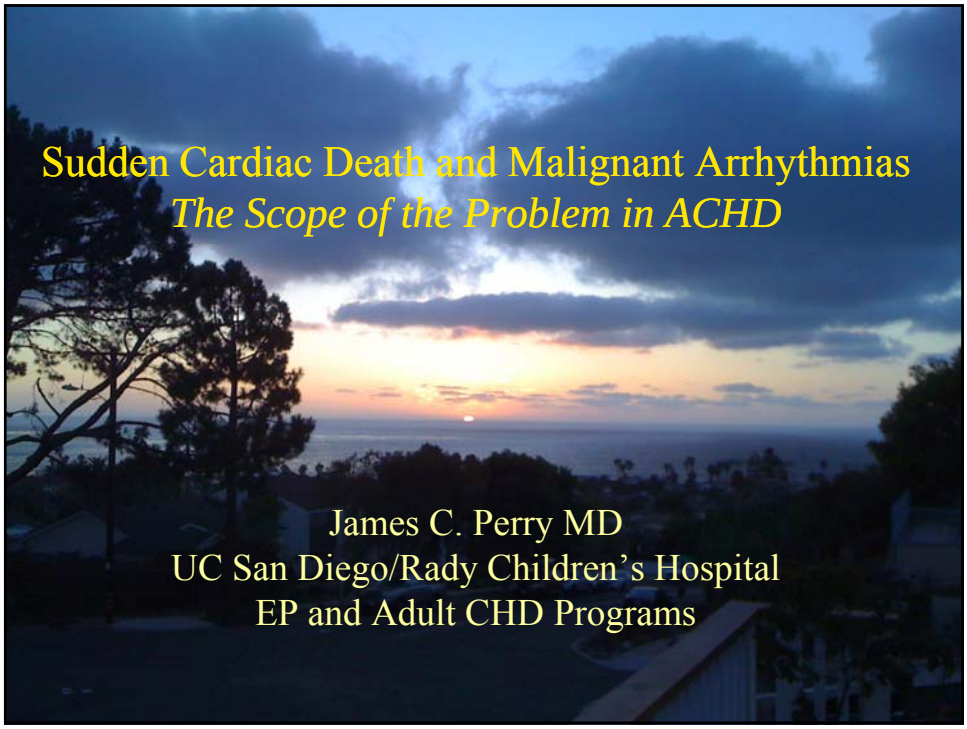



## Arrhythmia, SCD in ACHD
## here's a starting point…

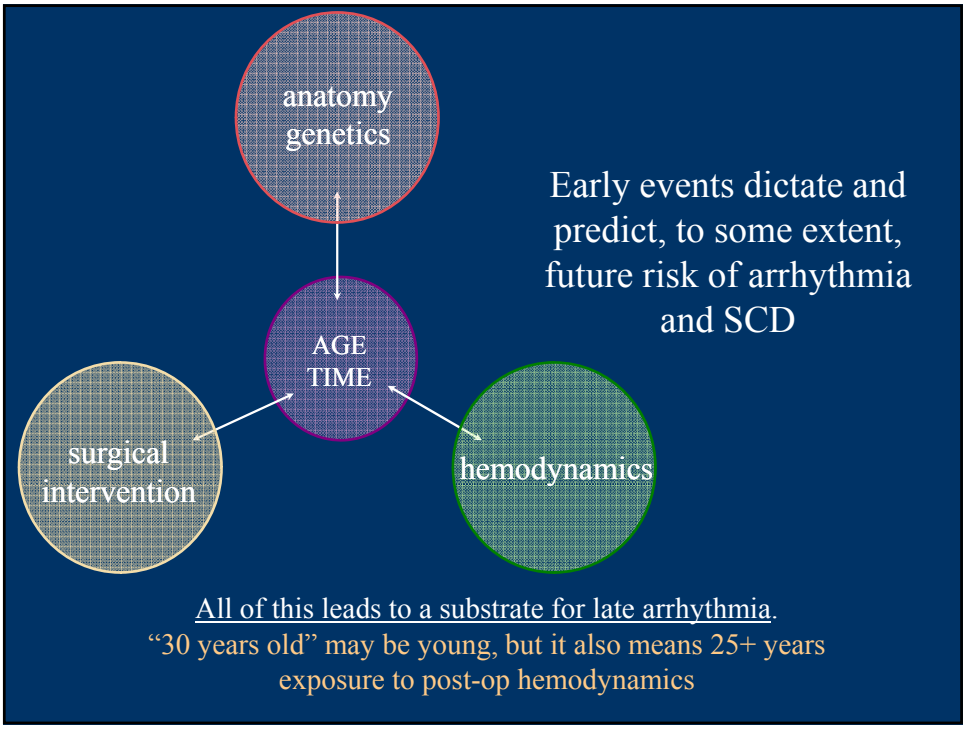
Oechslin et al. *Am J Cardiol* 2000
2609 ACHD pts: 199 deaths (~8%) - sudden death (26%) and CHF (21%)
CoA (EF<40%), Ebstein (not WPW), ccTGA (EF<45%), AoV abnl, TOF

We haven't understood this population very well…

Since late 1990's, more CHD patients over age 18 than under in the U.S.

