Evaluation of Ventricular Arrhythmias in Repaired Tetralogy of Fallot

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THE OHIO STATE UNIVERSITY
COLLEGE OF MEDICINE

NATIONWIDE CHILDREN'S
When your child needs a hospital, everything matters.

Objectives

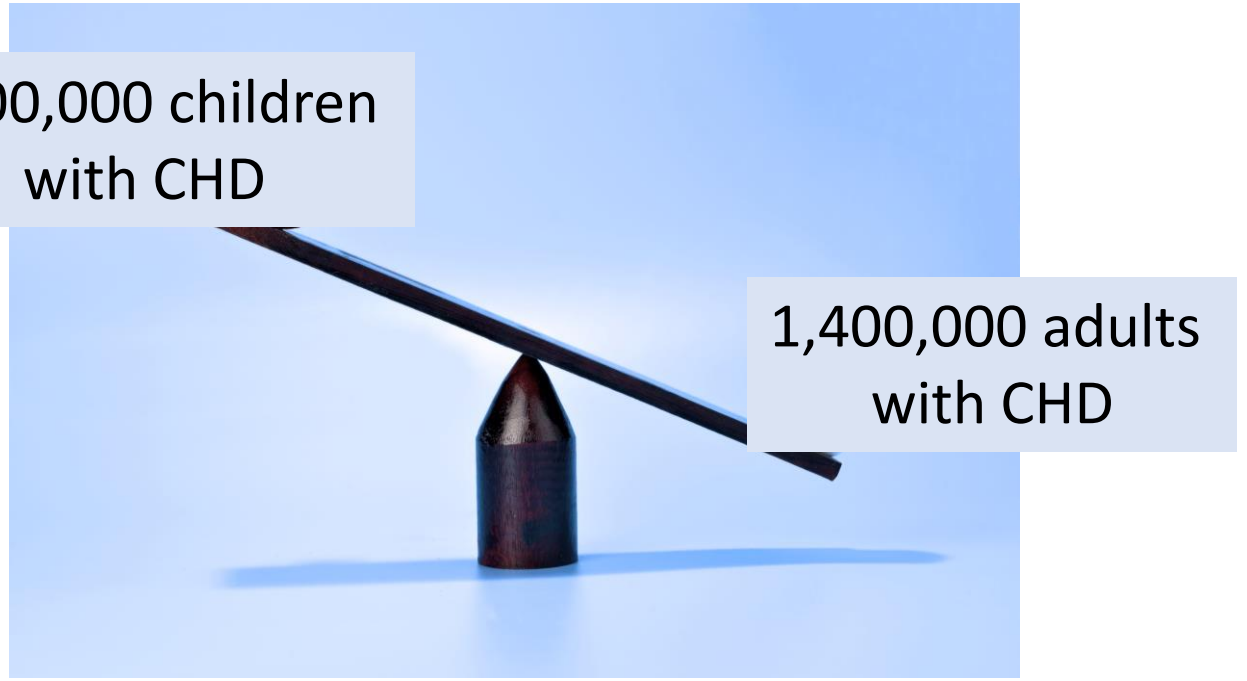
Think Like An Electrophysiologist

- Understand ventricular arrhythmogenesis in repaired Tetralogy of Fallot
- Outline arrhythmia management strategies and be able to apply to your specific patients

Who takes care of ACHD Arrhythmia Patients?

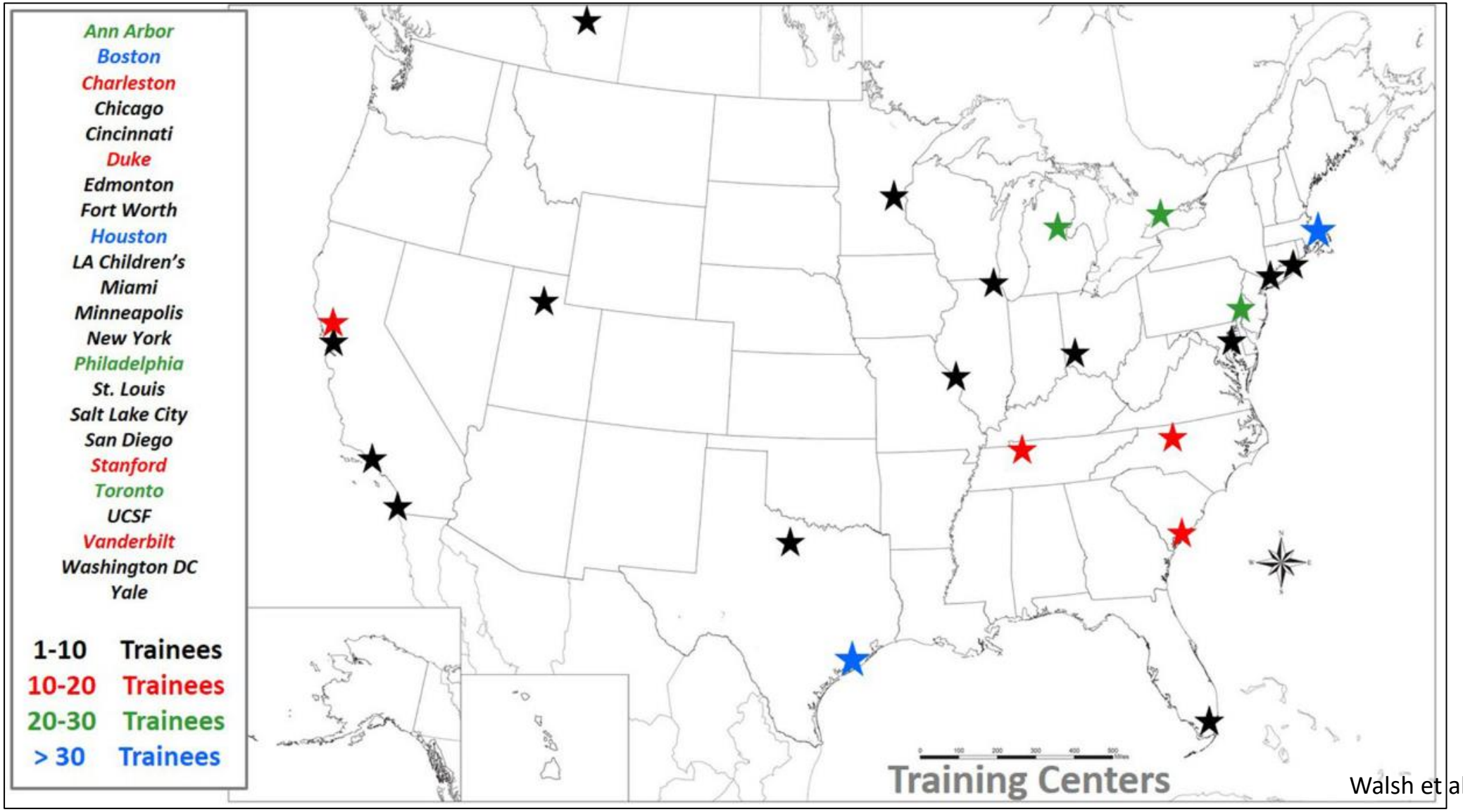
2010 US Census data:

