

# Adult Congenital Heart Disease for the Internist

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## 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

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### EDITORIAL

## Congenital Heart Disease

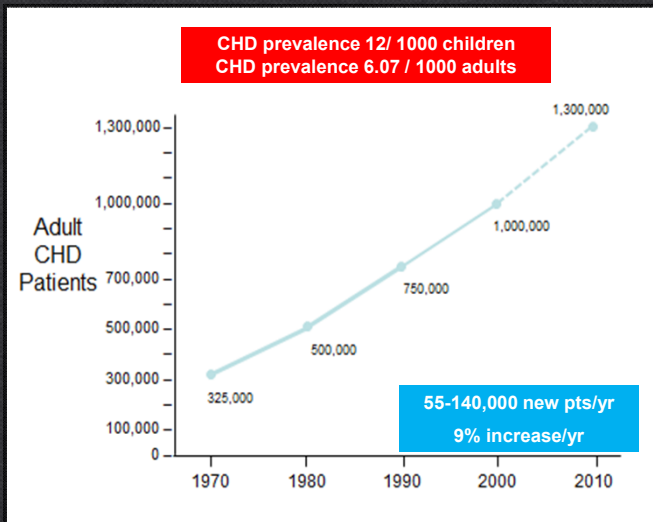
The Remarkable Journey From the "Post-Mortem Room" to Adult Clinics

Ali J. Marian

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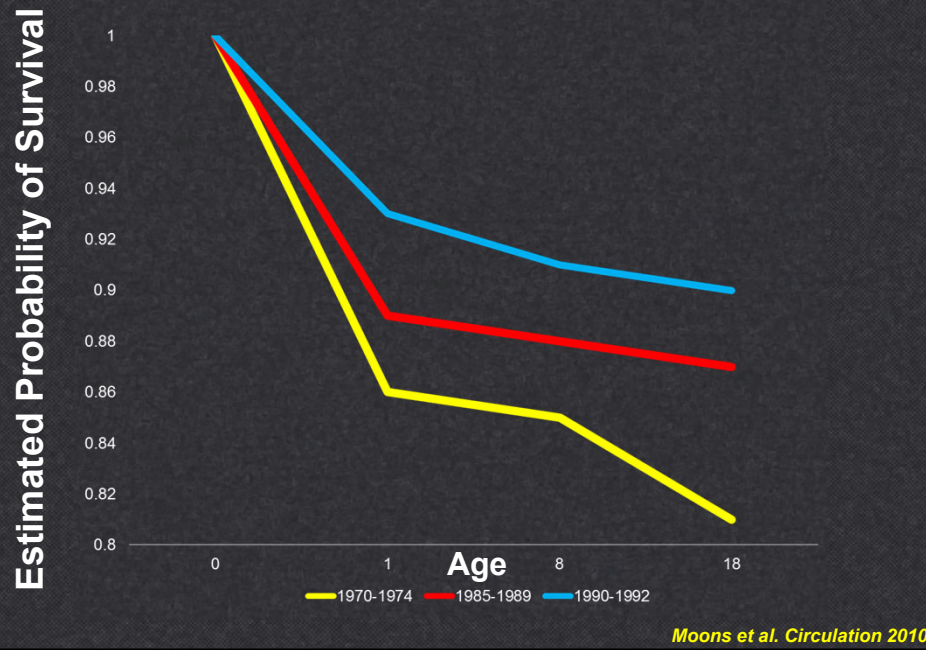
**DOI** <https://doi.org/10.1161/CIRCRESAHA.117.310830>  
Circulation Research. 2017;120:895-897  
Originally published March 16, 2017

# EXPLOSION of a "Life-Long" Disease Burden



Hoffman J et al. *Am J Cardiol*. 1979; 641-647; Fyler D. 1980; 375-464; Ferencz C et al. *Am J Epidemiol*. 1985;31-5. 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



## Trends in ACHD

	2005-2007	2008-2009	2014
<b>Specialized ACHD Centers</b>	68	94	126
<b>Patient Visits</b>	55777	5568	110740
<b>Publications</b>	4373	6281	6281