Now ~ 1,000,000 ACHD'ers

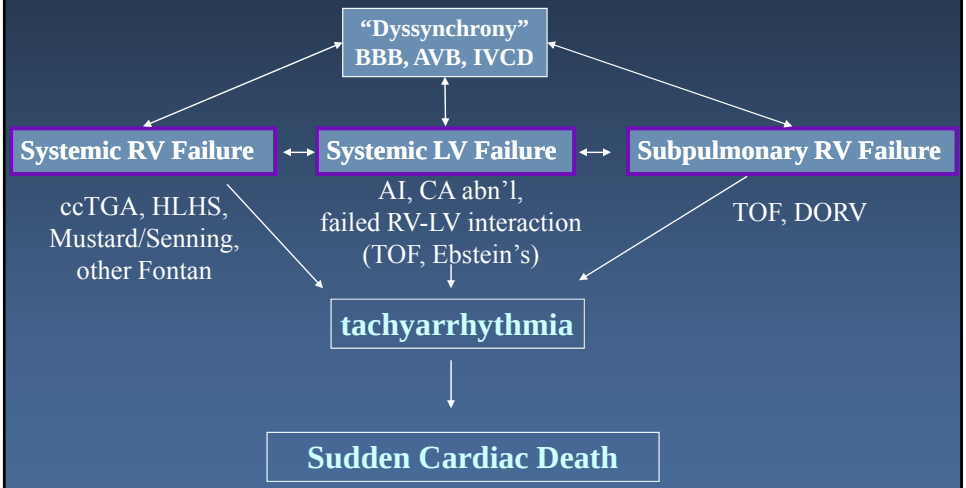
## *The SCD Problem in ACHD is a Continuum*




Early events dictate and predict, to some extent, future risk of arrhythmia and SCD

All of this leads to a substrate for late arrhythmia.
"30 years old" may be young, but it also means 25+ years exposure to post-op hemodynamics

## Categorizing the many different anatomies of ACHD: Heart Failure leads to VT/SCD



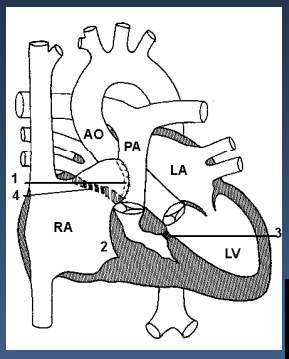

## "Old-style Fontan" RA-PA connection

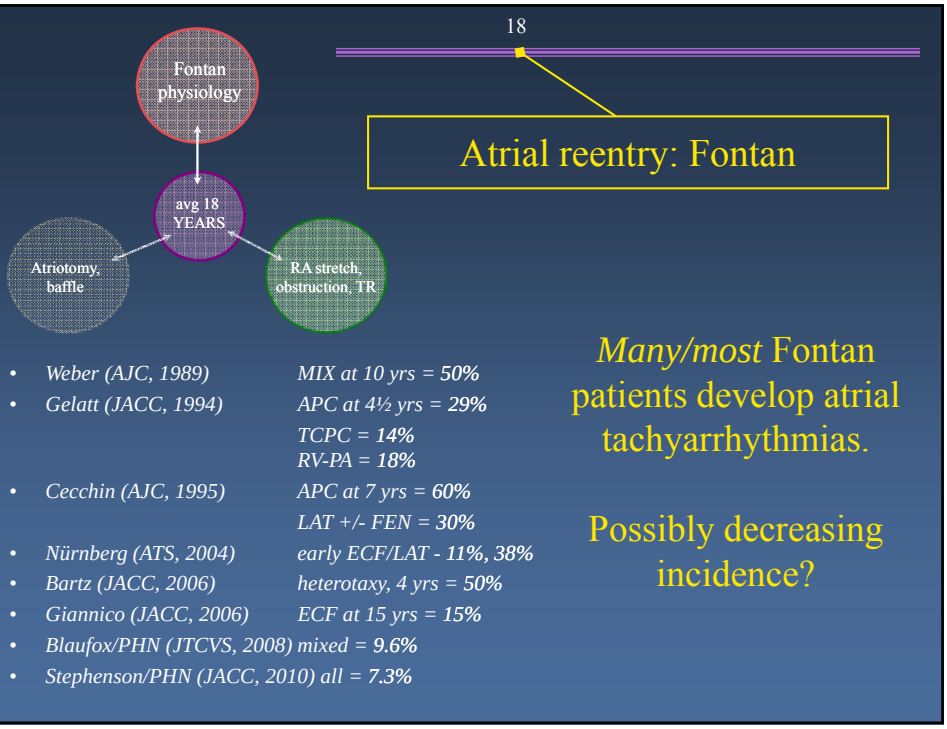
Current surgical management of "single ventricle physiology"

1- Ao-Pulm shunt: imposes a volume load on the ventricle ("chronic" stretch)

2 - Bidirectional Glenn shunt: reduces volume, persistent low sats for myocardium

3 - Fontan: multiple suture lines in atria, atrial stretch

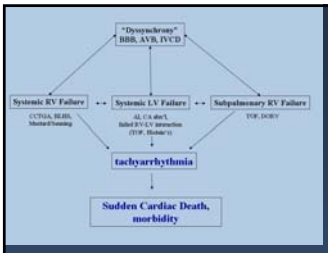



- *Weber (AJC, 1989)* — *MIX at 10 yrs = 50%*
- *Gelatt (JACC, 1994)* — *APC at 4½ yrs = 29%*; *TCPC = 14%*; *RV-PA = 18%*
- *Cecchin (AJC, 1995)* — *APC at 7 yrs = 60%*; *LAT +/- FEN = 30%*
- *Nürnberg (ATS, 2004)* — *early ECF/LAT - 11%, 38%*
- *Bartz (JACC, 2006)* — *heterotaxy, 4 yrs = 50%*
- *Giannico (JACC, 2006)* — *ECF at 15 yrs = 15%*
- *Blaufox/PHN (JTCVS, 2008) mixed = 9.6%*
- *Stephenson/PHN (JACC, 2010) all = 7.3%*

*Many/most* Fontan patients develop atrial tachyarrhythmias.

Possibly decreasing incidence?