Adults with CHD >>> Children with CHD



Gilboa et al. Circ 2016

Who takes care of ACHD Arrhythmia Patients?
Congenital EP Training Centers



Walsh et al. Ped Card 2022

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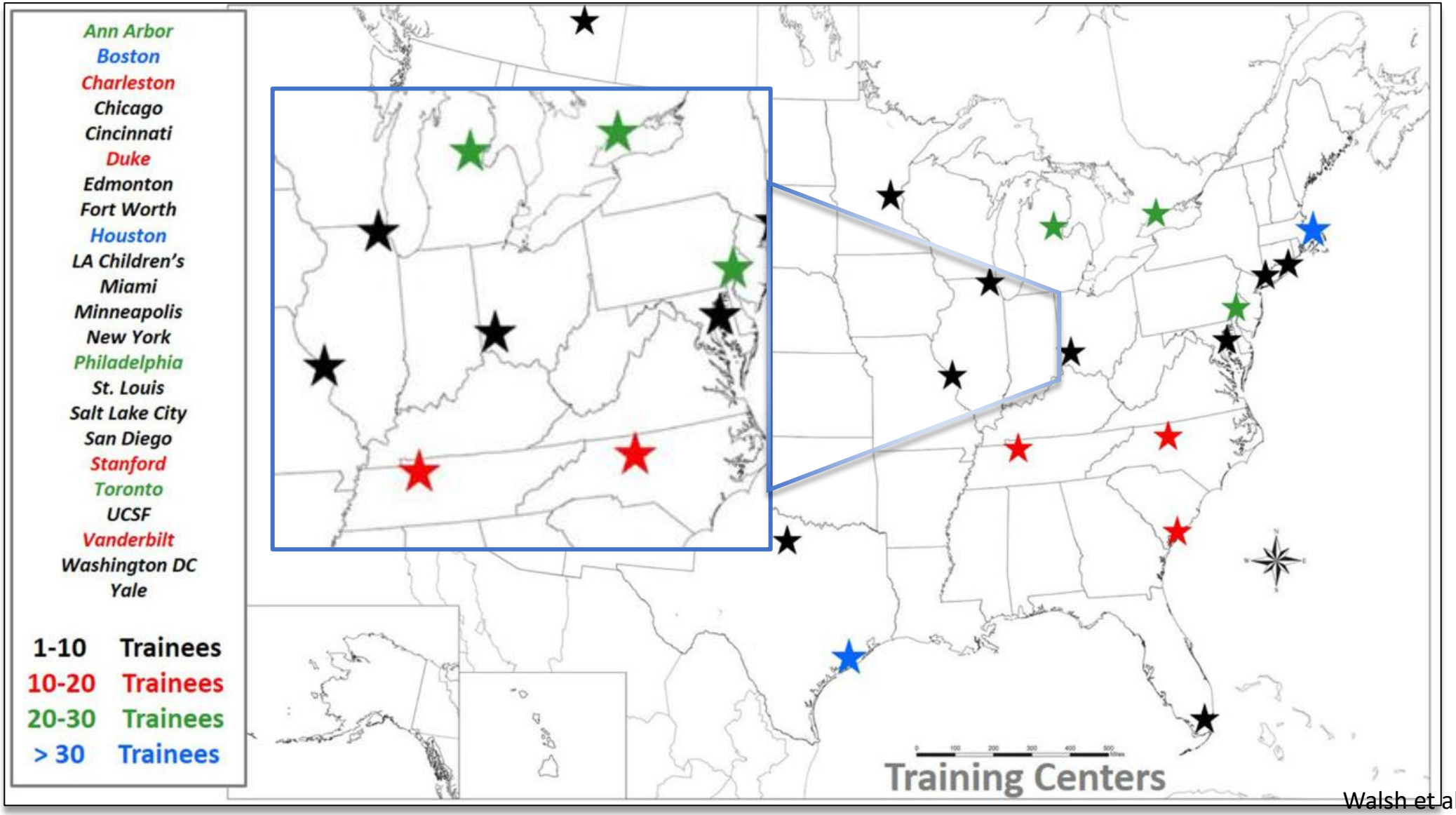
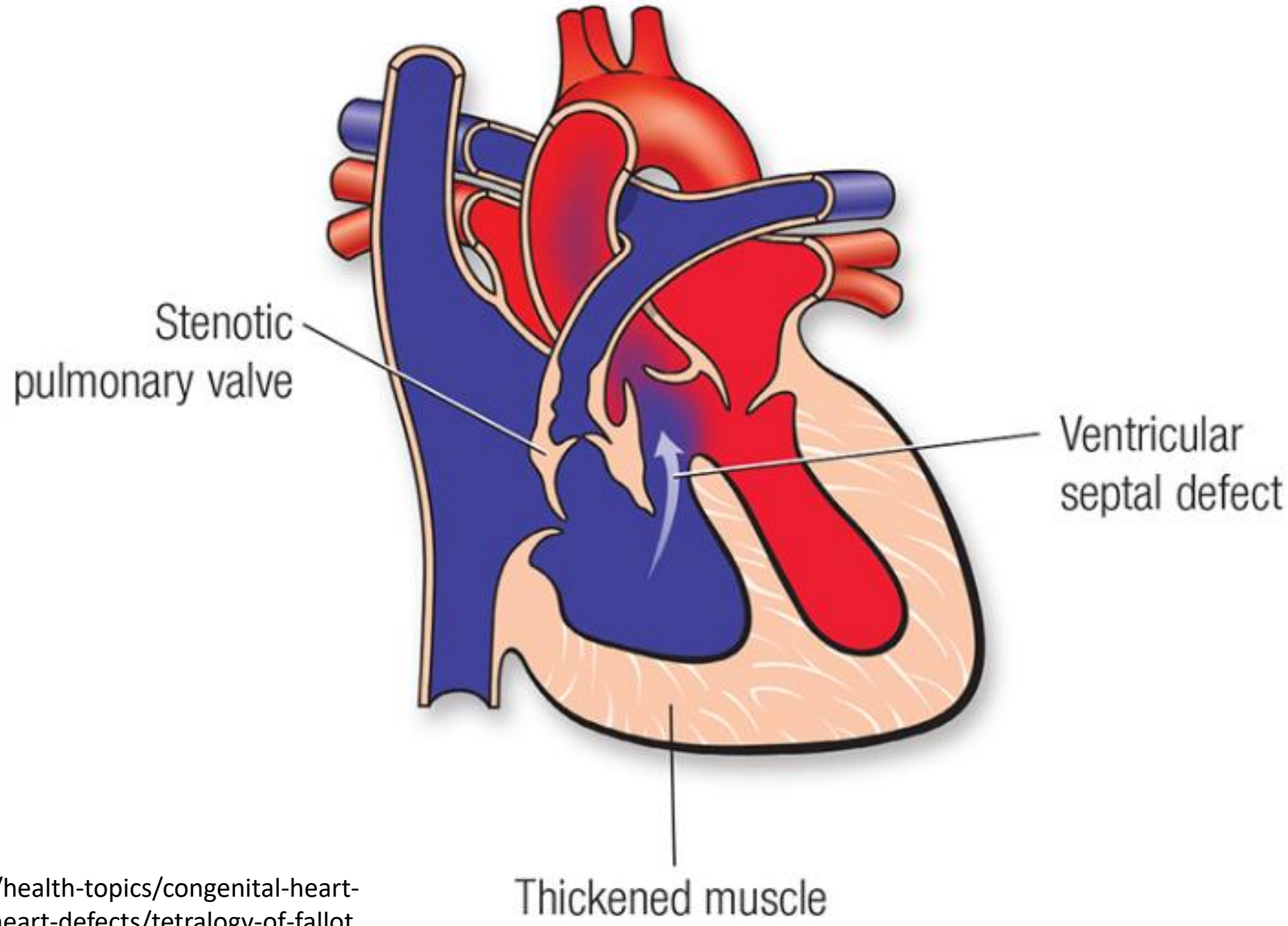