Richard A. Krasuski, and Thomas M. Bashore Circulation. 2016;134:110-113

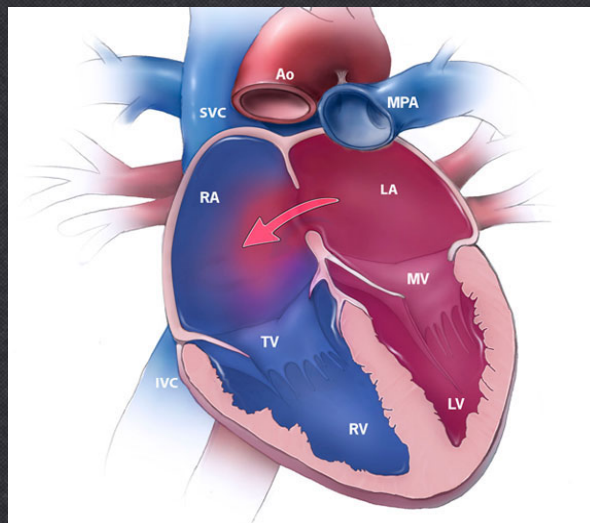


## ACHD Patients- Common Issues

- 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

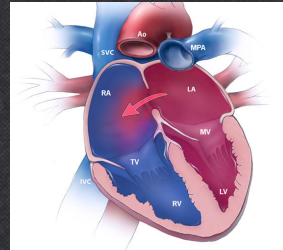
## Atrial Septal Defect



<https://www.cdc.gov/ncbddd/heartdefects/AtrialSeptalDefect-graphic2.html>

# Physical Exam – Atrial Septal Defect

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

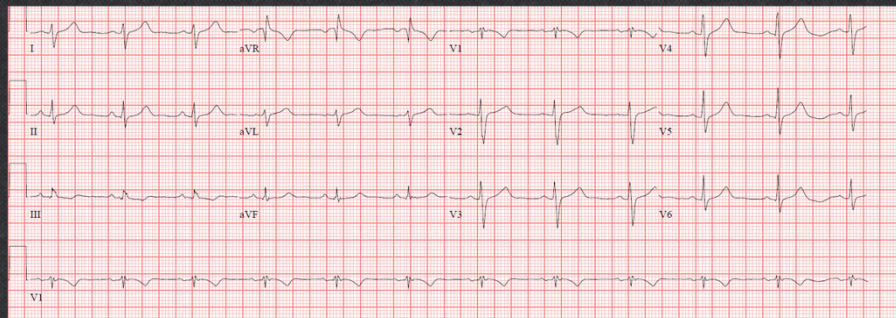


## Murmurs in ASD

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

<https://www.cdc.gov/ncbddd/heartdefects/AtrialSeptalDefect-graphic2.html>

## EKG



### Secundum ASD

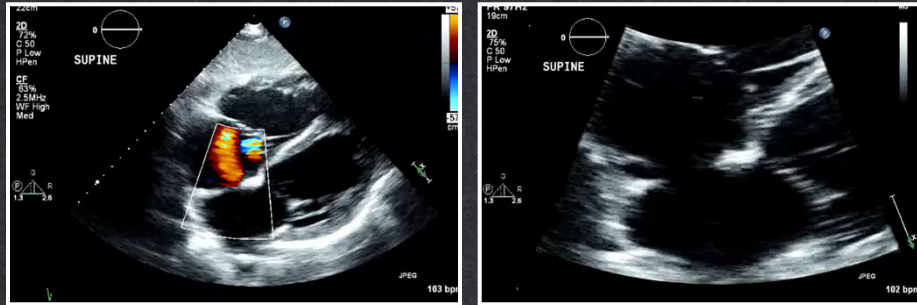
Incomplete RBBB  
Right Axis Deviation

### Primum ASD

Incomplete RBBB  
Left Axis deviation

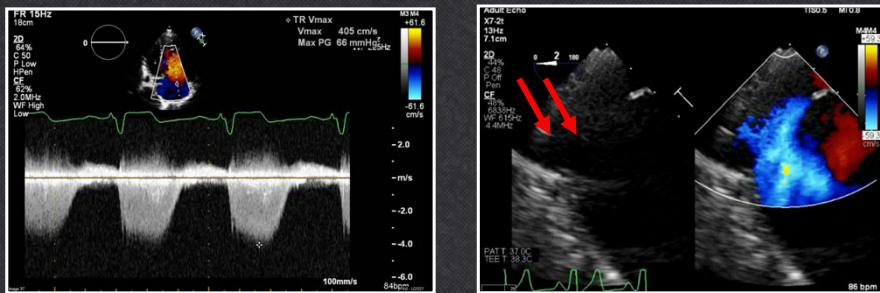


# ASD – Paradoxical Embolism



**Myocardial Infarction and PE after IVDU**

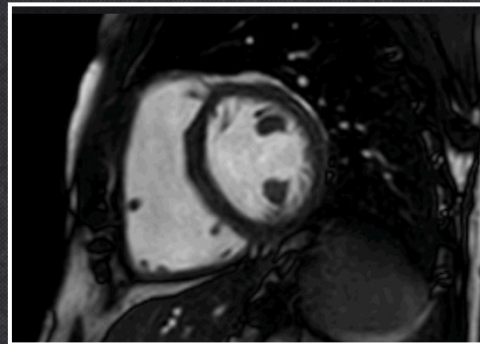
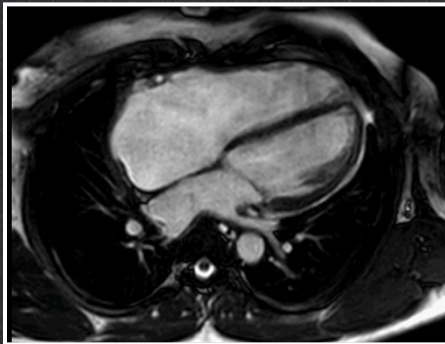
# ASD and Pulmonary Hypertension



