

Adult Congenital Heart Disease for the Internist

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The Ohio State University

Nationwide Children's Hospital



COACH

Columbus Ohio Adult Congenital
Heart Program



THE OHIO STATE UNIVERSITY

WEXNER MEDICAL CENTER

Objectives

- To discuss the increasing prevalence of adult congenital heart disease
- To discuss the common congenital heart disease diagnoses encountered in primary care clinics
- To discuss strategies for lifelong care of adult patients with congenital heart disease

Why Should the Internists Care?



Circulation Research

HOME ABOUT THIS JOURNAL ▼ ALL ISSUES SUBJECTS ▼ BROWSE FEATURES ▼

EDITORIAL

Congenital Heart Disease

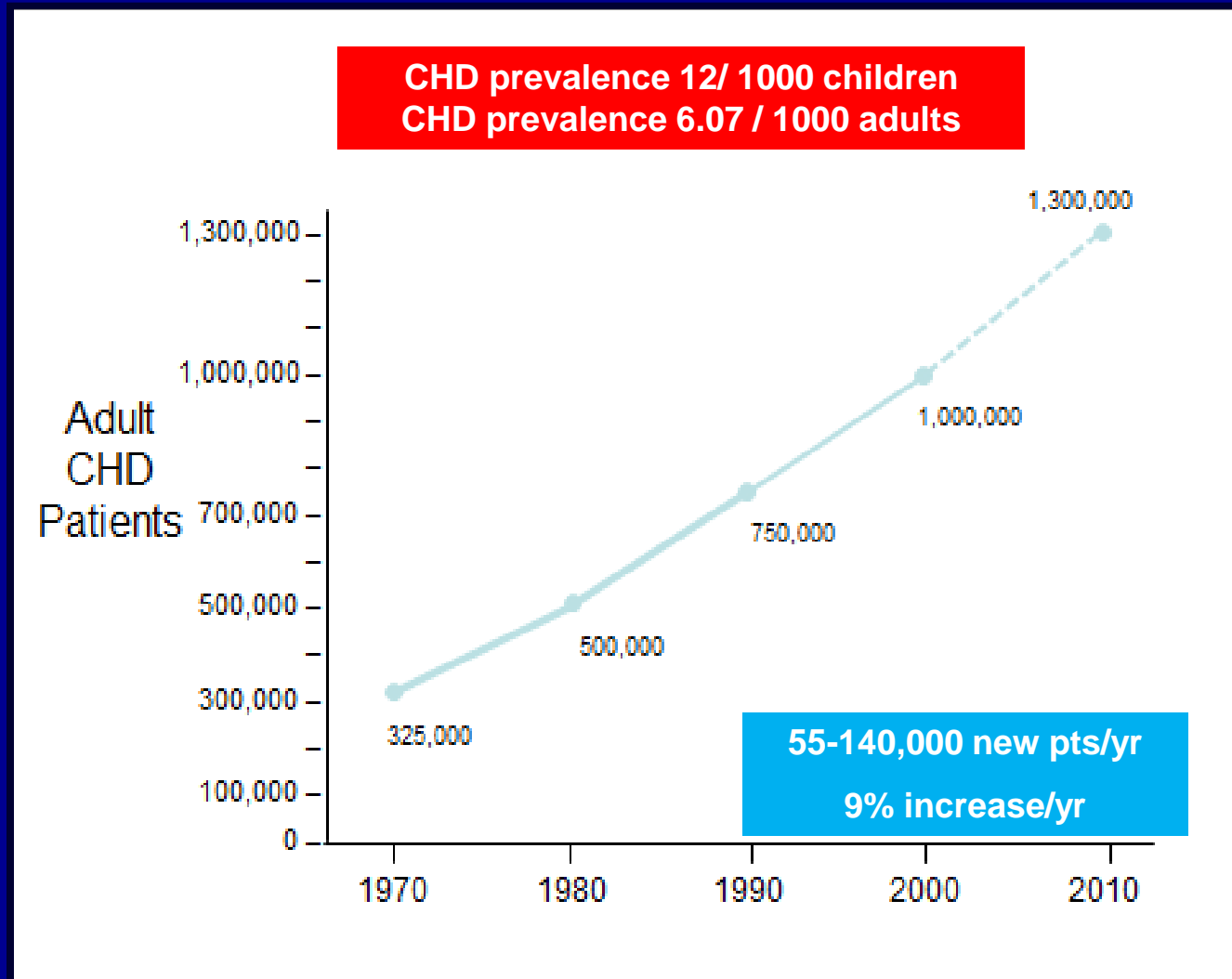
The Remarkable Journey From the “Post-Mortem Room” to Adult Clinics

Ali J. Marian

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DOI <https://doi.org/10.1161/CIRCRESAHA.117.310830>
Circulation Research. 2017;120:895-897
Originally published March 16, 2017

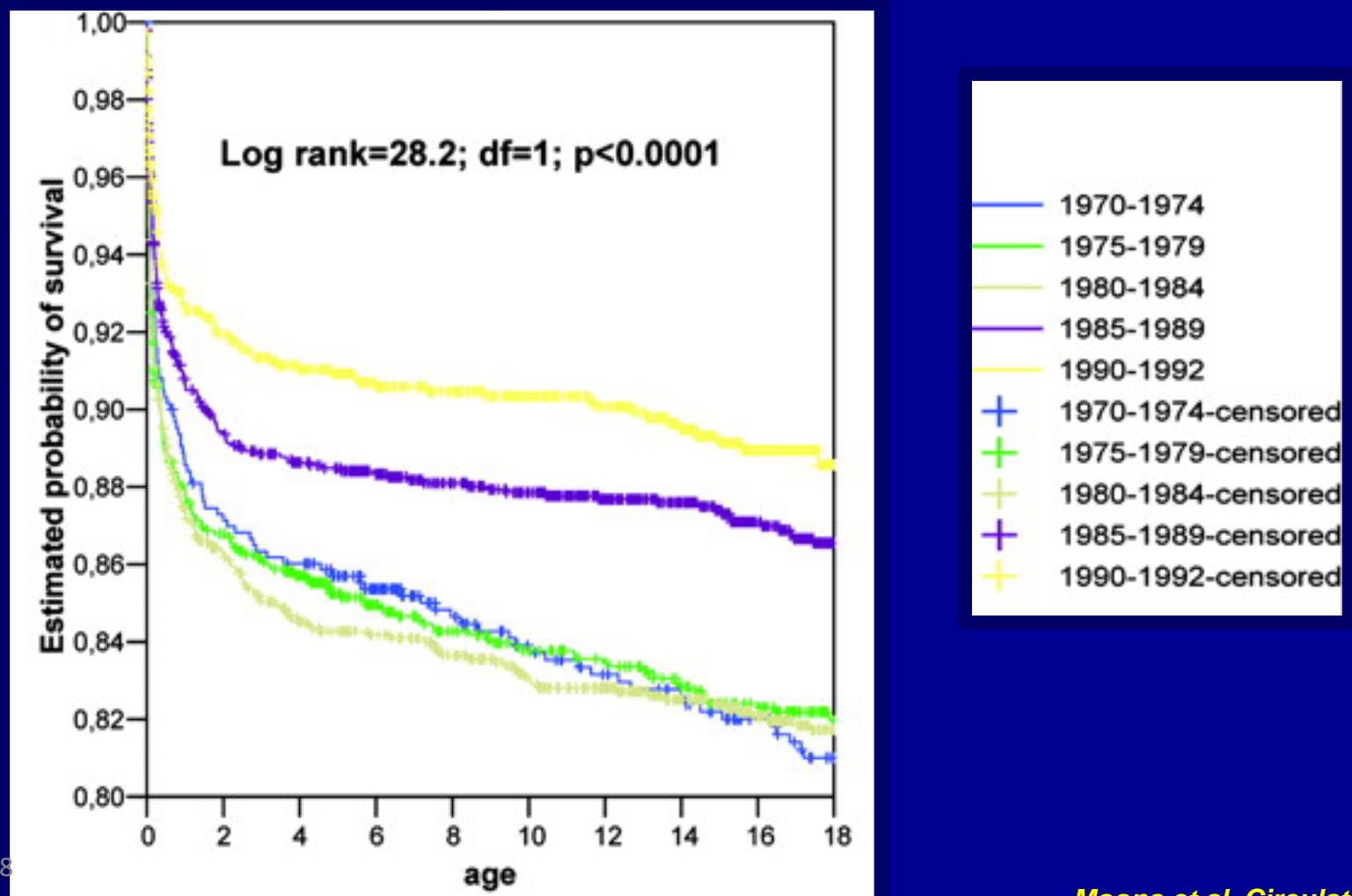
EXPLOSION of a “Life-Long” Disease Burden



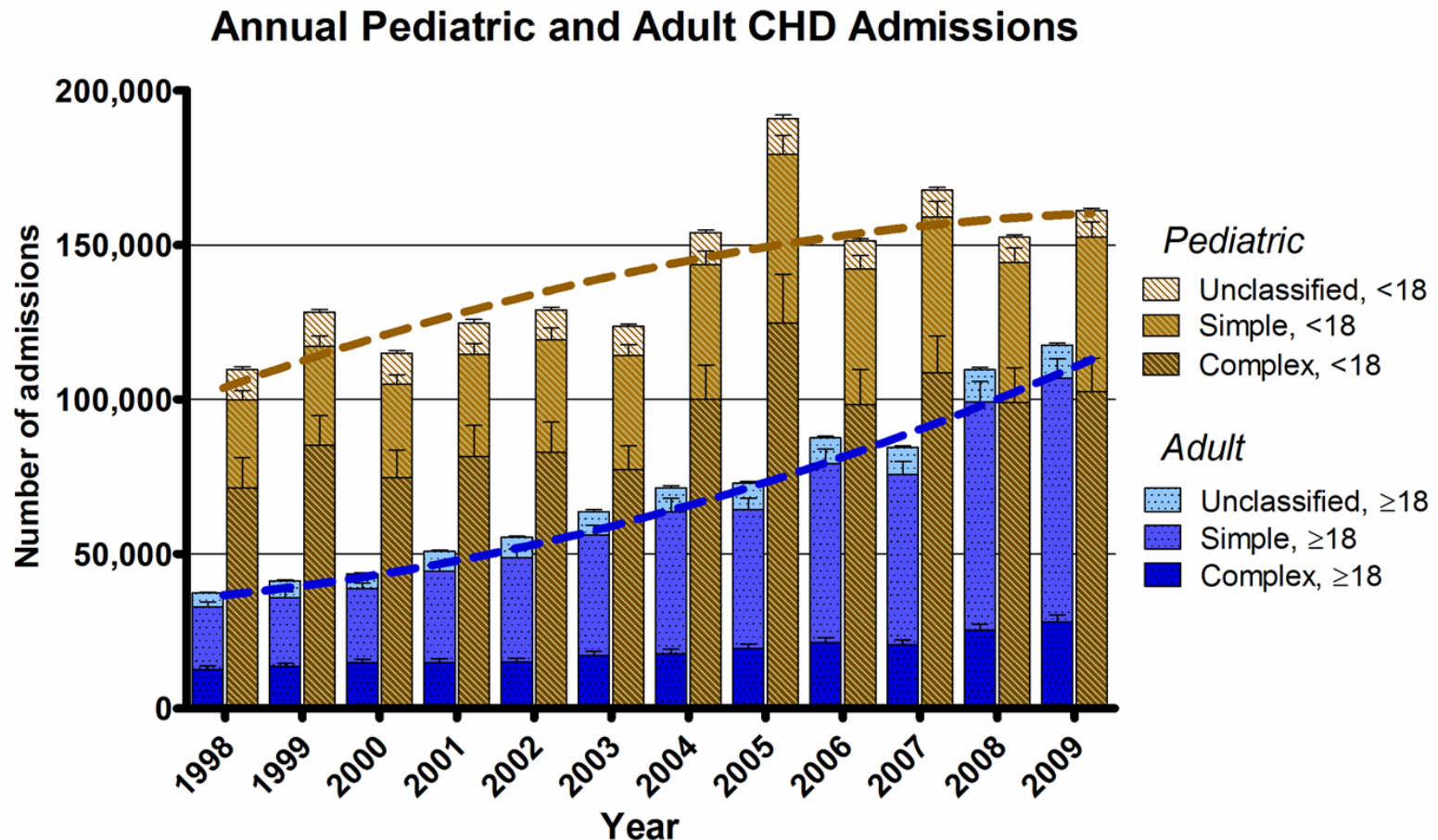
Hoffman J et al. *Am J Cardiol*. 1978; 641-647. Fyler D. 1980. 375-461. Ferencz C et al. *Am J Epidemiol*. 1985;31-6. Congenital heart disease after childhood: an expanding patient population. 22nd Bethesda Conference, Maryland, October 18-19, 1990. *J Am Coll Cardiol* 1991;18(2):311-342. Warnes CA et al. *J Am Coll Cardiol* 2001;1170-1175. Warnes CA et al. *J Am Coll Cardiol* 2008;52(23):1890-1947; Marelli A et al. *Am Heart J*. 2009; 1-8. Karouache M et al. *J Am Coll Cardiol* 2013; 61; E505..