Walsh et al. Ped Card 2022

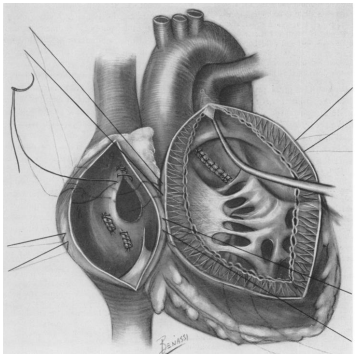
Tetralogy of Fallot

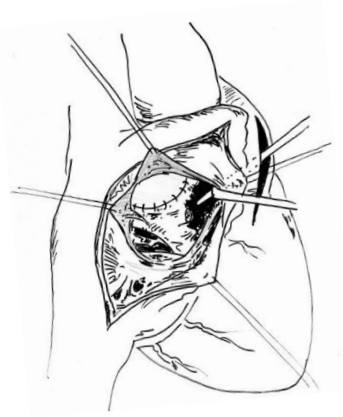


https://www.heart.org/en/health-topics/congenital-heart-defects/about-congenital-heart-defects/tetralogy-of-fallot

Surgical Evolution of Tetralogy of Fallot

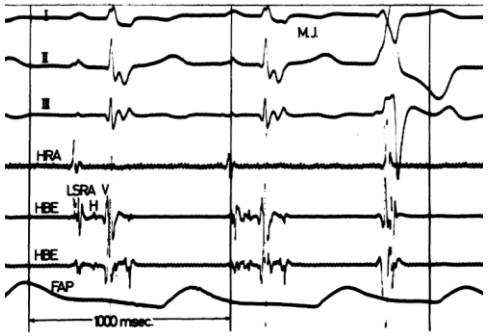
"la maladie bleue"
Etienne-Louis Fallot





1673 — 1888 — 1944 — 1955 — 1962 — 1977 — 1981 — 2008

Nils Stensen



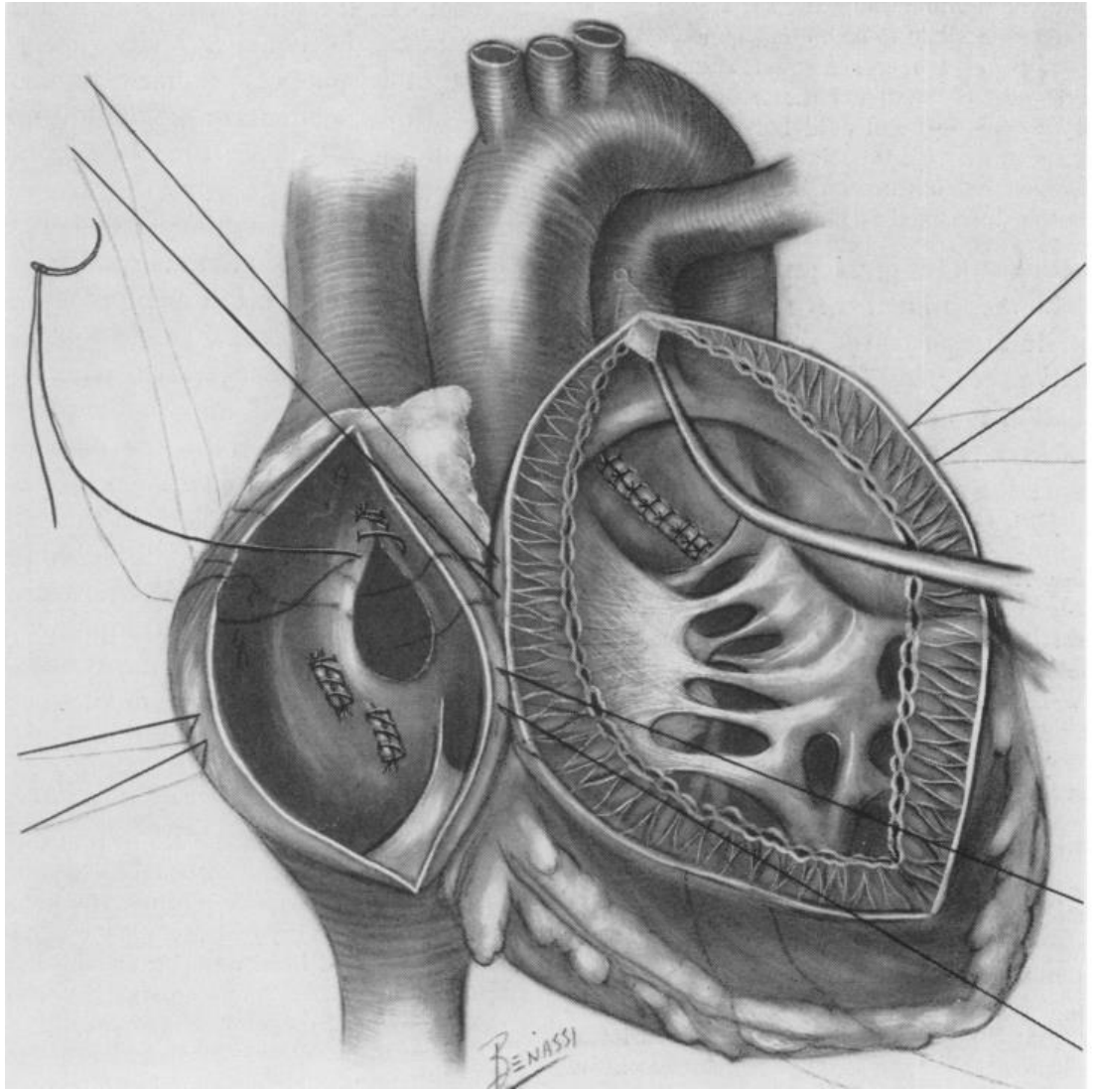
Loukas et al, Clinical Anatomy 2014; https://www.hopkinsmedicine.org/about/history;
Lillehei et al, Annals of Surgery 1955; Gillette et al. Circ 1977
Padalino, Vida, and Stellin, Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 2009

1955 – Intracardiac Correction of TOF

Direct Vision Intracardiac Surgical Correction of the Tetralogy of Fallot, Pentalogy of Fallot, and Pulmonary Atresia Defects

Report of First Ten Cases*

C. Walton Lillehei, M.D., Morley Cohen, M.D., Herbert E. Warden, M.D., Raymond C. Read, M.D., Joseph B. Aust, M.D., Richard A. DeWall, M.D., and Richard L. Varco, M.D.

