

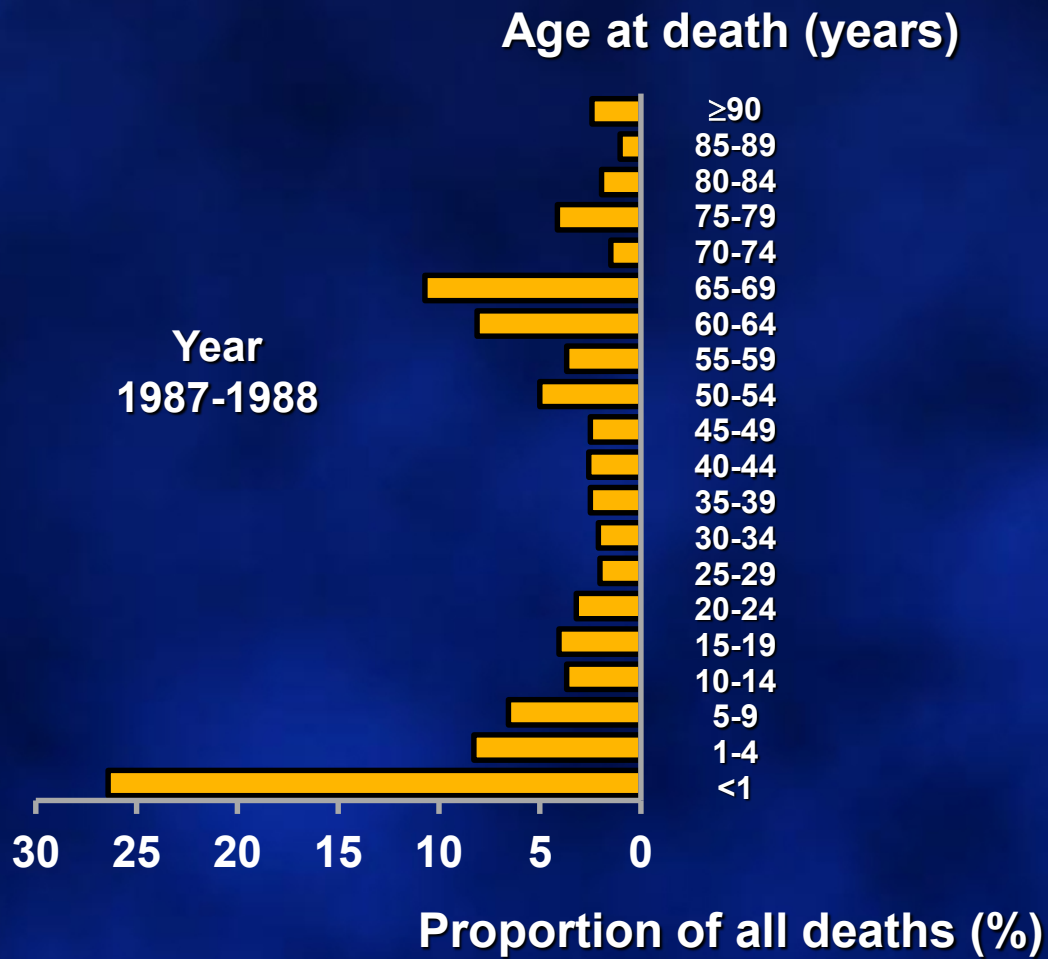
# Management of Heart Failure in Adult Congenital HD

**XXVIII GIORNATE CARDIOLOGICHE TORINES**  
**October 13-15, 2016, Turin**

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**Vice Chair, Clinical Practice, Cardiovascular Division**  
**Mayo Clinic, Rochester**

# Distribution of Age at Death

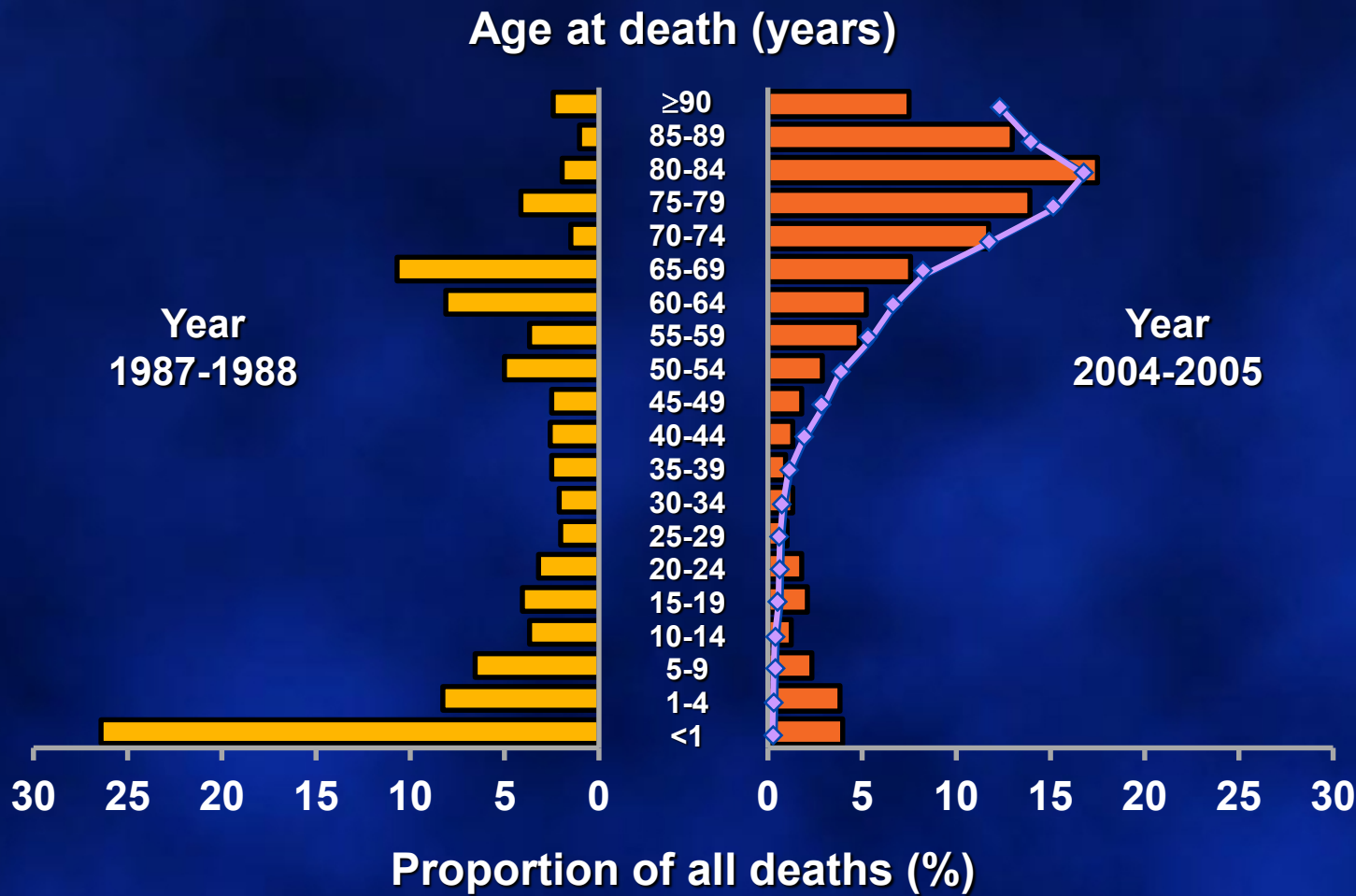
## 8,561 deaths occurred in 71,686 patients with CHD 1987-1988 and 2004-2005



# Distribution of Age at Death

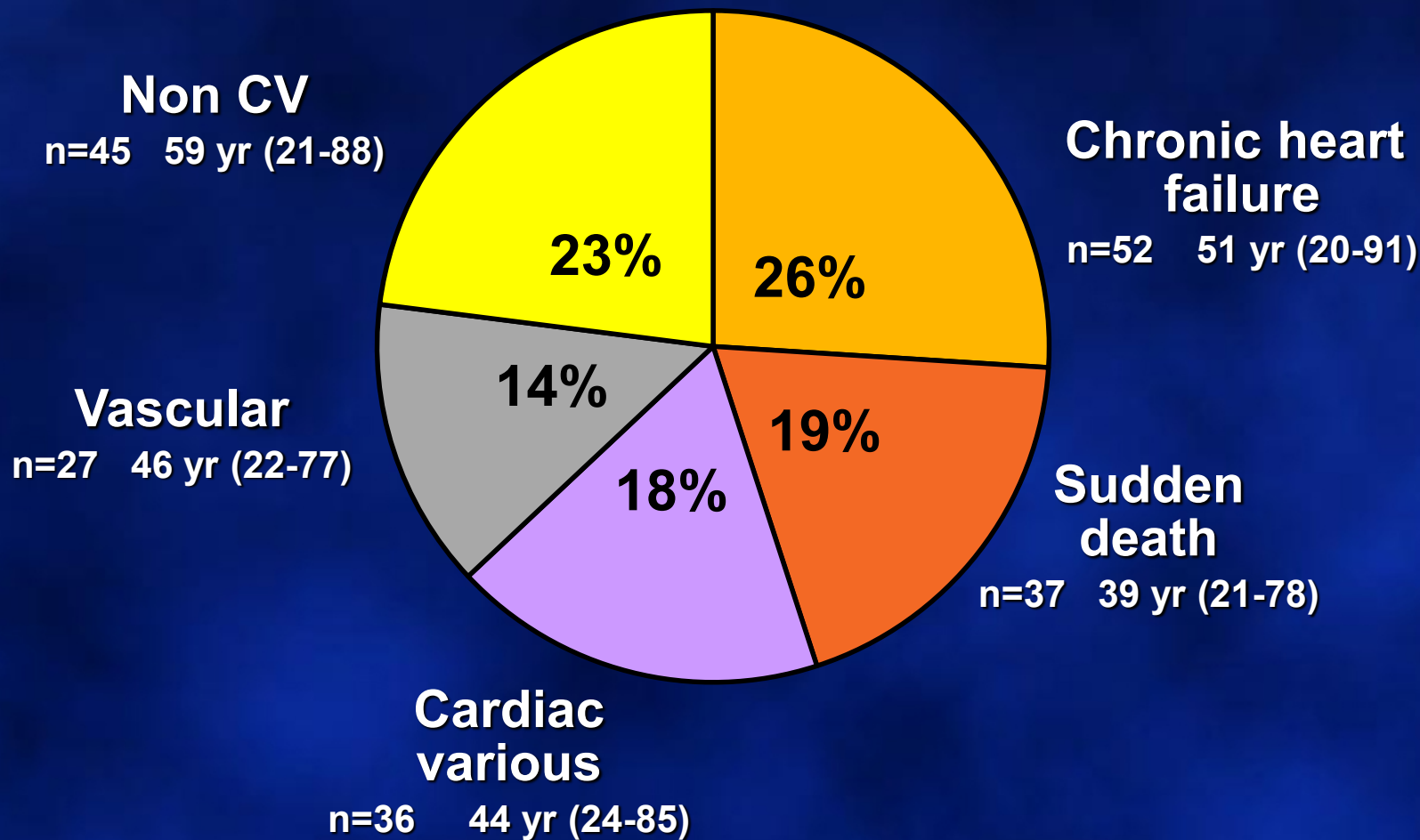
## 8,561 deaths occurred in 71,686 patients with CHD