Estimated Survival – Era Effect

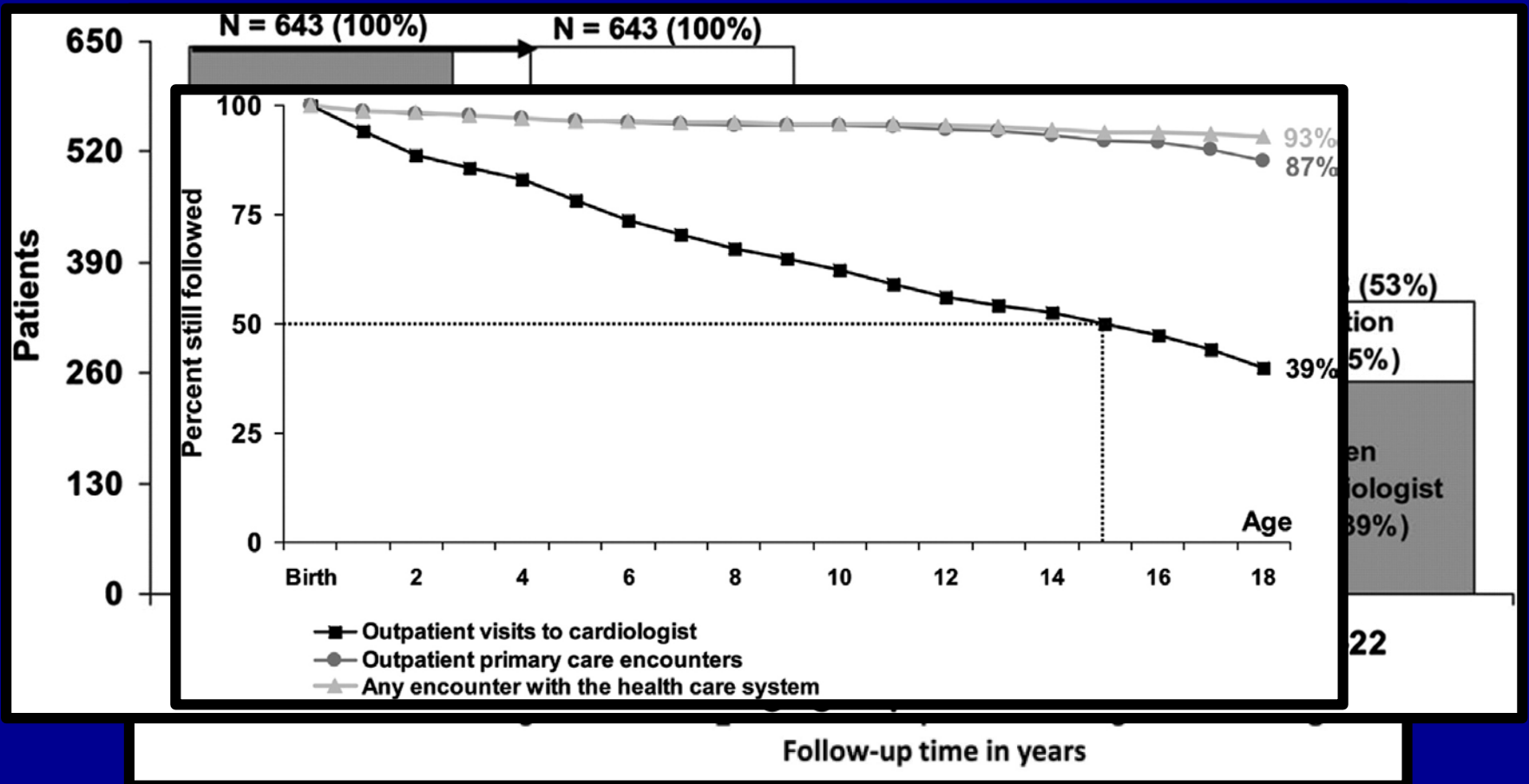
Birth - 18 years



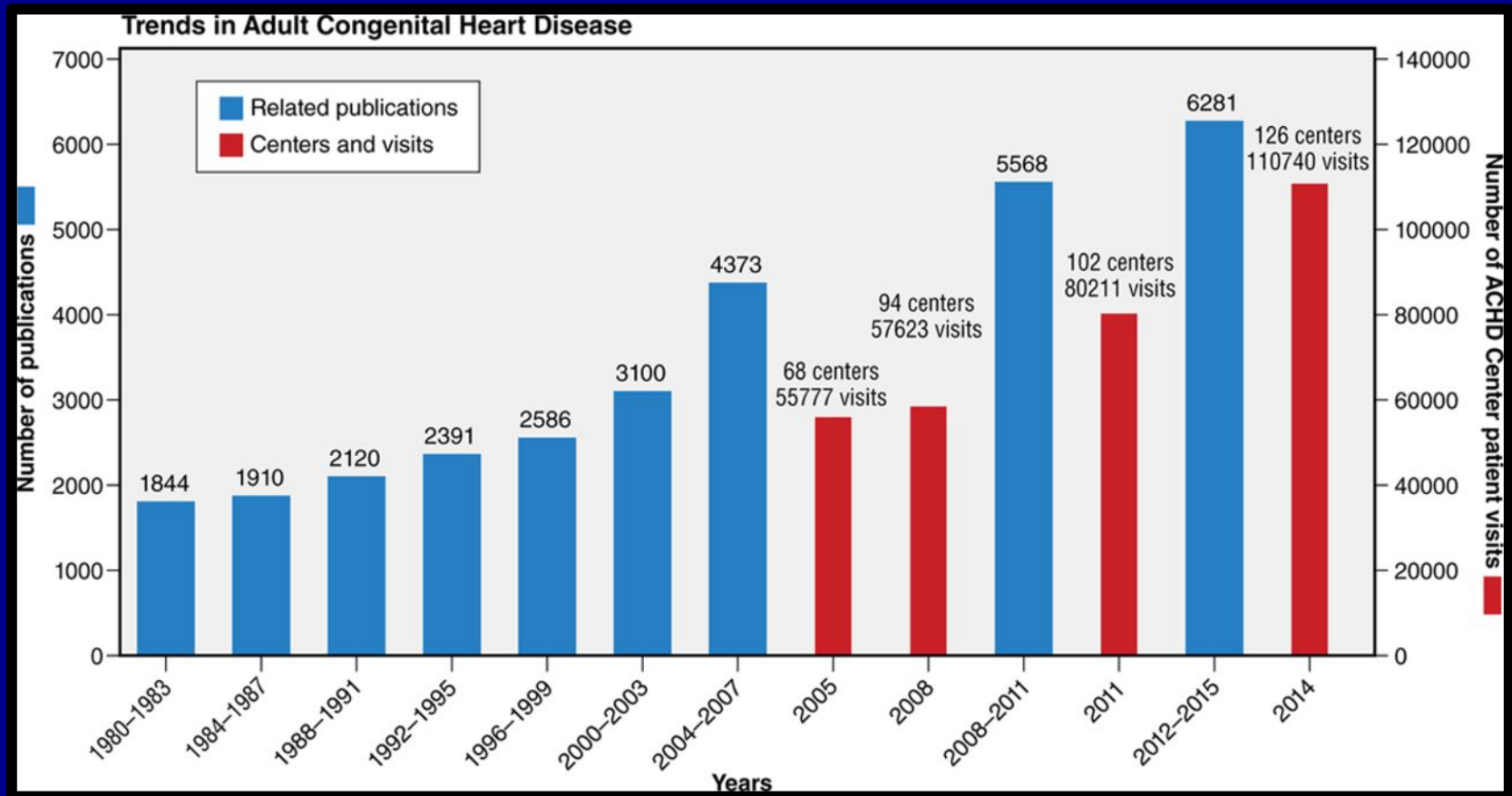
Hospitalizations for Adults With Congenital Heart Disease in the U.S.



Trends in ACHD



Trends in ACHD

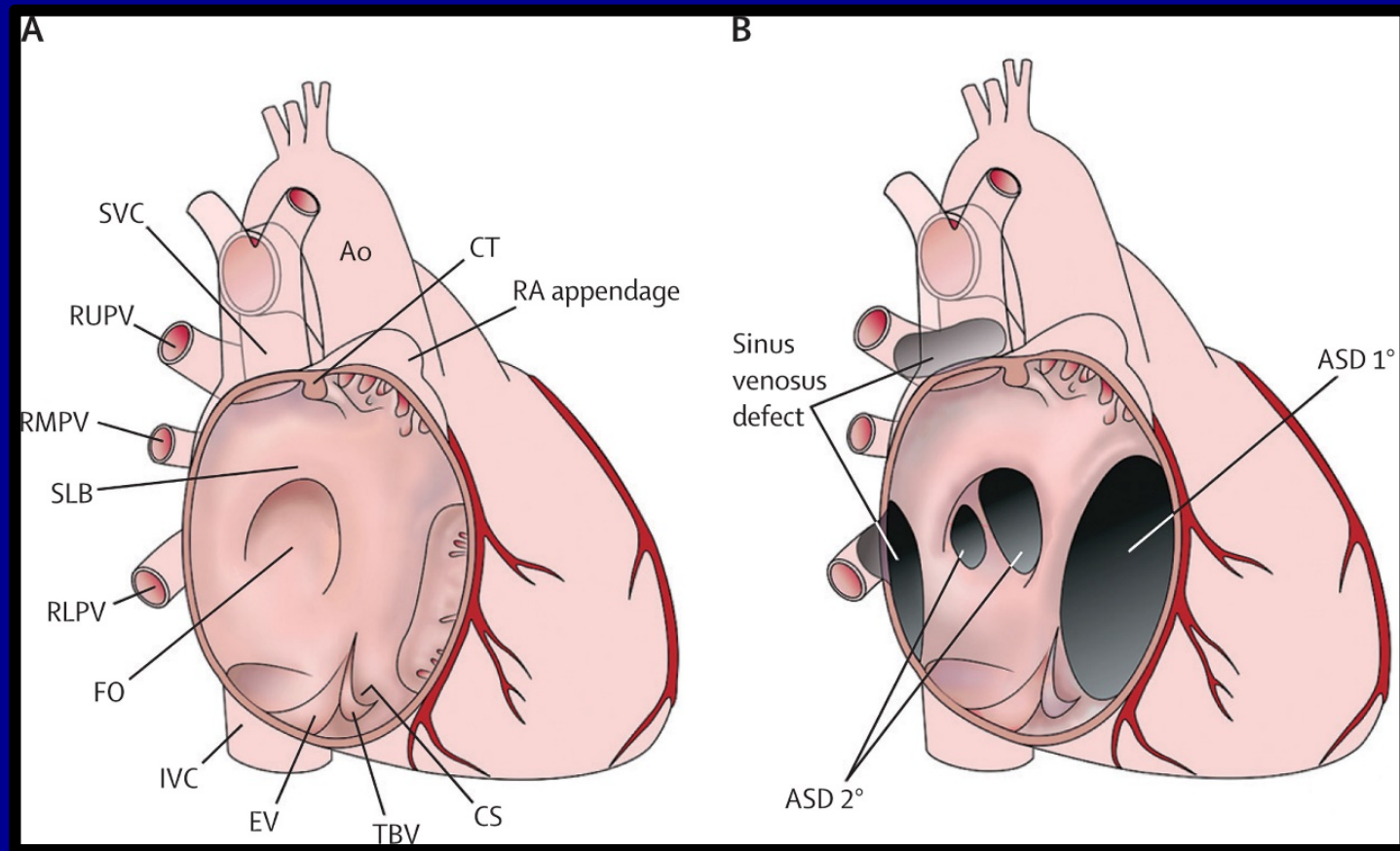


ACHD Patients- Common Issues

- Quality of life
- Transition
- Birth Control
- Pregnancy and CHD
- Dental Issues
- Exercise
- Hep C

- Pulmonary Hypertension
- Heart Failure
- Arrhythmias
- Neurocognitive issues
- Advance care planning and advanced directives

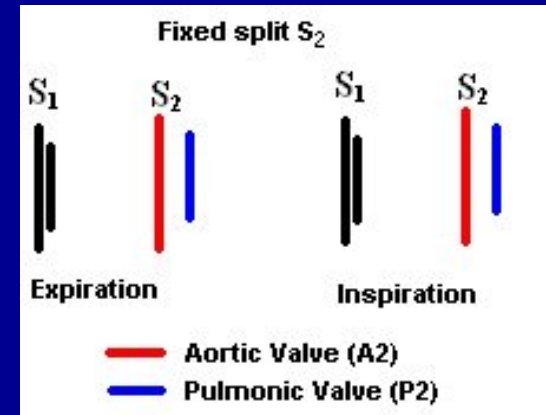
Atrial Septal defect



Geva et al. Lancet 2014

Physical Exam – Atrial Septal Defect

- Hyperdynamic precordium
- Loud P2- Pulm HTN
- Signs of RHF rare
- Widely split and fixed S2

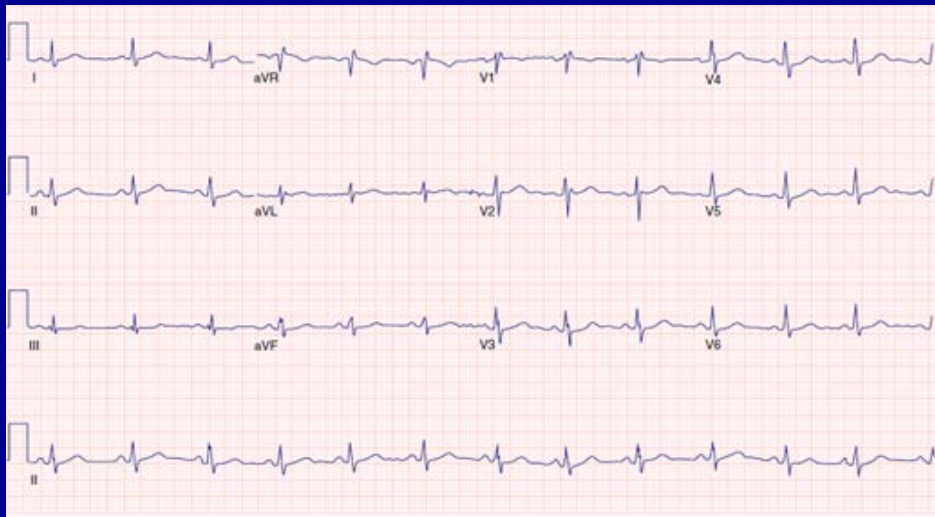


Murmurs in ASD

- Soft SEM- LUSB
- Diastolic rumble over LLSB- increased flow TV
- HSM at LLSB- TR