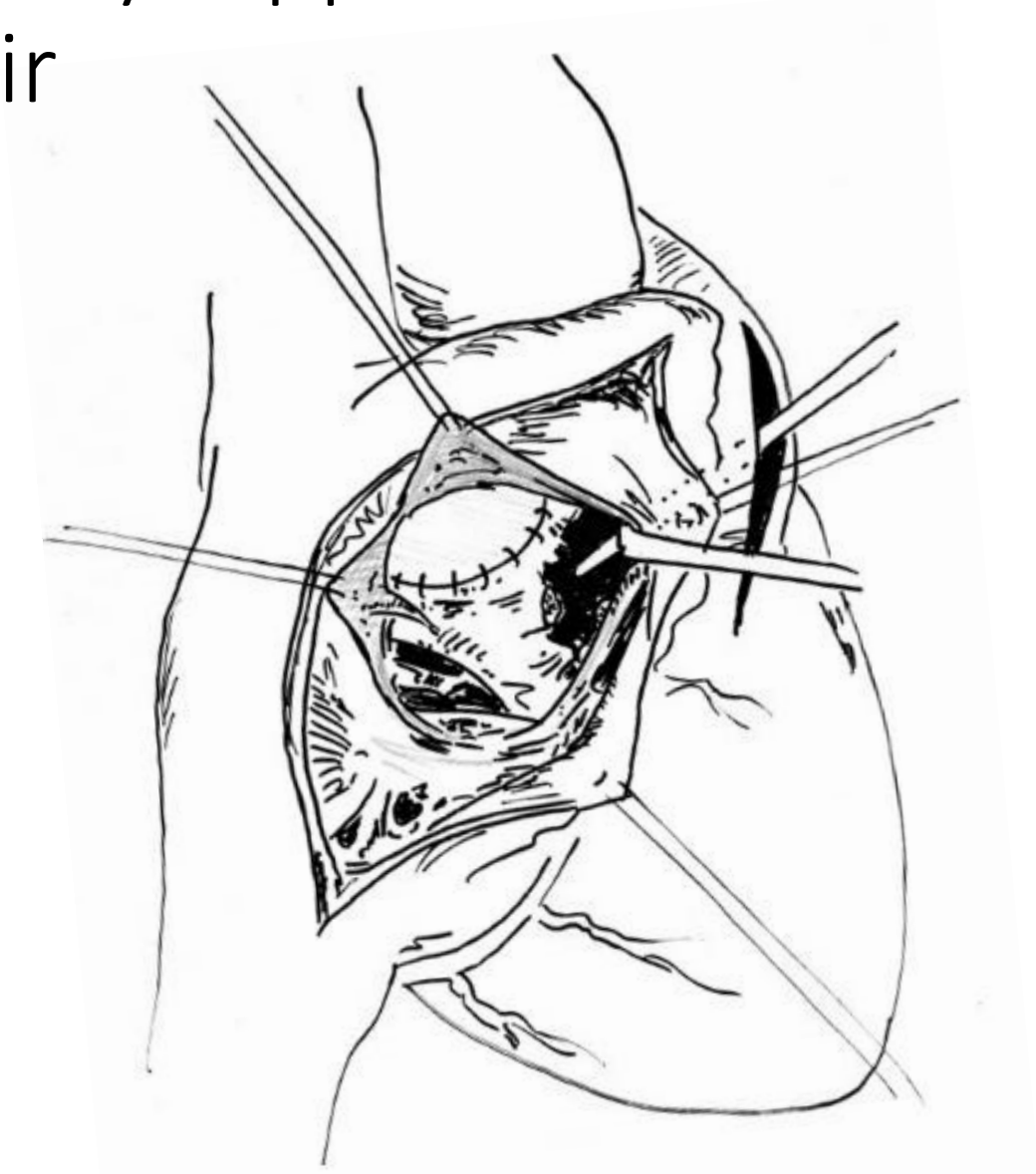


Lillehei et al, Annals of Surgery 1955

Transatrial – Transpulmonary Approach to TOF Repair

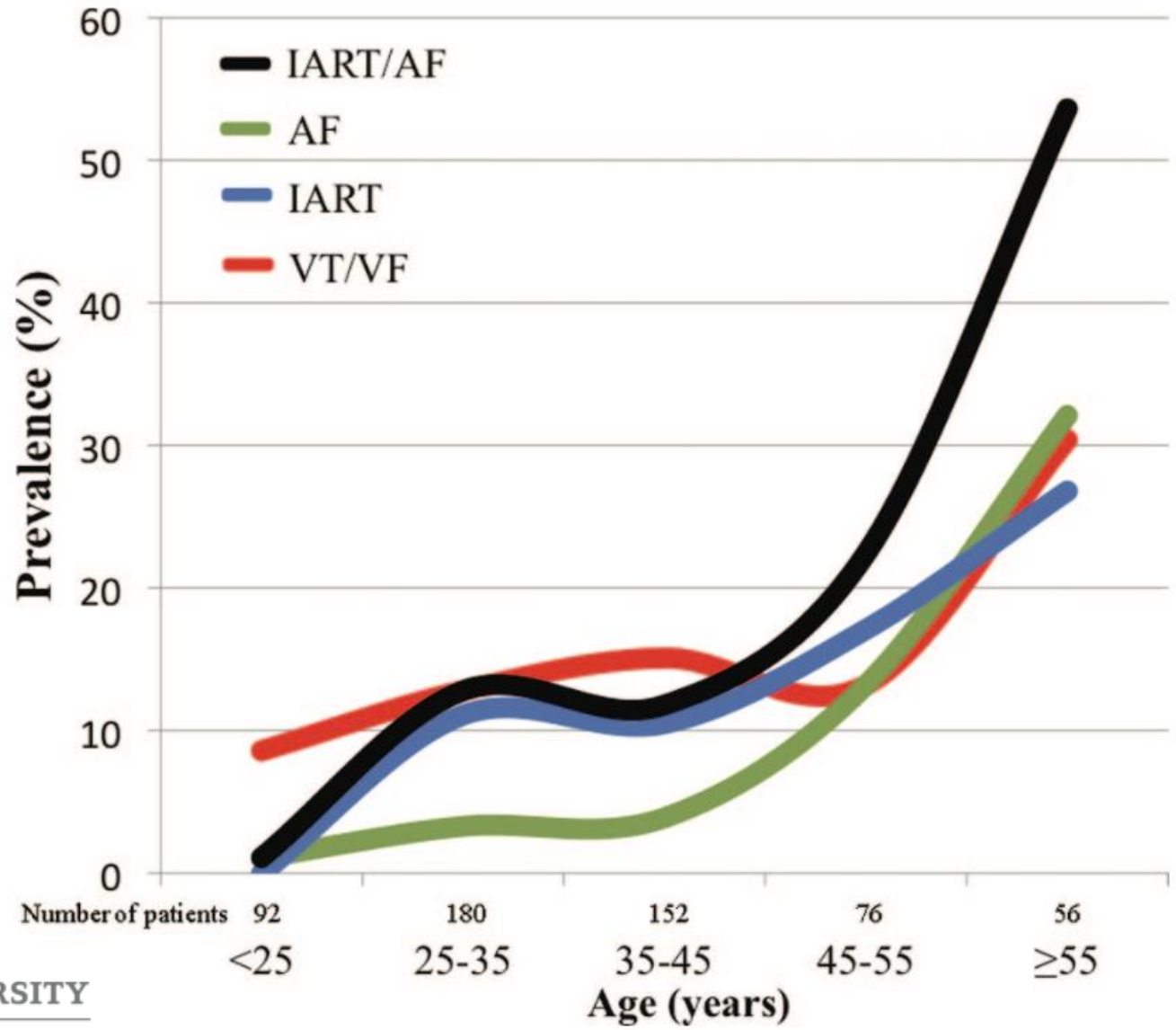
Corrective Surgery for Tetralogy of Fallot Without or With Minimal Right Ventriculotomy and with Repair of the Pulmonary Valve

YASUNARU KAWASHIMA, M.D., SOICHIRO KITAMURA, M.D., SUSUMU NAKANO, M.D., AND TOSHIKATSU YAGIHARA, M.D.