# Systemic SV Failure

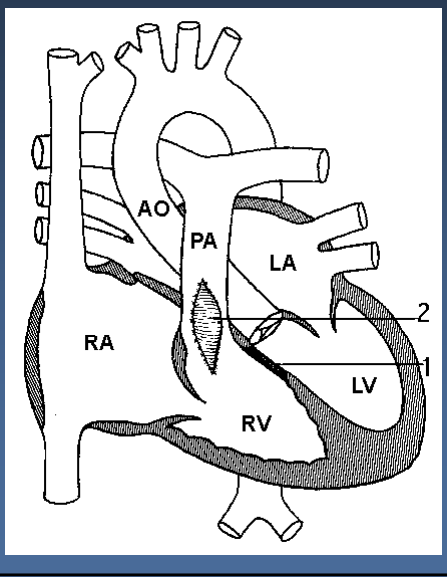
Piran et al. *Circ* 2002;105:1189
long-term f/u of 188 adult CHD

Adult Fontan patients:

- 45% had sx's of tachyarrhythmia or obvious CHF
- 18% died in 16 year follow-up
- 33% died 2° to HF
- ~90% of those dying with CHF had significant arrhythmia, mostly IART, VT

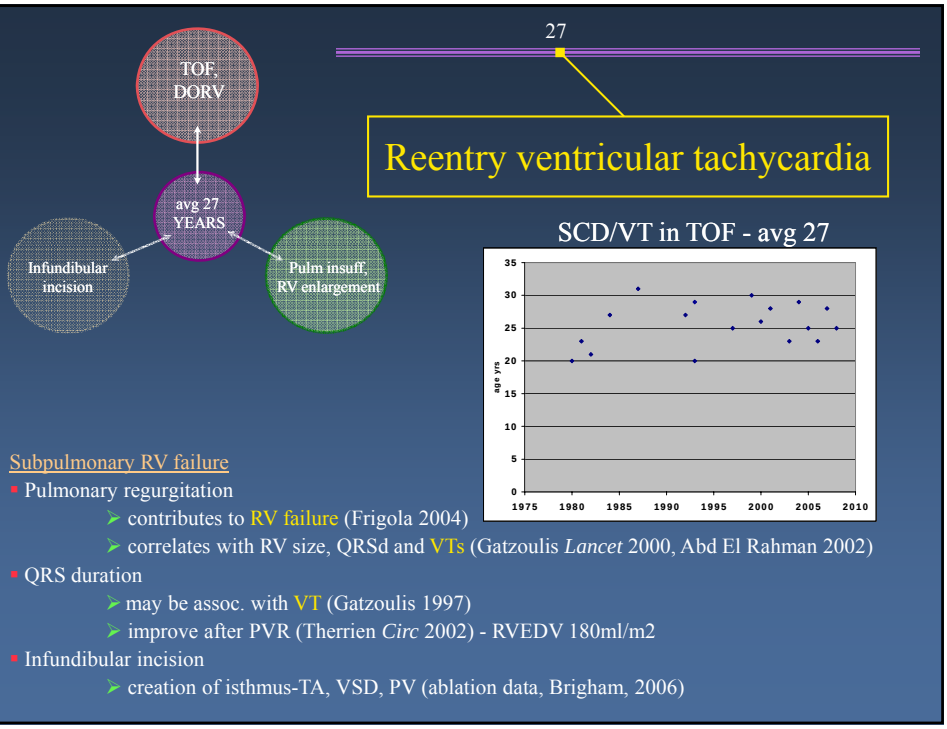
# Tetralogy of Fallot
# Subpulmonary RV Failure

"tetralogy"
1. RV outflow obstruction
2. VSD
3. Aortic override
4. RVH




"monology"

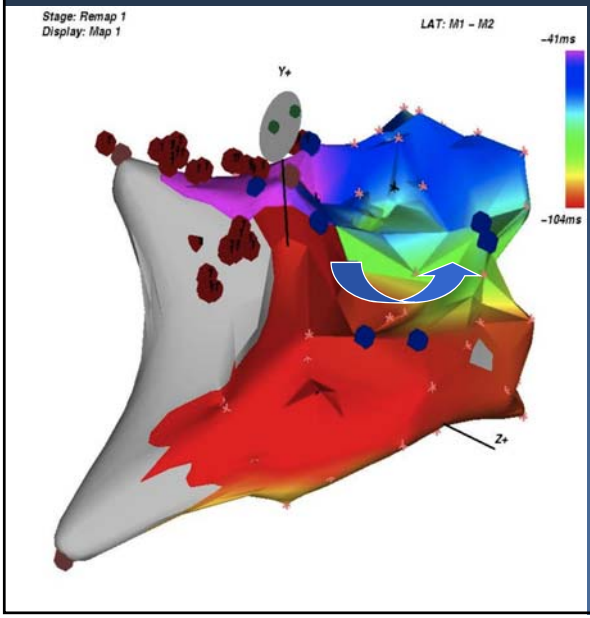
Anterior deviation of the infundibular septum *in utero*




Subpulmonary RV failure

- Pulmonary regurgitation
  - contributes to RV failure (Frigola 2004)
  - correlates with RV size, QRSd and VTs (Gatzoulis *Lancet* 2000, Abd El Rahman 2002)
- QRS duration
  - may be assoc. with VT (Gatzoulis 1997)
  - improve after PVR (Therrien *Circ* 2002) - RVEDV 180ml/m2
- Infundibular incision
  - creation of isthmus-TA, VSD, PV (ablation data, Brigham, 2006)