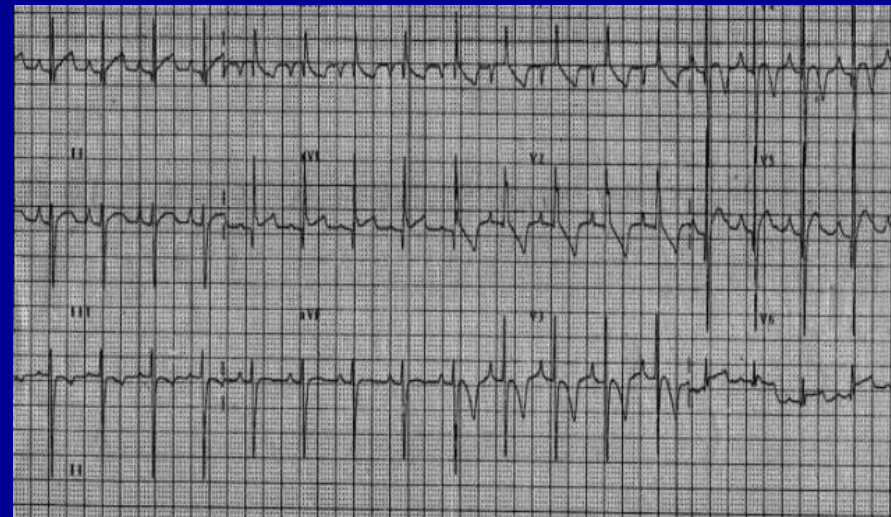
EKG

Secundum ASD



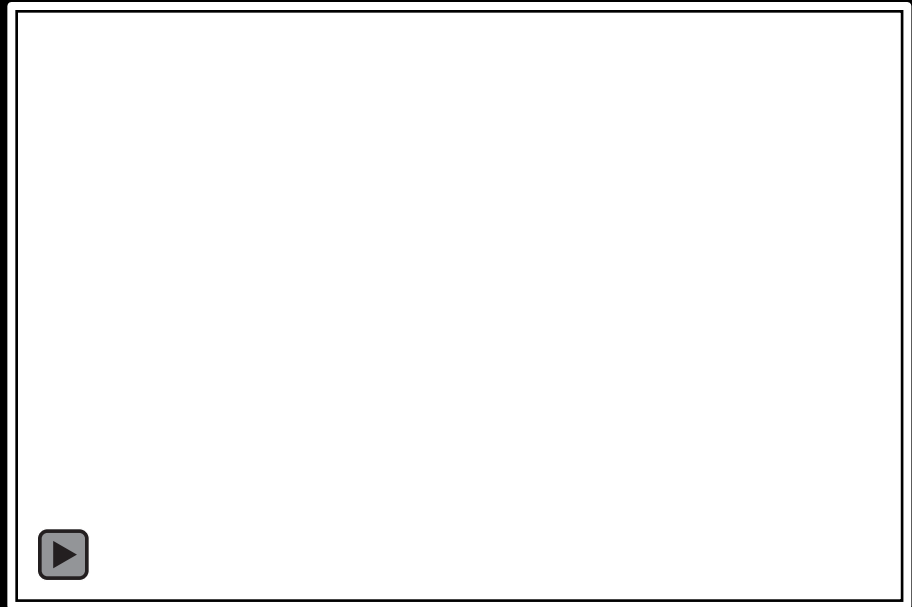
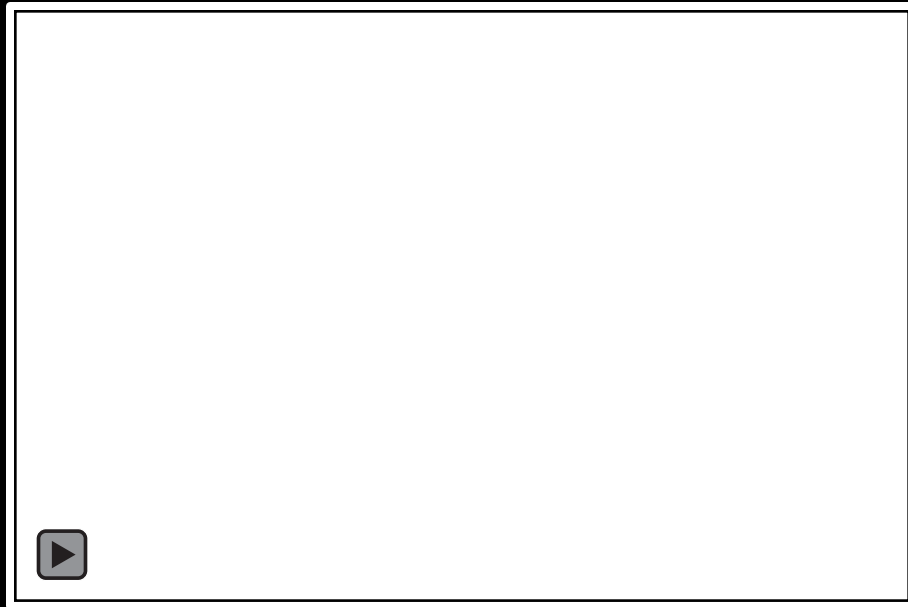
Incomplete RBBB
Right Axis Deviation

Primum ASD



Incomplete RBBB
Left Axis deviation

ASD – Myocardial Infarction and PE after IVDU



Associated Anomalies

- Anomalous pulmonary veins
- VSD
- Mitral regurgitation with primum ASDs due to cleft valve

RV Dilation



RVEDV_i: 145 ml/m²
RVEF: 61%

LVEDV_i: 102 ml/m²
LVEF: 56%

Qp/Qs : 1.1
TR fraction: 49%

Etiologies of RV Dilation

Tricuspid regurgitation

Pulmonary regurgitation

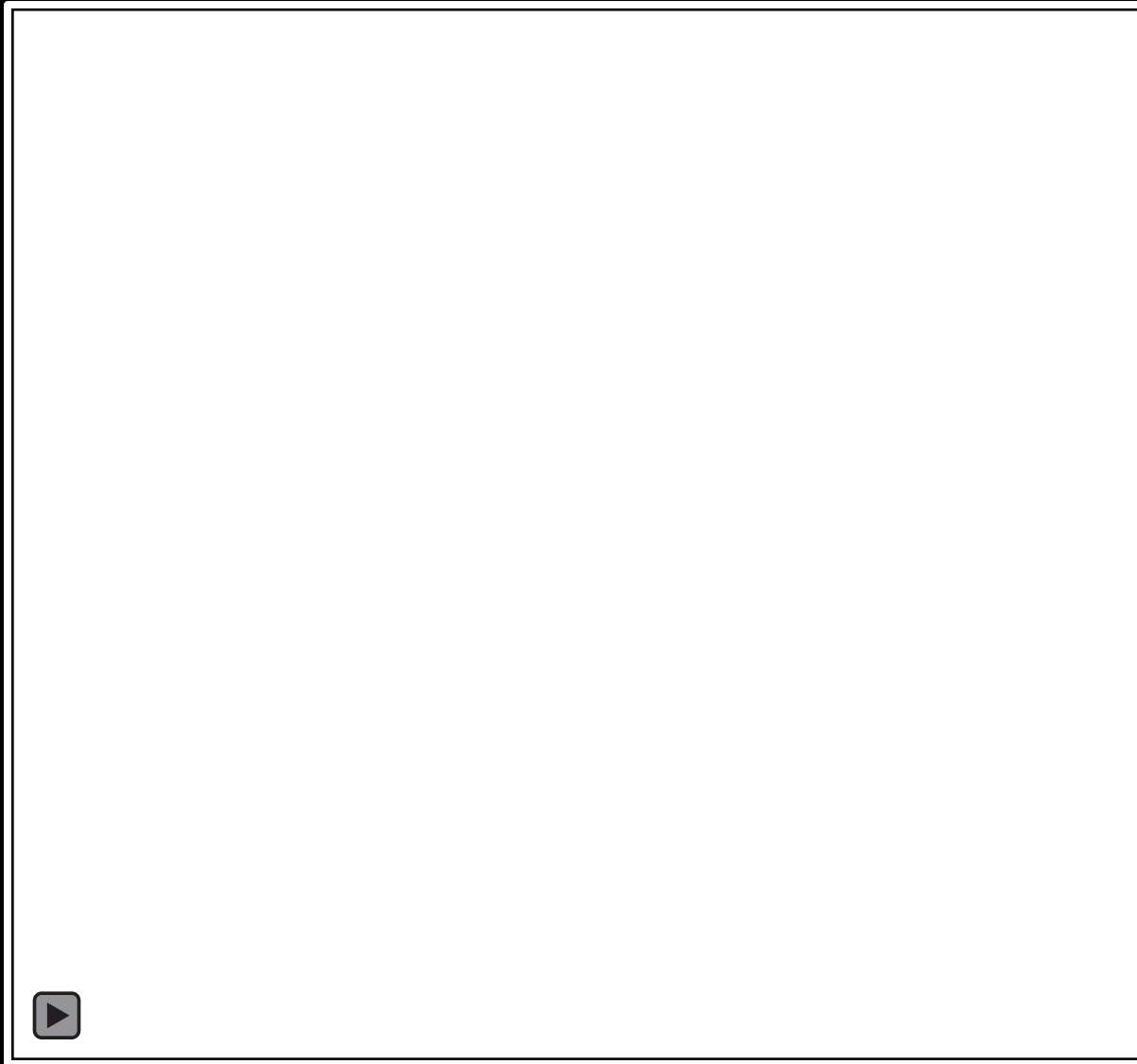
Pulmonary artery hypertension

Shunt Lesions

Myocardial abnormalities

- Uhl's anomaly
- ARVC
- Ventricular dysfunction

Inferior Sinus Venosus – ASD

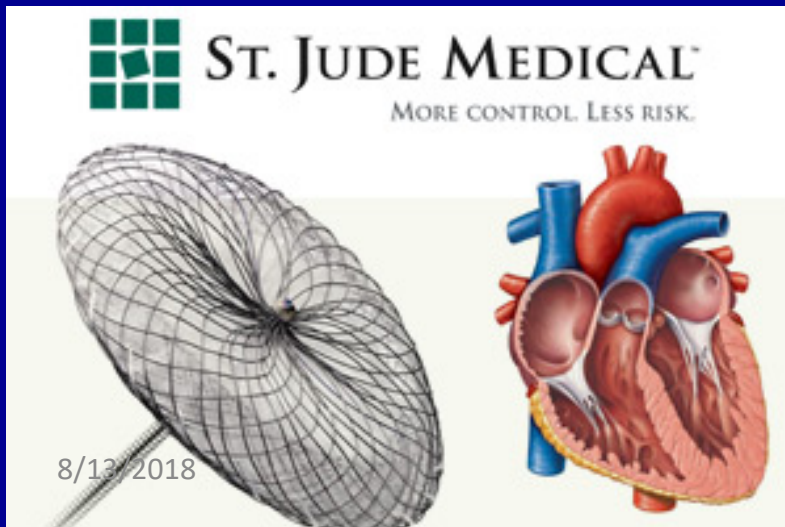


ASD- Indications for Closure

- **Significant left-to-right shunt**
 - right ventricular volume overload
 - with or without symptoms
 - without pulmonary hypertension*
- **Orthodeoxia-platypnea**
- **Paradoxical embolism**
- **At the time of another cardiac surgery**