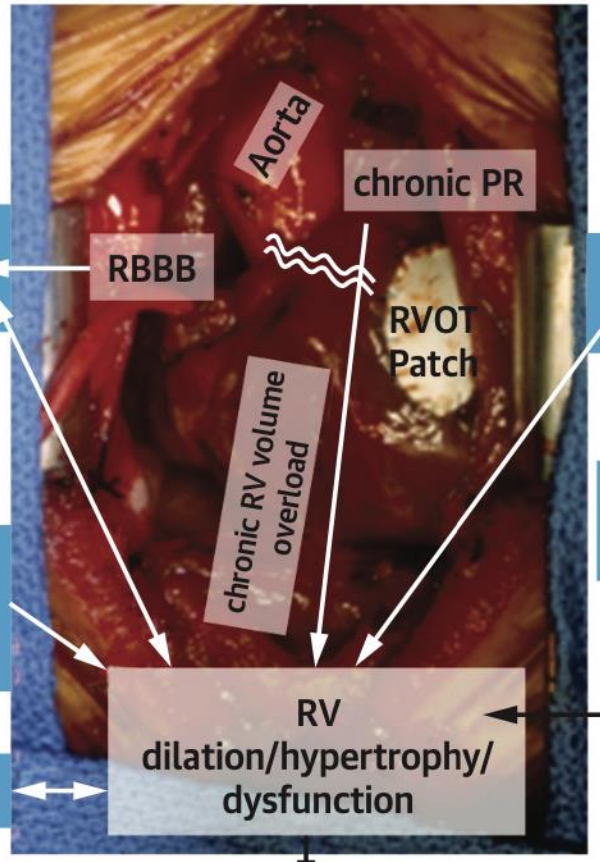
Hudspeth J Thorac CV Surg 1962; Kawashima Circ 1981; Padalino, Vida, and Stellin, Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 2009

Arrhythmia Burden in Adults with rTOF



Khairy et al. Circ 2010

Factors Contributing to Arrhythmias in rTOF



RVOT Sequelae

Ventricular Dyssynchrony

Increased RV Pressure

Localized degenerative remodeling and fibrosis

cyanosis

volume overload

RVOT aneurysm

RVOT patch/dyskinesia

Slow conducting zones

Diffuse myocardial fibrosis

RV dilation/hypertrophy/ dysfunction

LV Dysfunction

Adverse Electrophysiologic Remodeling

VT/VF

Sudden Cardiac Death

Cohen et al. JACC 2021

Predictors of ICD therapies
Long-term follow-up of rTOF with ICD
Nationwide French Registry of All rTOF with ICD

Table 3. Predictors of Appropriate Implantable Cardioverter Defibrillator Therapies in Primary Prevention

	Univariable analysis			Multivariable analysis		
	Hazard ratio	95% CI	*P*	Hazard ratio	95% CI	*P*
Age at implantation, per year	0.96	0.93–0.99	0.018		—	
History of atrial arrhythmia	0.39	0.13–1.22	0.110		—	
History of NSVT or VT/VF	2.21	0.95–5.65	0.066	1.58	0.66–3.76	0.310
QRS duration ≥180 ms	1.32	0.55–3.15	0.530		—	
QRS fragmentation	3.75	1.36–10.40	0.011	3.47	1.19–10.11	0.022
Left ventricular ejection fraction ≤35%	0.39	0.12–1.23	0.110	0.39	0.15–1.07	0.066
Positive programmed ventricular stimulation	1.18	0.45–3.07	0.740		—	

Waldmann et al. Circ 2020

Invasive Arrhythmia Assessment Statements from Professional Organizations

PACES/HRS Expert Consensus Statement on the Recognition and Management of Arrhythmias in Adult Congenital Heart Disease 2014

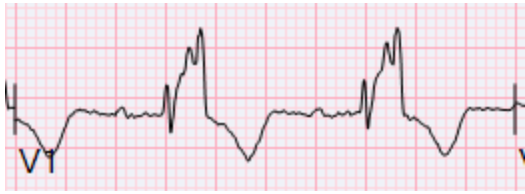
- **Class IIa**: PVS* useful in risk stratifying adults with TOF who have additional risk factors for SCD
- **Class III**: PVS* is not indicated as a screening tool to routinely risk stratify patients with TOF

AHA/ACC Guidelines for the Management of Adults with Congenital Heart Disease 2018

- **Class IIa**: PVS* can be useful to risk stratify adults with TOF and additional risk factors for SCD

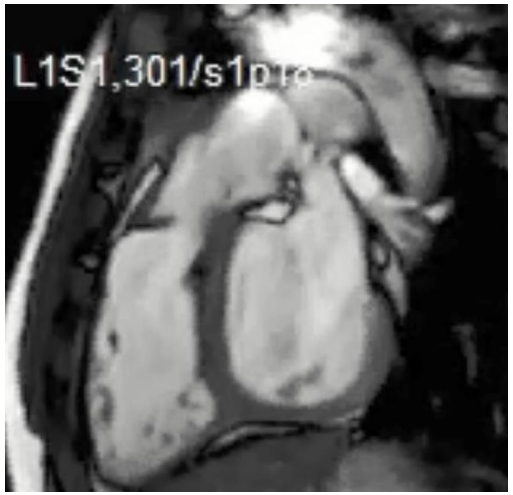
* Programmed ventricular stimulation

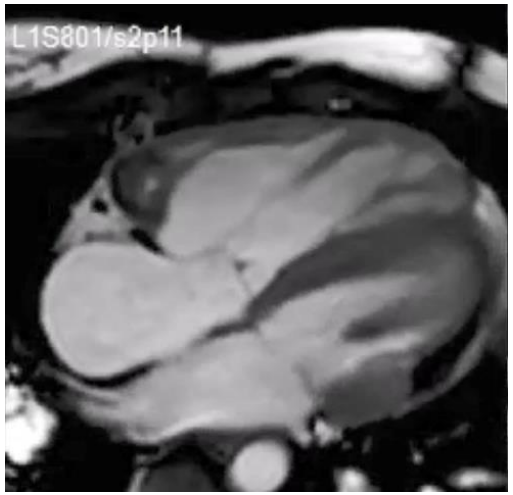
Not all TETS are the same



21 year old man

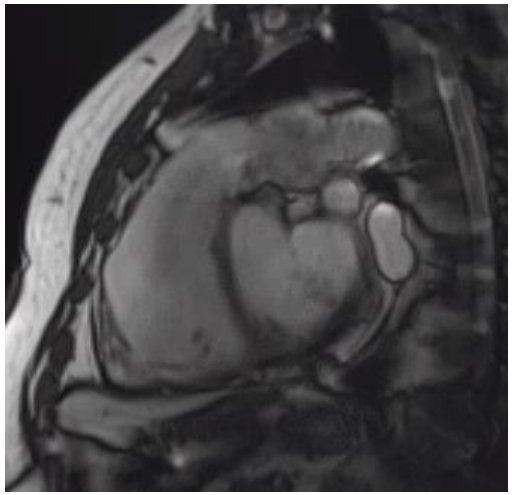
- Repair age 6 mos, TAP, free PI
- Exertional dyspnea
- No arrhythmias, no arrhythmic symptoms
- Normal biventricular function

