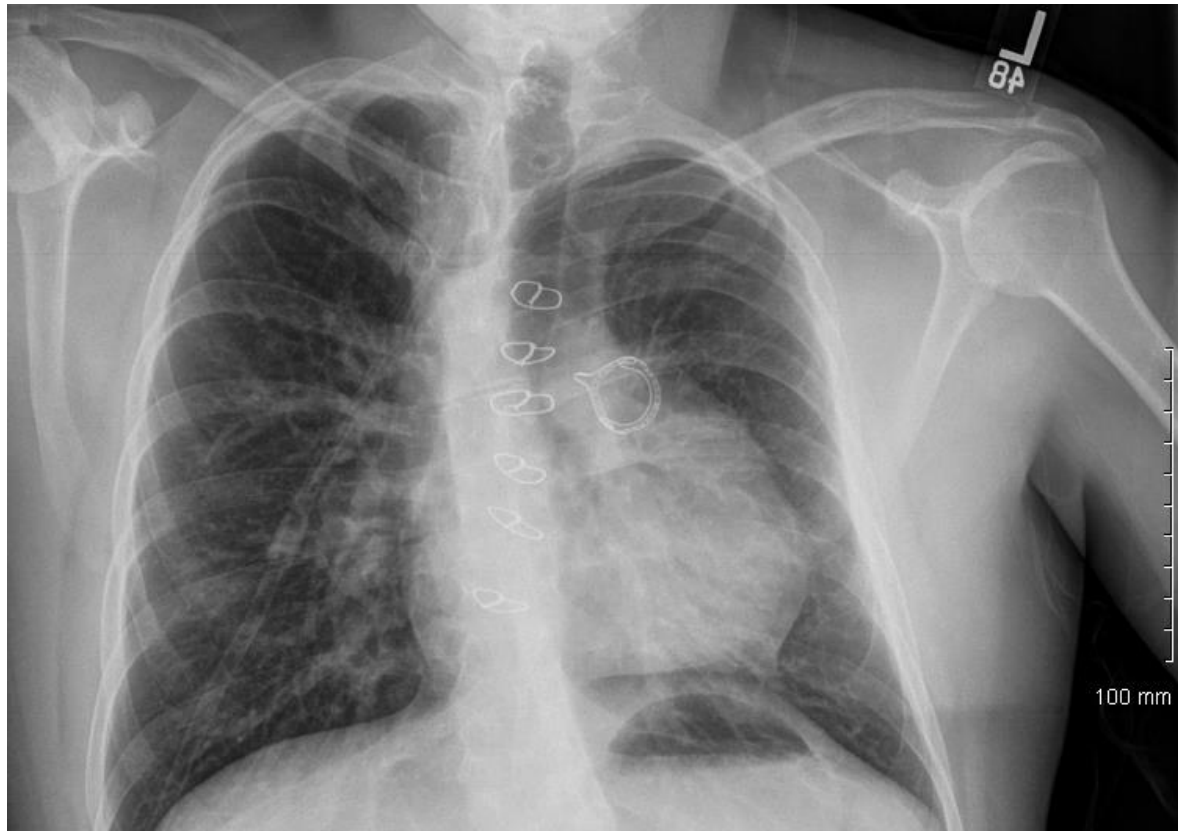
# Tetralogy of Fallot: Transventricular repair



27 y/o s/p ToF repair at 9m.

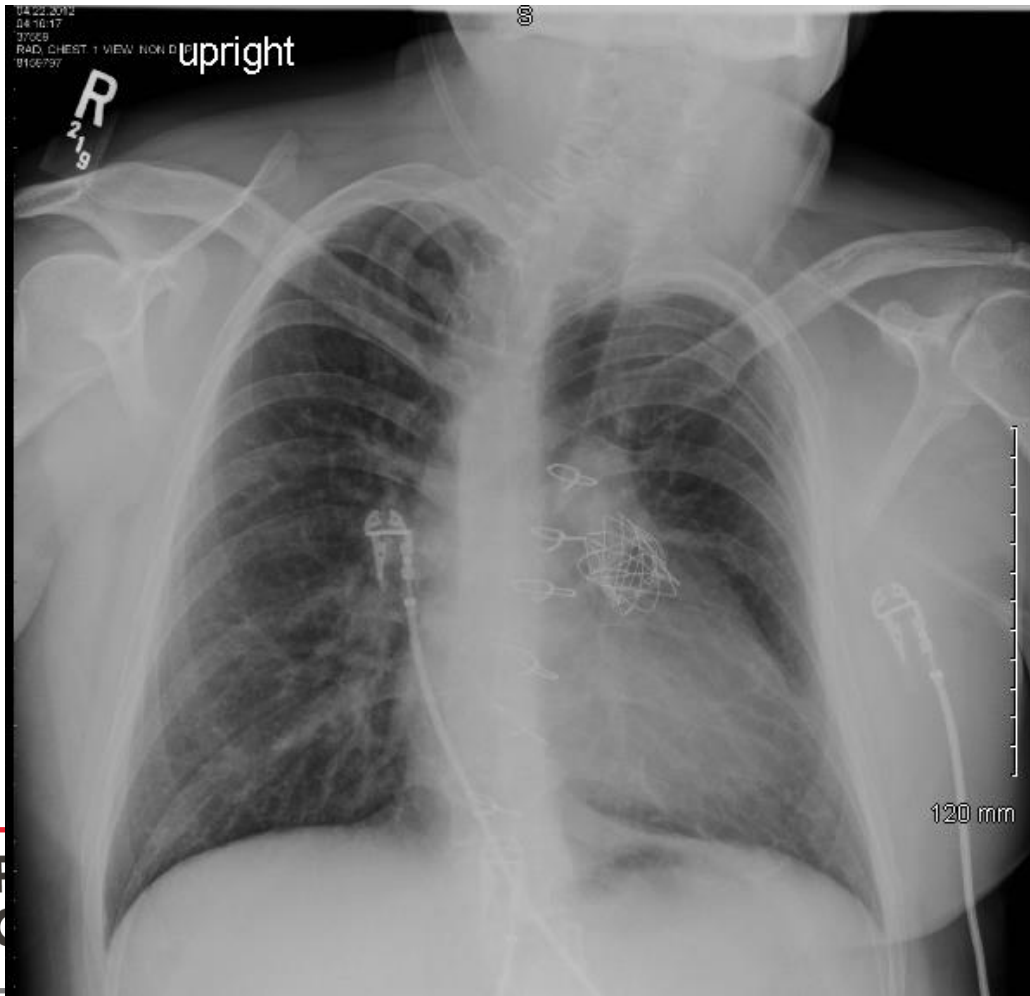
-Surgical Pulm Valve Replacemt (sPVR) 8y ago.

-Now with PVR dysfxn.



# S/p Transcatheter Valve

(percutaneous pulmonary valve implantation)



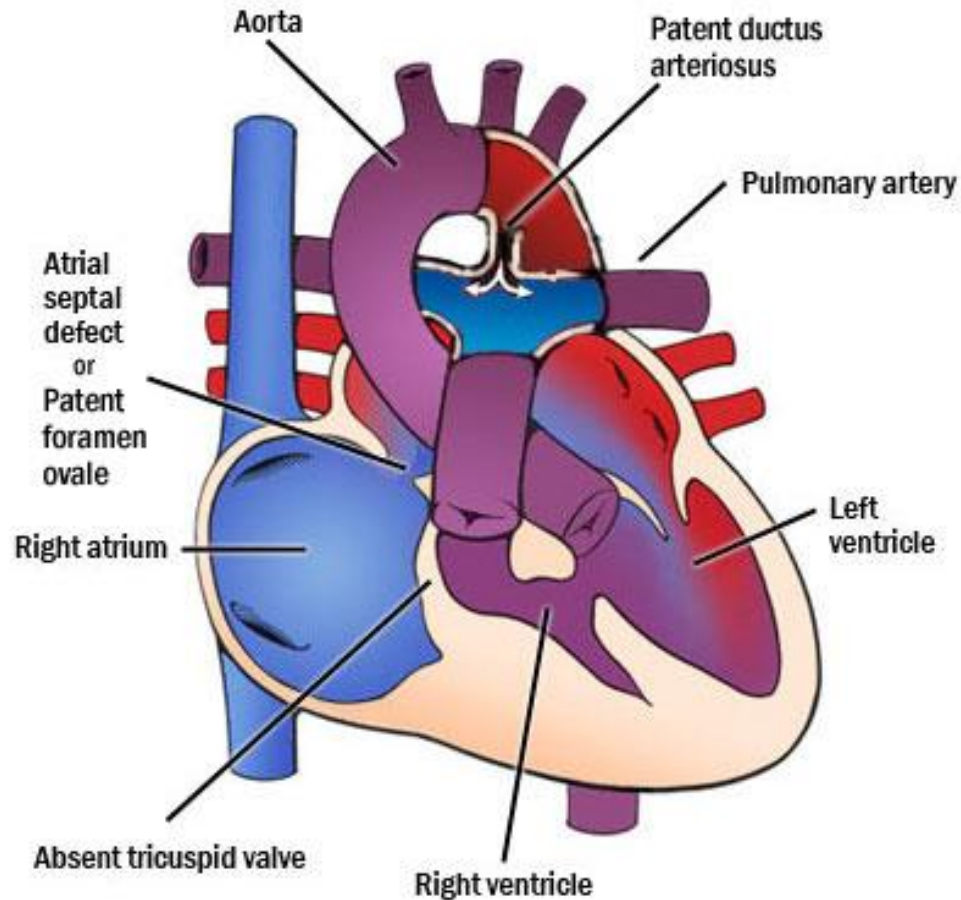
# Tough cases...