## Associated Anomalies

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

## RV Dilatation



RVEDV<sub>i</sub>: 145 ml/m<sup>2</sup>  
RVEF: 61%

LVEDV<sub>i</sub>: 102 ml/m<sup>2</sup>  
LVEF: 56%

Qp/Qs : 1.1  
TR fraction: 49%



## Etiologies of RV Dilation

Tricuspid regurgitation

Pulmonary regurgitation

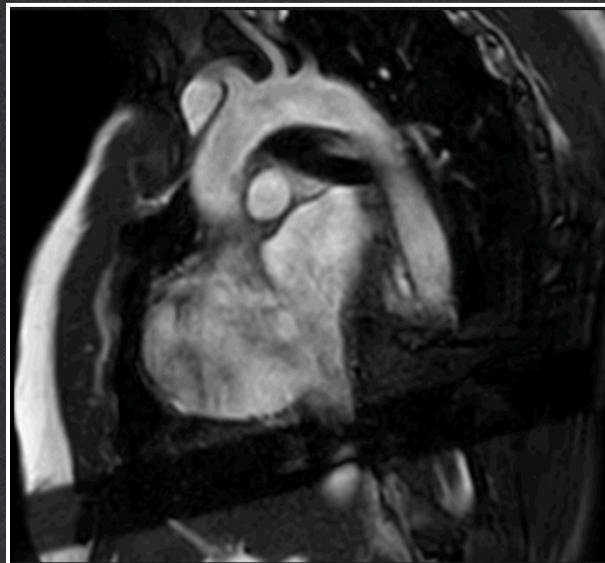
Pulmonary artery hypertension

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

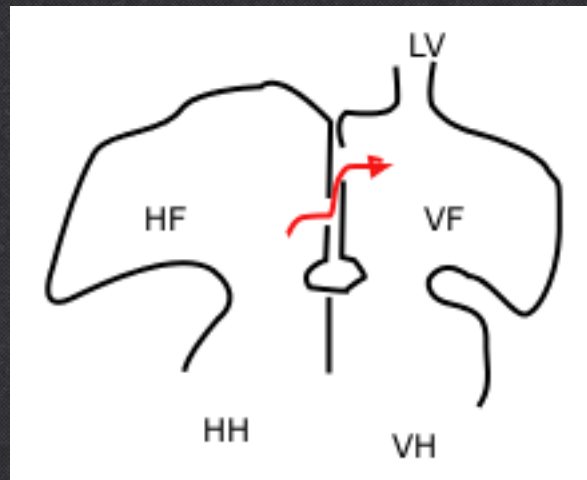
## ASD Closure

- Surgical
- Transcatheter

## Other Issues with ASDs

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

## Patent Foramen Ovale



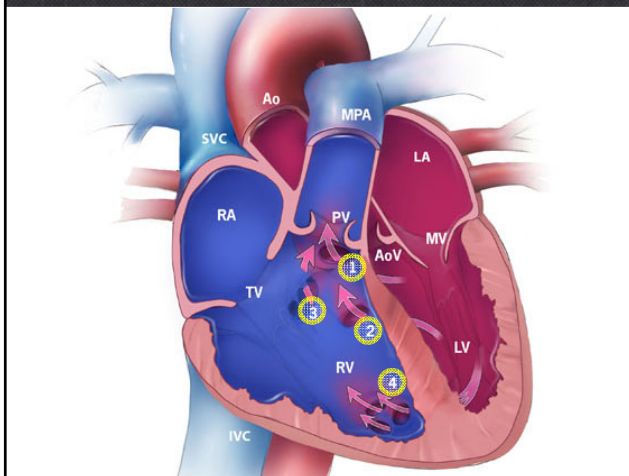
Drawn by: Kjetil Lenes