### 1987-1988 and 2004-2005

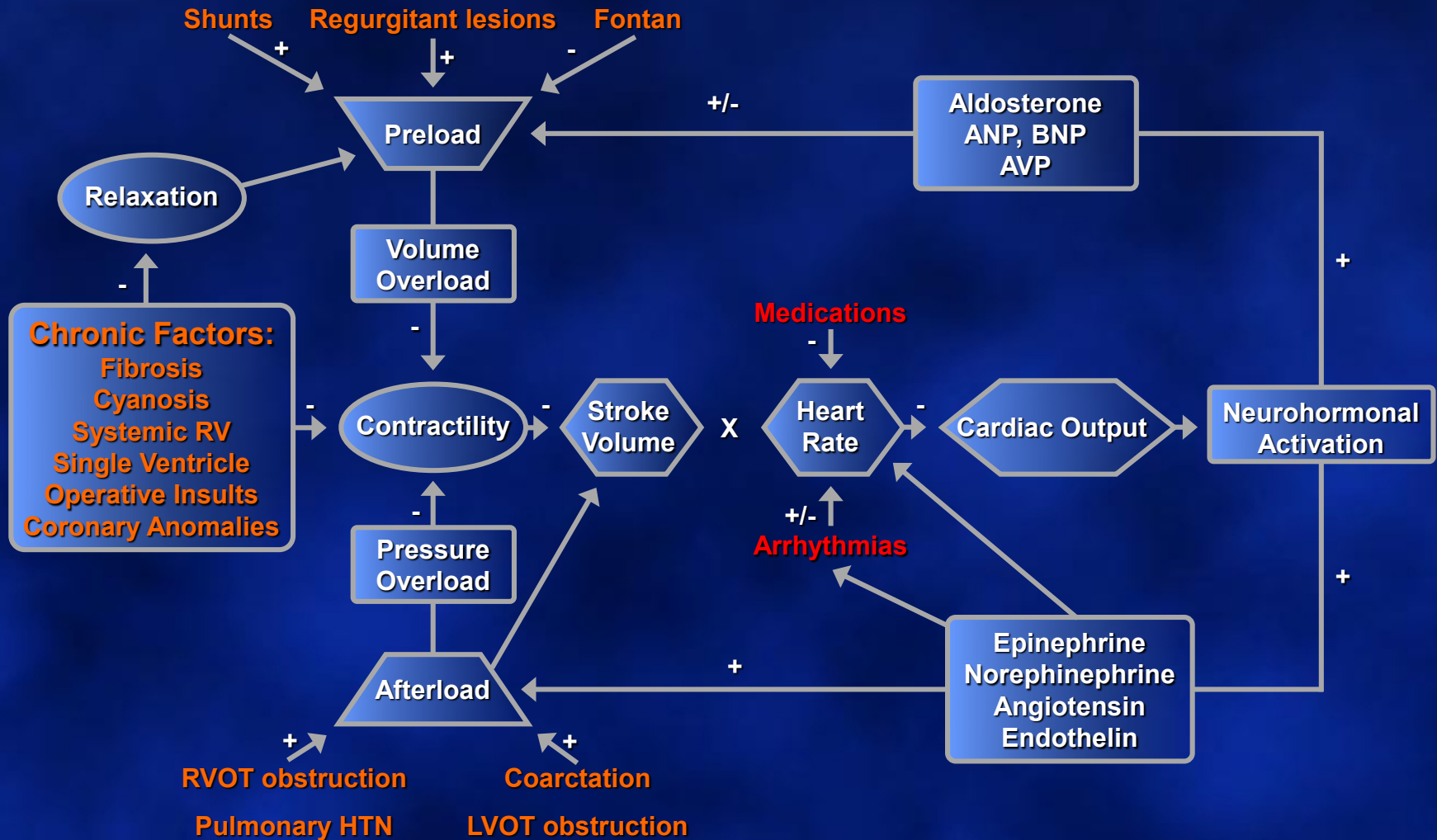


# Causes of, and Age at, Death in ACHD Patients

CONCOR Dutch Registry: 6933 ACHD, 24865 pt-yr FU



# Mechanisms of Heart Failure in Adults with Congenital Heart Disease

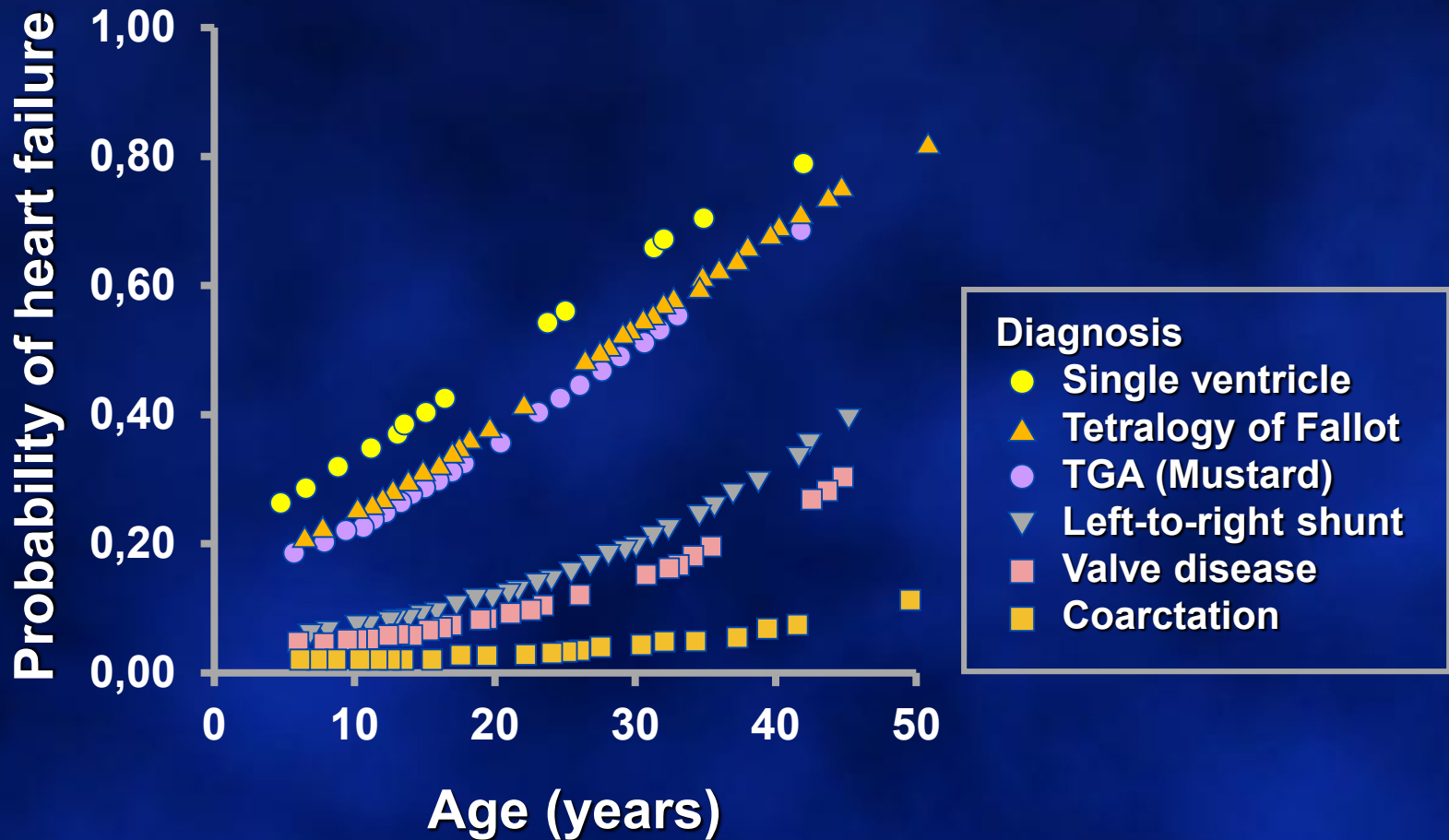


# Management of HF in Adult CHD

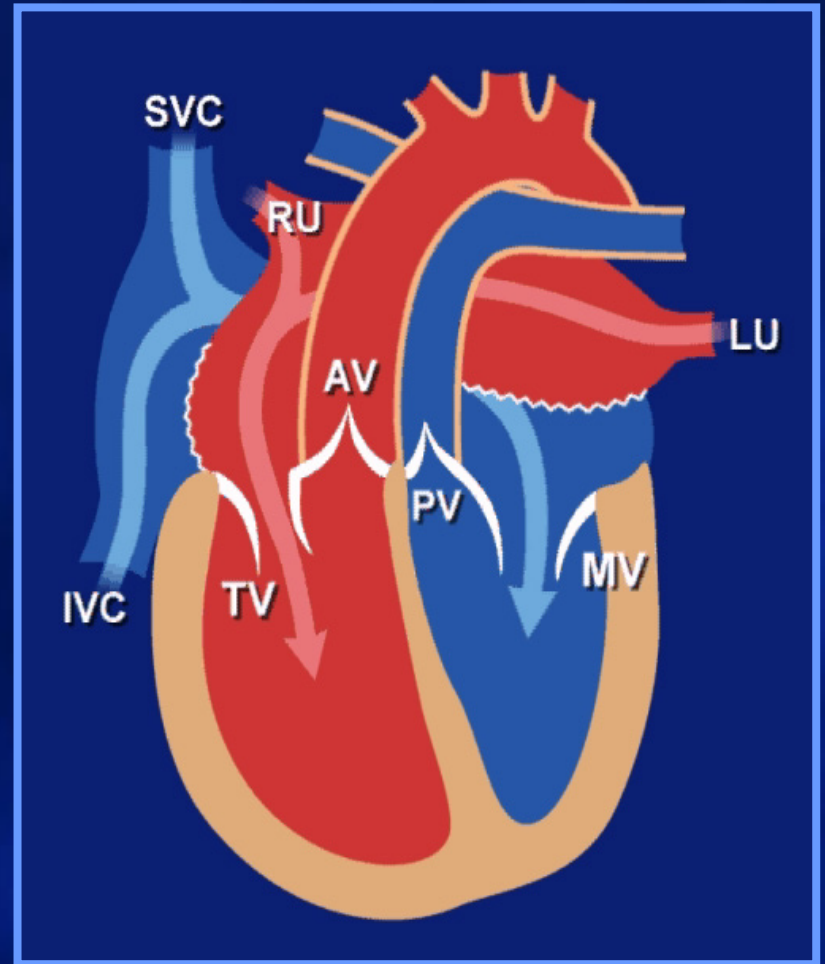
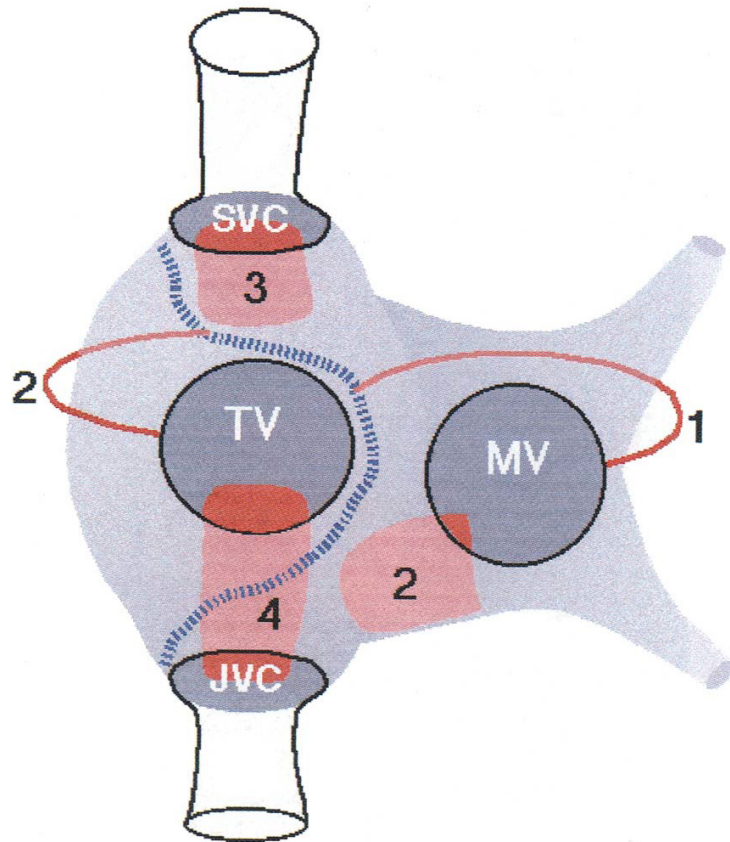
## Challenges

- **Diverse patient population with multiple mechanisms**
- **RV and/or LV can be the problem**
  - **Systolic and diastolic dysfunction**
- **Presenting signs/symptoms could be subtle**
- **Laboratory monitoring is suboptimal**
- **Major determinant of outcome**
- **Probability of HF increases with age**