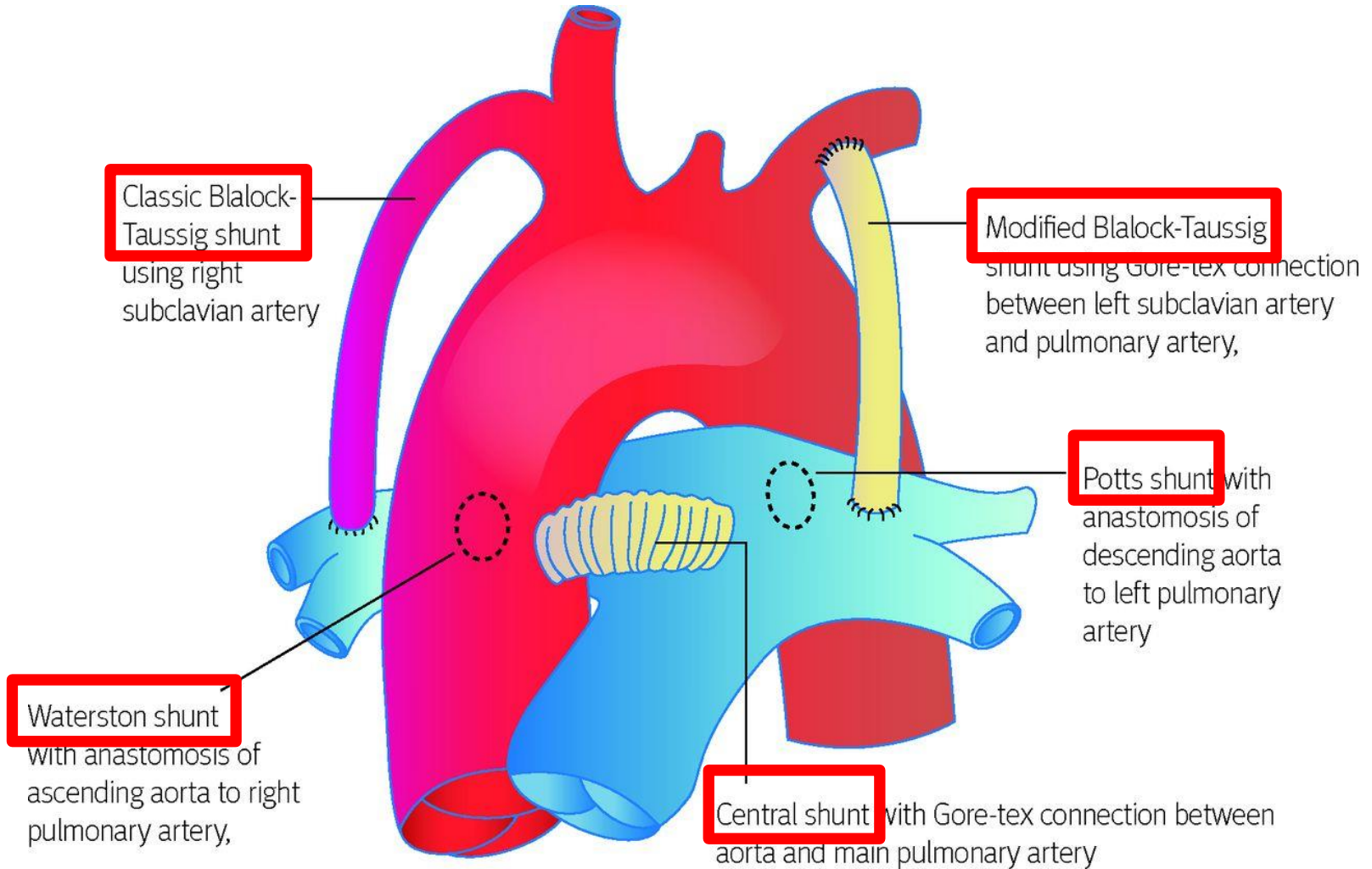
# Complex Congenital Heart Lesions

**#7: Single Ventricle/Fontan pts are increasing in number**

# Single Ventricle Patients



# Surgical Shunts in Cyanotic Single Ventricles

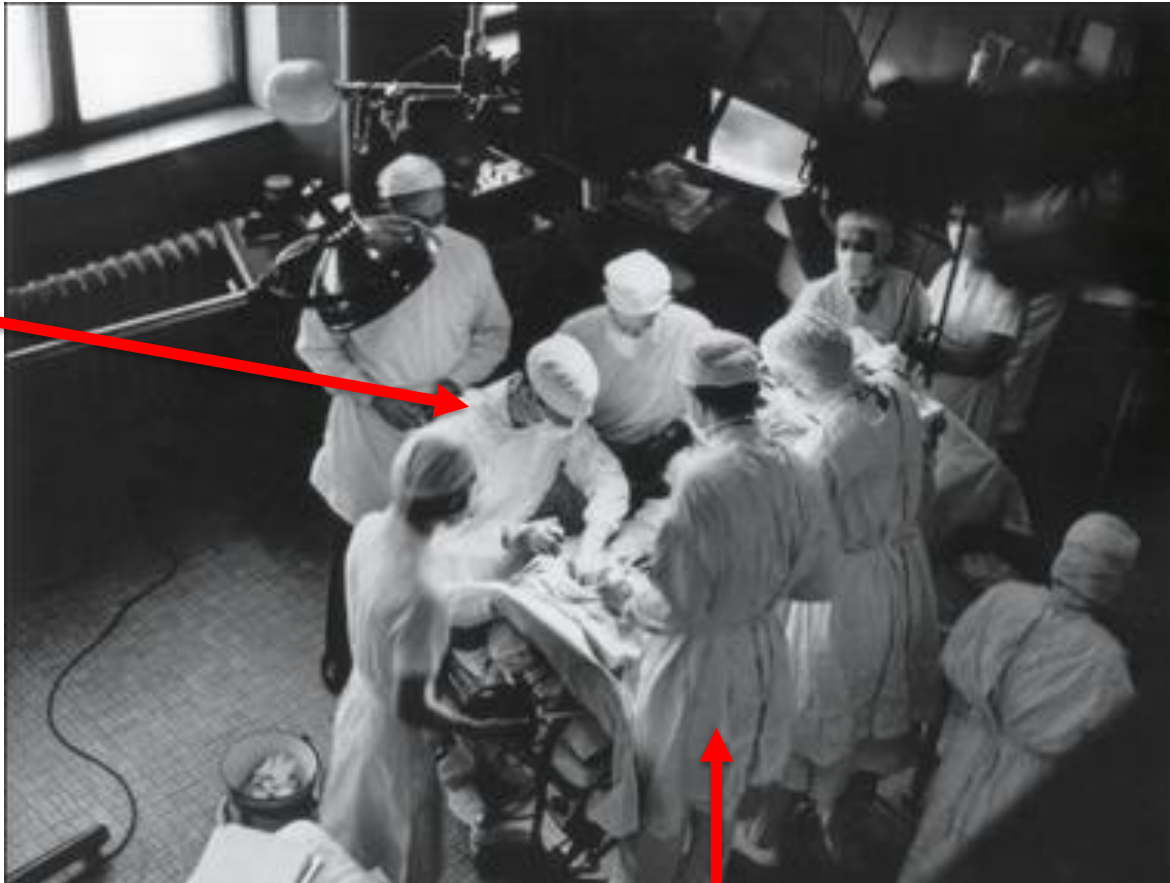


# **Pop Quiz:** Why is this picture important?



# 1944: Johns Hopkins – The first congenital heart operation

**Dr. Alfred  
Blalock**



**Dr. Denton Cooley**

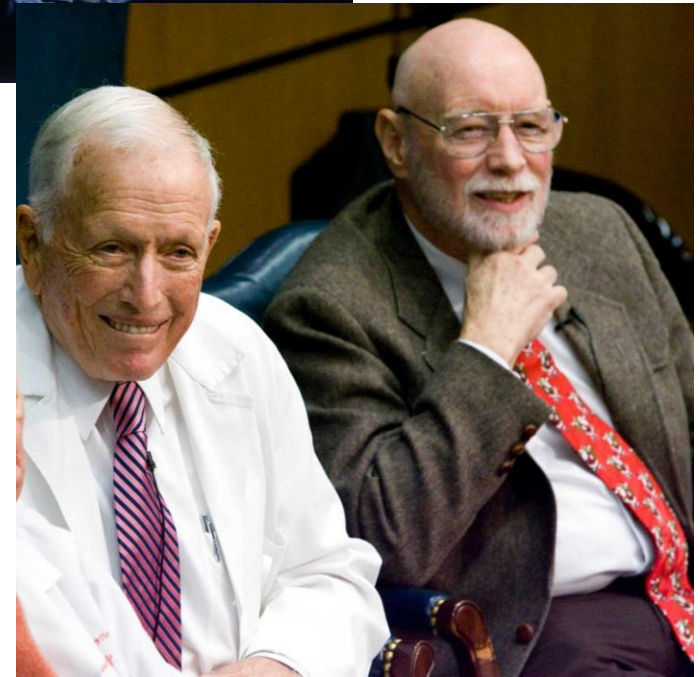


# Heart Disease History!

*(with a nod to Texas)*



1968



1980s

2015

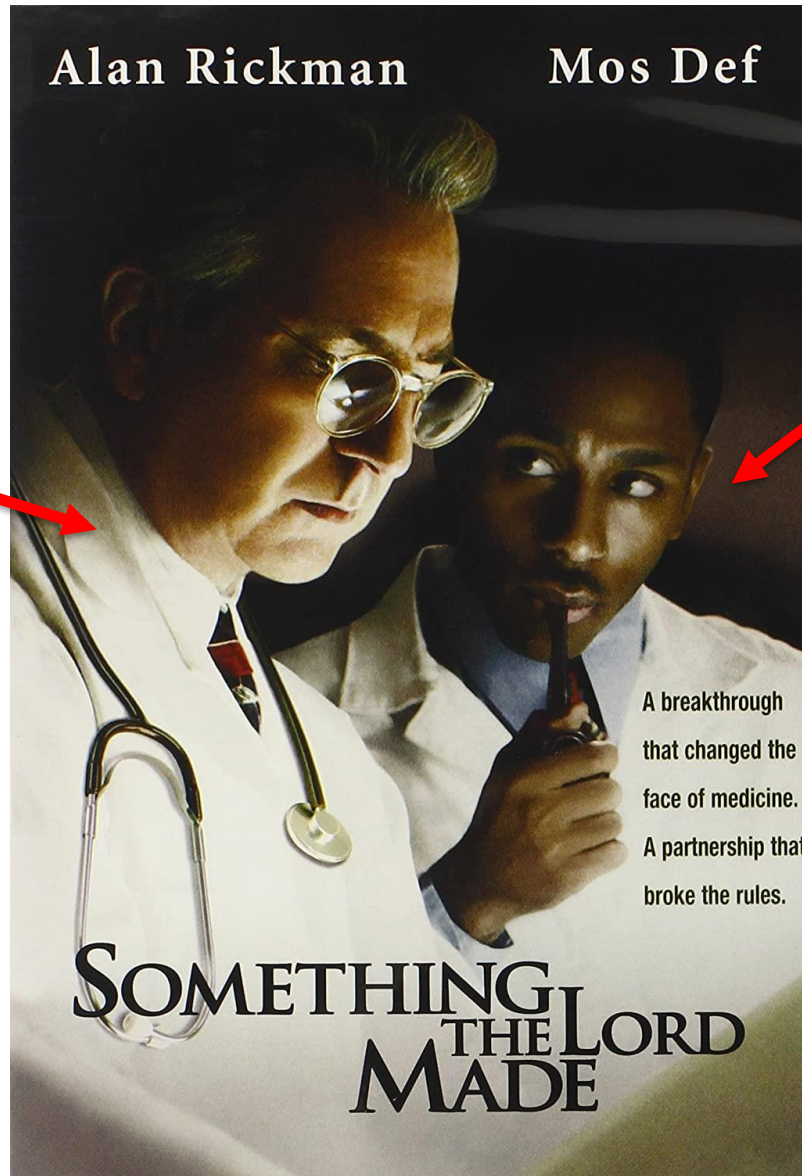


Alan Rickman

Mos Def

Dr. Alfred  
Blalock

Vivien Thomas



Phoenix  
Children's

HBO Films, 2004

# Drs. Thomas, Blalock, Taussig



Phoenix  
Children's

Source: Johns Hopkins University

# Dr. Francis Fontan



1971

*Thorax* (1971), 26, 240.

## Surgical repair of tricuspid atresia

F. FONTAN and E. BAUDET

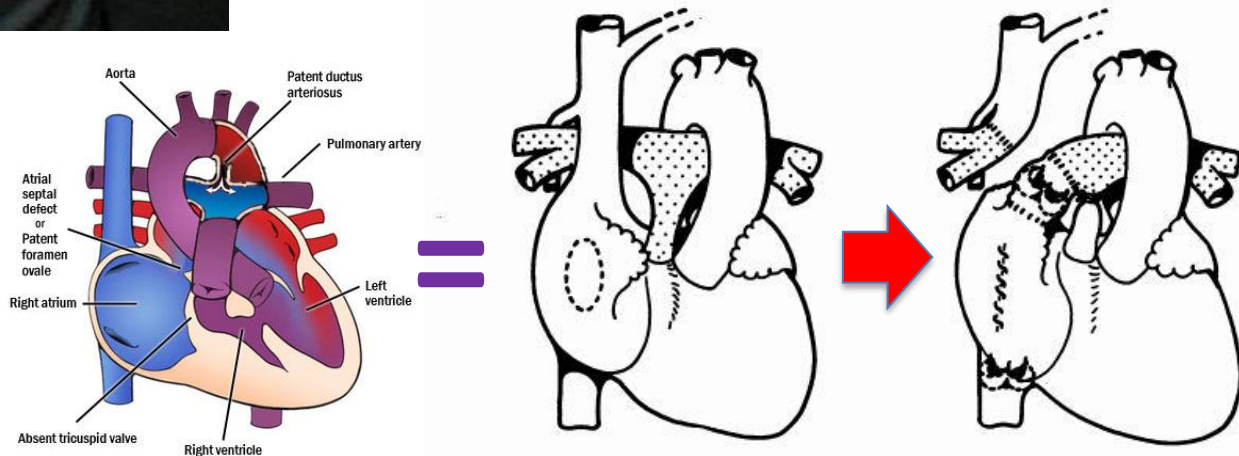
*Centre de Cardiologie, Université de Bordeaux II, Hôpital du Tondu, Bordeaux, France*

Surgical repair of tricuspid atresia has been carried out in three patients; two of these operations have been successful. A new surgical procedure has been used which transmits the whole vena caval blood to the lungs, while only oxygenated blood returns to the left heart. The right atrium is, in this way, 'ventriclized', to direct the inferior vena caval blood to the left lung, the right pulmonary artery receiving the superior vena caval blood through a cava-pulmonary anastomosis. This technique depends on the size of the pulmonary arteries, which must be large enough and at sufficiently low pressure to allow a cava-pulmonary anastomosis. The indications for this procedure apply only to children sufficiently well developed. Younger children or those whose pulmonary arteries are too small should be treated by palliative surgical procedures.