## 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



1. Conoventricular
2. Membranous
3. Inlet
4. Muscular

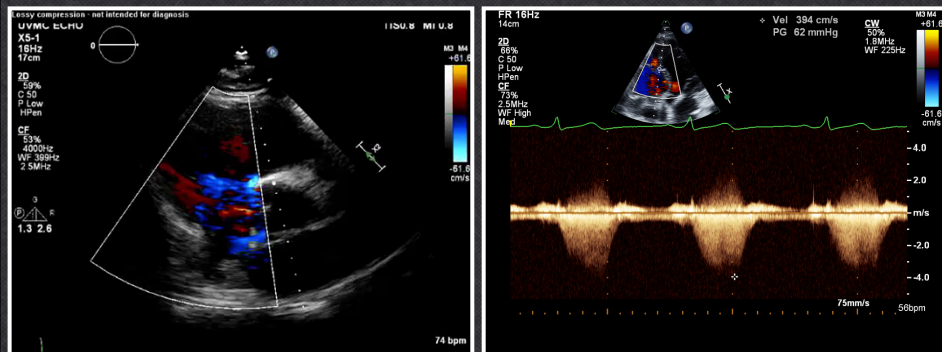
Source: Centers for Disease Control and Prevention



# Physical Exam and EKG

- Smaller the VSD, louder the murmur
- Holosystolic plateau-shaped murmur at LLSB
- Majority of patients with isolated ventricular septal defect have normal EKG
- 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

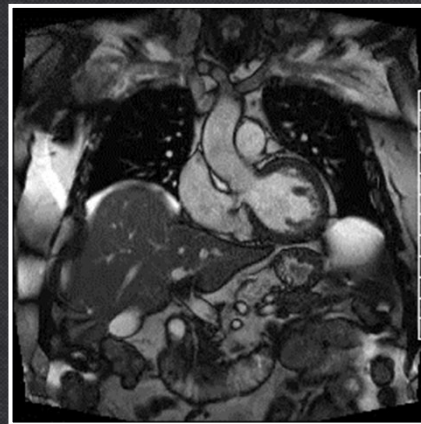
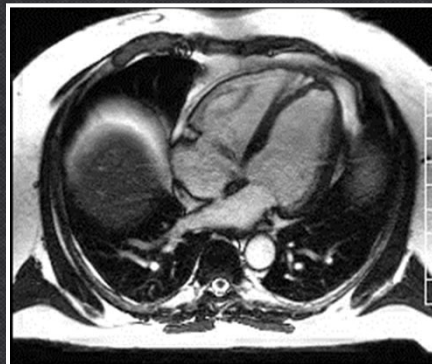
- Intervention is rarely required
- Small VSDs do not create a clinically important shunt
- Adult patients with large VSDs and irreversible pulmonary vascular disease
  - Need expert assessment