# The Probability of Heart Failure Over Age and Type of Heart Defect



# d-TGA post Mustard/ Senning

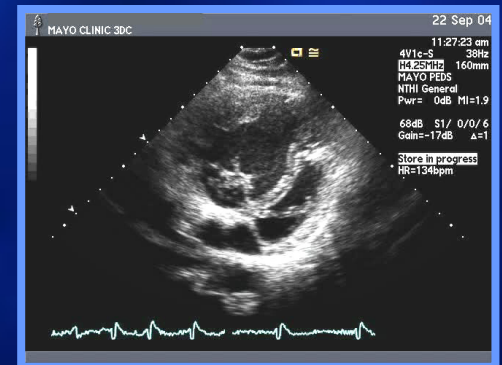


**Systemic RVD responsible for death in 40%**

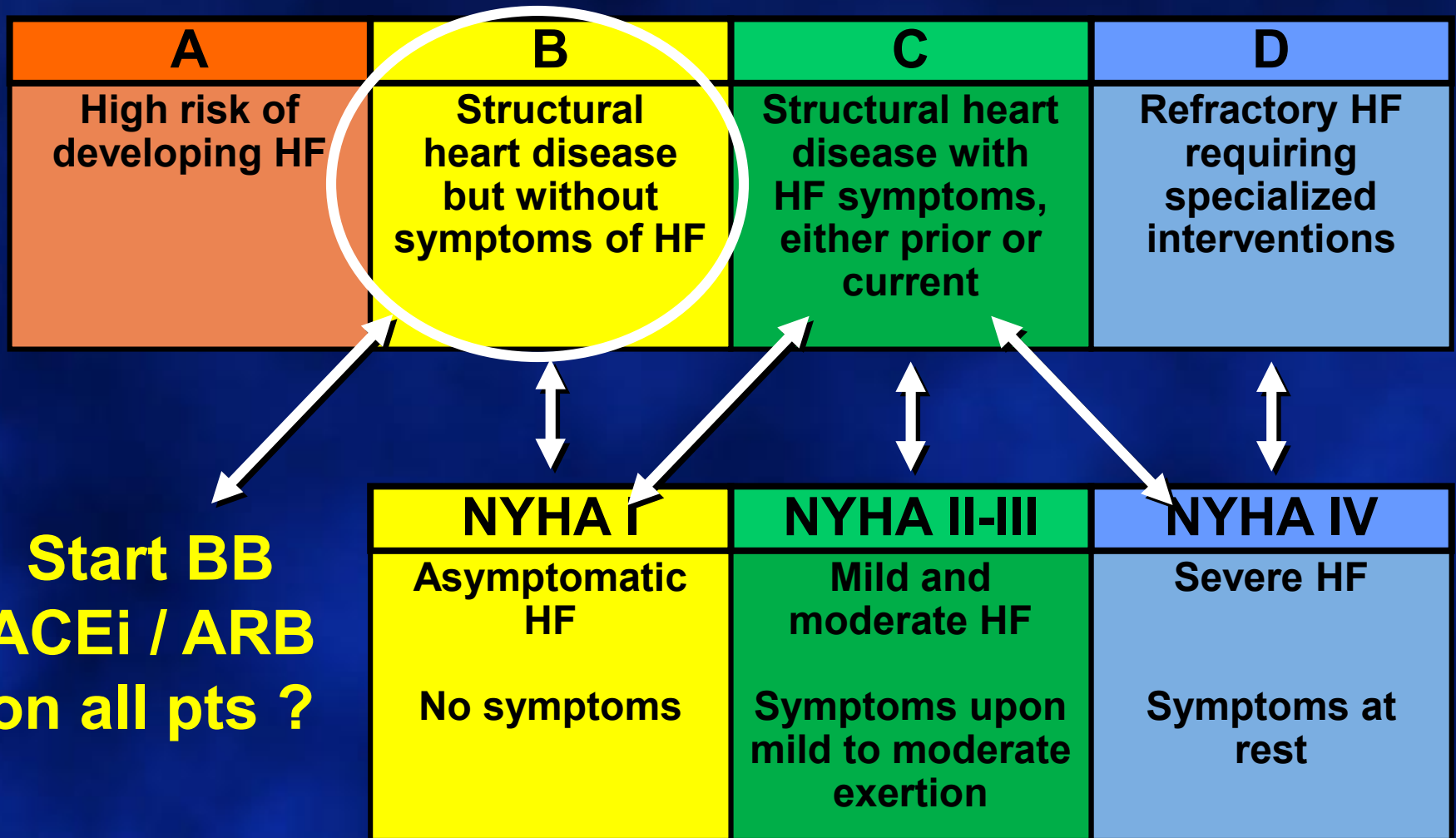


# d-TGA post Mustard/ Senning Systemic RV Dysfunction

- Poor structural /geometric adaptation to pressure overload
- Cyanosis and damage from operation
- Myocardial fibrosis
- Abnormal myocardial perfusion
  - Single RCA for systemic RV
- Tricuspid Reg. and Pulmonary HTN
- Fixed SV through stiff atrial baffles/ SND
- Atrial arrhythmias



# Classification of HF: Comparison Between ACC/AHA HF Stage and NYHA Functional Class



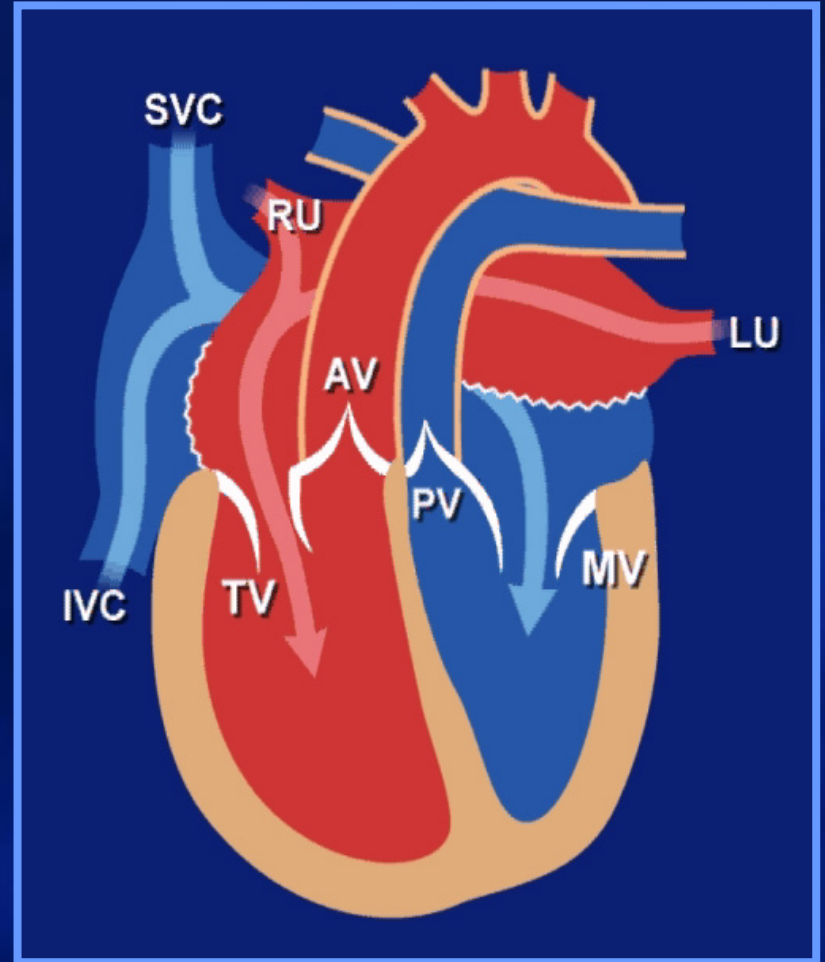
# Summary of Trials with $\beta$ -Blockers, ACE Inhibitors, ARB in Patients with a Systemic Right Ventricle