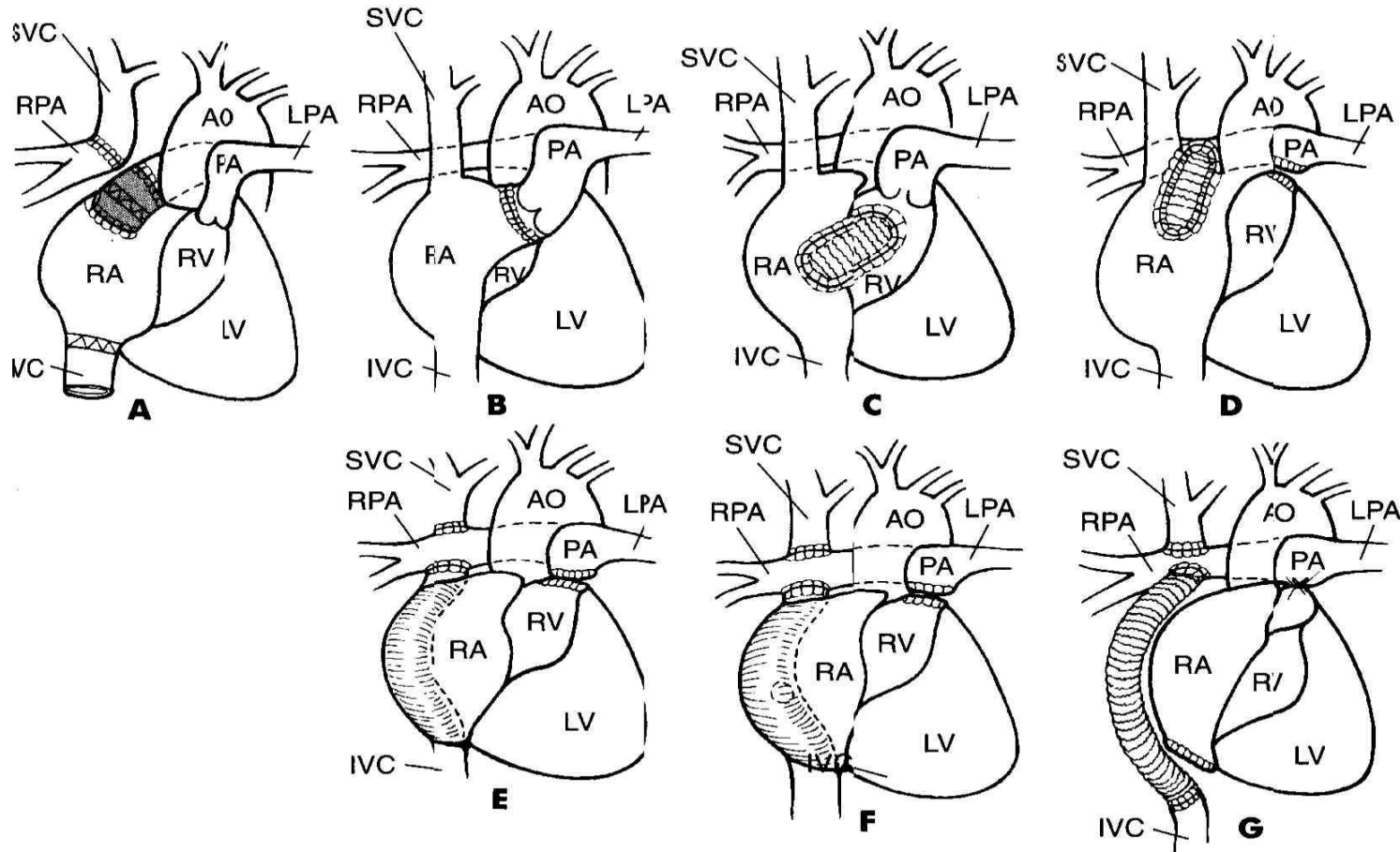
Only palliative operations (systemic vein to pulmonary artery anastomosis; systemic artery to pulmonary artery anastomosis) have been performed in tricuspid atresia. Although these procedures are valuable, they result in only a partial clinical improvement, because they do not suppress the mixture of venous and oxygenated blood.

We have initiated a corrective procedure for

tricuspid atresia, which completely suppresses blood mixing. The entire vena caval return under goes arterialization in the lungs and only oxygenated blood comes back to the left heart. This procedure is not an anatomical correction, which would require the creation of a right ventricle but a procedure of physiological pulmonary blood flow restoration, with suppression of right an-



# The *Fontan* Procedure— minimal change from 1971



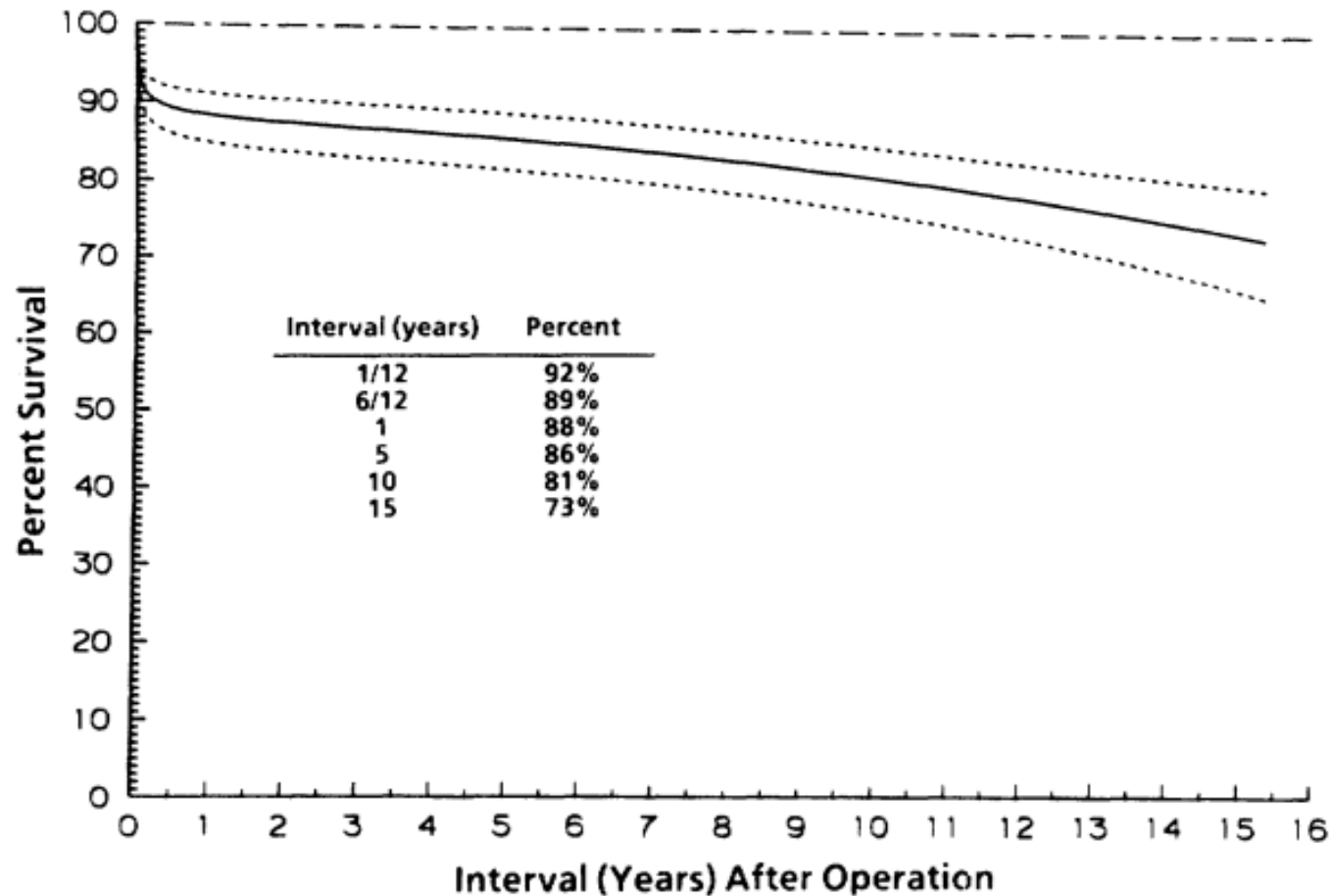
The *Fontan Operation* –  
Short and Intermediate-term results  
are very good...

## **Outcome After a “Perfect” Fontan Operation**

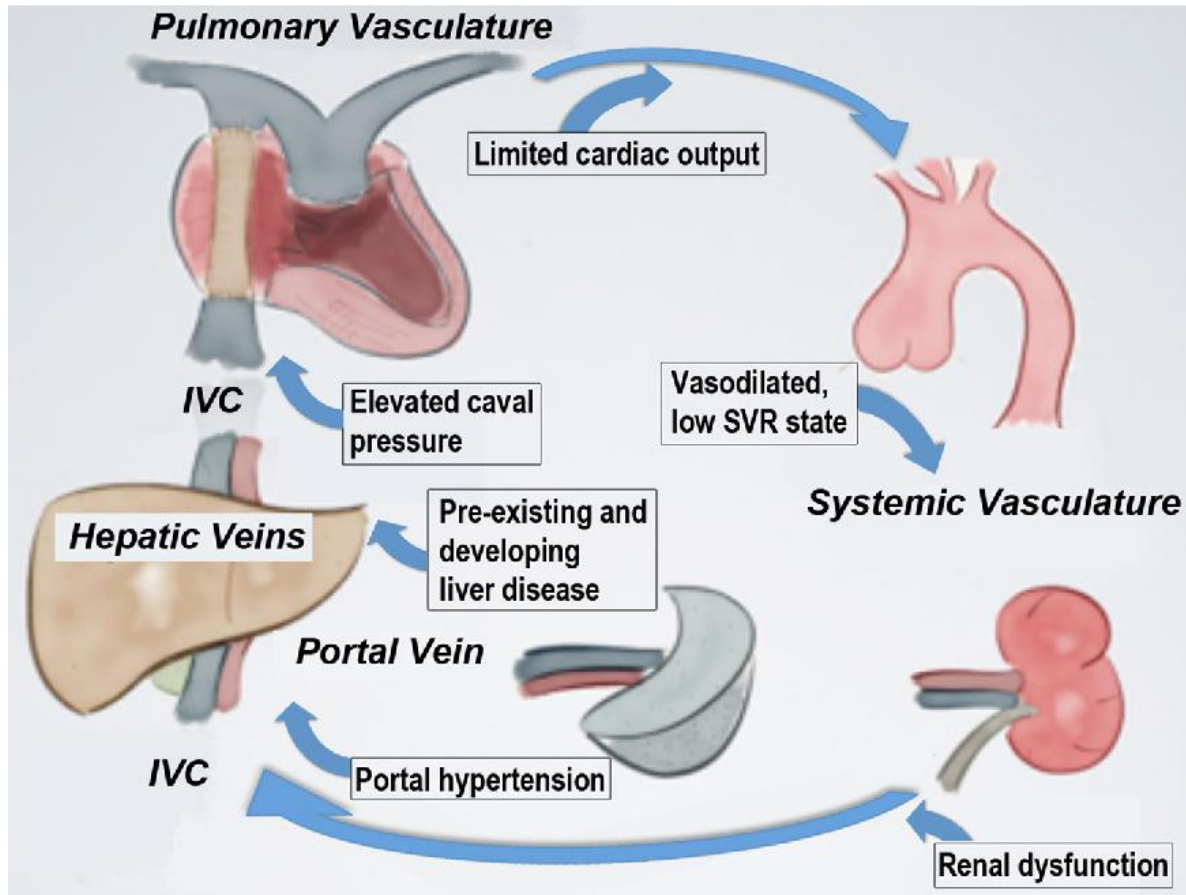
Francis Fontan, MD, John W. Kirklin, MD, Guy Fernandez, MD, Francisco Costa, MD,  
David C. Naftel, PhD, Francesco Tritto, PhD, and Eugene H. Blackstone, MD