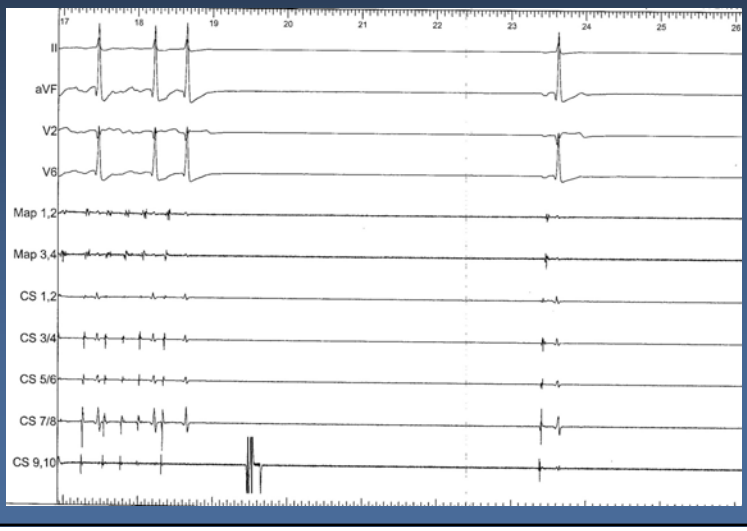
## VT in TOF



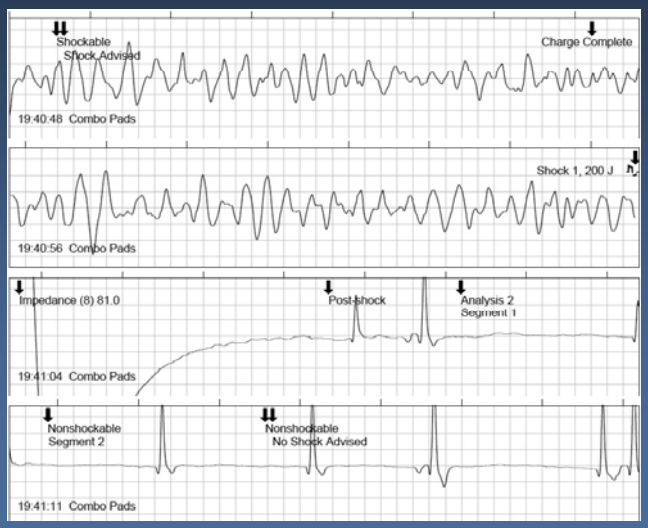

**Counterclockwise VT circuit around the anastomosis of the RV and conduit.**

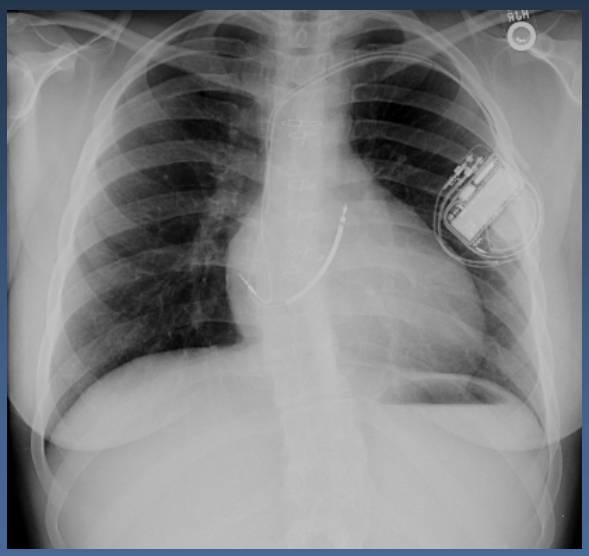
**Successful RF line created, posterior aspect of anastomosis and the tricuspid annulus.**

Extreme bradycardia, sinus arrest on cessation of rapid atrial tachycardia. 25 yo with complex pulm atresia, failed 1 1/2 ventricle approach, atriopulmonary Fontan



## Bradycardia-induced VF converted by AED at a gym

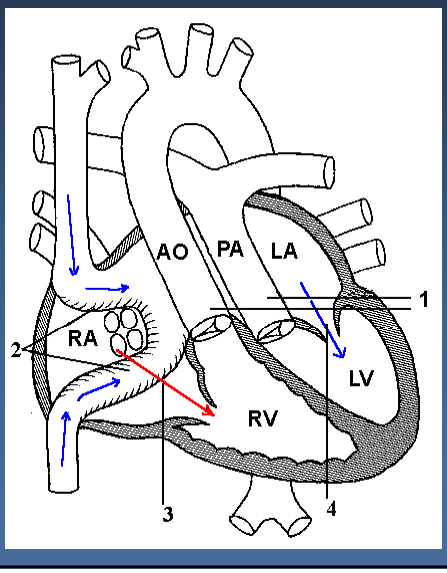


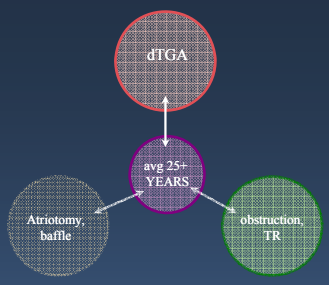



## Systemic RV Failure - Atrial Switch (Mustard, Senning) d-transposition of the great arteries

One operation. Good early result.