	Agent	TGA/ ccTGA	No.	Follow-up (mo)	MRI	VO <sub>2</sub> max	NYHA	Pro/retro
<b><math>\beta</math>-blocker</b>								
Lindenfeld et al	Carvedilol	ccTGA	1	7	↑	ND	ND	Pro
Giardini et al	Carvedilol	Both	8	12	↑	=	↑	Pro
<div style="border: 2px solid orange; padding: 10px; background-color: yellow;"> <p><b>Widely used conventional HF therapy might not apply and are not justified</b></p> </div>								
<b>ATII antagonist</b>								
Hecnter et al	various	TGA	14	24	=	=	ND	Retro
Robinson et al	Enalapril	TGA	9	12	ND	=	ND	Pro
Therrien et al	Ramipril	TGA	17	12	=	=	ND	Pro
Dore et al	Losartan	Both	29	3.5	ND	=	ND	Pro
Lester et al	Losartan	TGA	7	2	↑	ND	ND	Pro

# Treat Co-morbidities + diuretics

- **Coronary artery disease**
- **Diabetes**
- **Hyperlipidaemia**
- **Hypertension**
- **Iron deficiency and anaemia**
- **Kidney dysfunction**
- **Lung disease**
- **Obesity and sleep apnea**

# 32 YO S/P Mustard with CHF

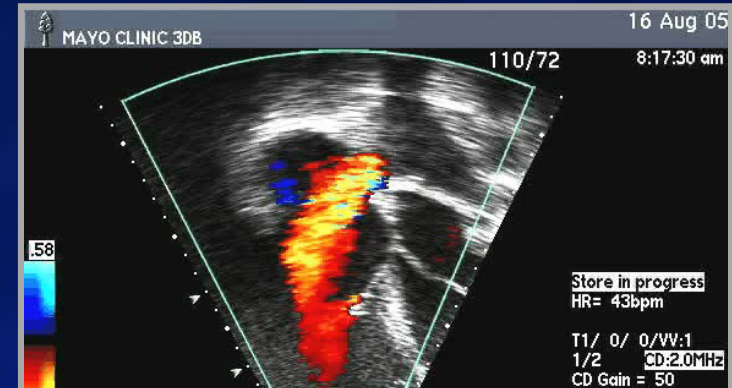
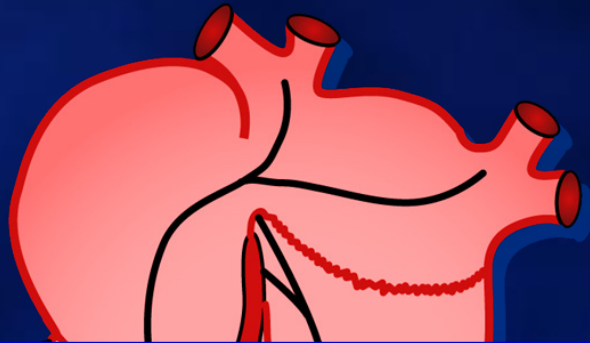


# d-TGA post Mustard/ Senning

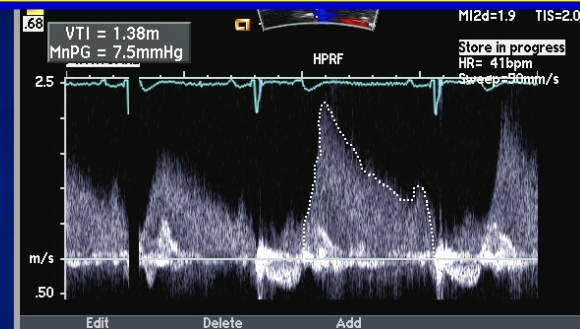
- Systemic RV dysfunction 90%
  - RV Failure (late) 40%
  - TV regurgitation 40%
- Pulmonary HTN 8-12%
- Atria arrhythmias 60%
  - IART, At Flutter
- Heart block/ AJ Rhythm 40%
- Baffle obstruction (early) 15- 30 %
  - SVC > IVC > Pul. Venous
- Baffle leak ( small) 25%
  - Paradoxical embolism ( SVT, PPM)

**Most can precipitate heart failure**

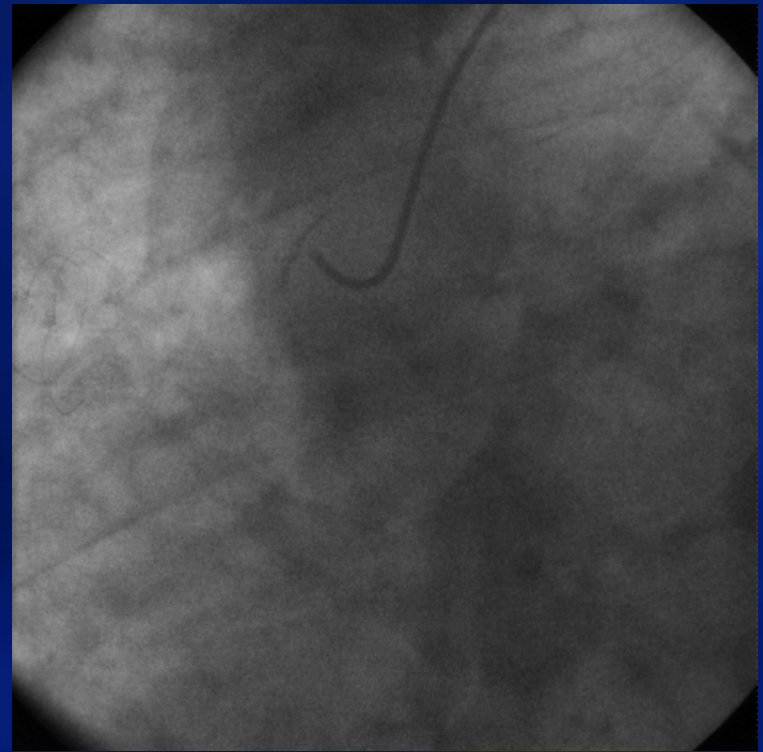
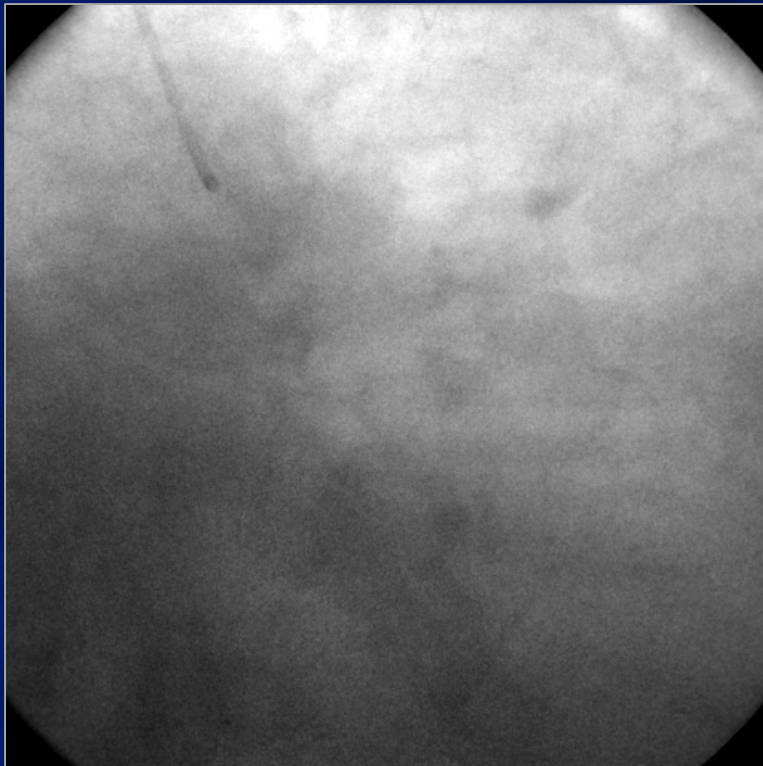
# 32 YO S/P Mustard with CHF



**Management of HF in Adult CHD:**  
**Look for lesion specific treatable and reversible causes of HF**



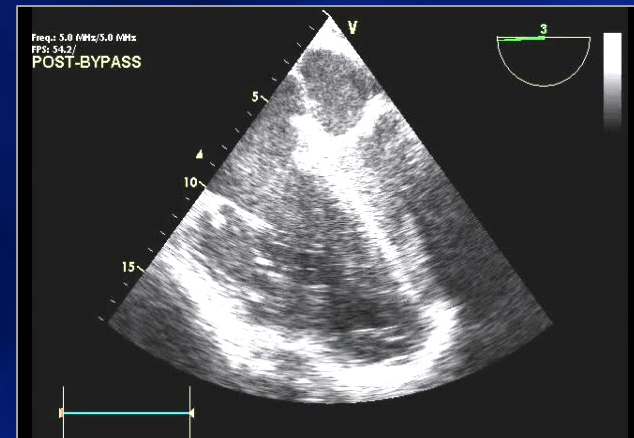
# 60 yr old survivor of repaired TOF Myocardial Infarction





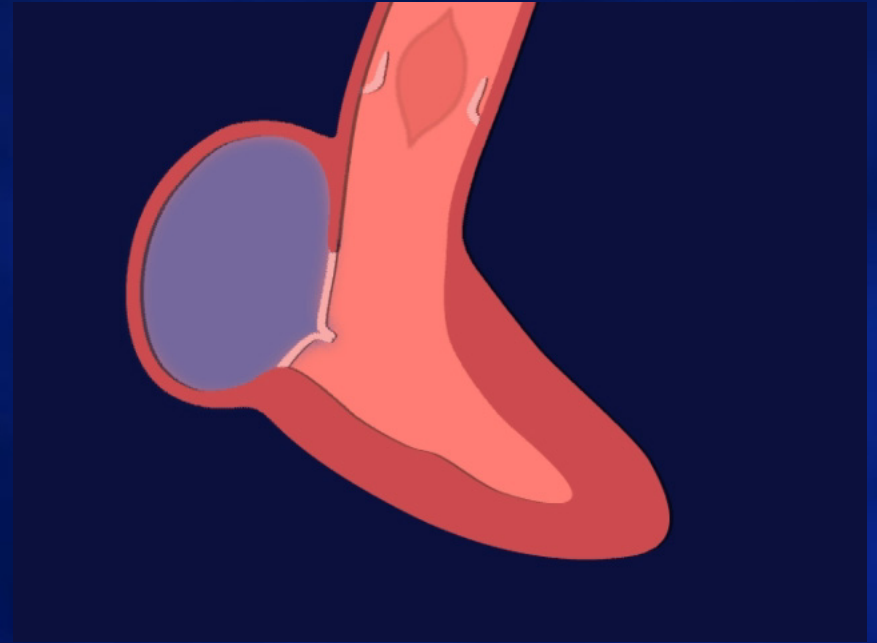
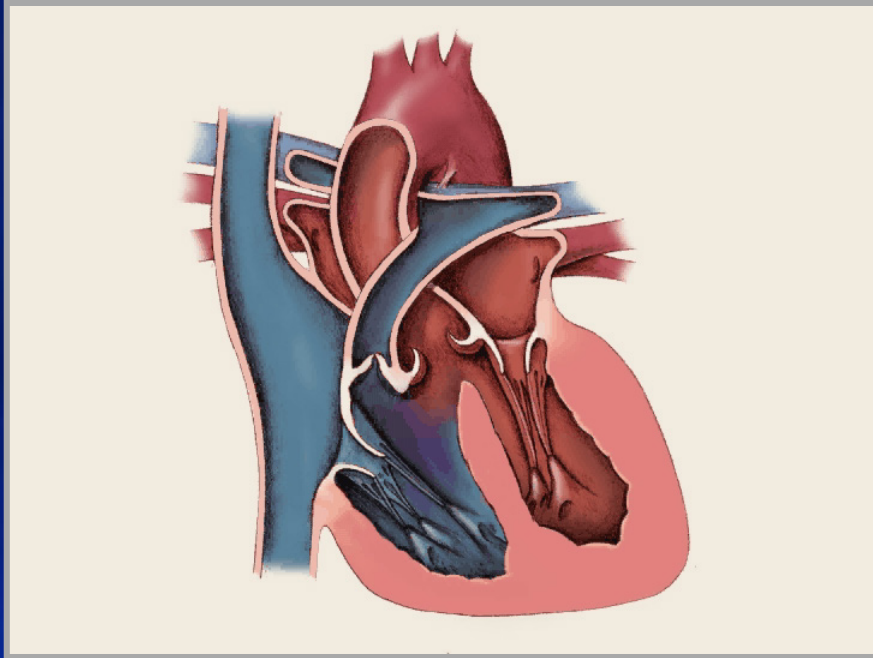
# 60 yr old survivor of repaired TOF CABG day 5