*Circulation, 1990*

# After a “Perfect” Fontan by Fontan



# Fontan Physiology



# Current Data on Fontans

## Contemporary Outcomes of the Fontan Operation: A Large Single-Institution Cohort



Carlos M. Mery, MD, MPH, Luis E. De León, MD, Daniel Trujillo-Diaz, MD, Elena C. Ocampo, MD, Heather A. Dickerson, MD, Huirong Zhu, PhD, Iki Adachi, MD, Jeffrey S. Heinle, MD, Charles D. Fraser, Jr, MD, and Peter R. Ermis, MD

ATS 2019

## The Adolescent and Adult With a Fontan Circulation

“Unnatural” Selection and Survival of the Fittest\*

JACC 2018

Jack Rychik, MD,<sup>a,b</sup> Yuli Kim, MD<sup>c,d</sup>

## **AHA SCIENTIFIC STATEMENT**

## Evaluation and Management of the Child and Adult With Fontan Circulation

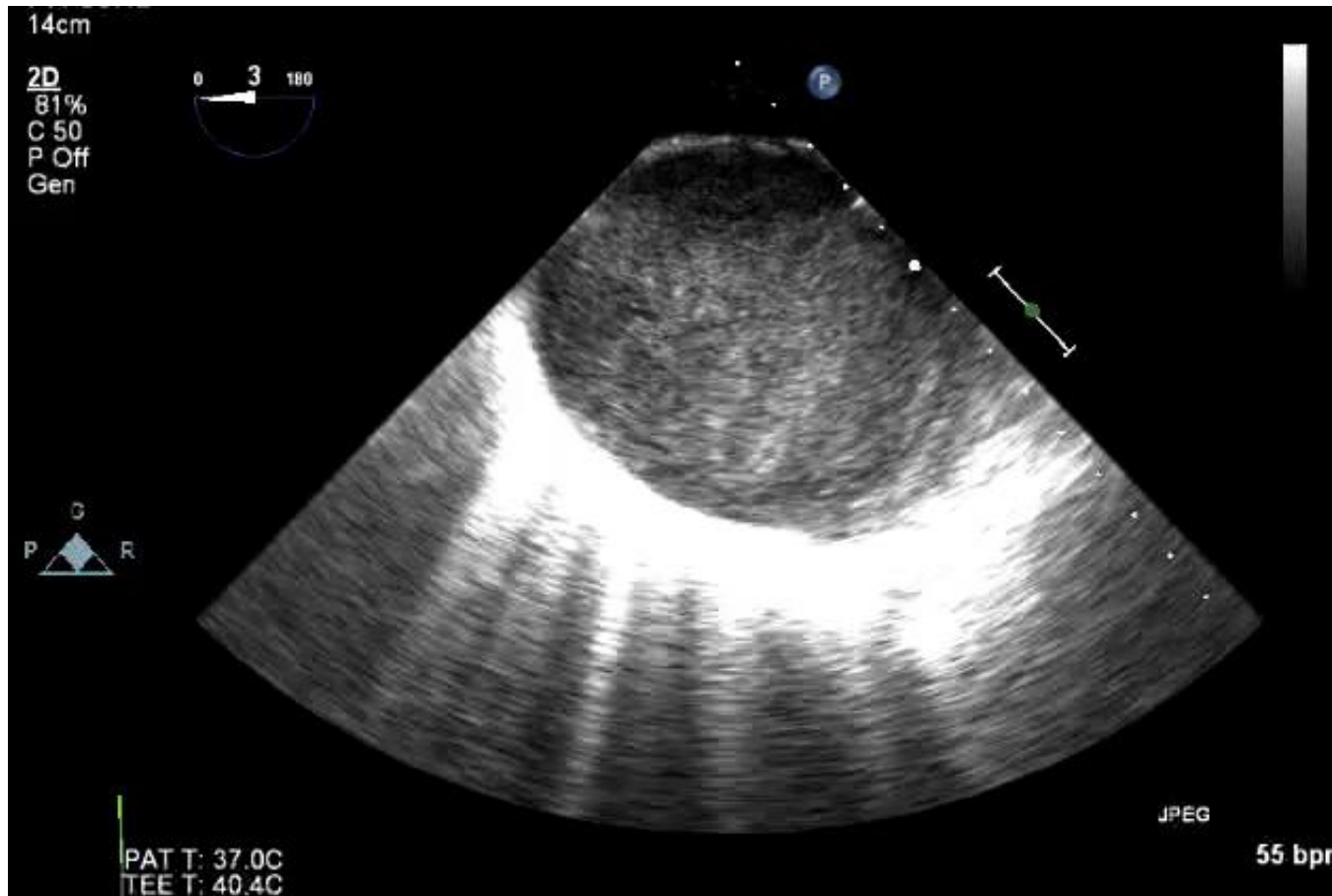
Circ 2019

# The Fontan Procedure – Long-term results are good, but mixed

- Complications
  - Arrhythmias (SVT: AFlutter/IART “Scar Flutter”), thromboembolism → PE,
  - ventricular failure,
  - pulm AVMs,
  - FALD: Fontan-Associated Liver Disease
- *“10 good years, 10 ‘so-so’ years, then TROUBLE!”*
  - *-from Professor Gary Webb*
- “Failing Fontan” Syndrome
  - protein-losing enteropathy,
  - SVT, ‘R-sided CHF’



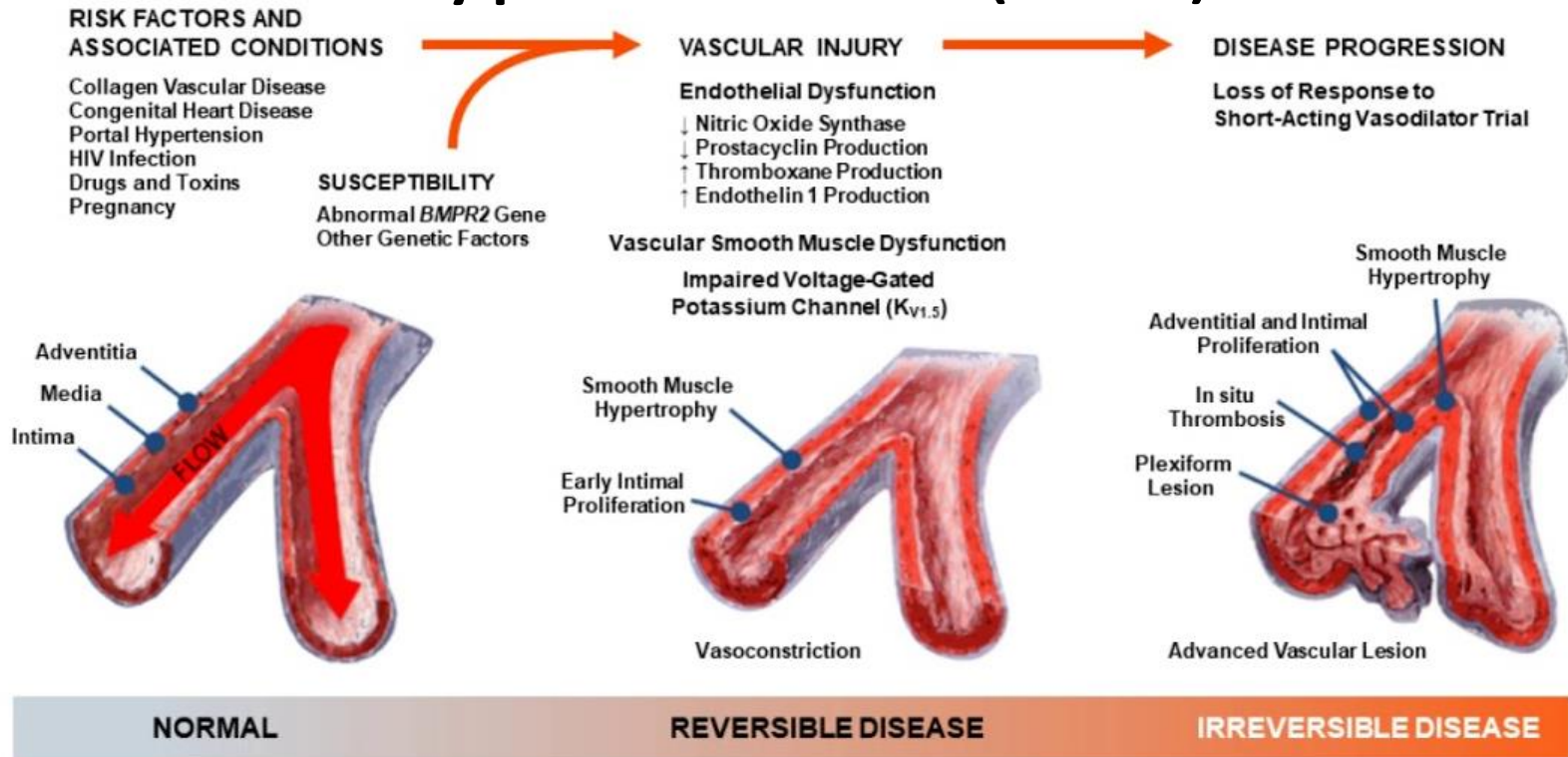
# Fontan TEE: R Atrial “smoke”