## Lifelong Follow Up -VSD

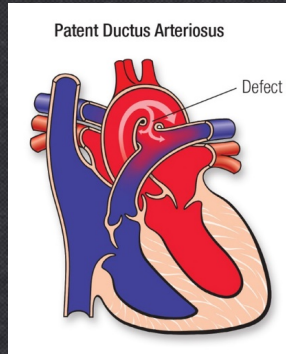
- More often repair of the VSD
  - Endocarditis
  - Aortic insufficiency
  - Pulmonary insufficiency
- Surgical repair remains the gold standard for treatment of VSD

## Gerbode Defect





## Patent Ductus Arteriosus



### Significant Left to Right Shunt

- Symptoms
- LA and LV enlargement

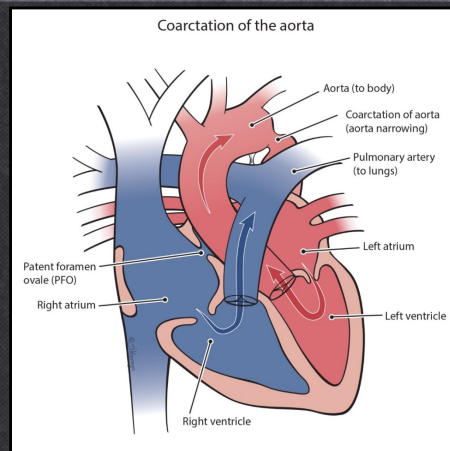
### Endarteritis

Source: Patent Ductus Arteriosus (PDA) - American Heart Association

<http://www.heart.org/en/health-topics/congenital-heart-defects/about-congenital-heart-defects/patent-ductus-arteriosus-pda>

## Coarctation of Aorta

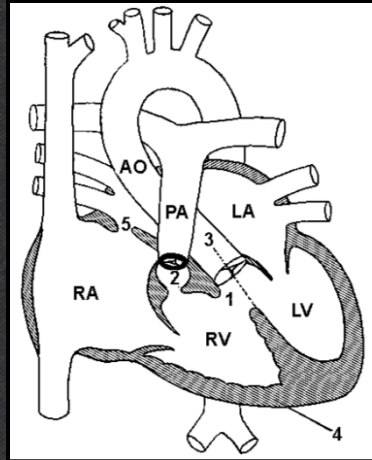
- Familial risk
- Turner syndrome
- Associated anomalies
  - ASD
  - VSD
  - Bicuspid aortic valve
- Hypertension
- Brachial-femoral delay
- Premature CAD, Stroke
- Intracranial aneurysms
- Surgery, Balloon Angioplasty, Stent



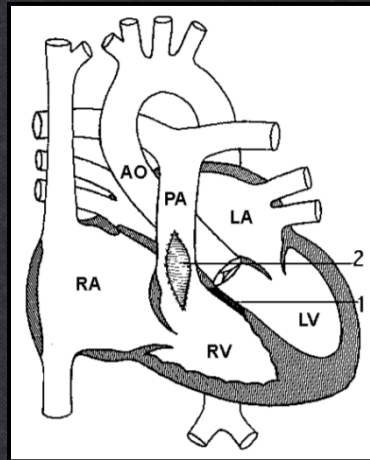
Courtesy: Ohio Fetal Medicine Collaborative