- Very dense adhesion
- Both IMAs were truncated at prior surgery
- SVG to LAD, OM, PDA
- IABP
- SVG to PDA
- Open chest



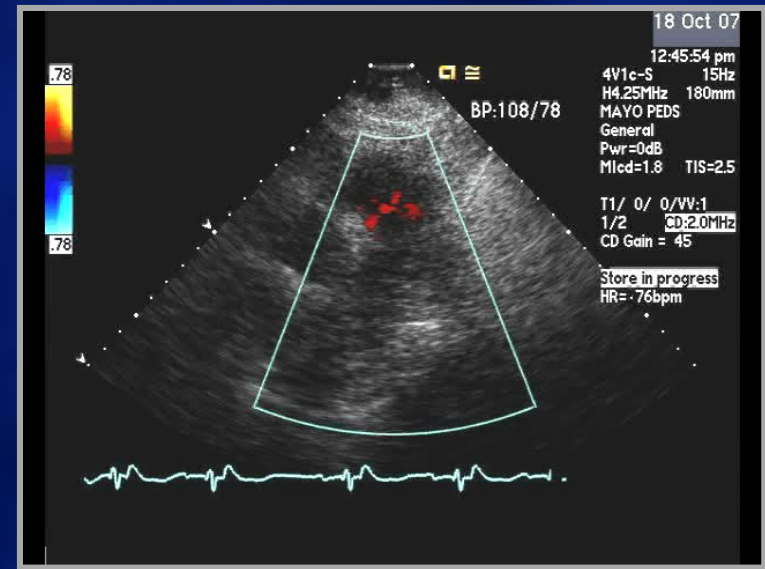
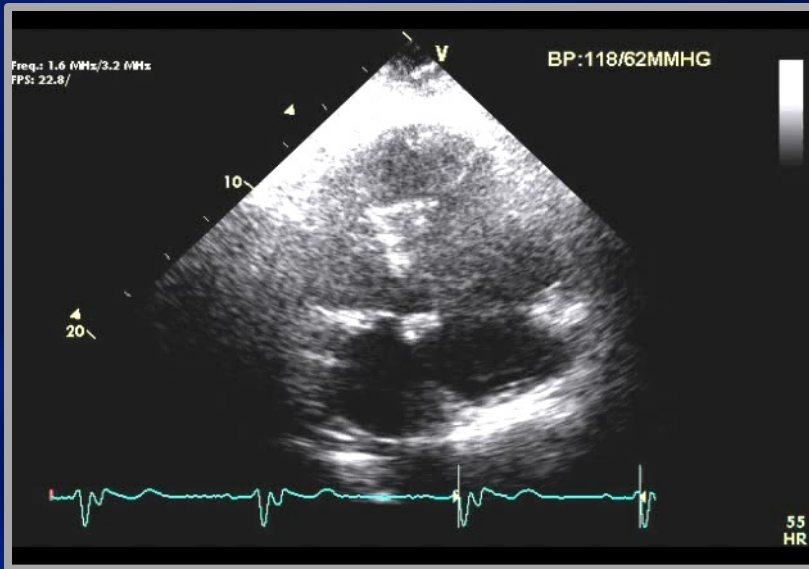
**Dismissed home on day 26**