Long-term:
Long atrial suture lines, conduction block
Systemic right ventricle

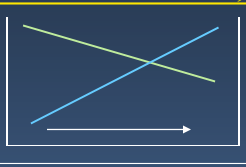



25 + ⇒

### Bradycardia/Tachycardias d-TGA: Mustard, Senning

NSR

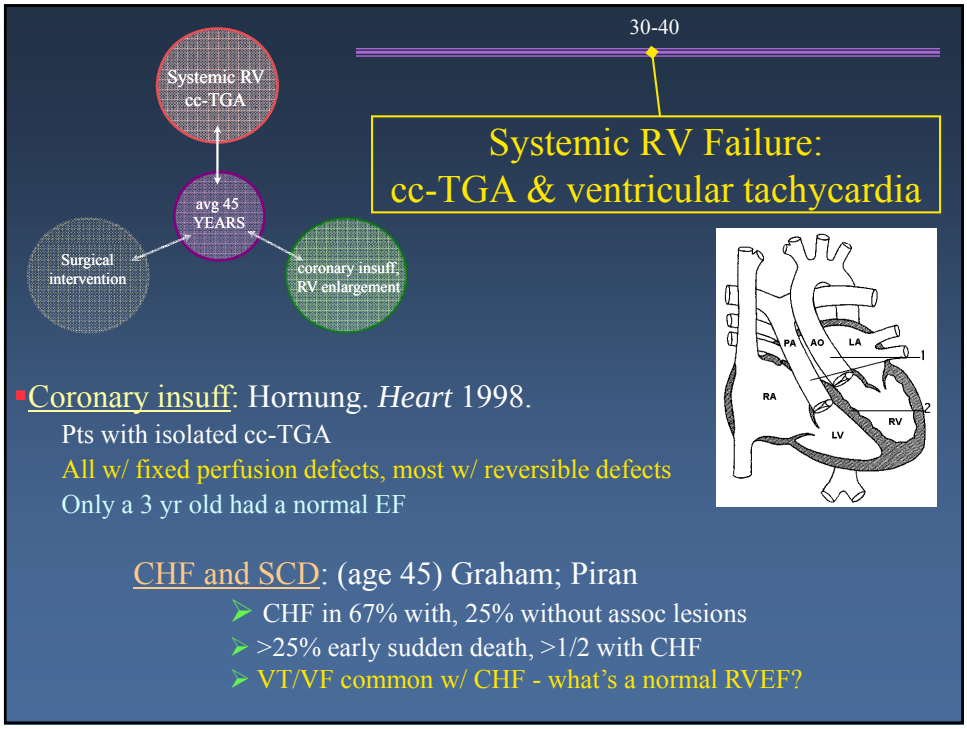


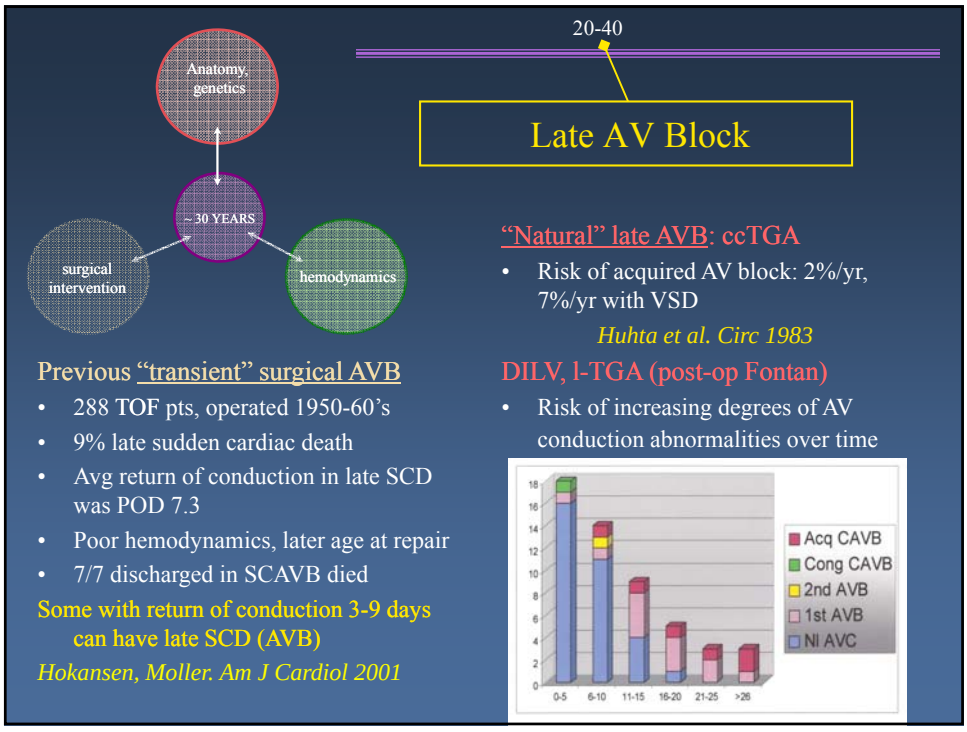
AtrTachy

**Bradycardia**
- "loss of sinus rhythm" - 20-60% at 20 yrs
- pacing and HF
- effects of slow HR on vent funct not well-studied
- dilation due to increased stroke volume contributes to HF, VA's

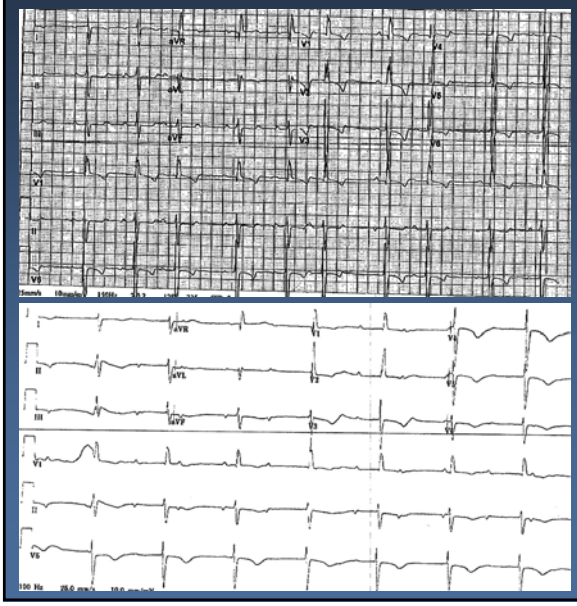
**Atrial reentry**
- most common tachy; can also 1:1 w/ ischemia, VT
- rare paroxysm of adv AV block
- uncontrolled IART: higher mortality risk

**1° VT/SCD**
- Most ACHD w/ dTGA have fixed or reversible CA perfusion defects (systemic RV/coronary supply)
- worse VT/SCD risk if Mustard with VSD
- 70% have HF symptoms, poorly Rx'd
- VT/VF documented in ~ *all* SCD, ~ 80% with exercise


30-40
Systemic RV
cc-TGA
avg 45
YEARS
Surgical
intervention
coronary insuff,
RV enlargement
Systemic RV Failure:
cc-TGA & ventricular tachycardia
PA
AO
LA
RA
RV
LV
1
2
Coronary insuff: Hornung. Heart 1998.
Pts with isolated cc-TGA
All w/ fixed perfusion defects, most w/ reversible defects
Only a 3 yr old had a normal EF
CHF and SCD: (age 45) Graham; Piran
CHF in 67% with, 25% without assoc lesions
>25% early sudden death, >1/2 with CHF
VT/VF common w/ CHF - what's a normal RVEF?