# Tetralogy Of Fallot (TOF)

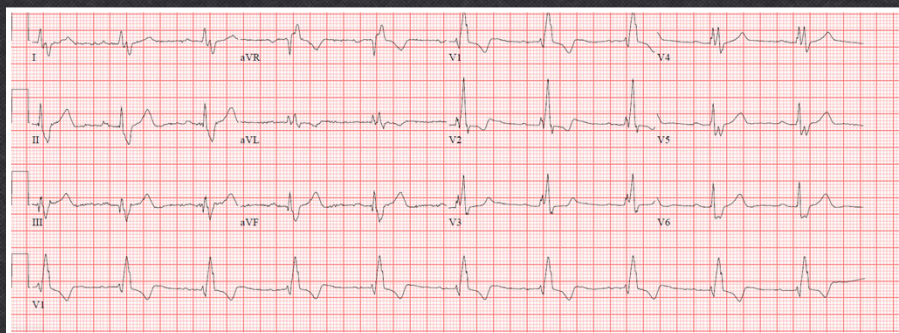
## Unrepaired



## Repaired



# EKG in Repaired TOF

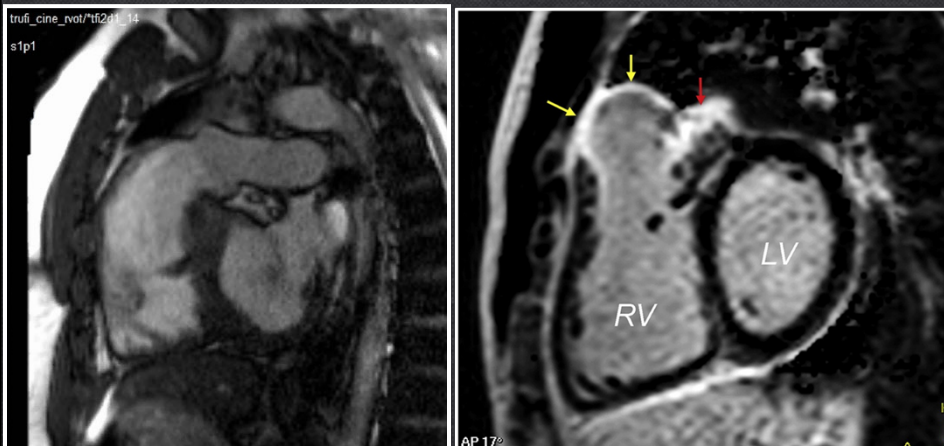




## Issues in Adults with Repaired TOF

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

## Tetralogy Of Fallot

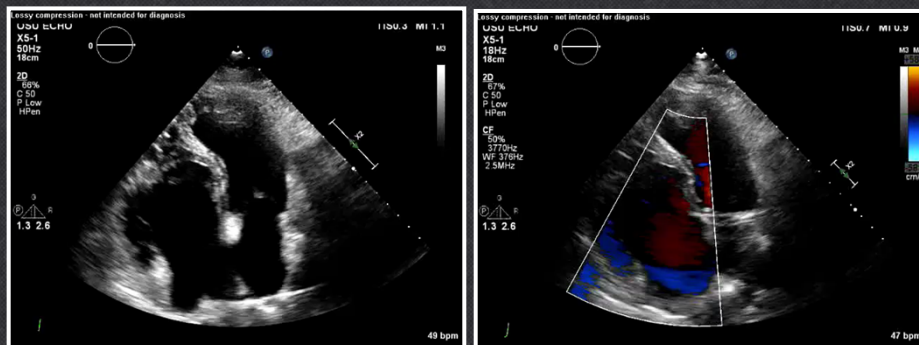




# Adults with Repaired TOF

- EP Procedures
- Pulmonary Valve Replacement
  - Transcatheter
  - Surgical
- Lifelong follow up with ACHD