# Tetralogy Of Fallot



**30 yrs survival 86%**

# 60 yr old survivor of repaired TOF CHF post CABG following MI

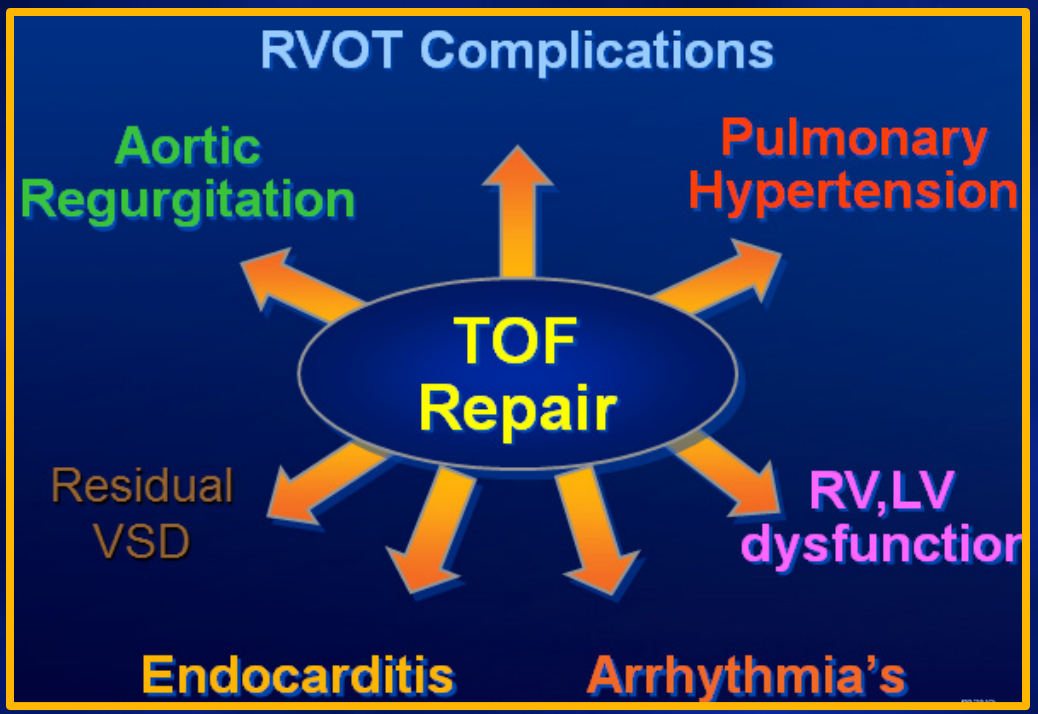


## Severe Pulmonary Regurgitation

# Clinical Pearls in Taking Care of CHD

## Post op CHD

Be aware of  
and look for  
post-operative  
residua and  
sequelae

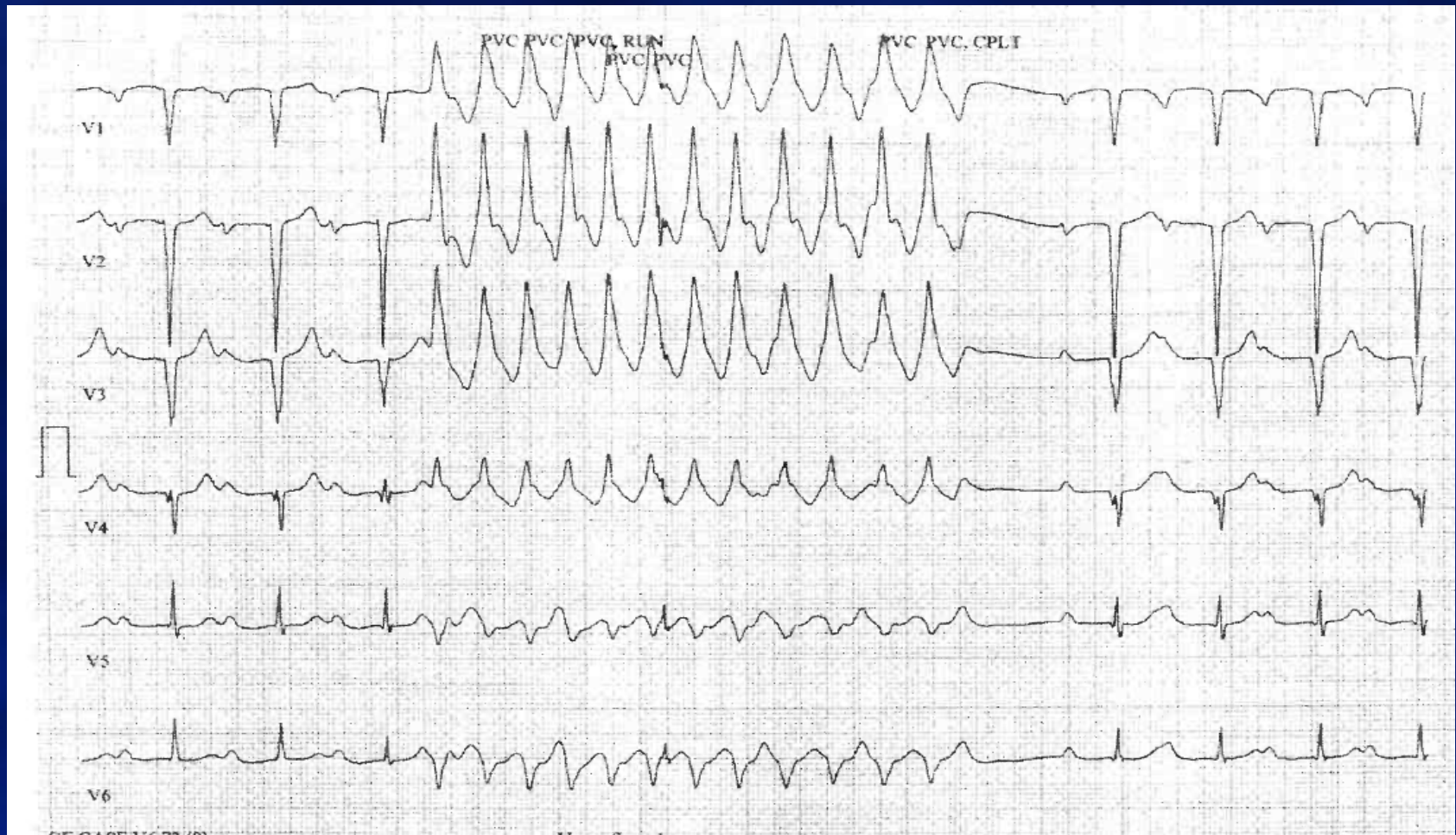


# HF Treatment in Adult CHD

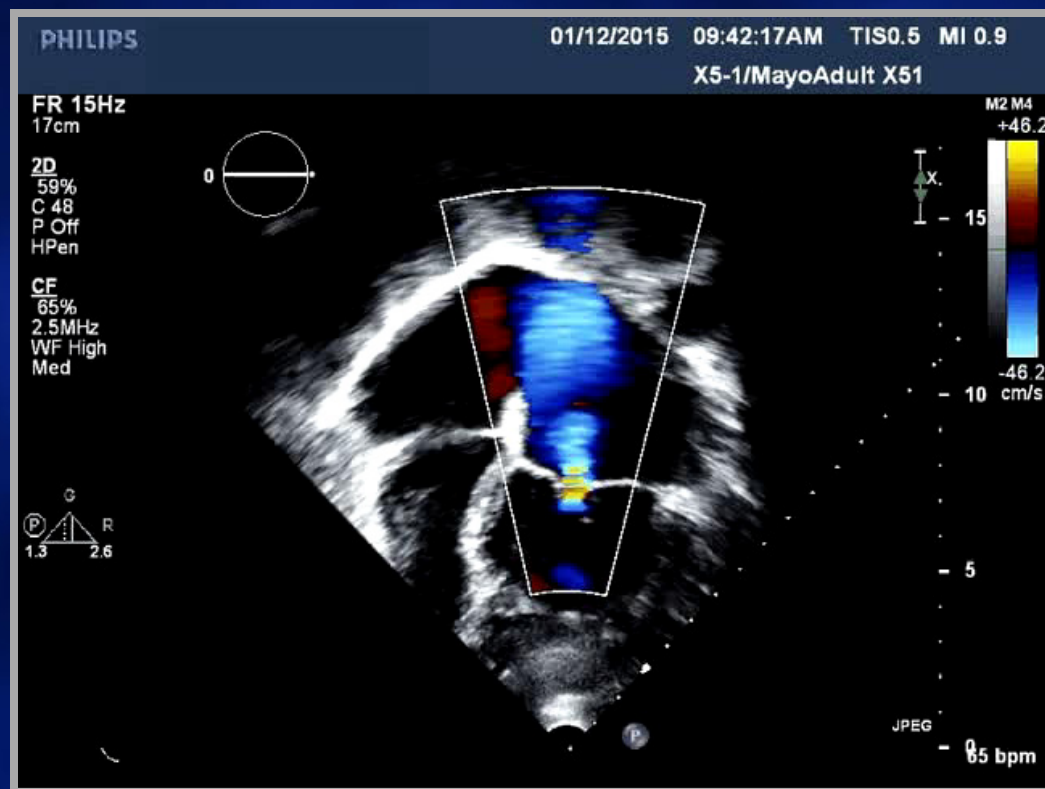
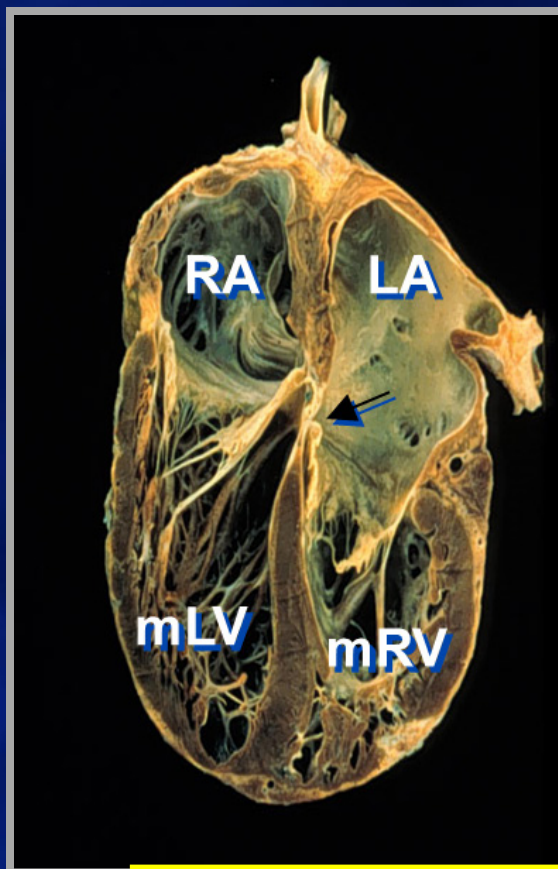
Evaluation of HF in adults with CHD must be

1. Individualized based on the underlying anatomy
1. Should include evaluation for lesions amenable to percutaneous or surgical interventions
  2. residual shunts
  3. baffle stenosis
  4. valvular or conduit dysfunction
  5. collateral vessels

# 29 YO with L-TGA and Heart Failure



# 29 YO with L-TGA and Heart Failure



30% develops CHB or 2%/yr

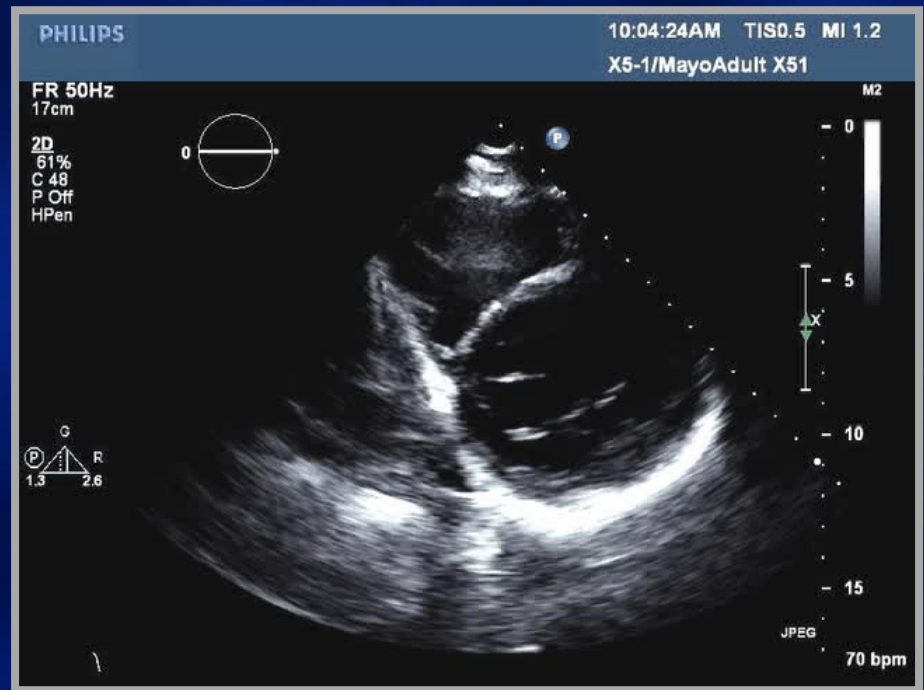
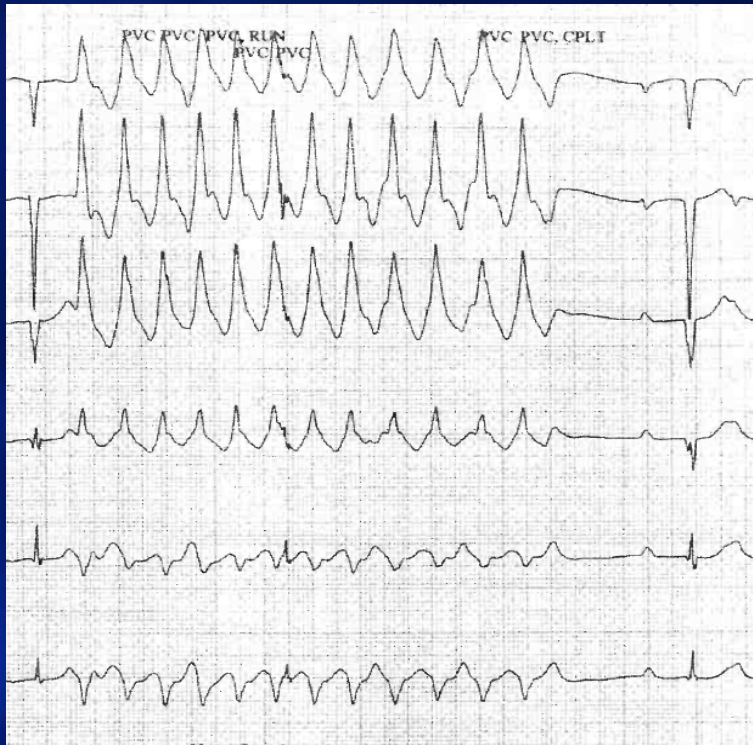
# Congenitally Corrected TGA or L-TGA

## Left AV Replacement Should be considered

- > moderate TV regurgitation
- onset of ventricular deterioration
  - Best when EF > 40%
- atrial arrhythmias
- ↓ exercise capacity