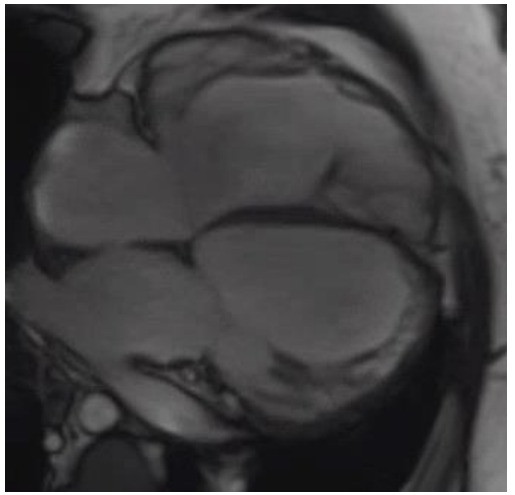




51 year old man

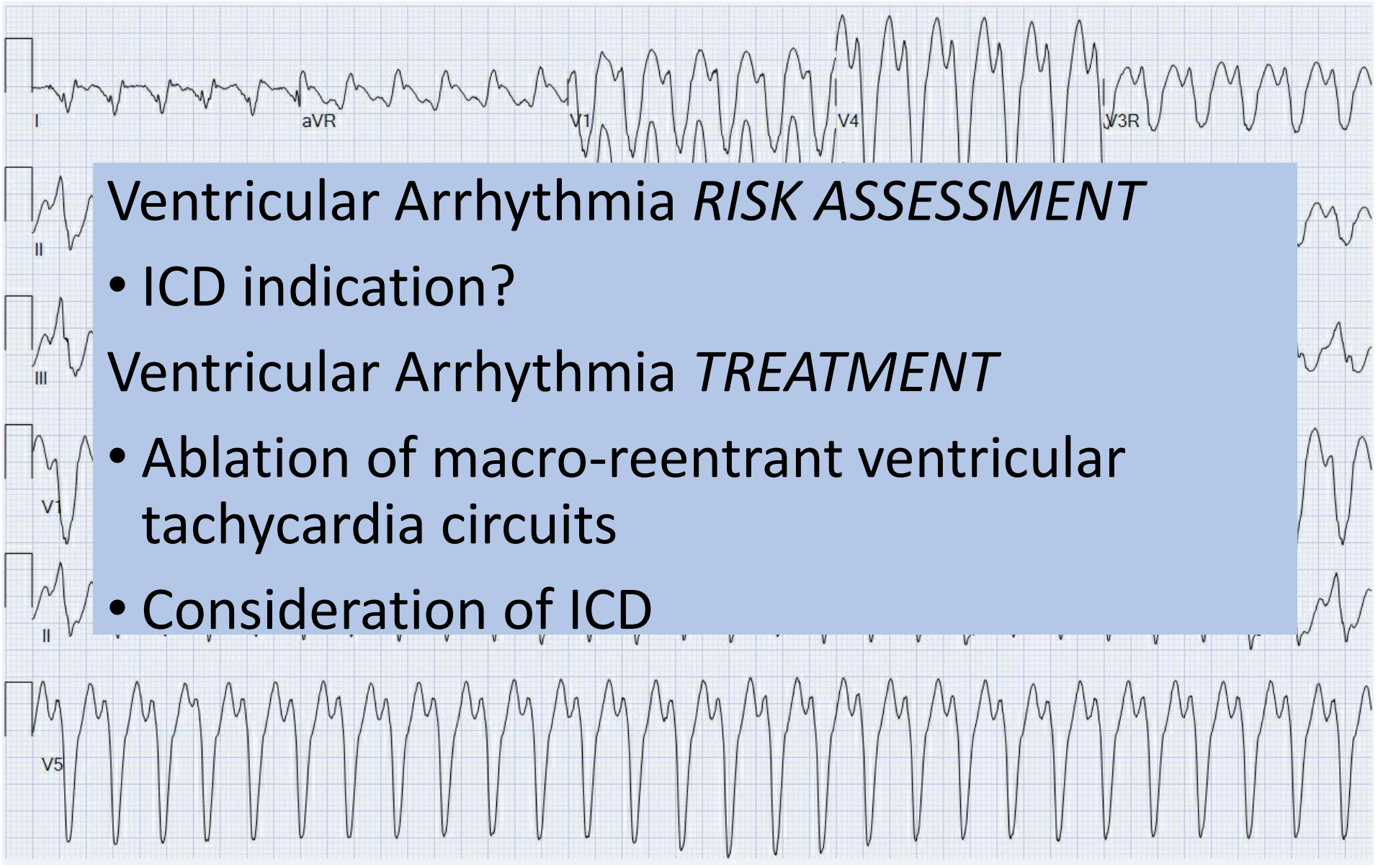
- BT shunt, repair age 6 years
- PVCs and NSVT on Holter
- Symptoms of palpitations and dizziness
- Biventricular dysfunction
- Extensive RV delayed enhancement





Images from Kan Hor, MD

Evolution of Ventricular Arrhythmias in rTOF



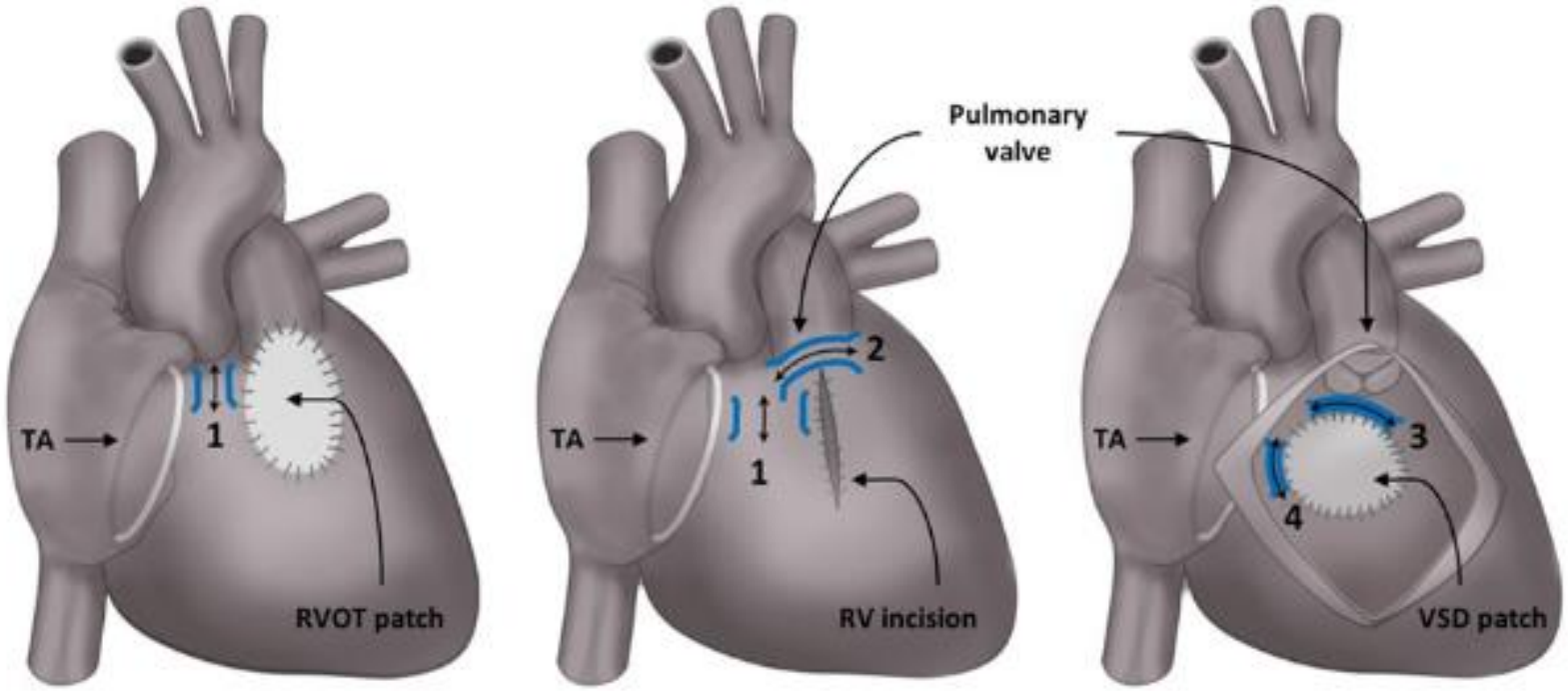
Ventricular Arrhythmia *RISK ASSESSMENT*

- ICD indication?