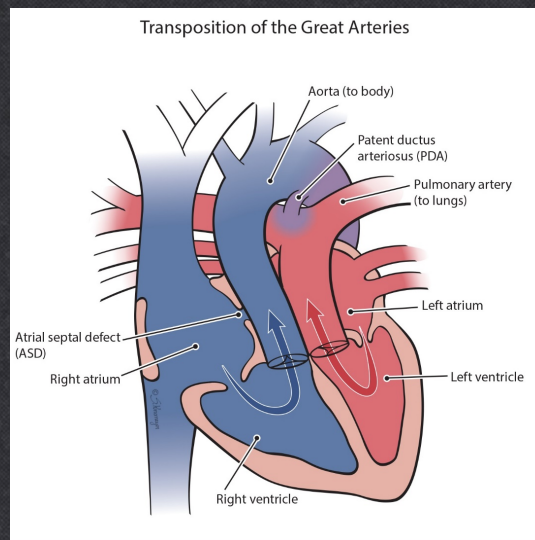
# Ebstein Anomaly



# Ebstein Anomaly

- Tricuspid regurgitation
- Right heart failure
- ASD - O2 desaturation
- Surgery-
  - Tricuspid valve replacement
  - Cone procedure
- Atrial arrhythmias
  - Atrial fibrillation
  - Atrial flutter
  - WPW
- Sinus node dysfunction
- Sudden cardiac death

# Transposition of Great Arteries



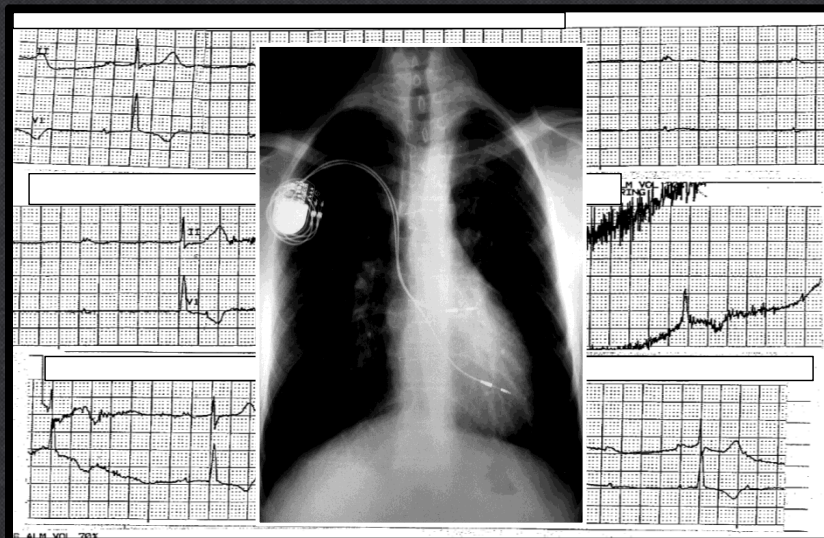
Courtesy: Ohio Fetal Medicine Collaborative



# Transposition of Great Arteries

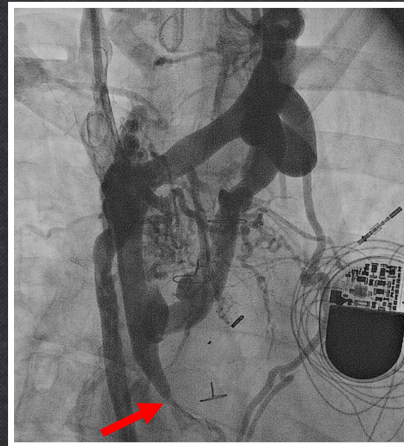
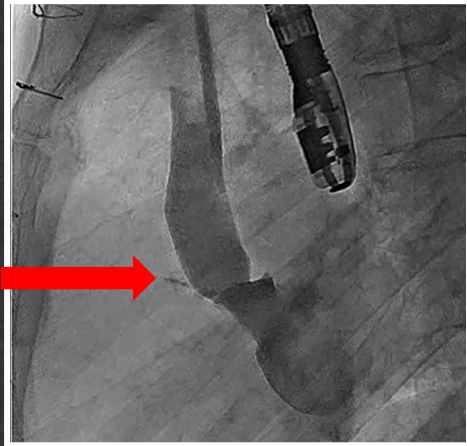