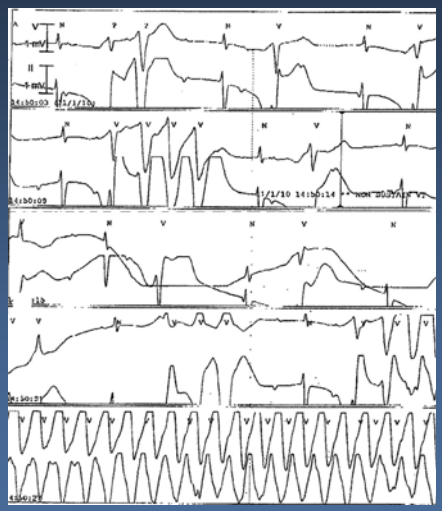

20-40
Anatomy,
genetics
~ 30 YEARS
surgical
intervention
hemodynamics
Late AV Block
Previous "transient" surgical AVB
288 TOF pts, operated 1950-60's
9% late sudden cardiac death
Avg return of conduction in late SCD was POD 7.3
Poor hemodynamics, later age at repair
7/7 discharged in SCAVB died
Some with return of conduction 3-9 days can have late SCD (AVB)
Hokansen, Moller. Am J Cardiol 2001
"Natural" late AVB: ccTGA
Risk of acquired AV block: 2%/yr, 7%/yr with VSD
Huhta et al. Circ 1983
DILV, l-TGA (post-op Fontan)
Risk of increasing degrees of AV conduction abnormalities over time
0-5
6-10
11-15
16-20
21-25
>26
Acq CAVB
Cong CAVB
2nd AVB
1st AVB
NI AVC

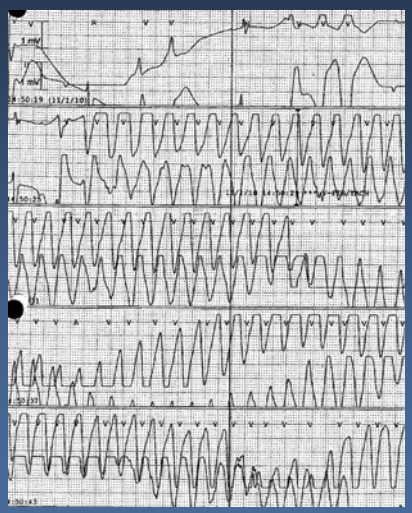
## New AV block

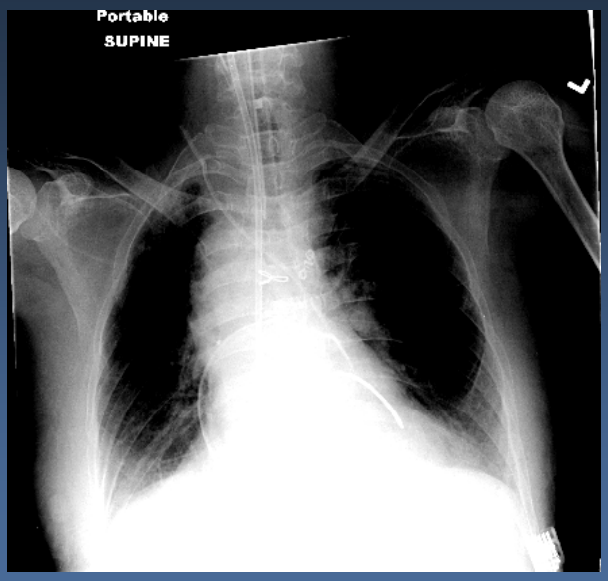


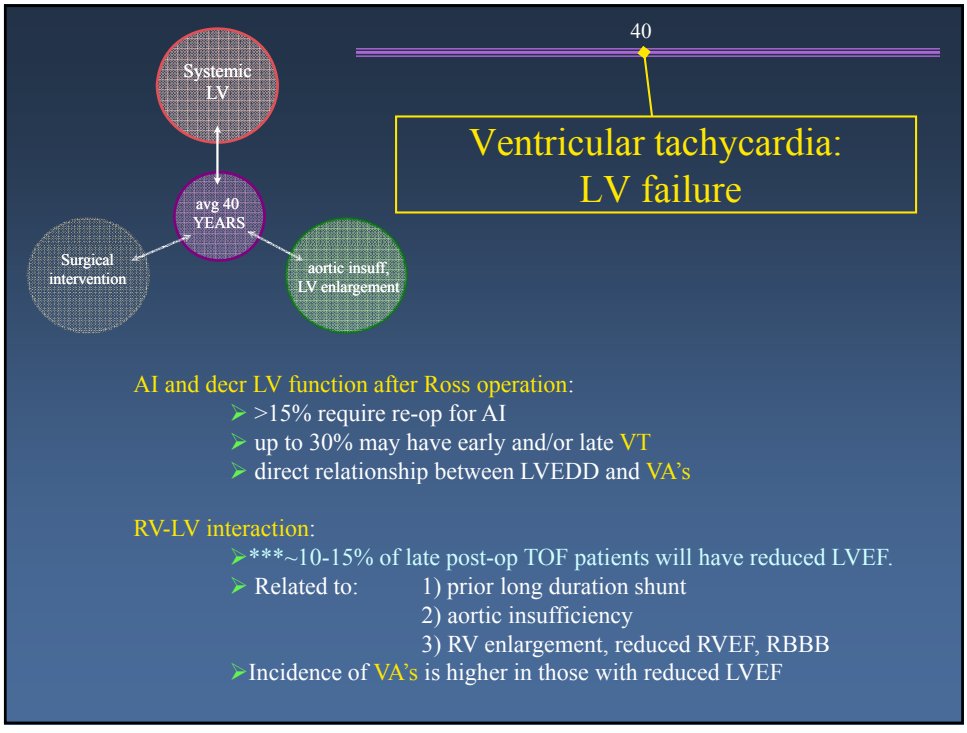
- 57 yo Fontan, complex anatomy, AV-VA discordance
- 1st ECG in September
- 2nd ECG in October