ASD Closure

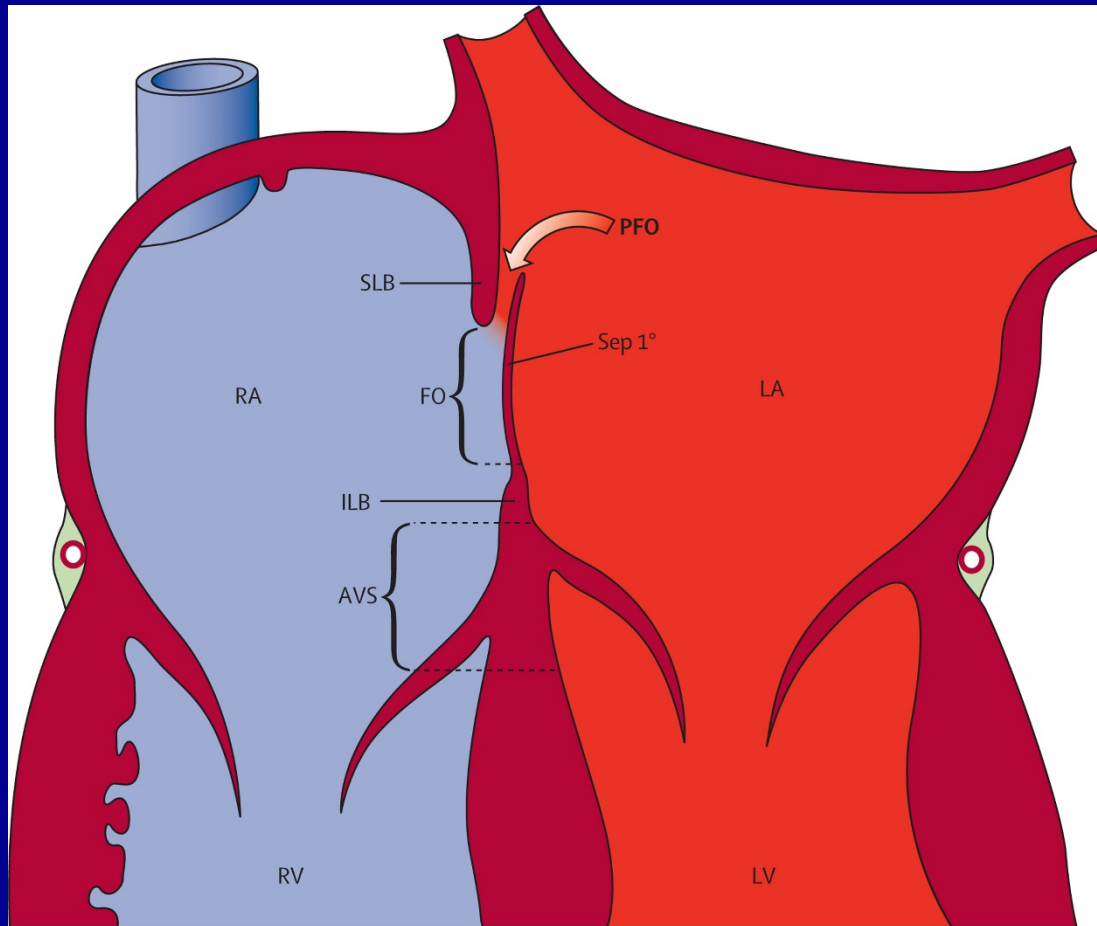
- Surgical
- Transcatheter



Other Issues with ASDs

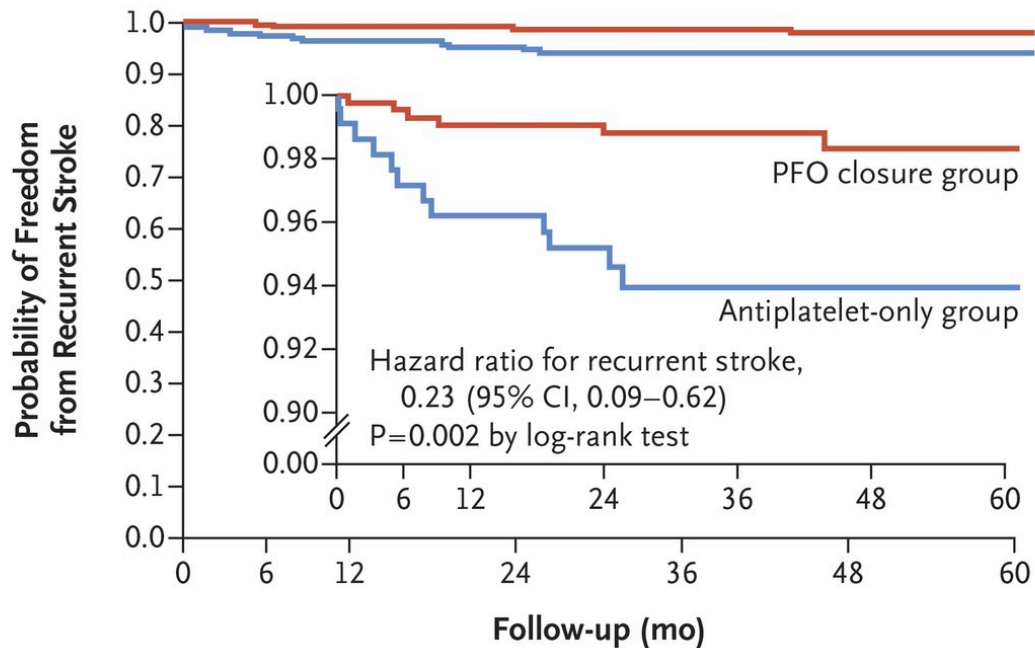
- Periodic follow up
- Arrhythmias (also with repaired)
- Pulmonary hypertension
- Scuba diving
- High altitude exposure

Patent Foramen Ovale



Geva et al. Lancet 2014

Patent Foramen Ovale

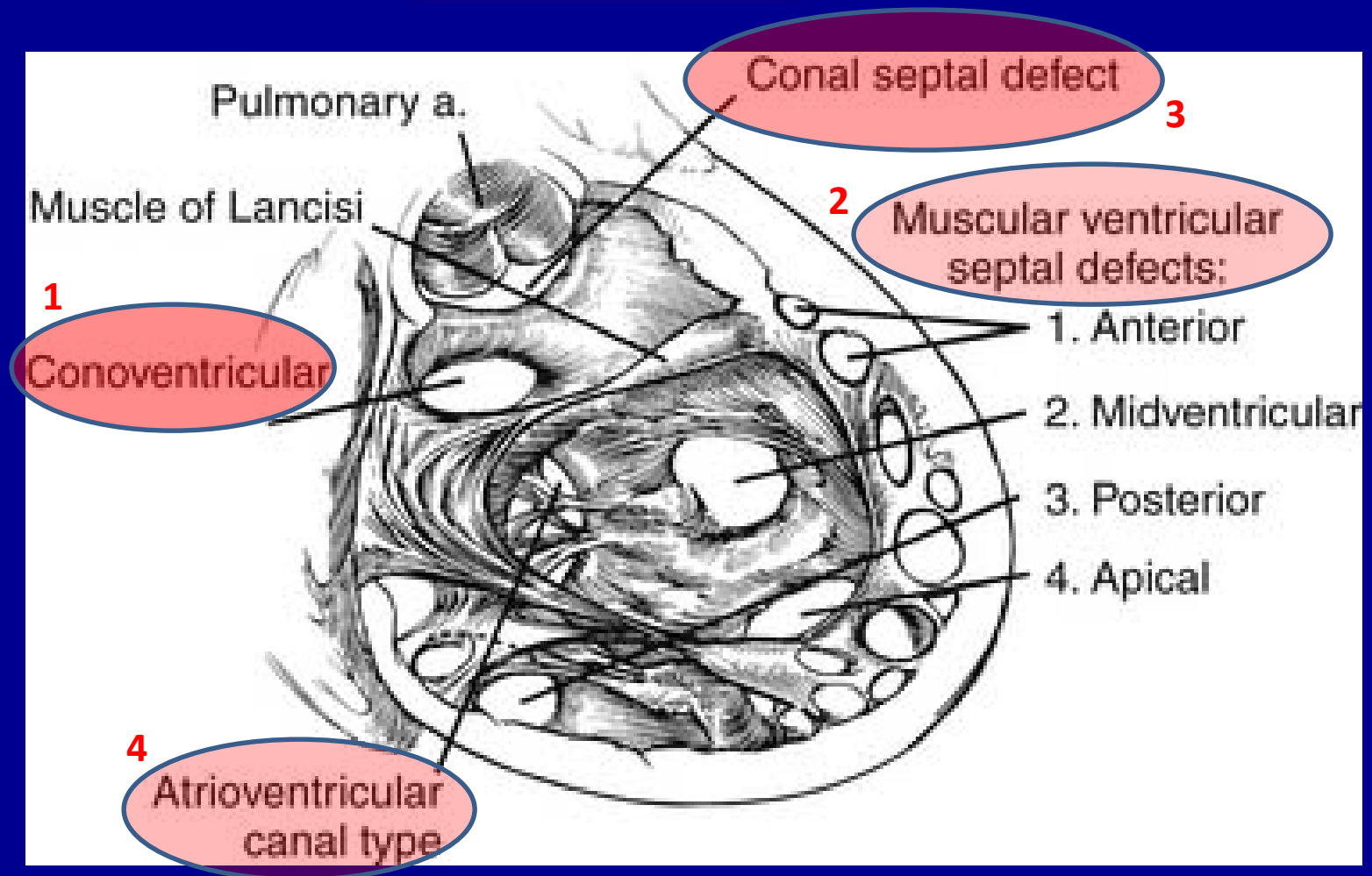


No. at Risk							
PFO closure group	441	422	417	398	278	182	102
Antiplatelet-only group	223	202	194	173	116	78	30

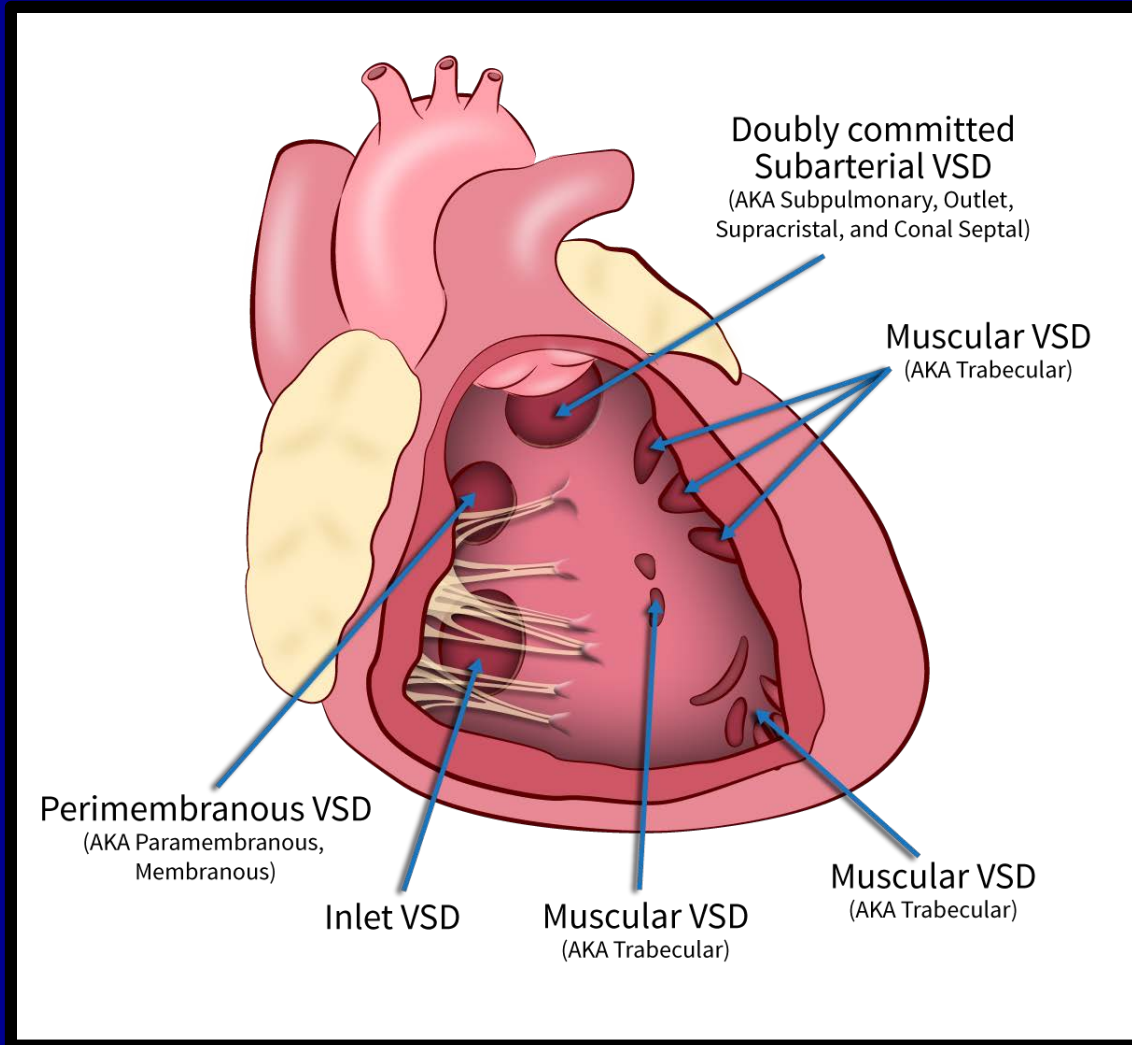
PFO - To close or not to close..

- Patient Factors
 - Hypercoagulable state
 - Atrial Fibrillation
 - ASCVD Risk Factors
 - Presence of devices in the RV
- PFO factors
 - Shunt size
 - Atrial Septal Aneurysm

Ventricular Septal Defect



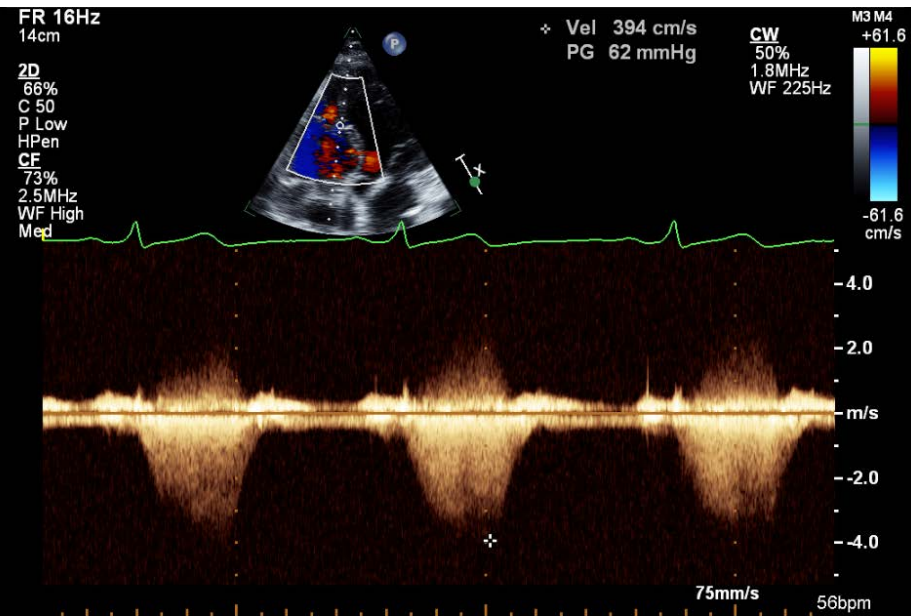
Ventricular Septal Defect



Physical Exam and EKG

- Smaller the VSD, louder the murmur
- Holosystolic plateau-shaped murmur at LLSB
- Majority of patients with isolated ventricular septal defect (VSD- Normal ECG)
- EKG signs of LAE and LVH – maybe