# #8: ACHD and Pulm HTN is Bad



# Pathophysiology of Pulmonary Arterial Hypertension (PAH)



Gaine S. 2009

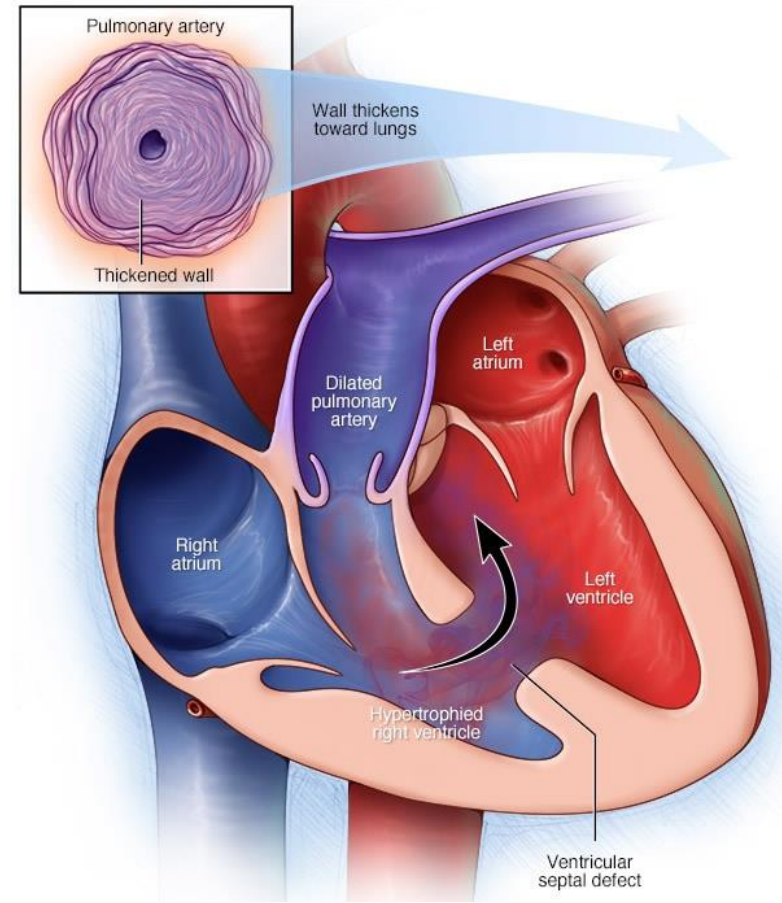
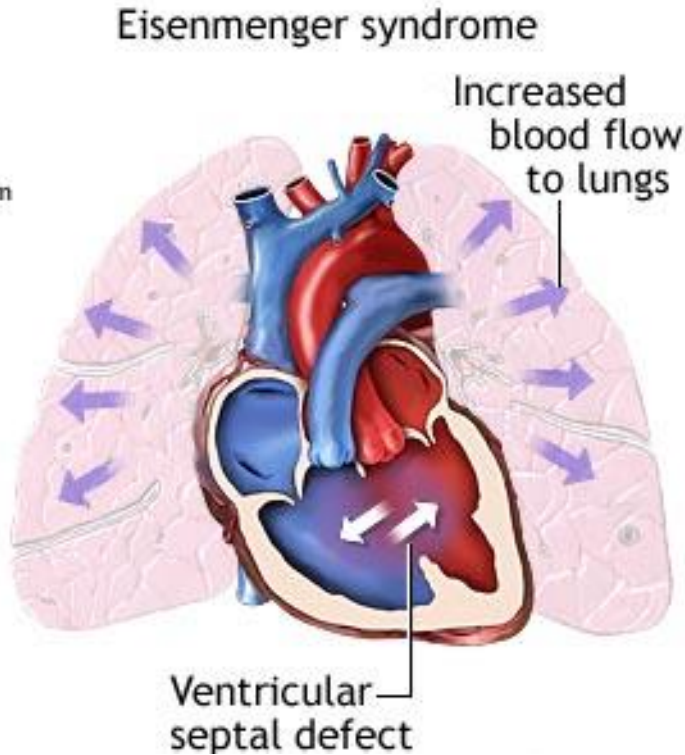
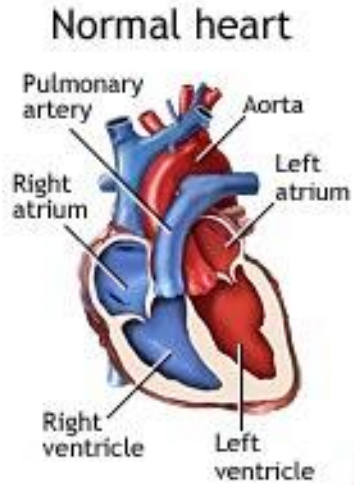
Franklin WJ. 2011

Brida M, Gatzoulis MA. 2018



**Phoenix  
Children's**

# Pathophysiology of Eisenmenger Syndrome



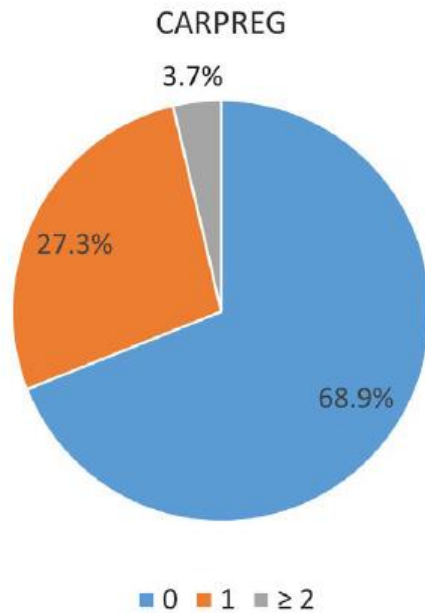
Findings in Eisenmenger Syn:  
Clubbing, cyanosis, O<sub>2</sub> sat<85%, HCT↑



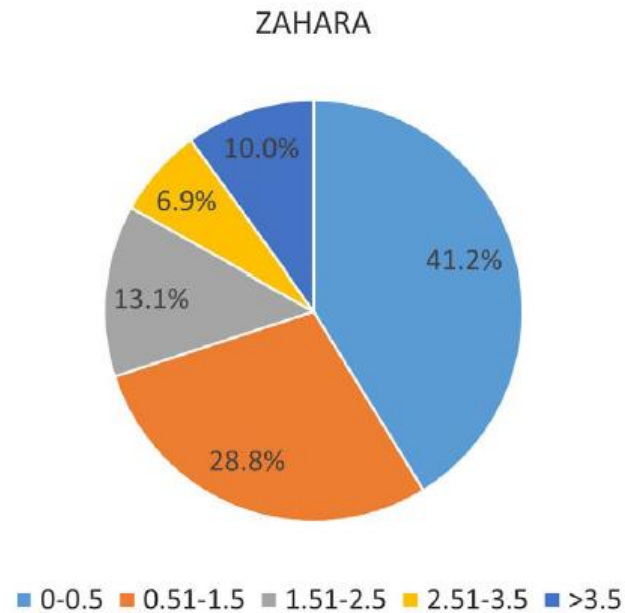
#9: Pregnancy with ACHD is risky, but  
can be successful



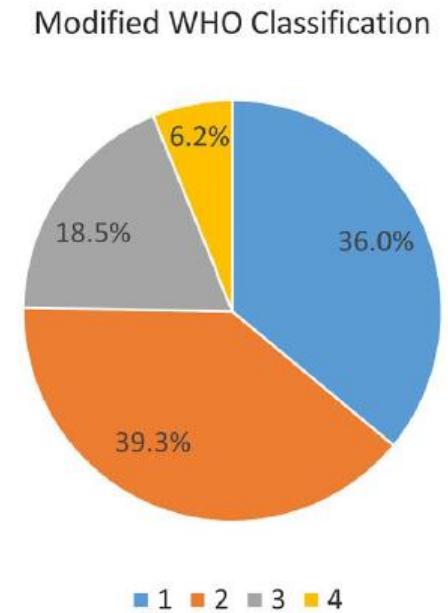
# CV Risk Prediction in Pregnancy: *WHO is better (=more Sn for CHD)*