Ventricular Arrhythmia *TREATMENT*

- Ablation of macro-reentrant ventricular tachycardia circuits
- Consideration of ICD

Anatomic Arrhythmogenic Isthmuses in rTOF

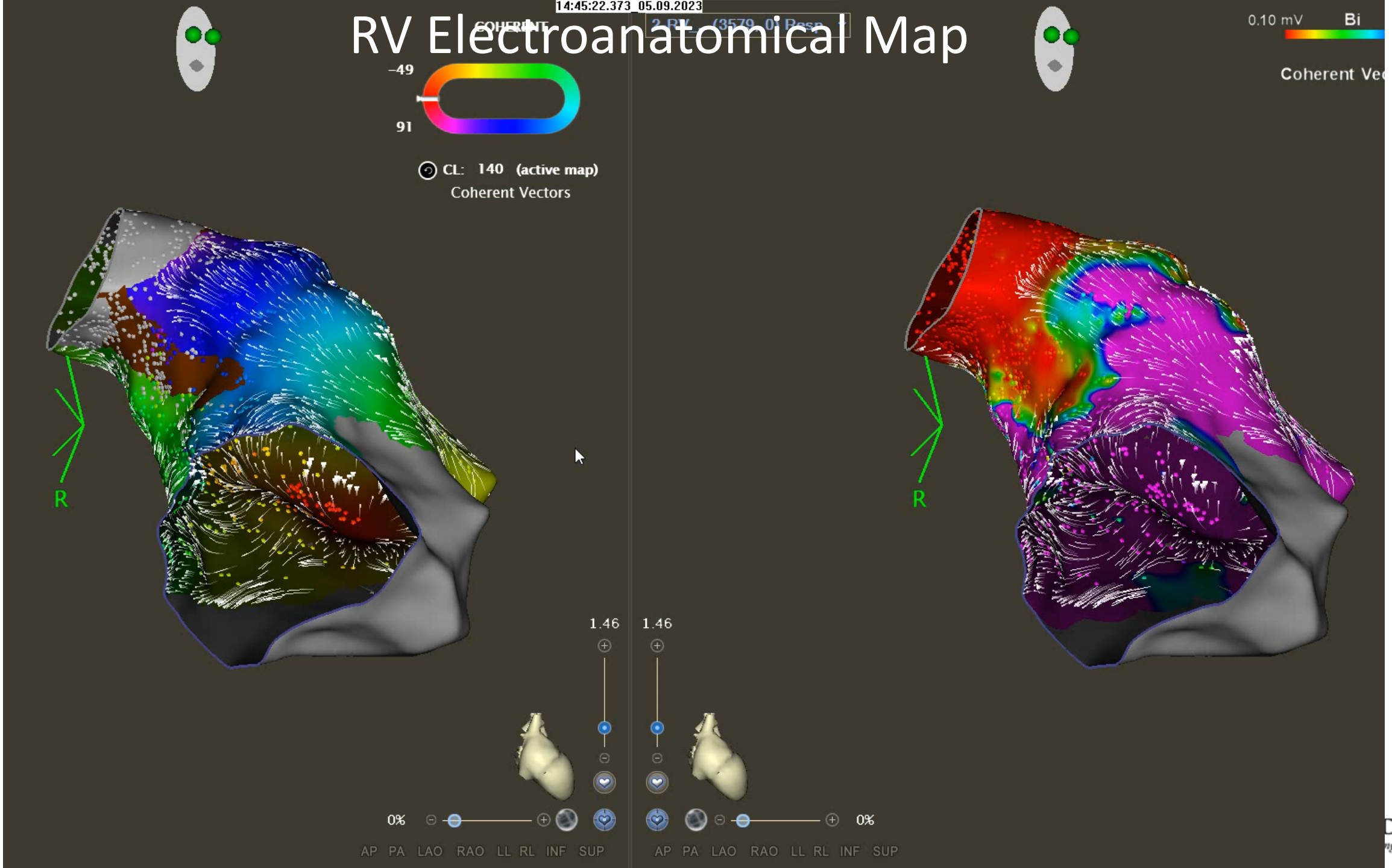


EP Study

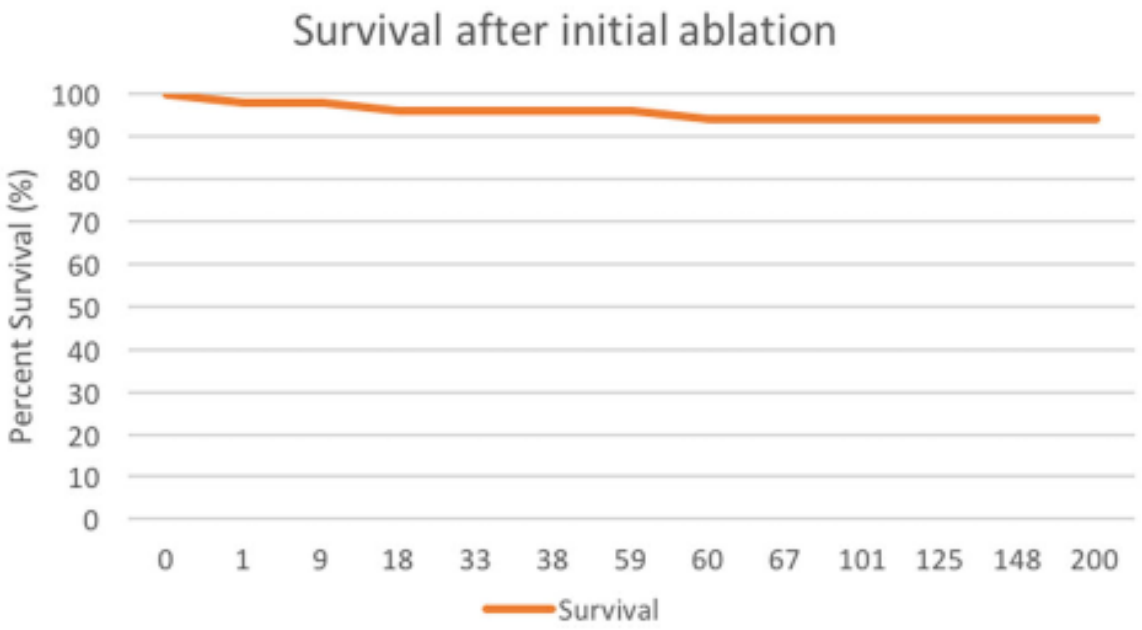
- RV electroanatomical map during sinus rhythm
- Assess for slow conduction through anatomic isthmus
  - Associated with inducible VT and ICD therapy