VSD



Indications for Closure of VSD

- Symptoms of heart failure
- Large LV
- Normal PVR

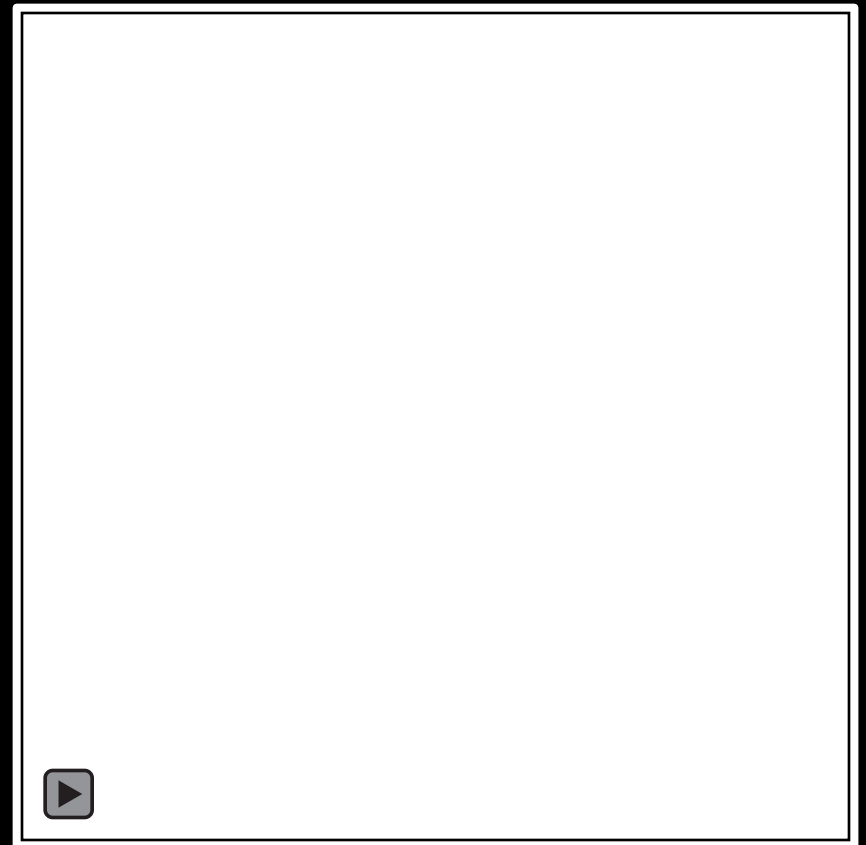
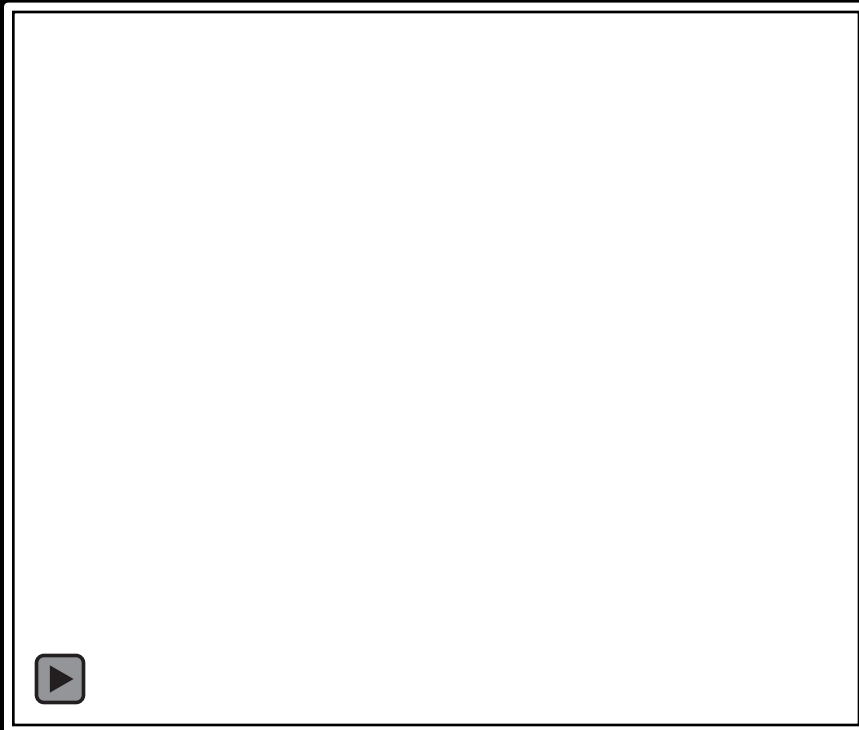
VSD Closure in Adults

- Intervention is rarely required
- Adult patients with large VSDs - irreversible pulmonary vascular disease - should never undergo intervention
- Small VSDs from childhood have usually closed spontaneously
- If small VSD's they remain open, do not create a clinically important shunt

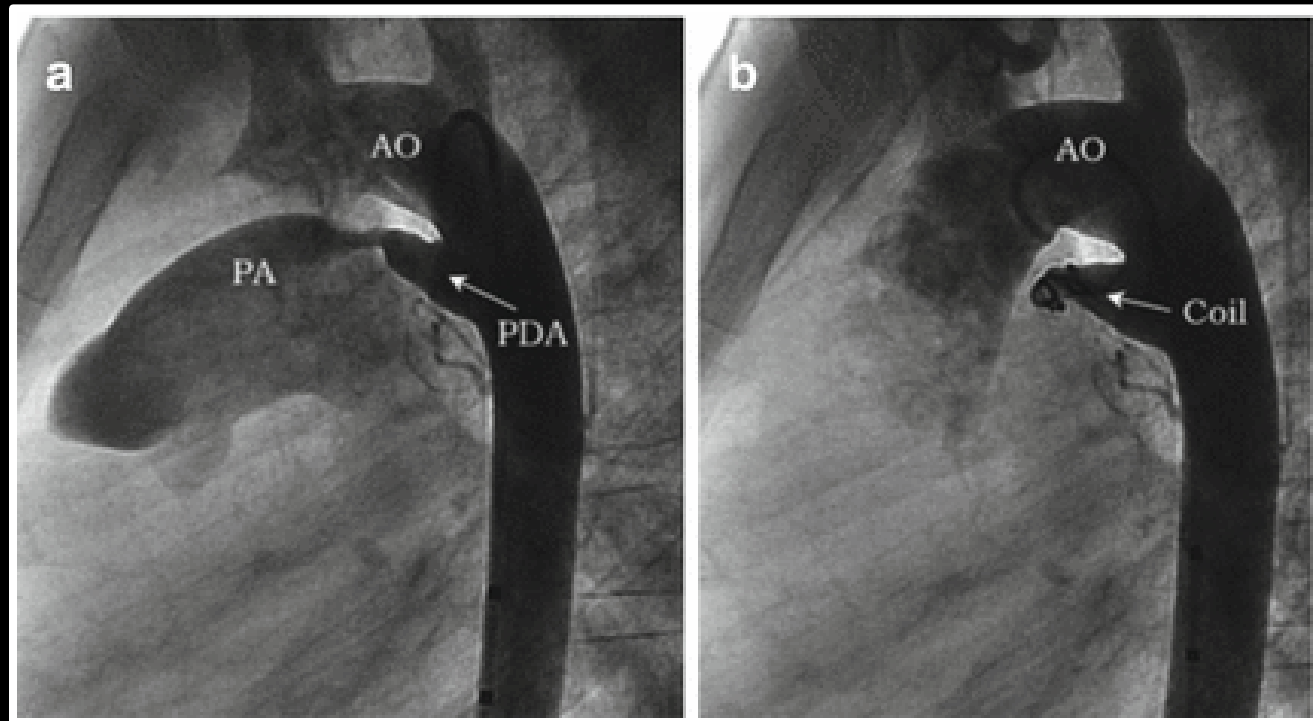
Lifelong Follow Up -VSD

- More often repair of the VSD is required for non–shunt related issues such as
 - Endocarditis
 - Aortic insufficiency
 - Pulmonary insufficiency
- Surgical repair remains the gold standard for treatment of VSD.
- Transcatheter closure of VSD- investigational

Gerbode Defect



Patent Ductus Arteriosus



Significant Left to Right
Shunt

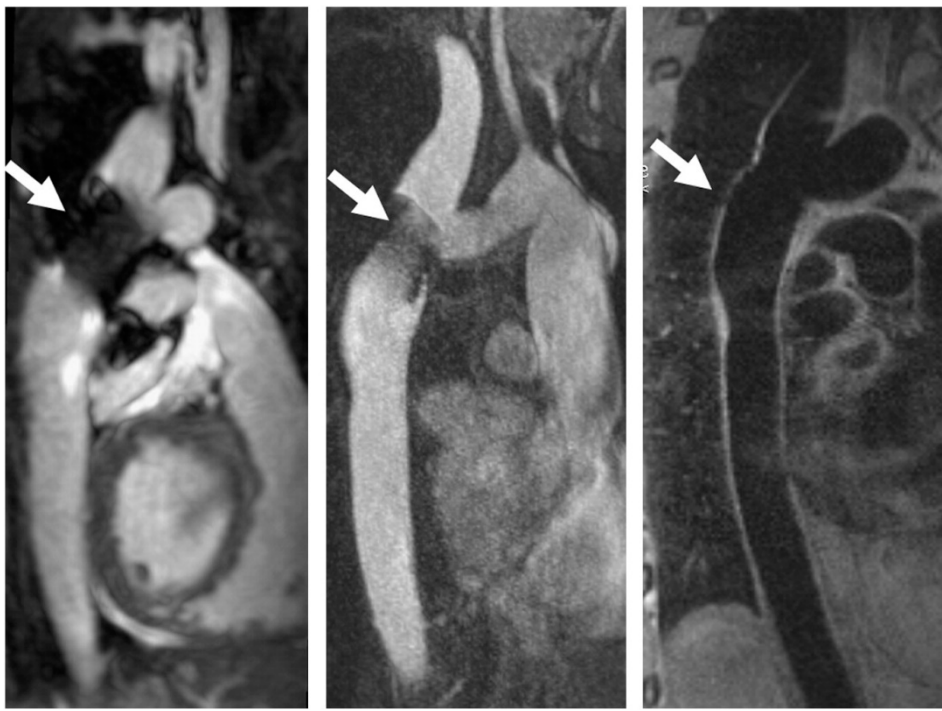
- Symptoms
- LA and LV enlargement

Endarteritis

Coarctation of Aorta

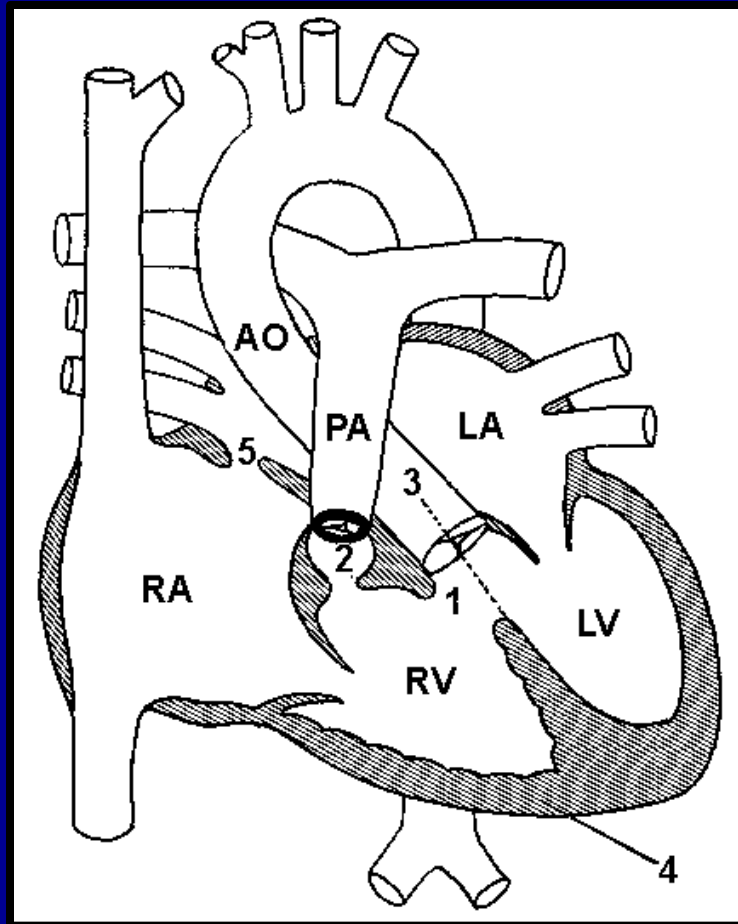
- Familial risk
- Turner syndrome
- Associated anomalies
 - ASD
 - VSD
 - Bicuspid aortic valve
- Hypertension
- Brachial-femoral delay
- Premature CAD, Stroke
- Intracranial aneurysms
- Surgery, Balloon Angioplasty, Stent

Coarctation of Aorta- 3D SPACE TSE

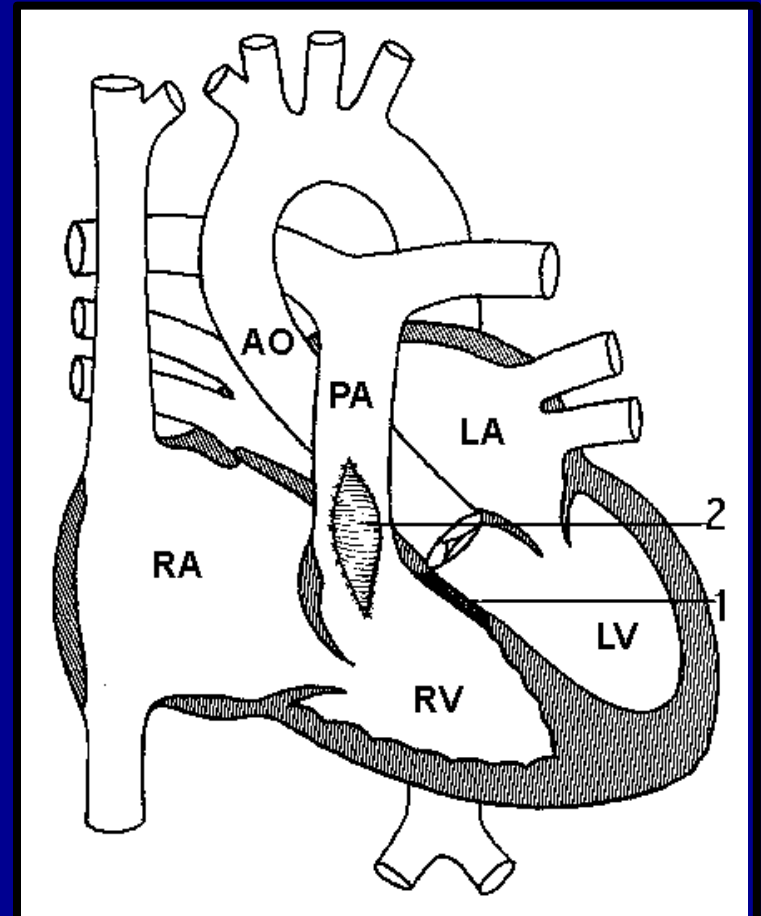


Tetralogy Of Fallot (TOF)