Siu 2001

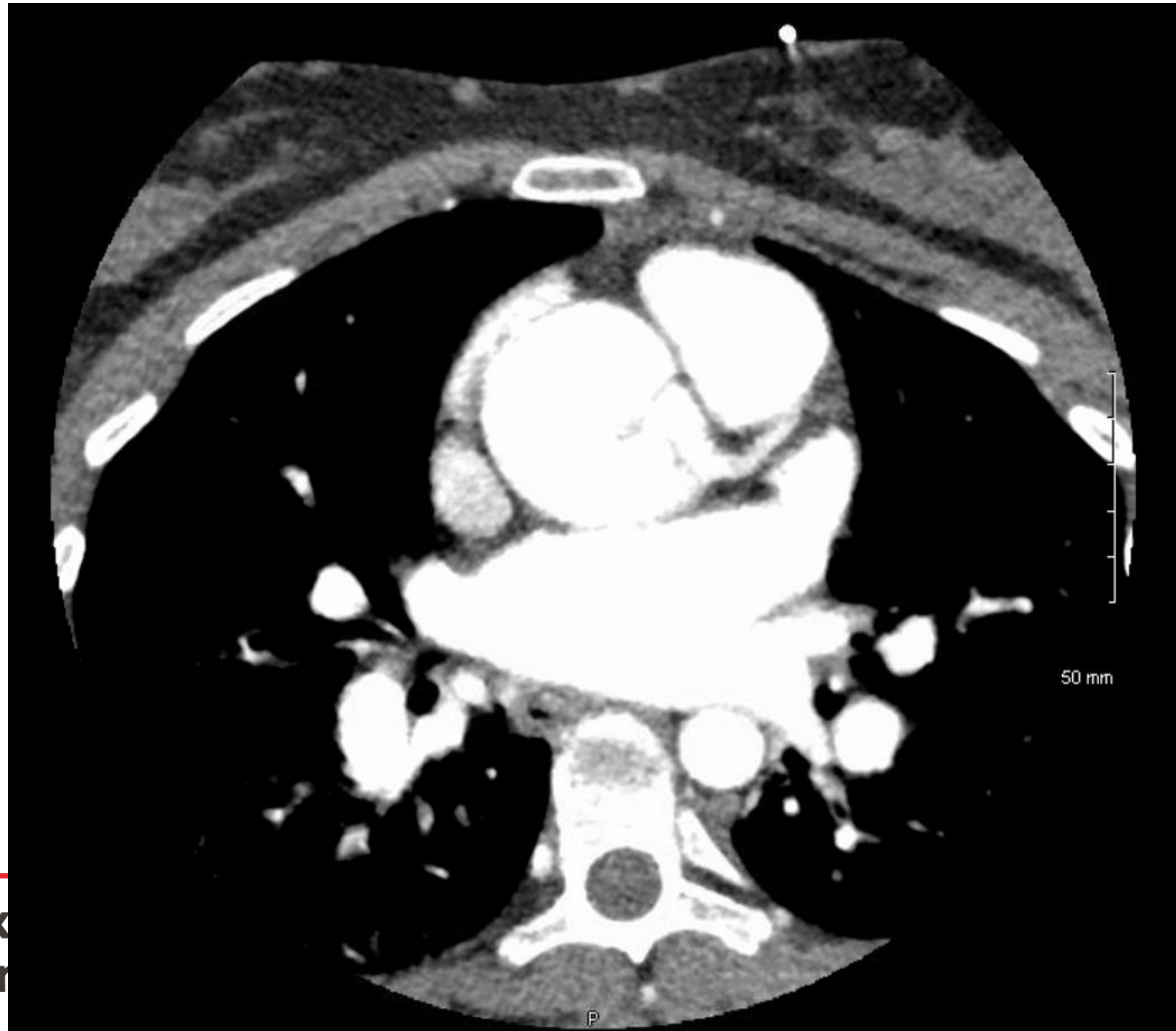


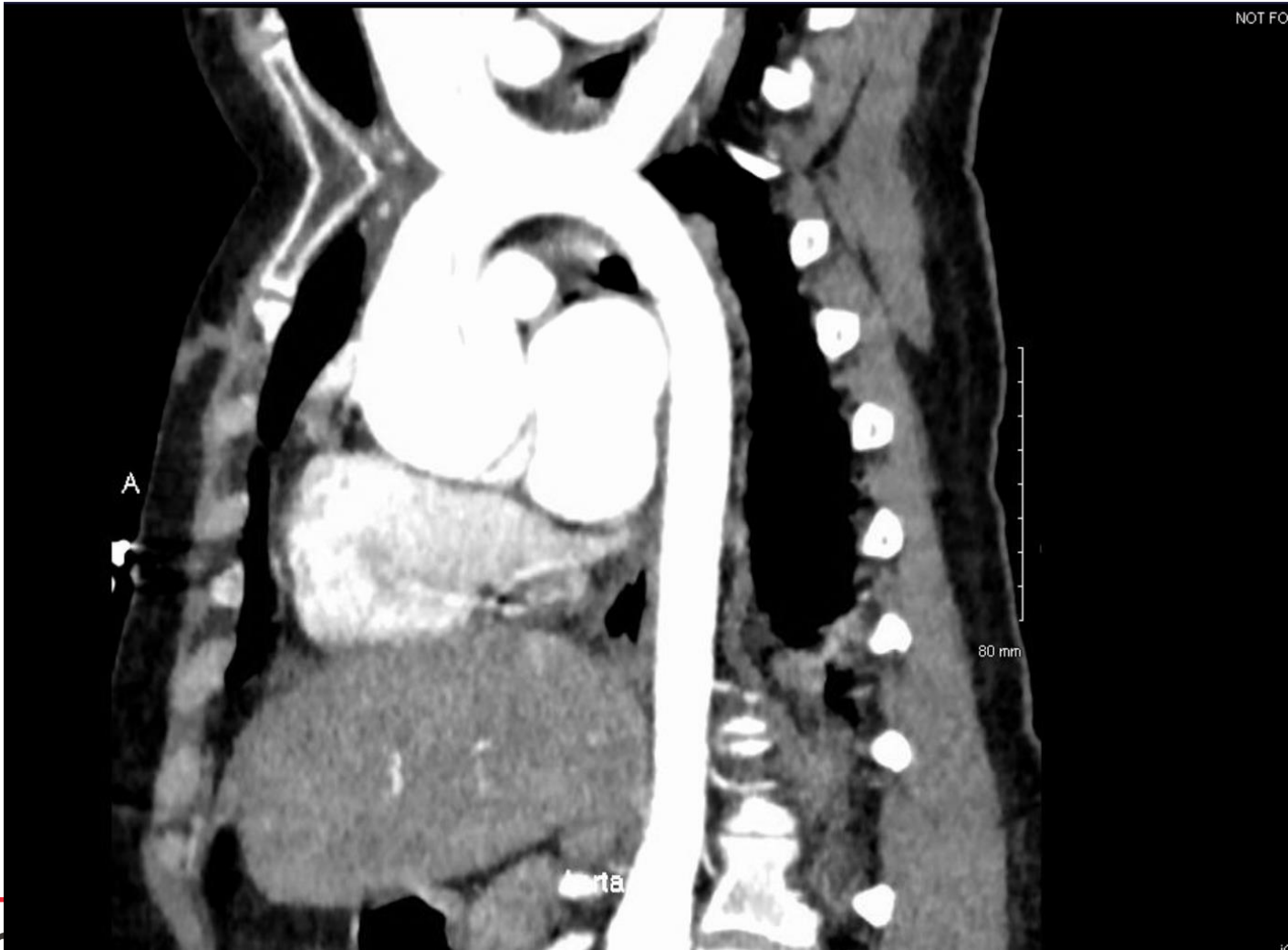
Drenthen 2010



Diller 2014

# 23 y/o pregnant Marfan pt





Phoenix  
Children's

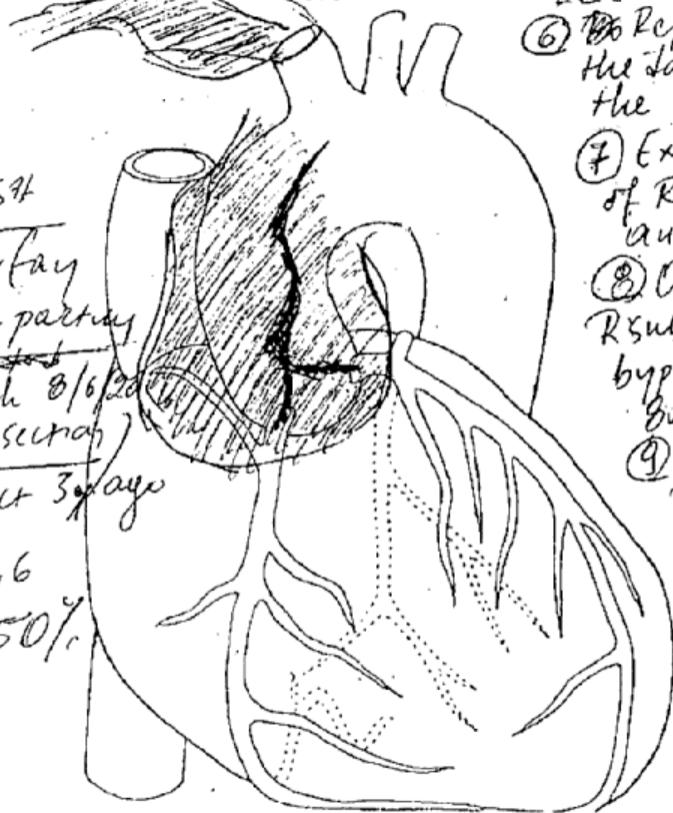
Antegrade cerebral perfusion: 5+

- innominate
- left common carotid

PHILIPSAH

- Maxillary
- Post partum
- ~~birth~~ birth 8/6/2006
- > C-section
- C-section 3y ago

CR = 0,6  
EF = 50%



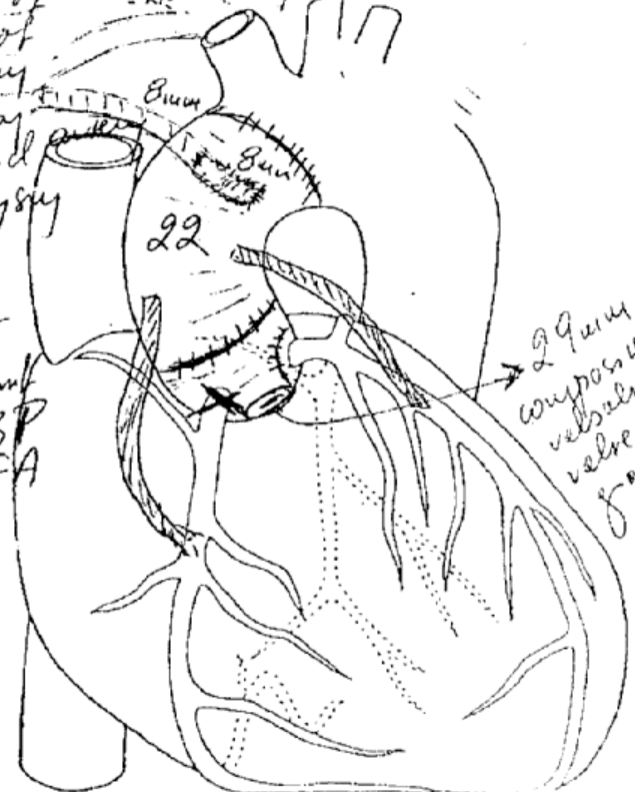
PREOPERATIVE DIAGNOSIS AND OPERATIVE FINDINGS

Harvesting LLE

- Repair of the base of the aorta
- Excision of R subcl aneurysm
- Aorta R subcl bypass
- IABP R CFA

Complications: ... / Drains: ...

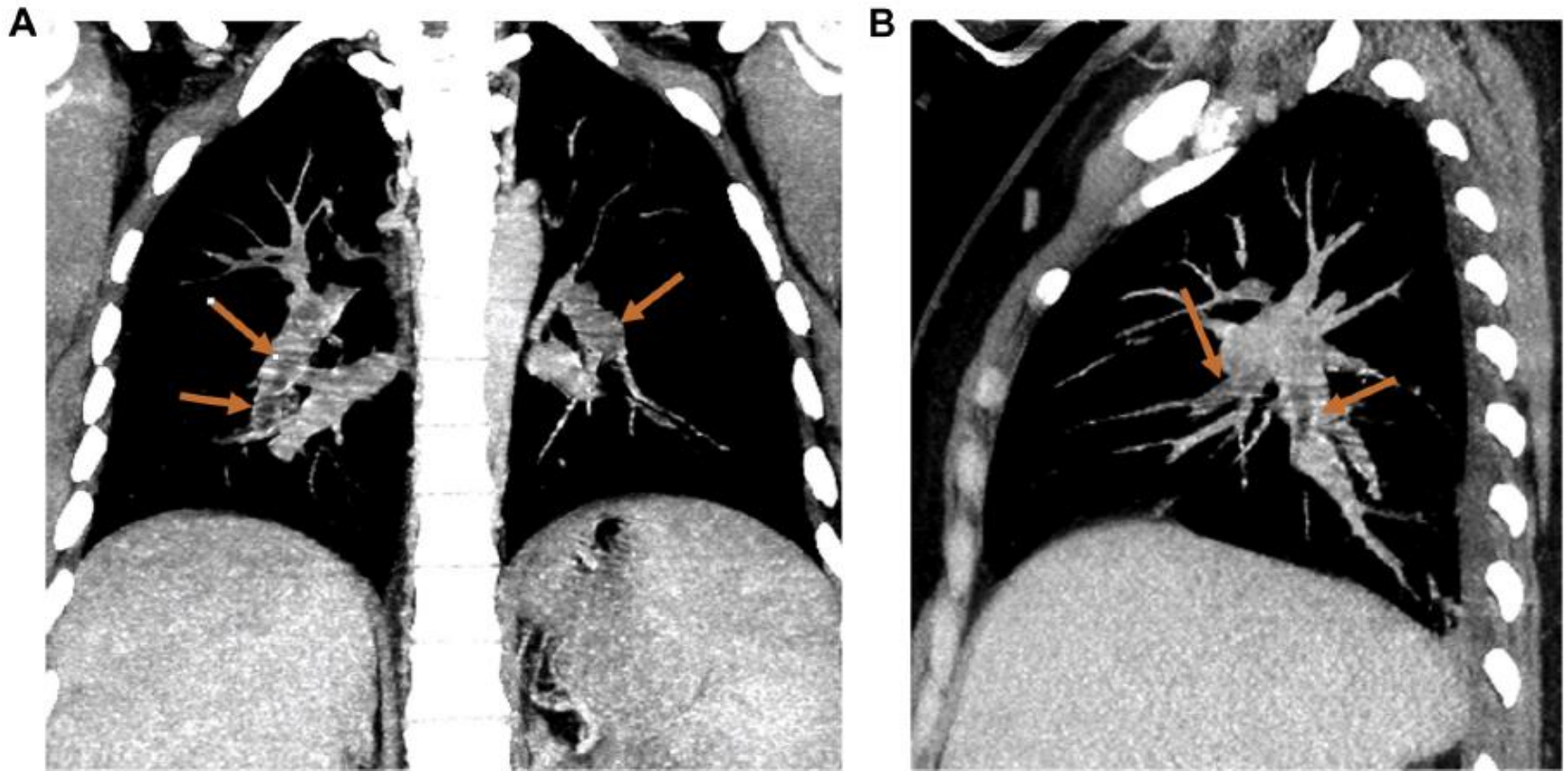
Specimens: AV leaflets wall - 1 L pleural