# 29 YO with L-TGA and Heart Failure

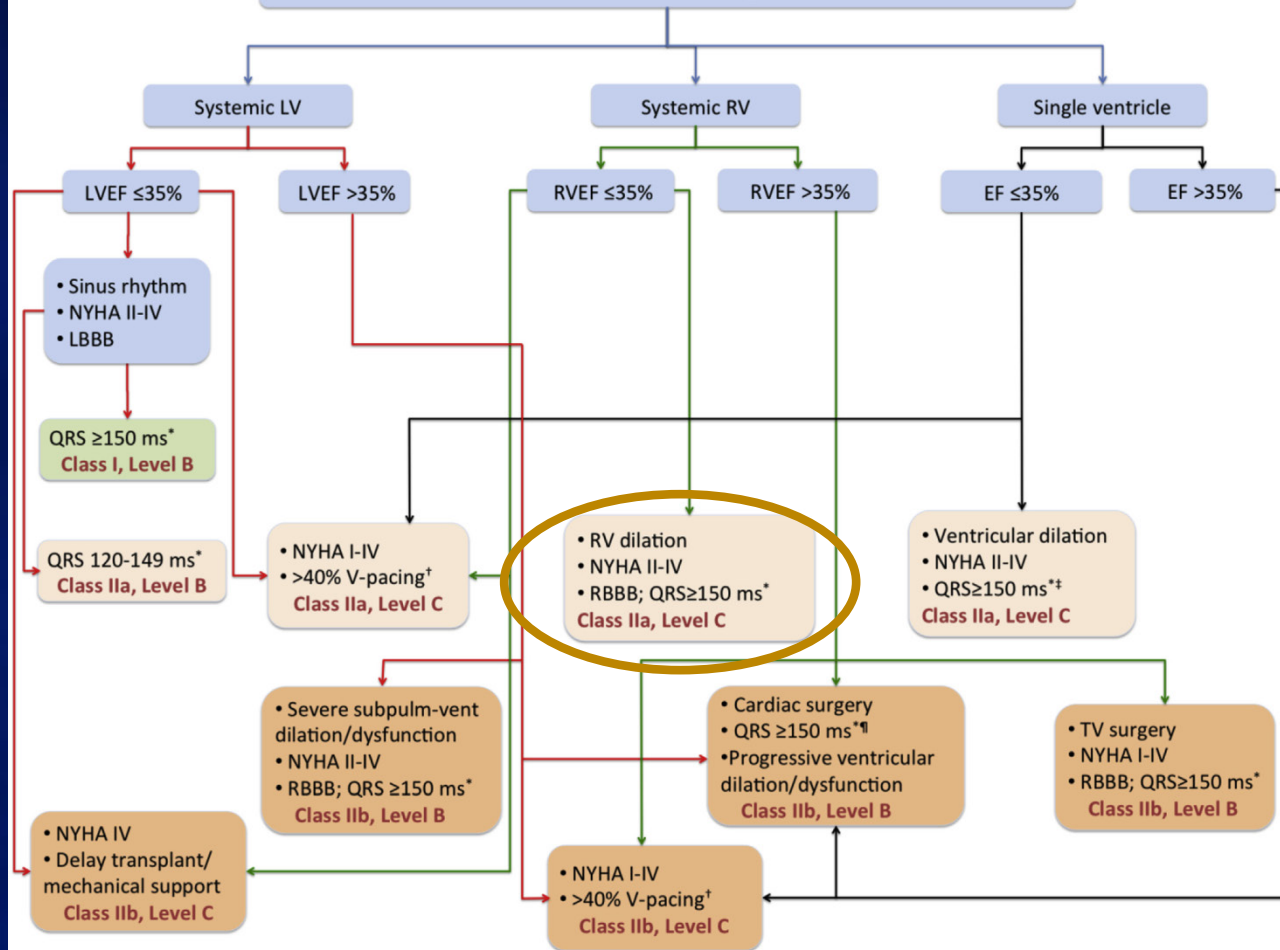


Systemic RV EF 34 %

# Class I ICD therapy in adults with CHD

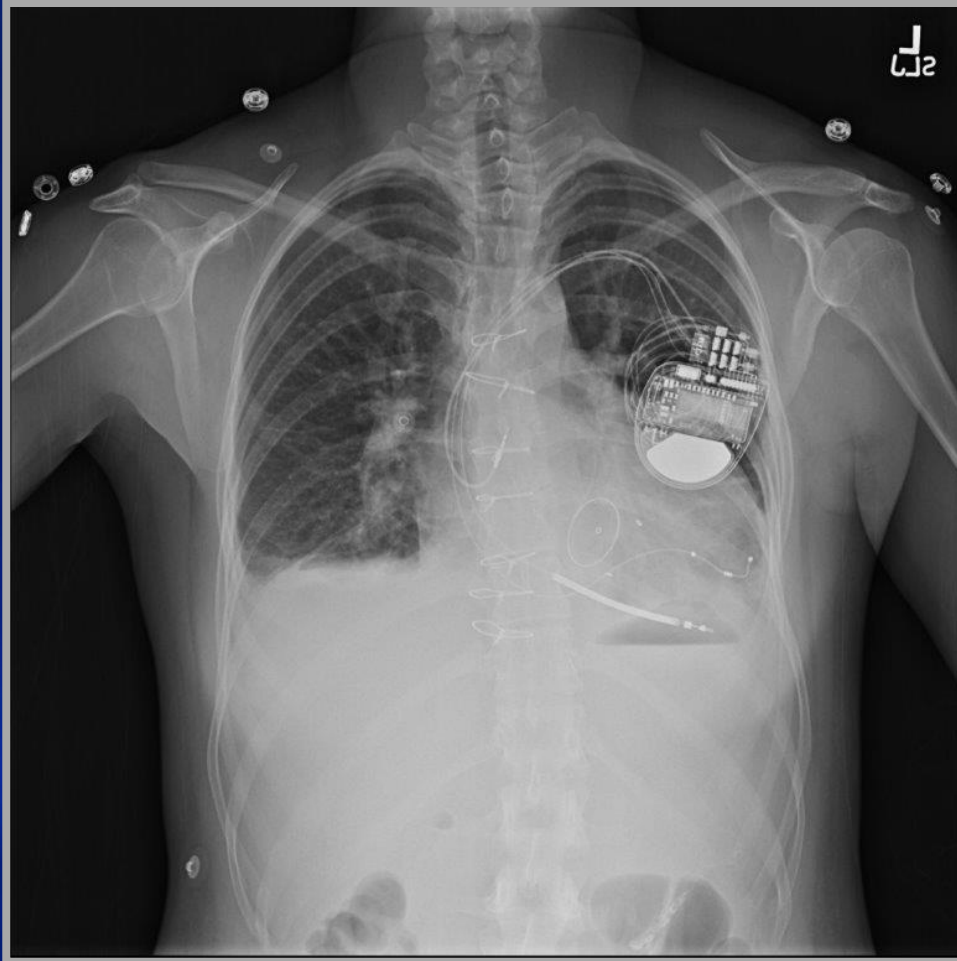
- Survivors of *cardiac arrest* due to VF/ sustained VF after evaluation to define the cause of the event and exclude any completely reversible etiology (LOE: B)
- *Spontaneous sustained VT* who have undergone hemodynamic and EP evaluation (LOE: B), RFA surgery may be alternative or adjunct to ICD in selected patient
- Systemic LV EF  $\leq 35\%$ , biventricular physiology, and NYHA class II or III symptoms (LOE: B)

## CRT indications in adults with congenital heart disease



LV denotes left ventricle; RV, right ventricle; EF, ejection fraction; NYHA, New York Heart Association functional class; LBBB, left bundle branch block; RBBB, right bundle branch block; V-pacing; ventricular pacing: subpulm-vent, subpulmonary ventricular; TV, tricuspid valve  
 \*Spontaneous or paced  
 †New or replacement device implantation with anticipated requirement for >40% ventricular pacing, intrinsically narrow QRS complex; single site pacing from the systemic ventricular apex/mid-lateral wall may be considered as an alternative  
 ‡RBBB or LBBB  
 †Complete bundle branch block ipsilateral to the systemic ventricle

# 29 YO with L-TGA + NSVT + EF 34%



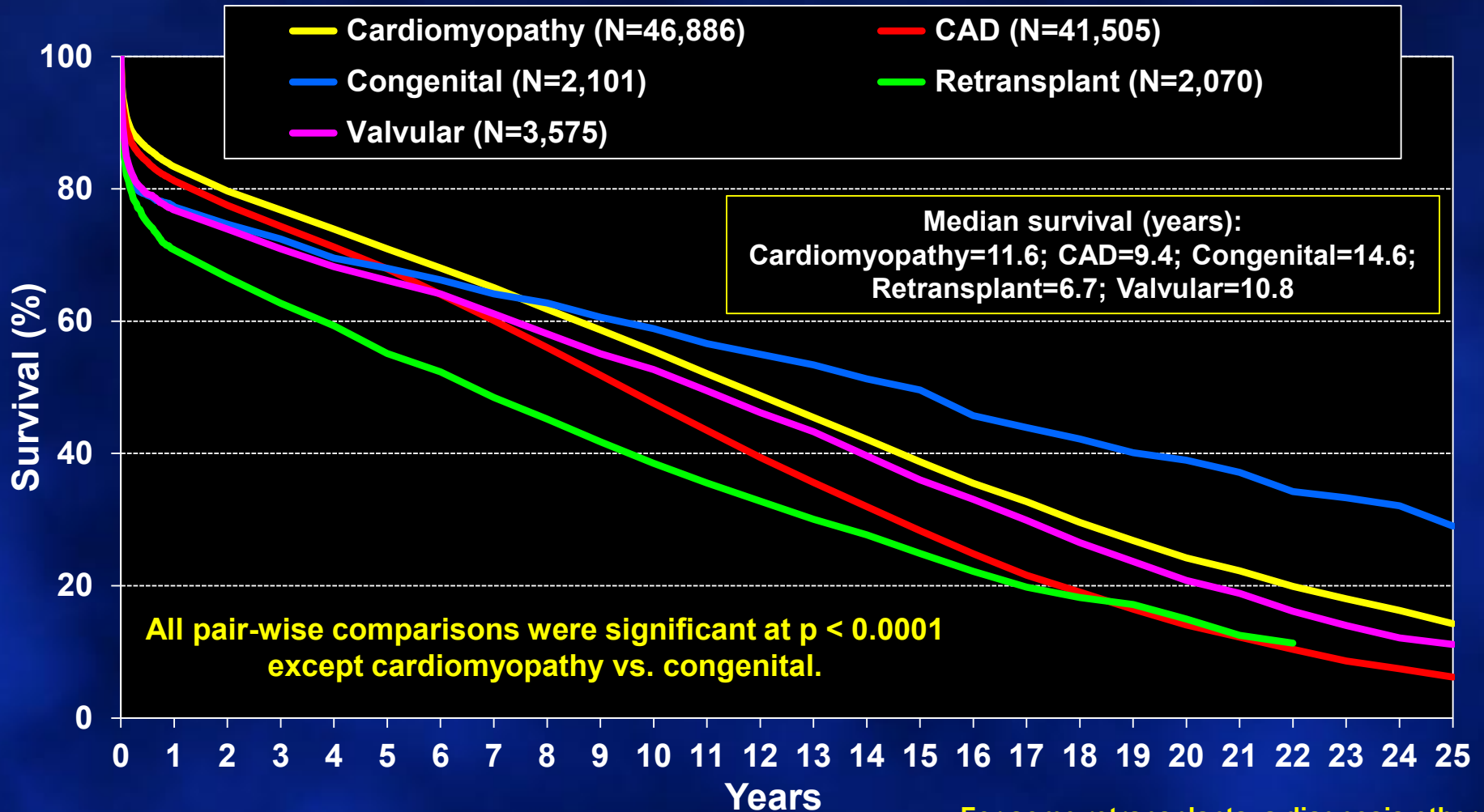
# CRT in Adult CHD

- Prevalence of dyssynchrony in ACHD unknown with no prospective studies or clear criteria
  - Used criteria: NYHA  $\geq$  II and QRS  $\geq$  150 msec
  - 9.3 % of Mustard and 6.1% of L-TGA will qualify
- Three retrospective studies of CRT in ACHD
  - Clinical response not uniformly defined and included improvement in EF or NYHA in 32–87%
  - rTOF with LV dysfunction and Systemic RV especially when needing RV pacing
- 40% awaiting OHT who received CRT were delisted