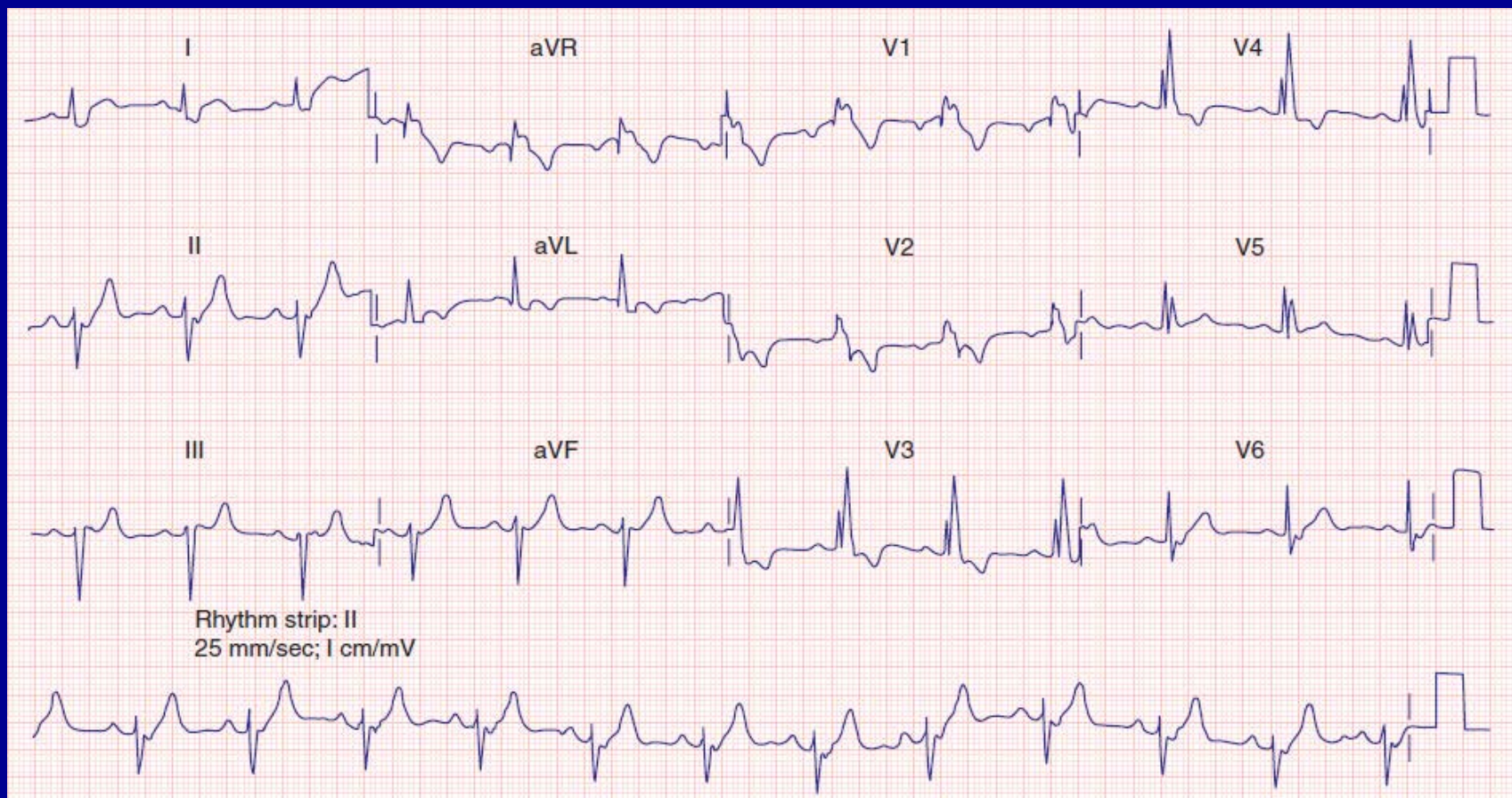
Unrepaired



Repaired



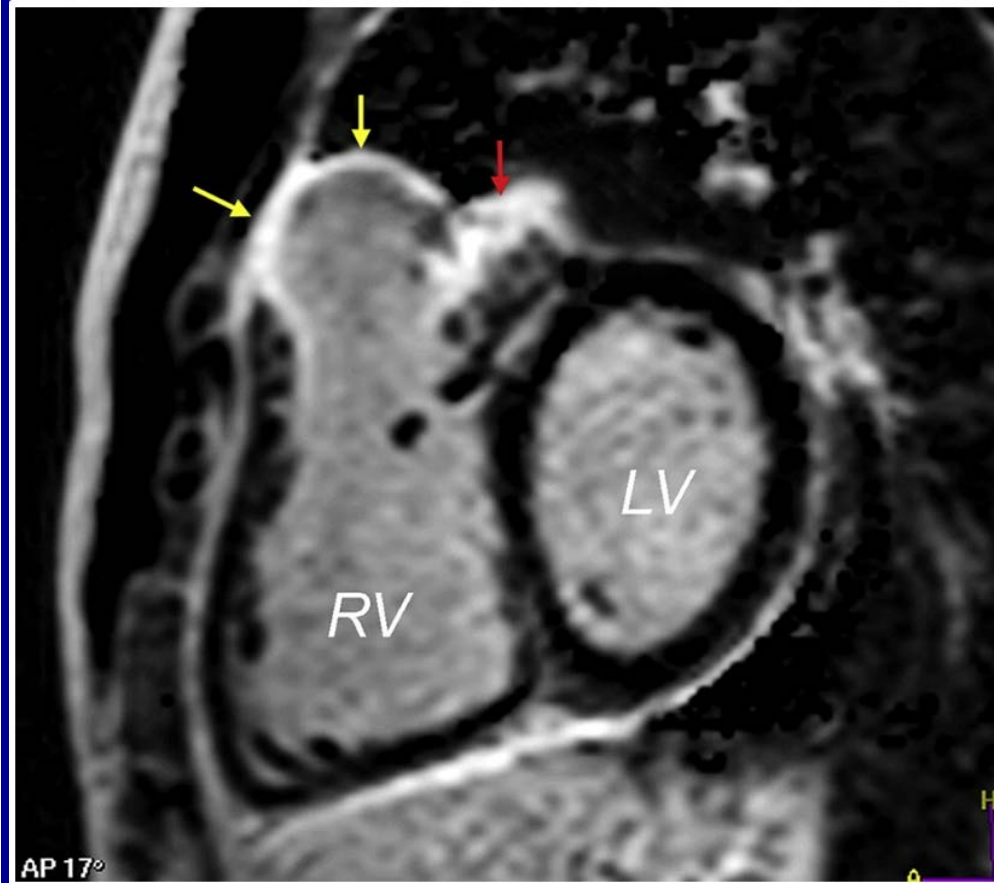
EKG in Repaired TOF



Issues in Adults with Repaired TOF

- Pulmonary Regurgitation
- Atrial Arrhythmias
- Ventricular Arrhythmias and Sudden Cardiac Death
- Residual VSD
- LV Dysfunction
- Right Heart Failure

Tetralogy Of Fallot



Adults with Repaired TOF

- EP Procedures
- Transcatheter Vs Surgical pulmonary valve replacement
- Lifelong follow up with ACHD Clinic

Ebstein Anomaly



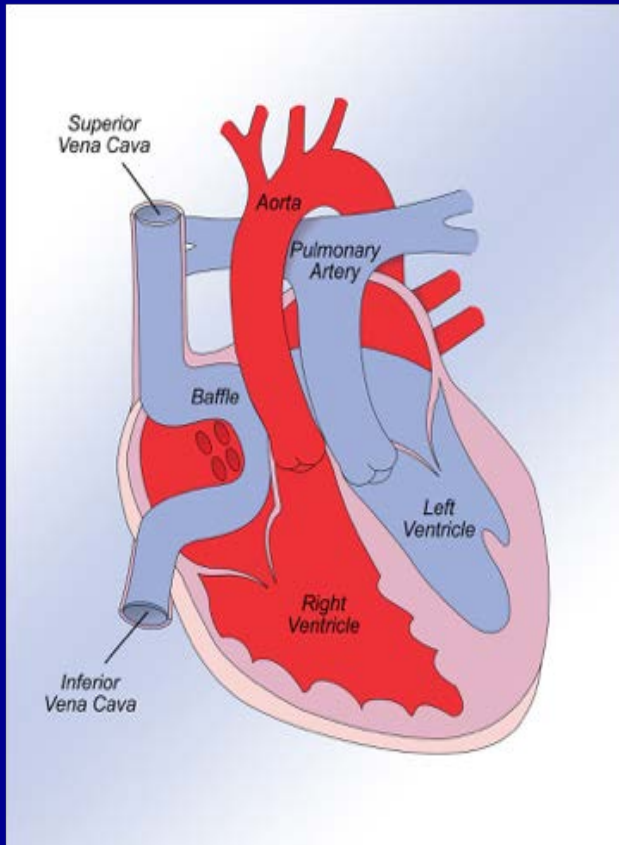
Ebstein Anomaly

- Tricuspid regurgitation
- RV Failure
- ASD - O2 desaturation
- Atrial Arrhythmias
- Surgery- TVR or Cone procedure