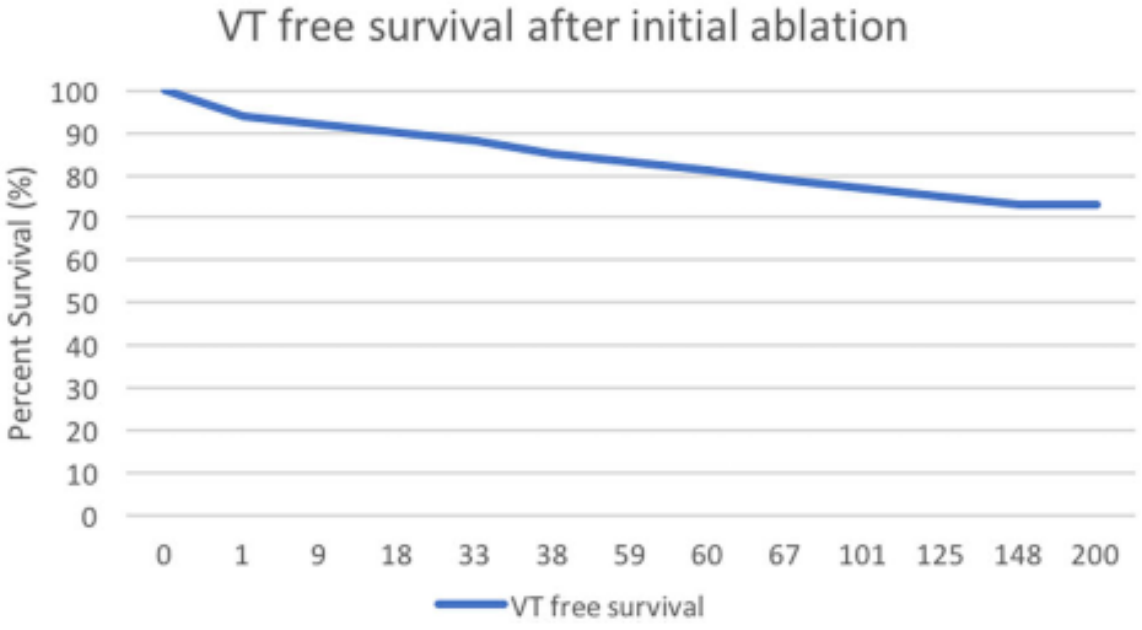
Kapel et al. Eur Hrt Jnl 2017

RV Electroanatomical Map



Does VT warrant an ICD?
ACHD VT ablation data





Yang et al. J CV EP 2019

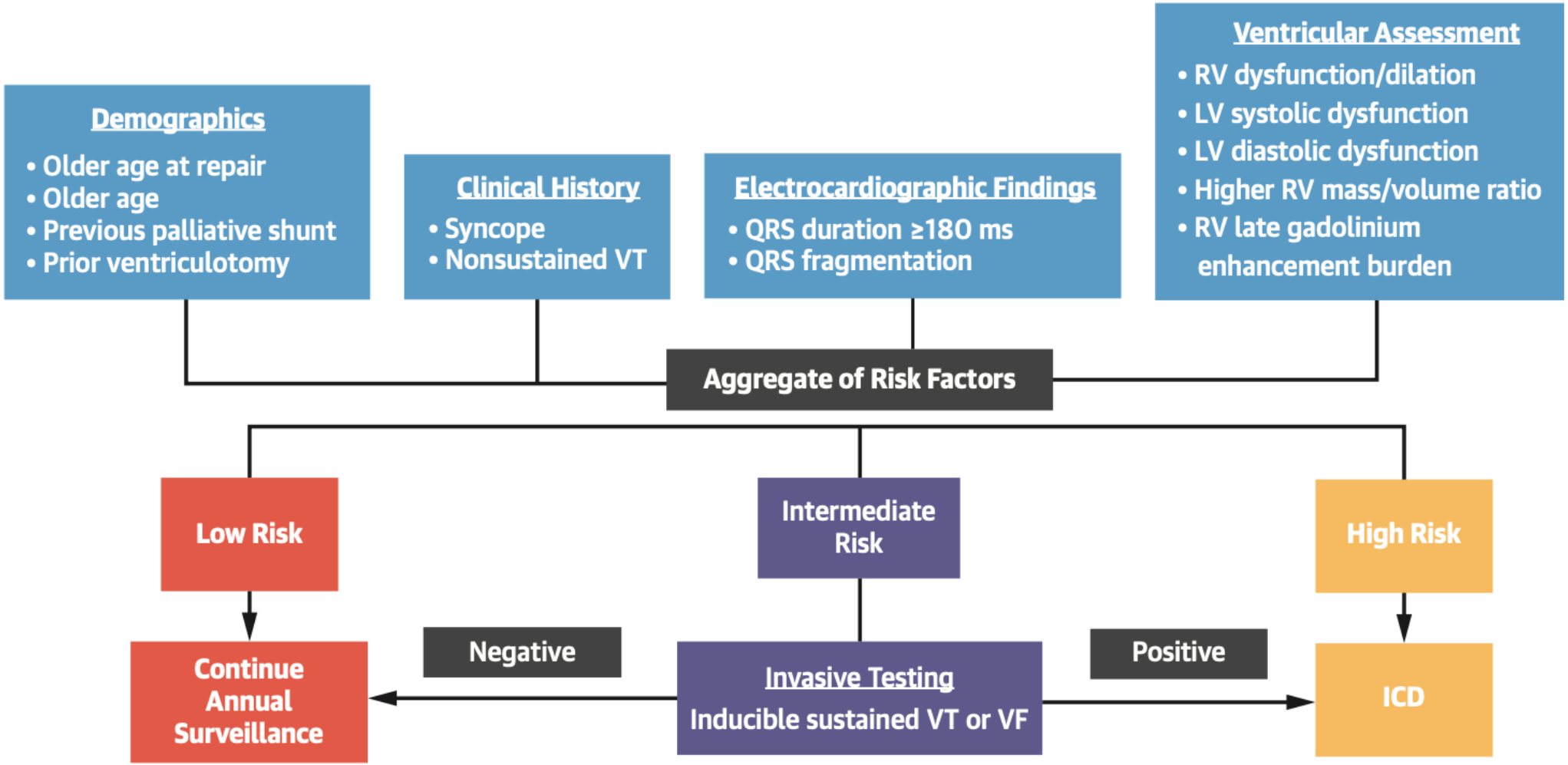
Does VT warrant an ICD? TOF VT ablation data

Univariate predictors of death, VT recurrence, and appropriate ICD therapies

Variable	Hazard ratio (95% CI	*P*
Age at surgical repair	1 (0.83–1.21)	0.9
Age at procedure	1 (0.93–1.10)	0.8
Time between first VT and VT CA	1 (0.94–1.16)	0.5
QRS complex duration	1 (0.99–1.04)	0.23
QRS complex duration ≥180 ms	0.42 (0.08–2.29)	0.3
RV systolic dysfunction	0.19 (0.03–1.04)	0.056
Moderate to severe PR	1.7 (0.2–14.65)	0.6
LVEF < 60%	16.35 (1.82–147)	0.013
>1 induced VT morphology	3.43 (0.4–29.4)	0.26

Laredo et al Arch of CV Disease 2017

Proposed rTOF VA Risk Assessment



Cohen et al. JACC 2021

Take Home Points
Ventricular Arrhythmias in Tetralogy of Fallot

- Surgical evolution has changed the arrhythmogenic substrate in rTOF
- Consider ventricular arrhythmia risk assessment at regular intervals
- Monomorphic VT – consider ablation
- Primary prevention ICD – shared decision making

THANK YOU

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Post Question

A 30 year old man with tetralogy of Fallot s/p complete repair at age 5 mos with VSD patch closure and transannular patch is being evaluated for pulmonary valve replacement with severe PI and exertional limitations. He has no known arrhythmias but has symptoms of palpitations. What is the most appropriate arrhythmia management for him?

A. Start amiodarone prior to PVR
B. Offer ICD due to possible risk of ventricular arrhythmias
C. Evaluate clinical risk factors for ventricular arrhythmias and consider usefulness of invasive EP testing
D. No further arrhythmia evaluation needed