## Bradycardia and AVB-induced VT/VF








AI and decr LV function after Ross operation:

- >15% require re-op for AI
- up to 30% may have early and/or late VT
- direct relationship between LVEDD and VA's

RV-LV interaction:

- ***~10-15% of late post-op TOF patients will have reduced LVEF.
- Related to: 1) prior long duration shunt
  2) aortic insufficiency
  3) RV enlargement, reduced RVEF, RBBB
- Incidence of VA's is higher in those with reduced LVEF


## Dyssynchrony

2° to Bundle Branch Block

Adult HF: (CRT) Bradley et al. *JAMA* 2003;289;730

1634 pts, meta-analysis: CONTAK, MUSTIC, MIRACLE, InSync ICD

LBBB 54-87%, LVEFs ≤ 30%; Ischemic CM 37-69%; Age ~ 65 yrs

ACHD: RBBB - operations: TOF, VSD, etc, etc
LBBB - operations: AS, subAS, some DORV, AV canal

2° to Pacing

Adult pts: DAVID Trial (2002); MOde Selection Trial (2003)

Mortality, hosp and atrial fib > with more RV pacing

⇒ Over 1 year+

Ped/ACHD pts (mult studies)

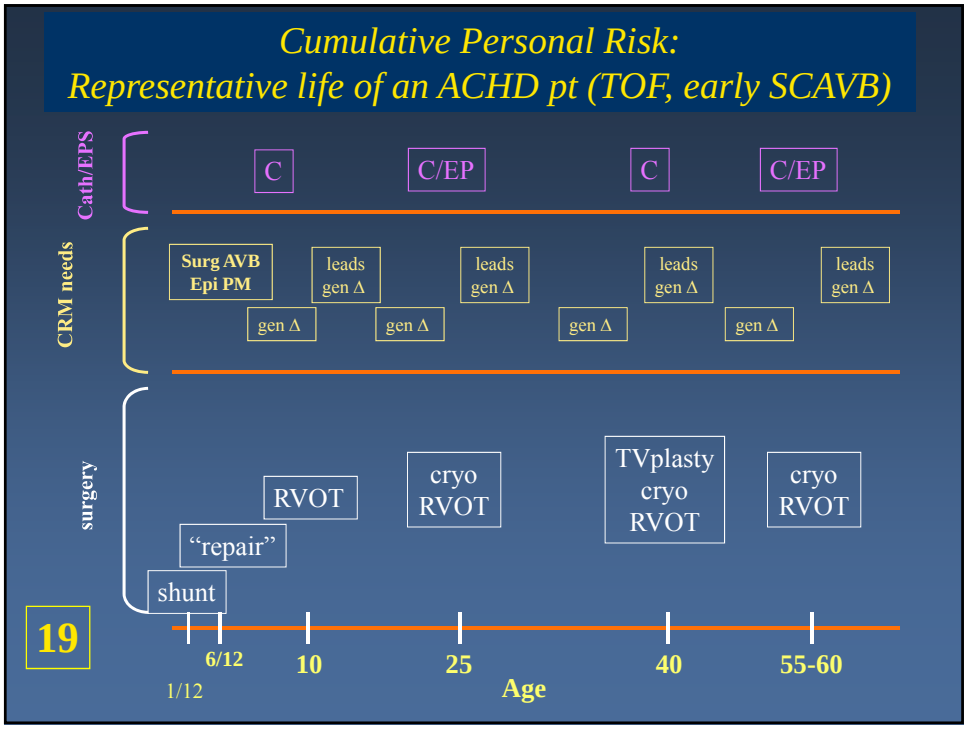
incr. RV paced QRS duration and age = poorer LV function

Location: septum better than apex - LV dP/dt and LVEDP

"Upgrade" from to DDDR: Ventricular funct worse

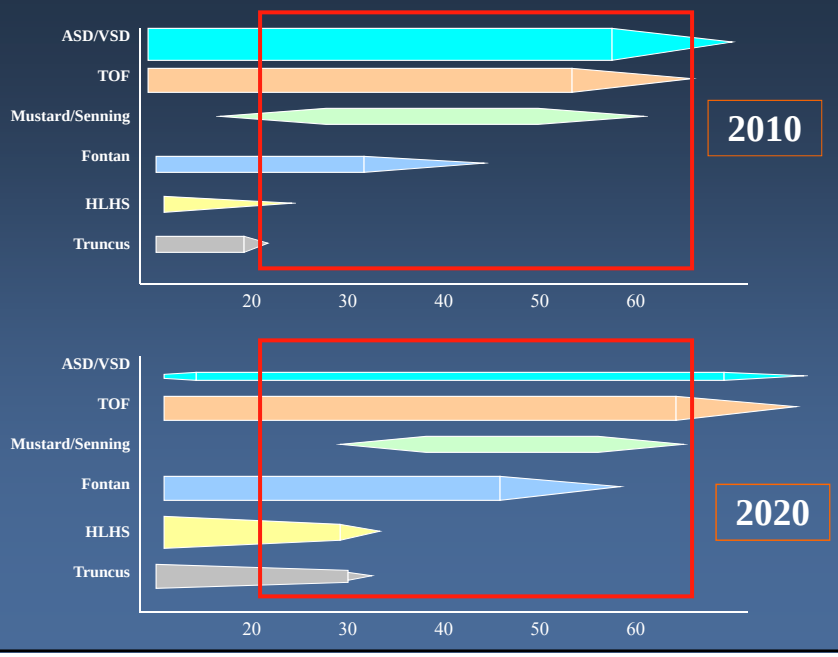
⇒ Over decades?