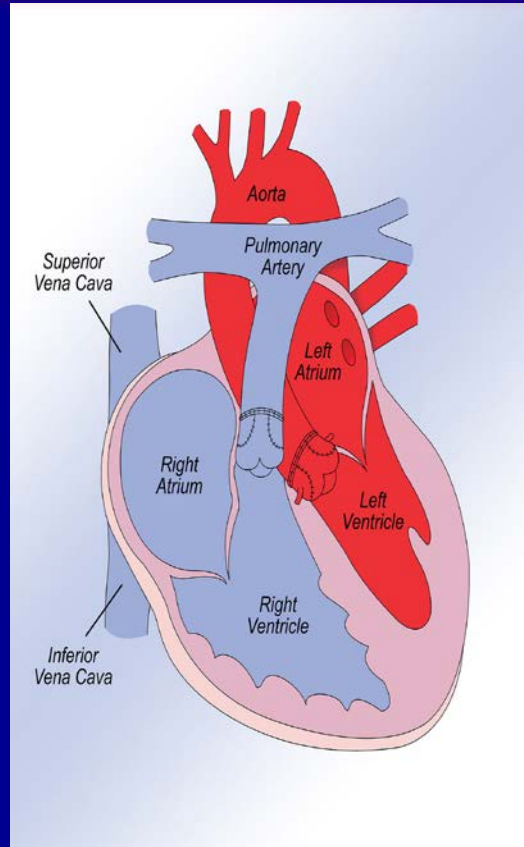
Transposition of Great Arteries



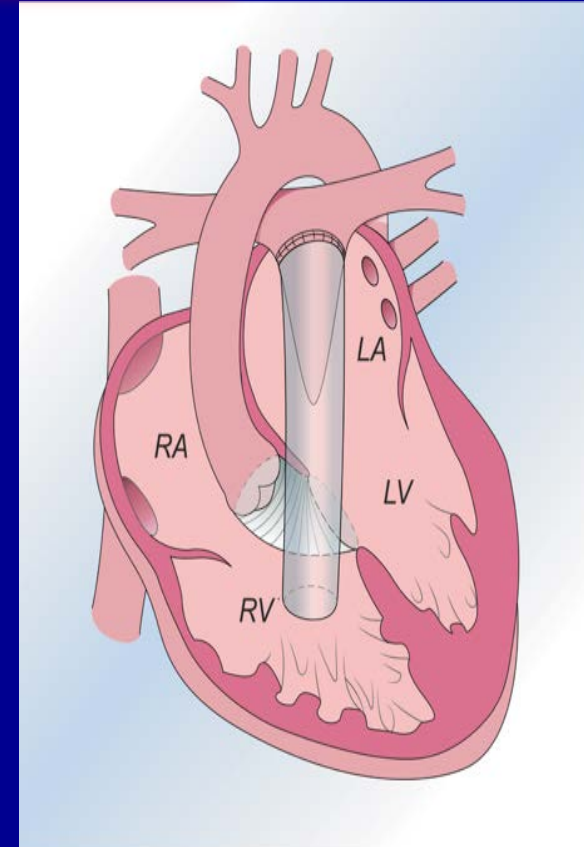
Surgical Repairs TGA



Atrial switch

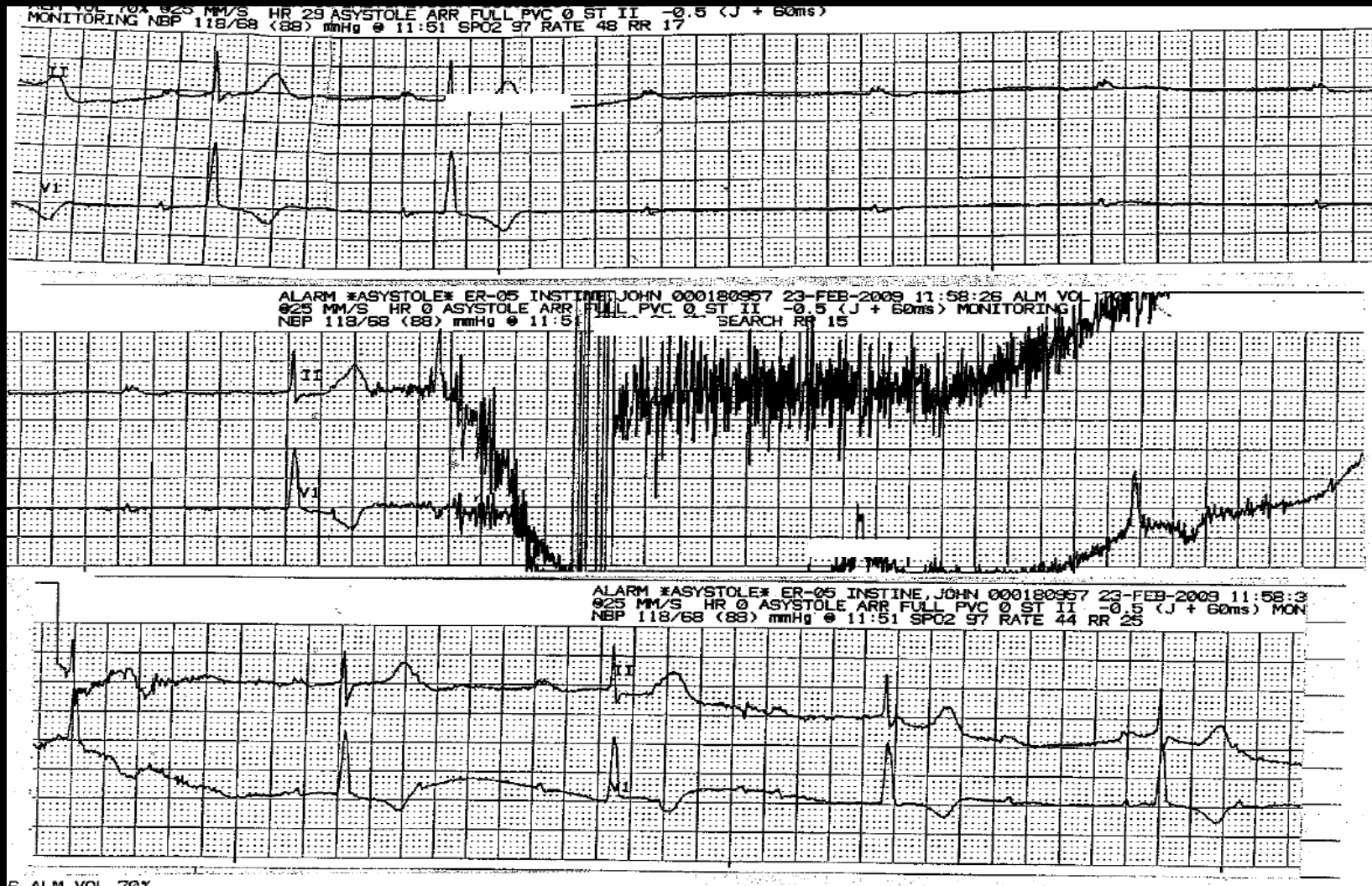


Arterial switch

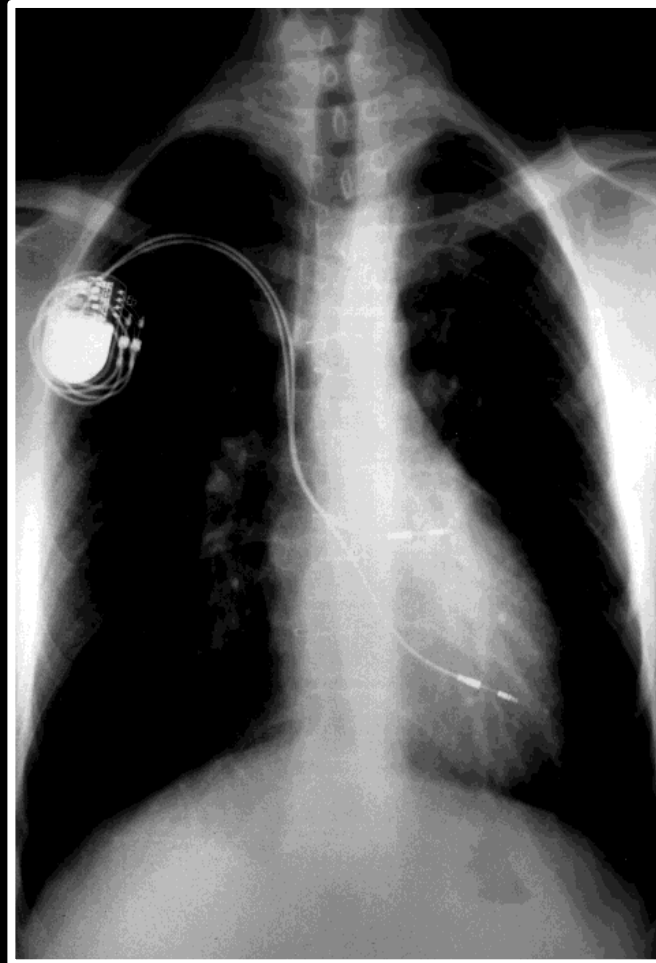
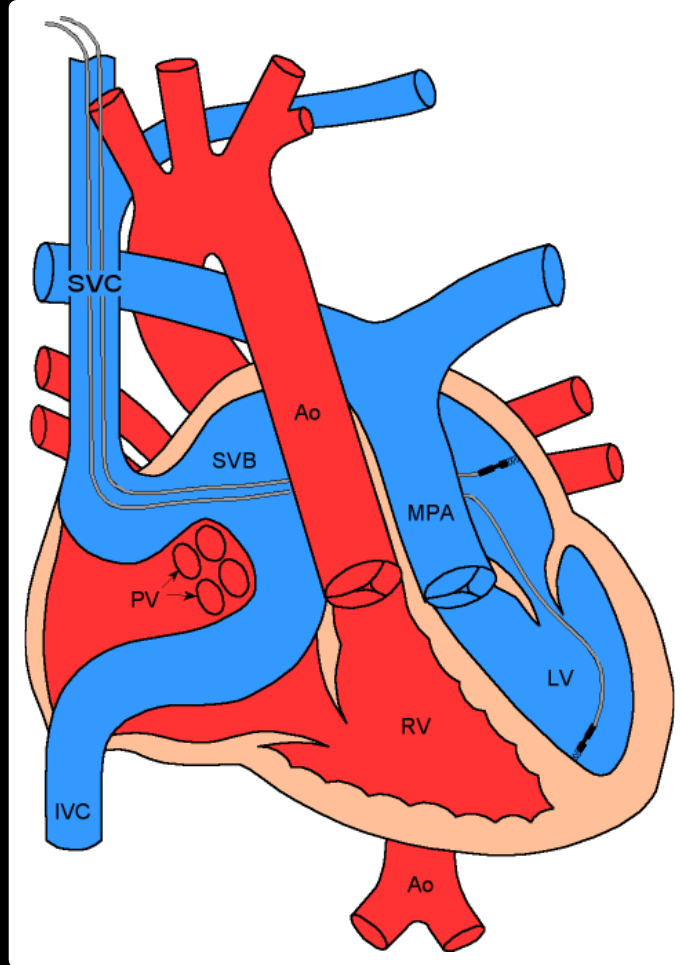


Rastelli procedure

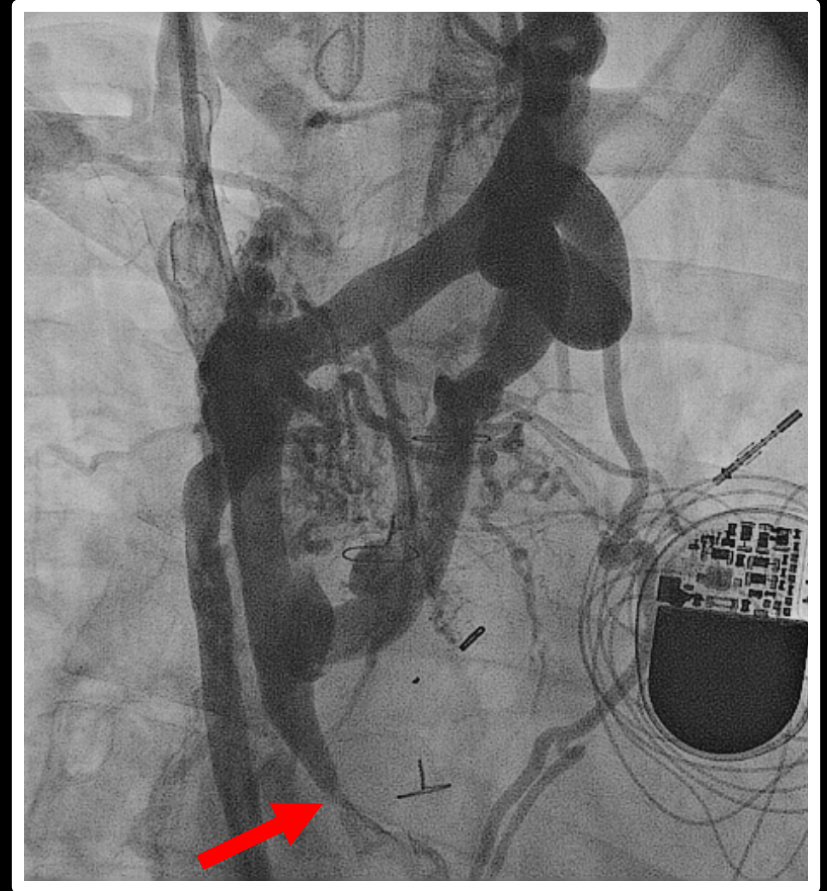
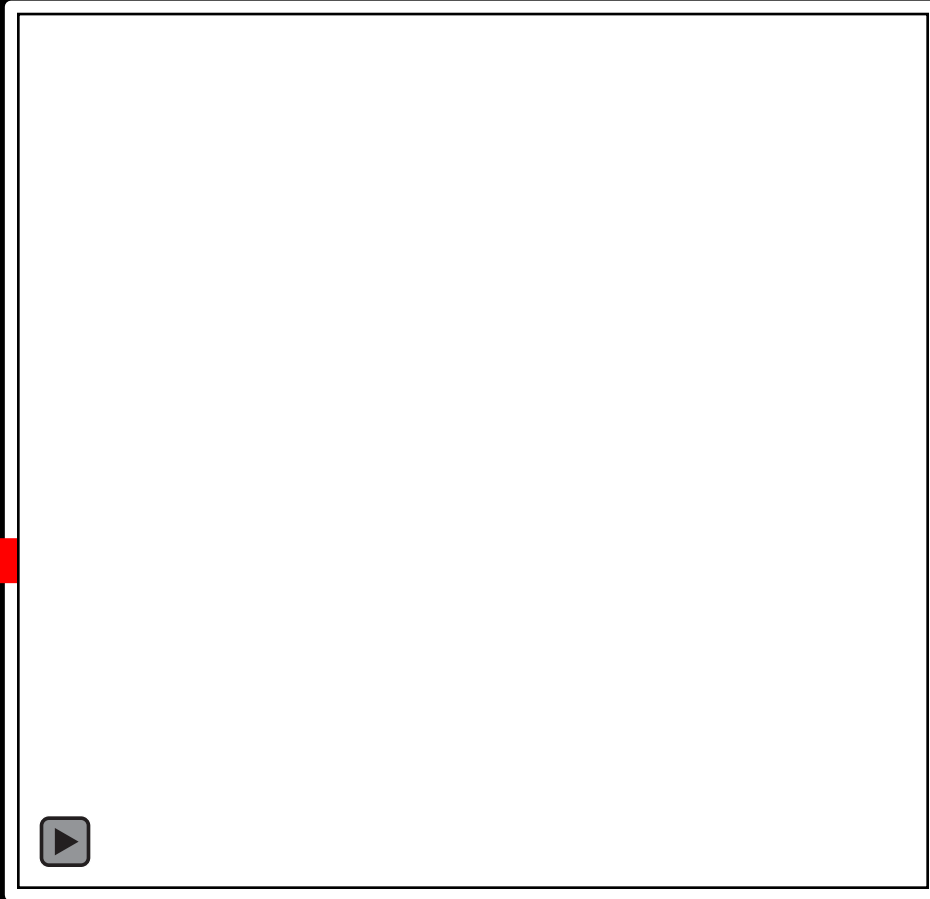
42 year old female with D-TGA s/p atrial switch w/syncope