PROCEDURE PERFORMED

# Pregnancy in Fontans – **HIGH RISK** and *must see ACHD and MFM*

**FIGURE 1** Maximum Intensity Projection Computed Tomography Images



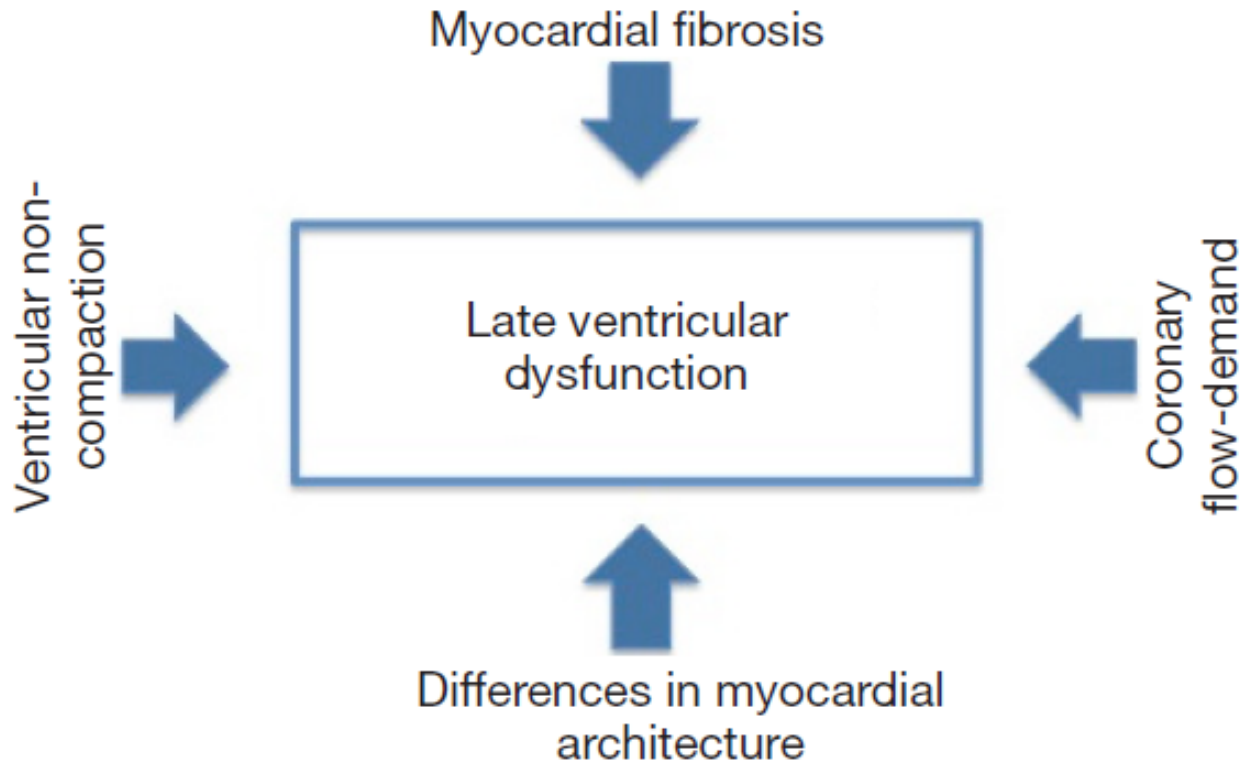
# Contraindications to Pregnancy

- 1) Pulmonary HTN (PVR > 4 or mPAP>45)
- 2) Cyanosis (O2 sat < 85%)
- 3) Marfan Syn with Dilated Ao Root (> 4.0 cm)
- 4) LVEF < 20%
- 5) Severe LVOTO (MS, AS, Coarctation)

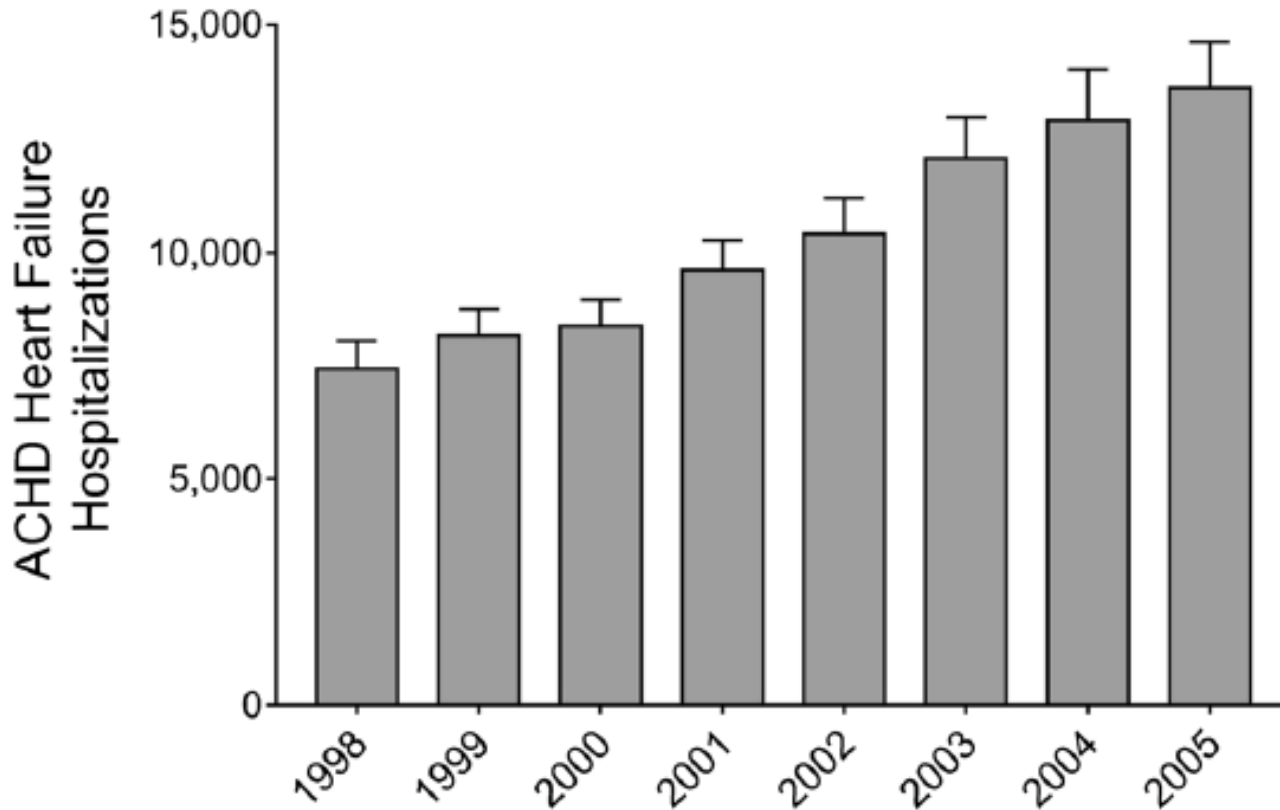
# #10: Heart Failure in ACHD is Common!