42 year old female with D-TGA s/p atrial switch w/syncope





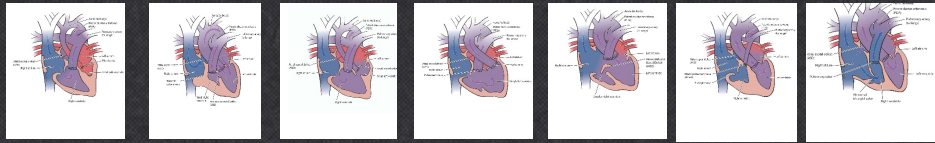
## D-TGA Atrial Switch



## Complex Congenital Heart Disease

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

# Single Ventricle Anatomy



HLHS

TA

DORV

DILV

Unbalanced  
AVC

PA

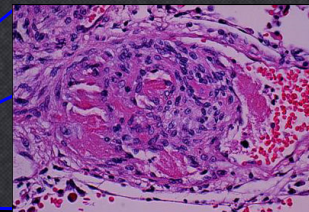
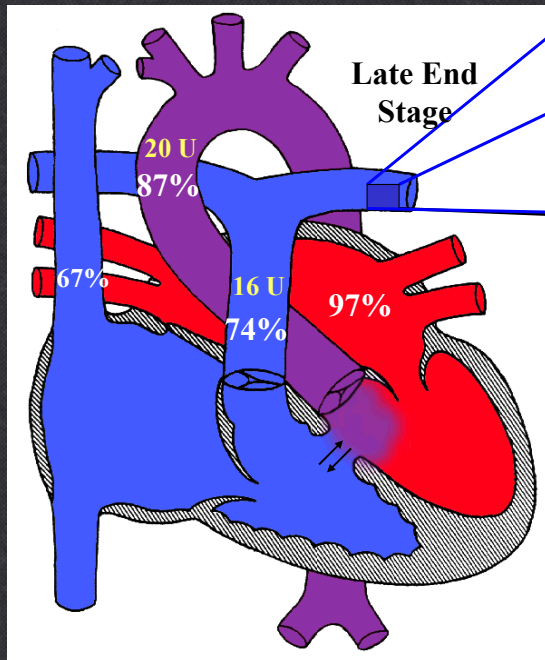
Ebstein

Surgical  
Shunts



- BT
- Central
- Waterston
- Potts

Courtesy: Ohio Fetal Medicine Collaborative



$PVR > SVR$

$Q_p:Q_s = 0.9:1$

**Eisenmenger  
Syndrome**

Courtesy : Curt Daniels, MD



## Complex Congenital Heart Disease

- 6 monthly 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
- Dental follow up
- High risk of stroke and brain abscess in cyanotic patients

## ACHD Patients- Common Issues

- |   |  |
|---|--|
| <ul style="list-style-type: none"><li>• Quality of life</li><li>• Transition</li><li>• Birth Control</li><li>• Pregnancy and CHD</li><li>• Dental Issues</li><li>• Exercise</li><li>• Hep C</li></ul> | <ul style="list-style-type: none"><li>• Pulmonary Hypertension</li><li>• Heart Failure</li><li>• Arrhythmias</li><li>• Neurocognitive issues</li><li>• Advance care planning and advanced directives</li></ul> |
|---|--|

## 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

## 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



Billy Rose Theatre Division, The New York Public Library. "Unidentified members exercising" *The New York Public Library Digital Collections*. 1930 - 1940. <http://digitalcollections.nypl.org/items/025b8e10-63aa-0132-0f5b-58d385a7bbd0>

## 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

Tobler et al Tobler Am J Cardiol 2012  
Tobler et al Palliative Medicine 2012



## Current Concerns Neurodevelopmental Outcomes

- 60 young adults with arterial switch operation re-evaluated at a mean age of  $16.9 \pm 1.7$  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

Heinrichs et al. *J Thorac Cardiovasc Surg* 2014

## New Guidelines

### 2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease

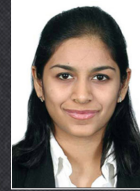
A Report of the American College of Cardiology/American Heart Association Task Force on  
Clinical Practice Guidelines

*Developed in Collaboration With the American Association for Thoracic Surgery, American Society of  
Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease,  
Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons*

JACC 2018

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