# Adult Heart Transplants

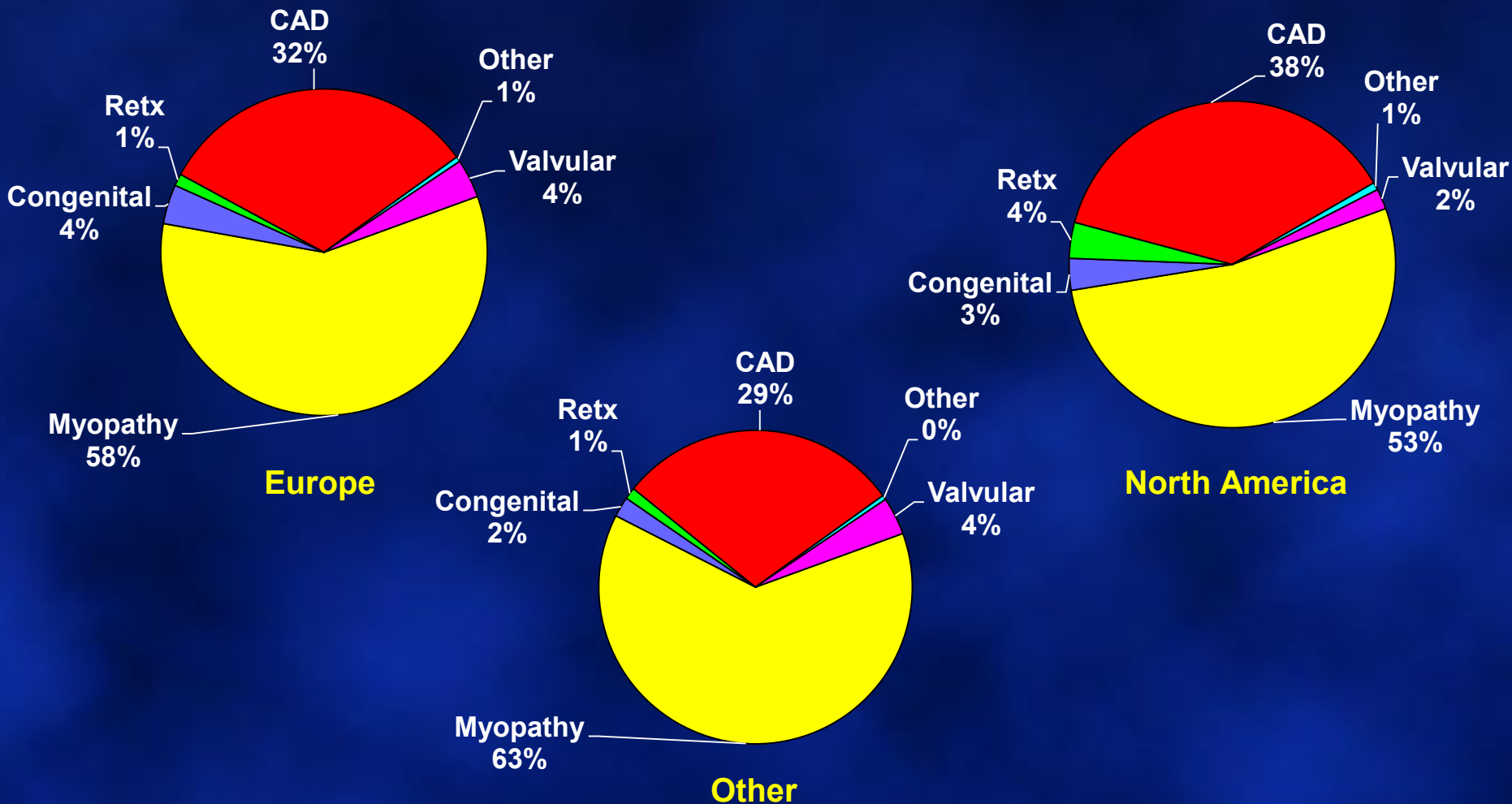
## Kaplan-Meier Survival by Diagnosis

(Transplants: January 1982 – June 2013)



For some retransplants, a diagnosis other than retransplant is reported, so the total number of retransplants may be greater.

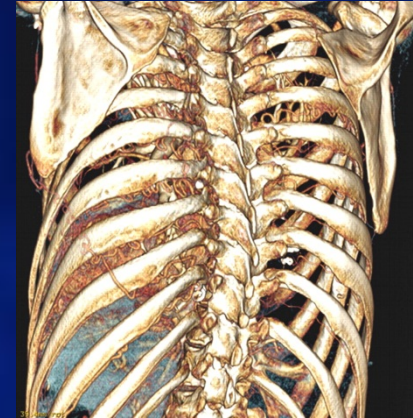
# Adult Heart Transplants Diagnosis by Location (Transplants: January 2009 – June 2014)



For some retransplants a diagnosis other than retransplant was reported, so the total percentage of retransplants may be greater.

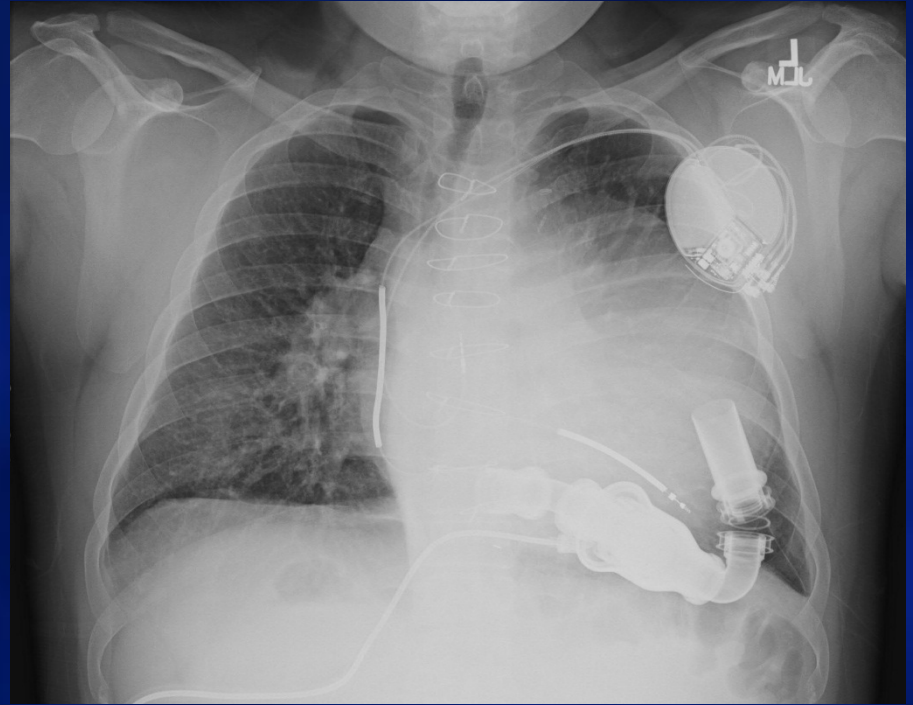
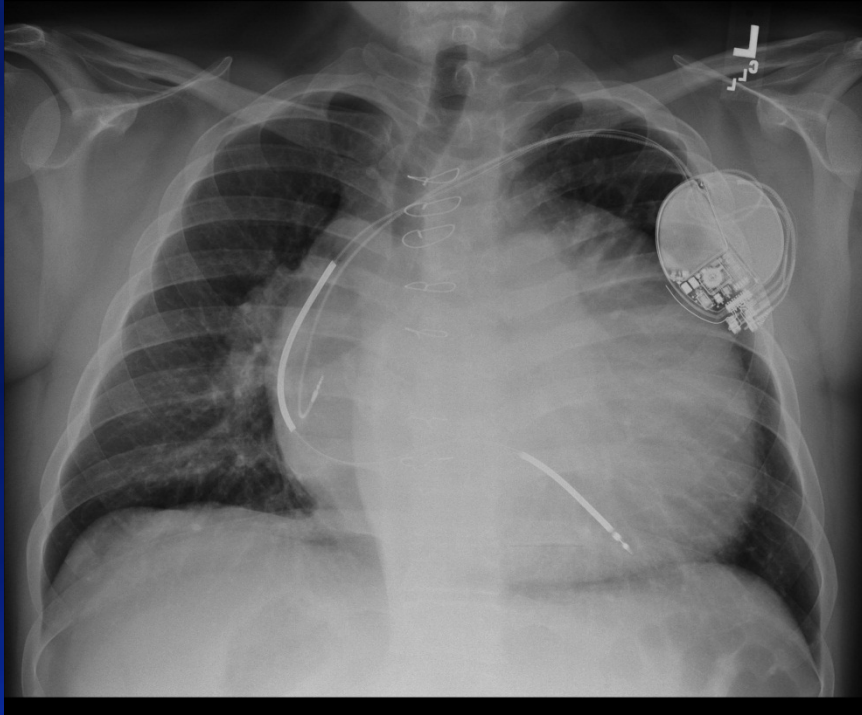
# Factors that discourage listing/OHT

- Need additional reconstructive surgery of great Art/Veins
- Multiple prior sternotomies/ thoracotomies
- Presence of aorto-pulmonary collaterals
- Level of pre-formed HLA and higher PRA
- Malnutrition and debilitation
- Active Hep C infection: 5x if prior operation before 1992
- Presence of renal or liver impairment and PHTN
  - Consideration for multi-organ transplantation





# LVAD in Adult CHD



**33 YO with repaired complex L-TGA  
S/P Heart Mate II May 2010**

# LVAD in Adult CHD

- Limited experience: L-TGA, D-TGA post Mustard, Fontan
- Challenges:
  - Anatomic complexity/sternotomies complicate insertion
  - Likely to have RV Dysfunction, PHTN, shunt
  - Different VAD type and configuration for different CHD
- Co-morbidities: Liver and renal dysfunction
  - Protein losing enteropathy
  - ↑↑ infection/ bleeding/ thrombosis

# Management of HF in Adult CHD

- ✓ Probability of HF increases with age and is the leading cause of death
- ✓ Multiple mechanisms for HF in this diverse cohort
  - ✓ RV and/or LV can be the problem
  - ✓ Renal and hepatic impairment not uncommon
- ✓ Limited benefits from conventional therapy
- ✓ Important to recognize and treat residual / post op sequelae
  - ✓ Consultation with ACHD specialist
- ✓ OHT is good option for few: consider CRT, LVAD in some

# Thank You!

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