D-TGA Atrial Switch

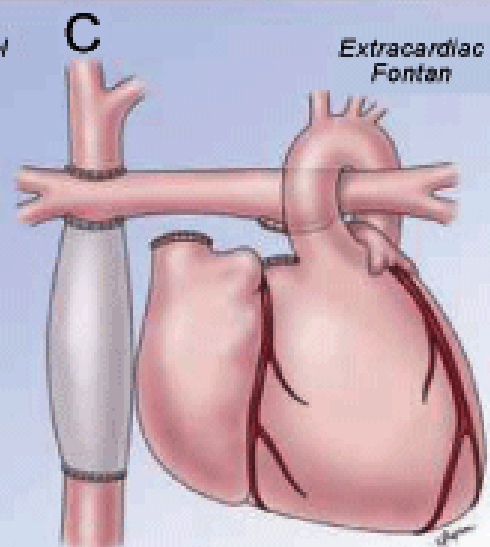
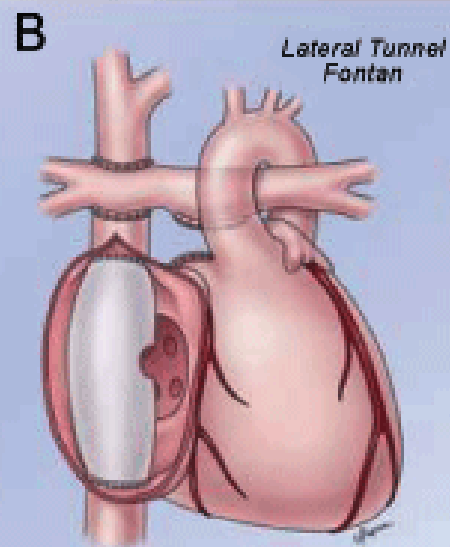
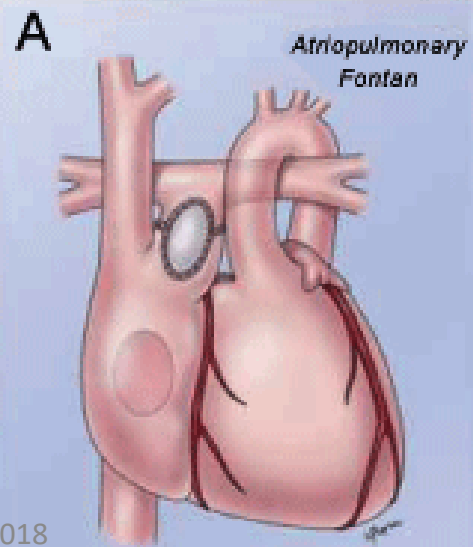
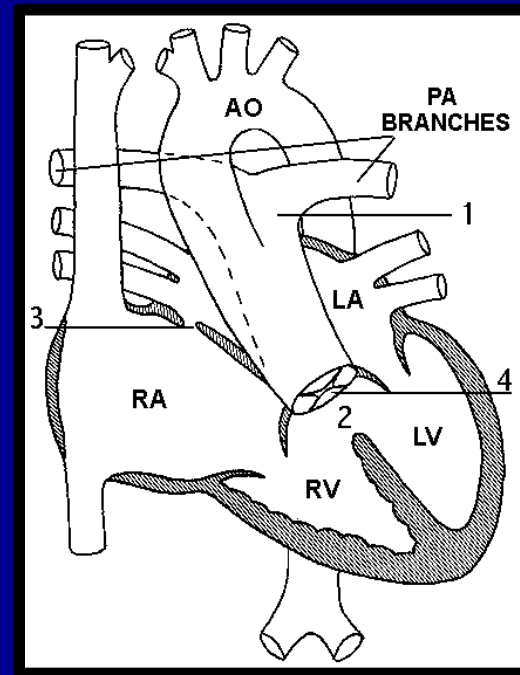
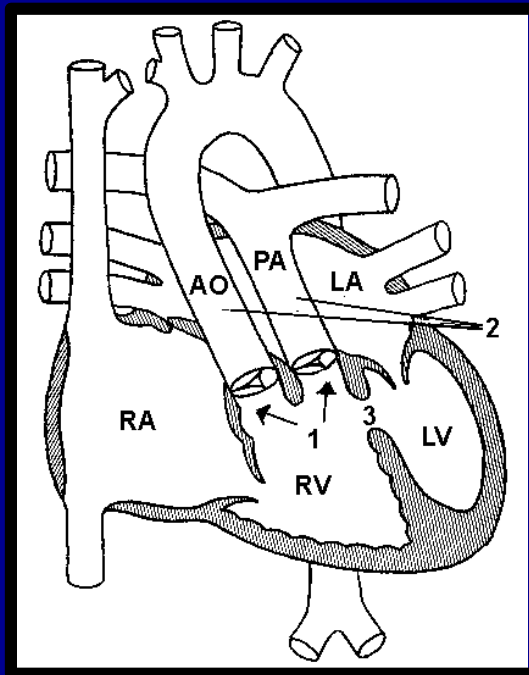


D-TGA Atrial Switch



Complex Congenital Heart Disease

- Eisenmenger Syndrome
- Unrepaired Cyanotic Congenital Heart Disease
- Fontan and Single Ventricle
- Patients Palliated with Systemic to Pulmonary Artery Shunts



Complex Congenital Heart Disease

- Must have 6 mthly to yearly ACHD follow-up
- Know and check O2 saturation regularly
- Check iron stores periodically
- Monitor renal and liver function
- Most have restrictive or other lung disease
- Must have dental follow up
- High risk of stroke and brain abscess in cyanotic patients

ACHD Patients- Common Issues

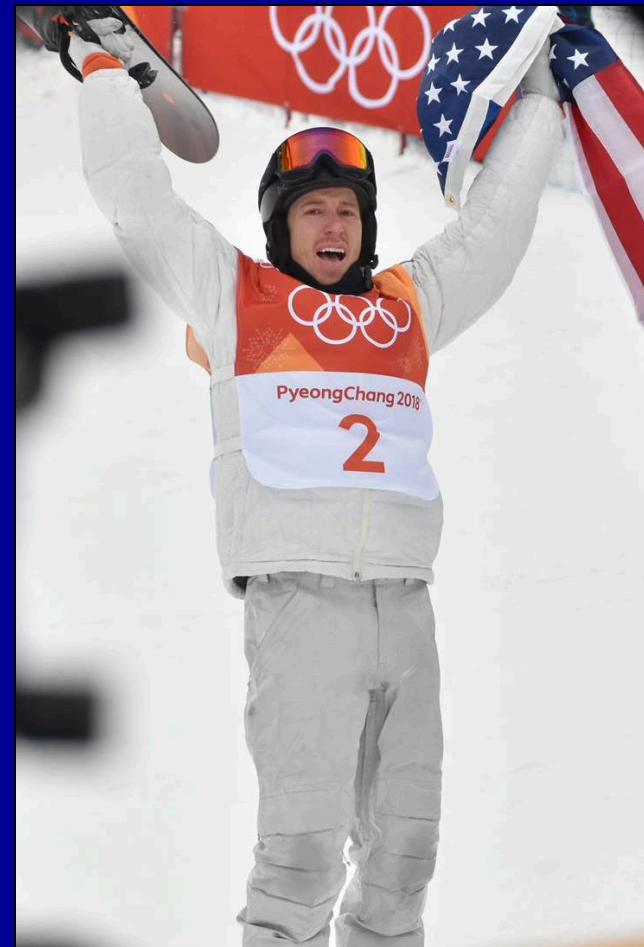
- Quality of life
- Transition
- Birth Control
- Pregnancy and CHD
- Dental Issues
- Exercise
- Hep C

- Pulmonary Hypertension
- Heart Failure
- Arrhythmias
- Neurocognitive issues
- Advance care planning and advanced directives

Quality of life

- Quality of a person's life is related to how satisfied they are with their life overall
- Functional status has to do with a person's ability to do normal daily activities and perform their roles in life
- Disability paradox
- Response shift
- Sense of coherence

Quality of life – Adults with Congenital Heart Disease



Pregnancy

- High Risk
 - Aortopathies including Marfan syndrome
 - Severe left sided obstructive lesions
 - Fontan
 - Eisenmenger
 - Pulmonary Hypertension
 - Severe LV Dysfunction

Birth Control

- Hypercoagulable states
- Low dose progestin pills
- IUDs
- Complex congenital heart disease patients should be evaluated in tertiary centers

Exercise



SBE Prophylaxis

- Prosthetic heart valves
- Prosthetic material used for cardiac valve repair
- Prior history of IE
- Unrepaired cyanotic congenital heart disease
- Repaired congenital heart disease with residual shunts or valvular regurgitation at the site or adjacent to the site of the prosthetic patch or prosthetic device
- Repaired congenital heart defects with catheter-based intervention involving an occlusion device or stent during the first six months after the procedure
- Valve regurgitation due to a structurally abnormal valve in a transplanted heart

Advance Care Planning and Advance Directives

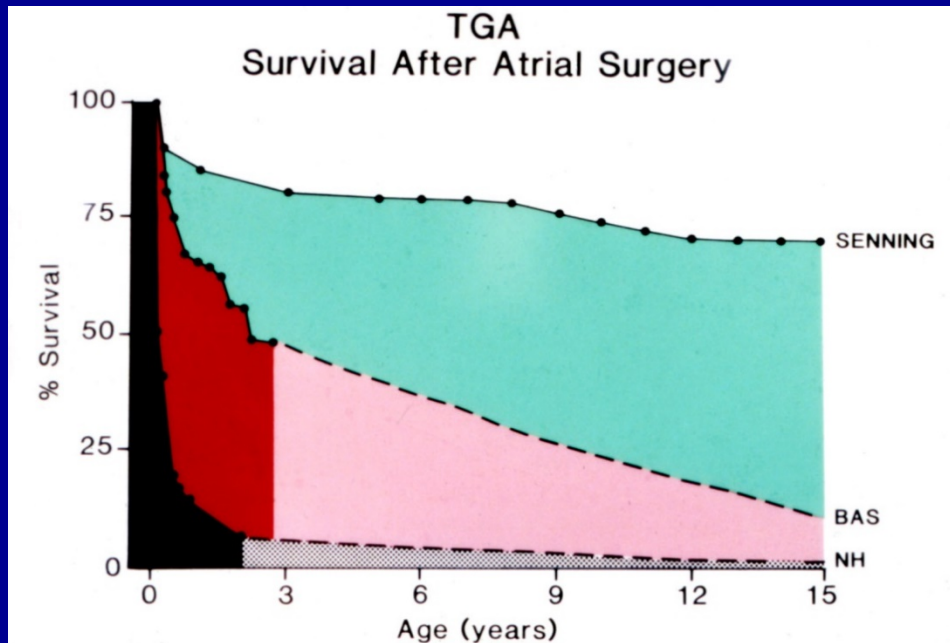
The place for these difficult conversations should **not** be in the Intensive Care Unit (ICU)

- 50% of ACHD patients die in the hospital
- Of these, two-thirds die in the intensive care setting and almost a half were on life support
- Only 10% of patients in ACHD care had an end-of-life discussion

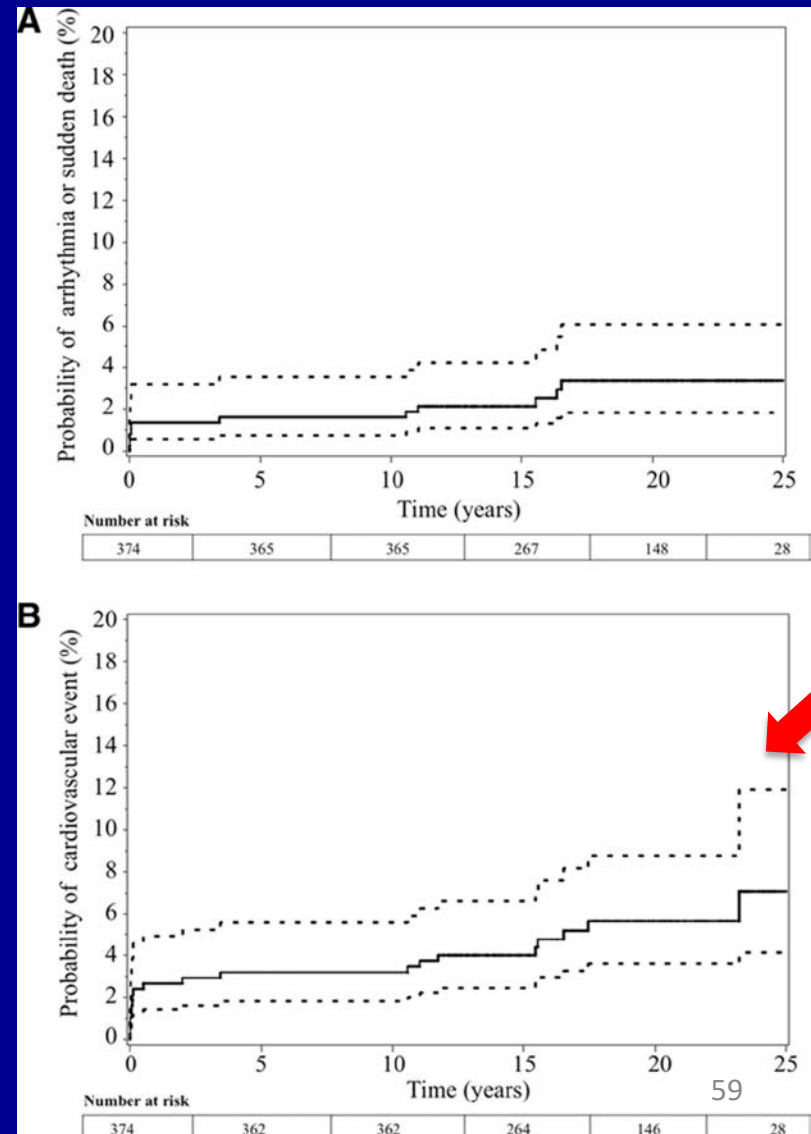
Current Concerns Neurodevelopmental Outcomes

- 60 young adults with **arterial switch operation** re-evaluated at a mean age of 16.9 ± 1.7 years
- **Neurologic impairment in 10%**
- Periventricular **leukomalacia** was detected in **>50%**; its severity correlated with the grade of neurologic impairment
- Magnetic resonance imaging demonstrated moderate or severe **structural brain abnormalities in 32%** of the patients

Congenital Heart Disease – The Journey



Courtesy: Peter Lang, MD



Adults with Congenital Heart Disease

Games

Hyper Light Drifter - how heart disease inspired one of 2016's great games

The central character in this brilliant new game is haunted by a deadly illness - something with which creator Alex Preston is all-too familiar

Chris Priestman

Thu 2 Jun 2016 06:14 EDT

The Guardian US edition ▼



▲ Hyper Light Drifter. Photograph: Heart Machine

COACH

Columbus Ohio Adult Congenital Heart Program



Thank you

- Thank you