# Heart Failure in ACHD: Pathophysiology

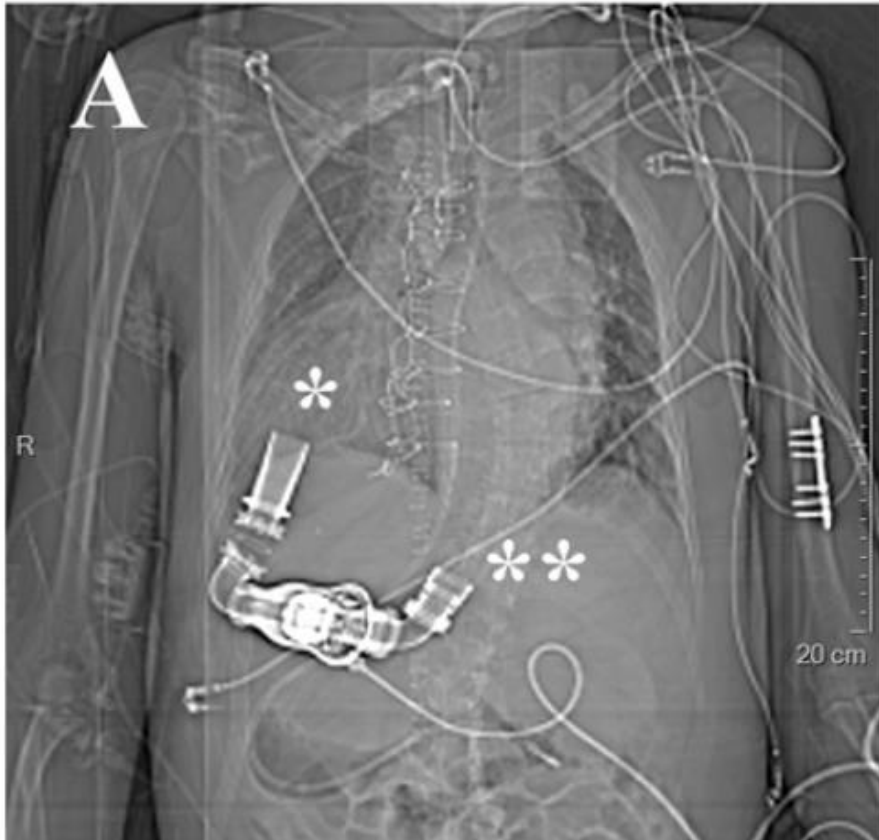


# Heart Failure in ACHD: *Increasing!*

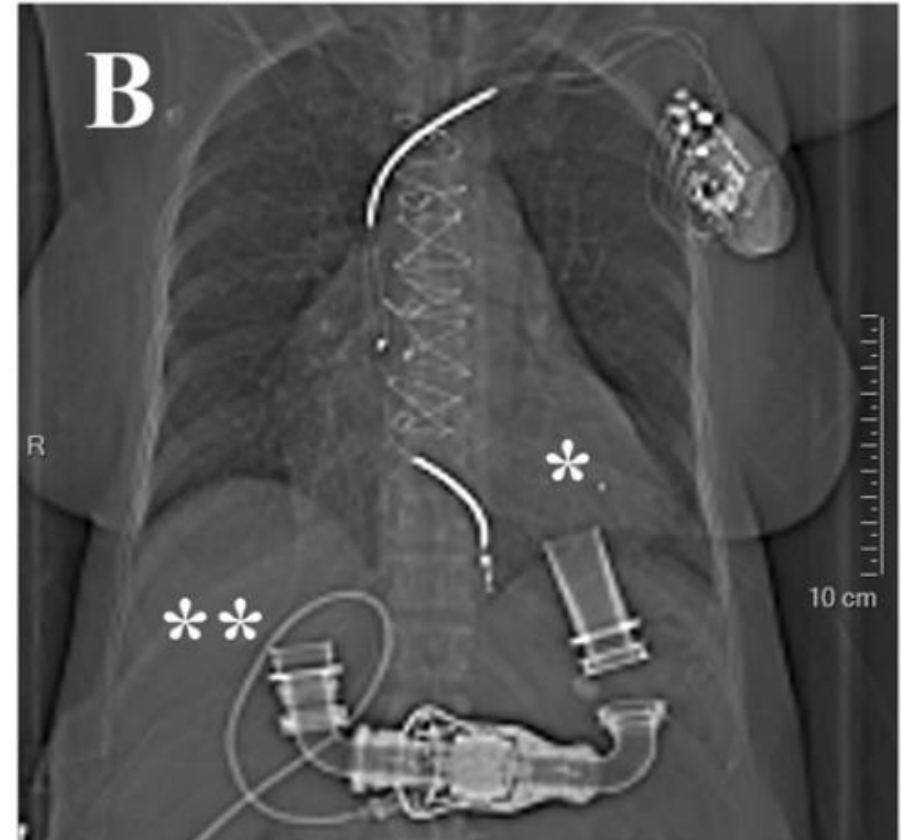


# Ventricular Assist Devices (VADs) in ACHD: *Geometric, Anatomic, and Bleeding Challenges!*

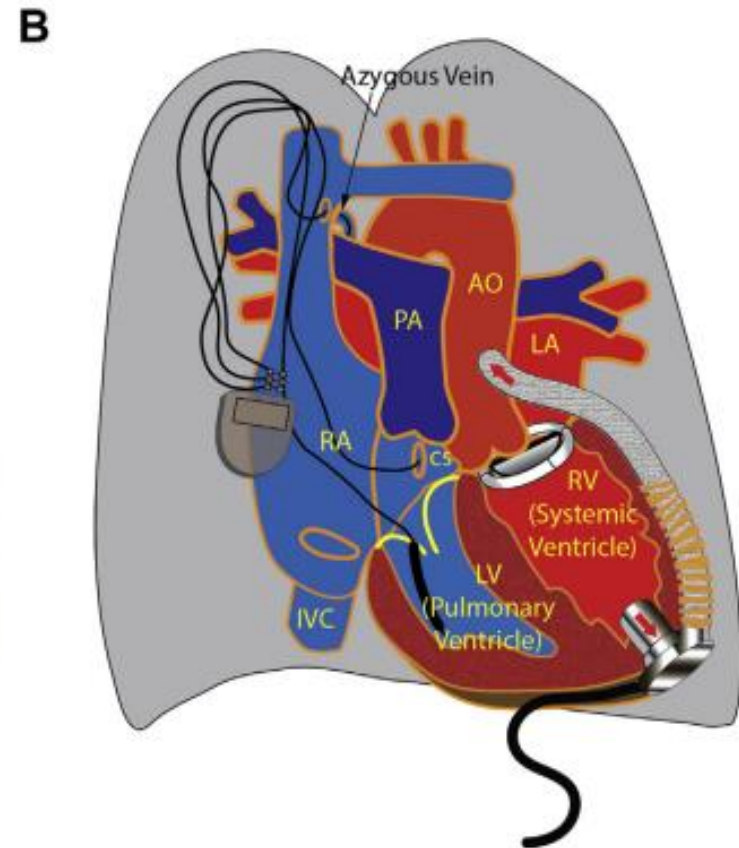
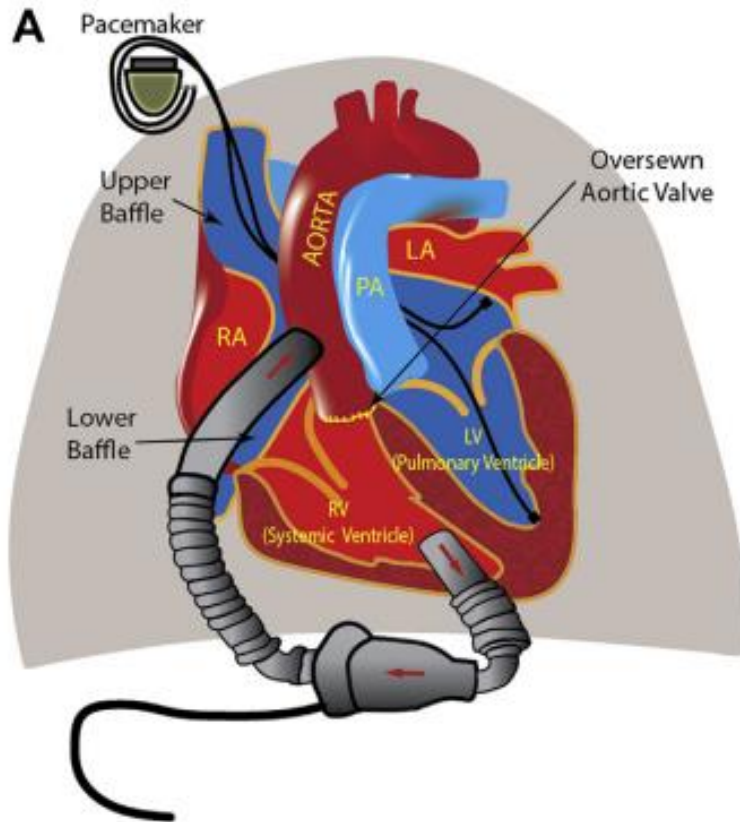
Dextrocardia



Normal heart, Levocardia

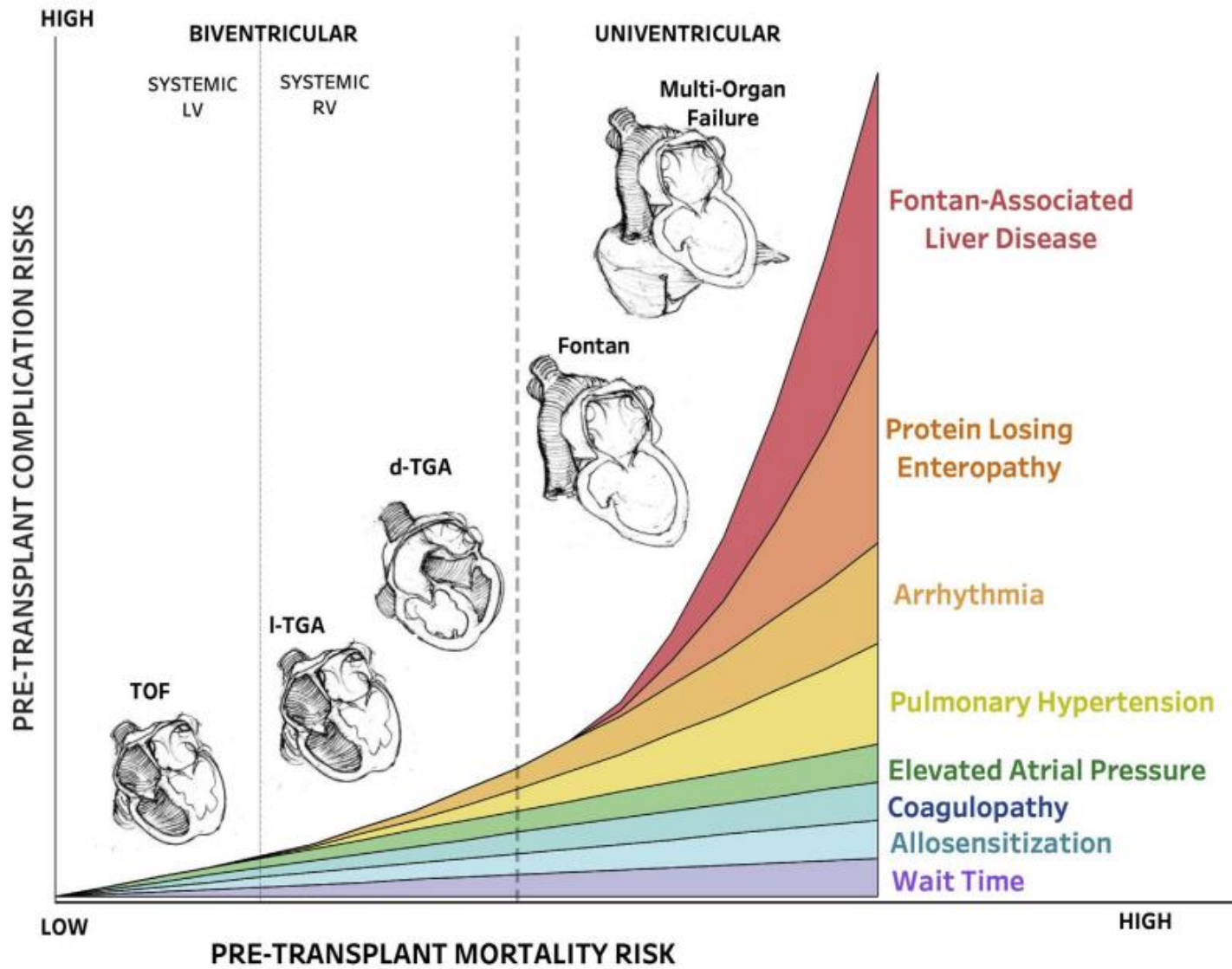


# VADs in D-TGA and L-TGA

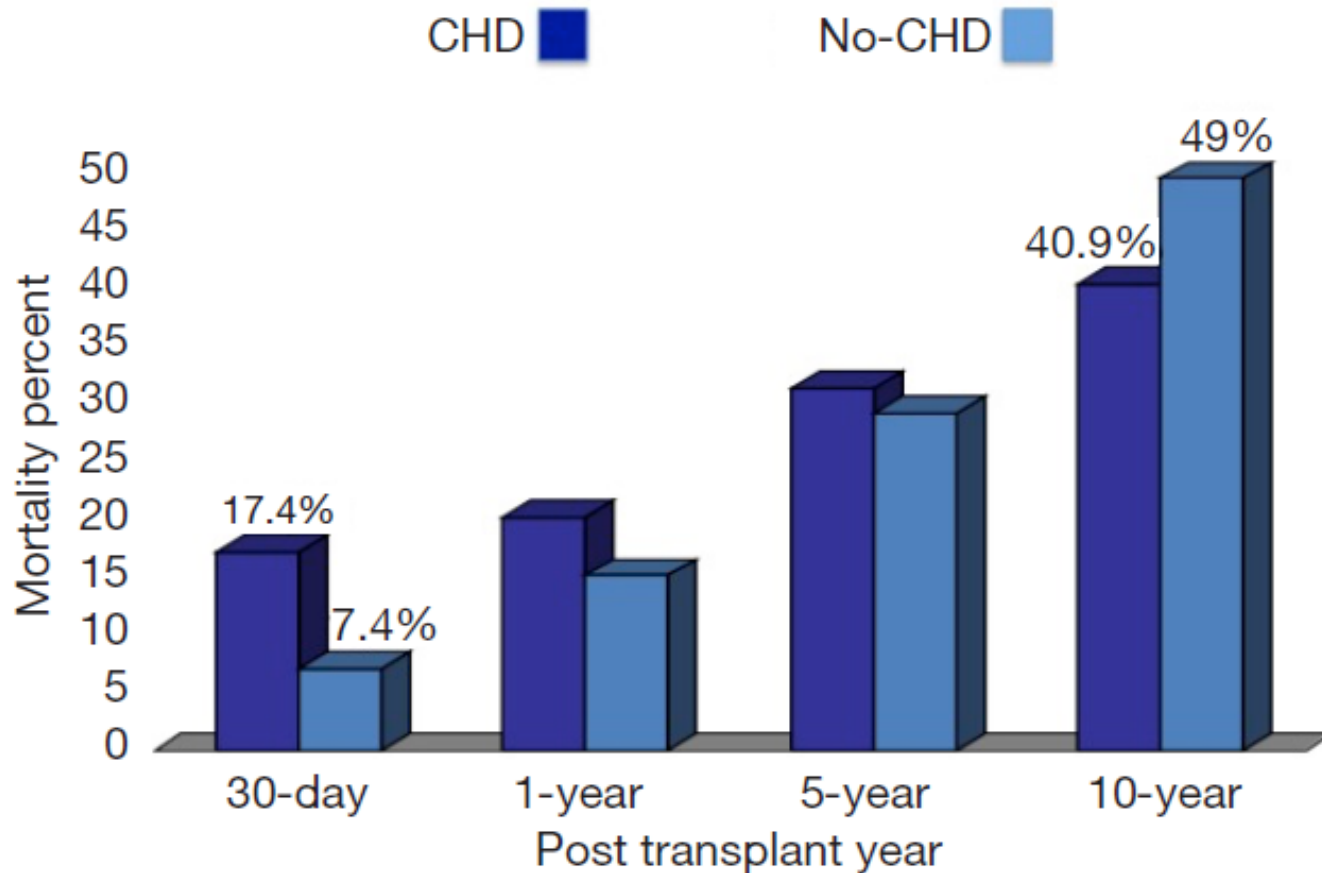


#10a: OHT is not a “good” option  
(but sometimes OHT is the ONLY option)





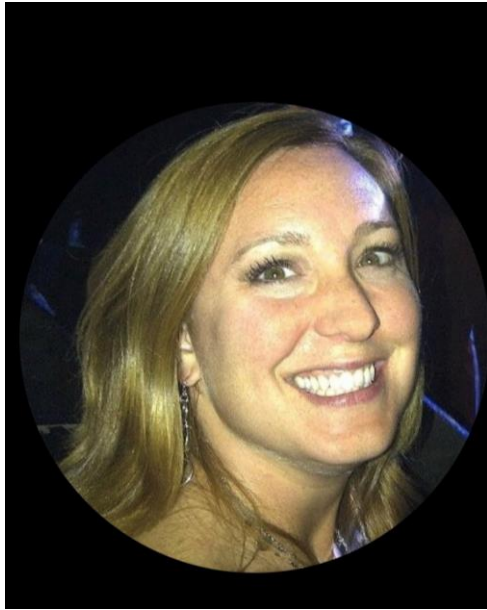
# AChD s/p Heart Transplant



# Need ACHD Help?

- ***Call us!!!***
- ***Cardiology on-call : 602-933-BEAT***

Susie Pepp, FNP



Jordan Awerbach, MD MPH



Wayne Franklin, MD





# THANK YOU! QUESTIONS?



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