*The SCD Problem in ACHD is Personal*




*The SCD Problem in ACHD is Changing*

## *Changing… increasing complexity of the ACHD population*




*The Problem of*
*Lifetime Arrhythmia Burden*
*in ACHD needs to be Quantified*

## CRM needs by CHD type

| DEFECT | Incidence (in CHD Population) |
|---|---|
| Patent Ductus Arteriosus | 7.3% |
| Pulmonary Stenosis | 6.6% |
| Aortic Coarctation | 6.2% |
| Ventricular Septal Defect | 30.7% |
| Atrial Septal Defect | 8.9% |
| Total Anomalous Pulmonary Venous Return | 1.8% |
| Atrioventricular Canal | 4.4% |
| Aortic Stenosis | 4.0% |
| Truncus Arteriosus | 1.9% |
| Pulmonary Atresia | 2.4% |
| Tetralogy of Fallot | 4.9% |
| Double Outlet Right Ventricle | 1.7% |
| Hypoplastic Left Heart | 4.0% |
| Transposition of the Great Arteries | 6.3% |
| OTHER - Single vent, ccTGA, heterotaxy, Ebstein's | 9.0% |

Estimate candidates:

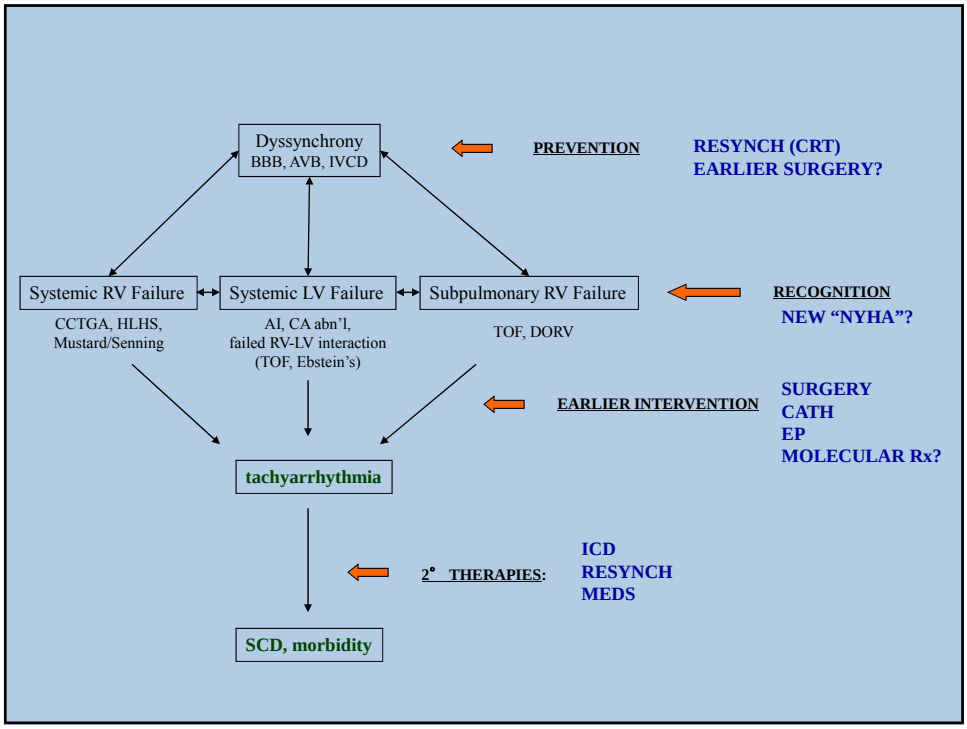
- Pacing: AV block, SND/AT
- ICD: primary, secondary
- CRT for HF

| DEFECT | CRM needs pediatric | CRM needs ACHD |
|---|---|---|
| Patent Ductus Arteriosus | 0.0% | 0.0% |
| Pulmonary Stenosis | 0.0% | 0.0% |
| Aortic Coarctation | 0.0% | 6.0% |
| Ventricular Septal Defect | 2.5% | 2.5% |
| Atrial Septal Defect | 6.0% | 25.0% |
| Total Anomalous Pulmonary Venous Return | 2.0% | 5.0% |
| Atrioventricular Canal | 5.0% | 8.0% |
| Aortic Stenosis | 5.5% | 38.0% |
| Truncus Arteriosus | 34.0% | 26.0% |
| Pulmonary Atresia | 8.0% | 29.0% |
| Tetralogy of Fallot | 28.0% | 42.0% |
| Double Outlet Right Ventricle | 14.0% | 42.0% |
| Hypoplastic Left Heart | 14.5% | 49.0% |
| Transposition of the Great Arteries | 34.0% | 100.0% |
| OTHER - Single vent, ccTGA, heterotaxy, Ebstein's | 32.0% | 75.0% |

Pediatric CHD: overall 9%
Adult CHD: overall 20%

< 10% are getting CRM Rx

*The SCD Problem in ACHD needs a Plan*




Take home messages
Heart Failure and Arrhythmia in ACHD:
ACHD is an *electroanatomic* defect

- Important component of antiarrhythmic therapy in ACHD patients with HF is *heart failure therapy*.
- Anticipate EP needs by each CHD defect, physiology
- Use all available opportunities, don't burn bridges
- Give the pediatric CHD patient a realistic appraisal of life as an adult CHD patient.