

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

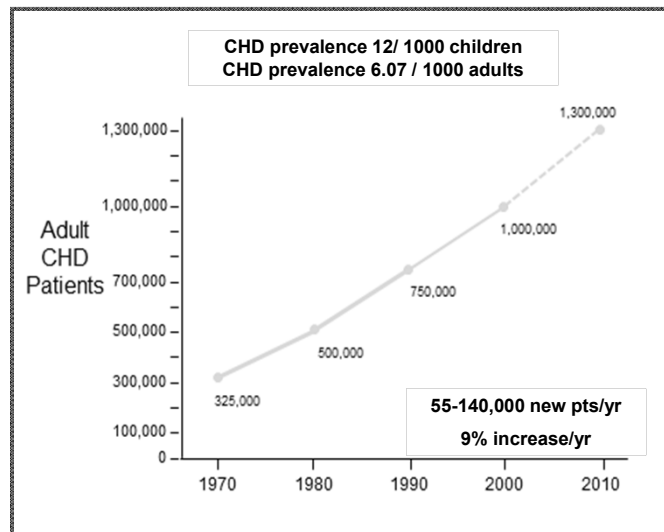
The Remarkable Journey From the “Post-Mortem Room” to Adult Clinics

Ali J. Marian



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



Trends in ACHD

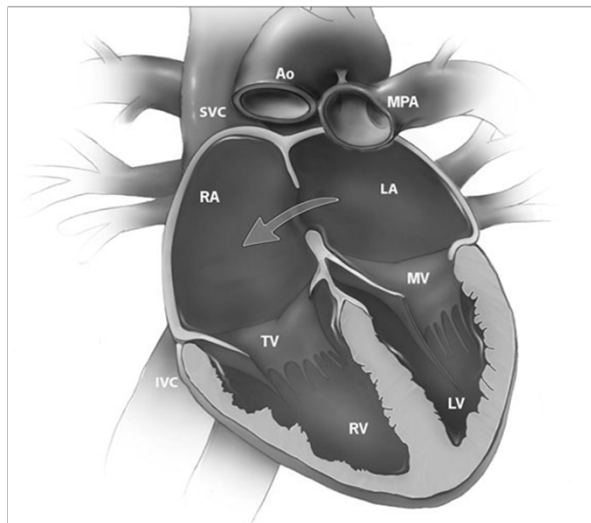
	2005-2007	2008-2009	2014
Specialized ACHD Centers	68	94	126
Patient Visits	55777	5568	110740
Publications	4373	6281	6281

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

ACHD Patients- Common Issues

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

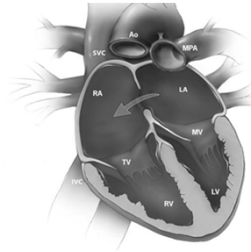
Atrial Septal Defect



<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

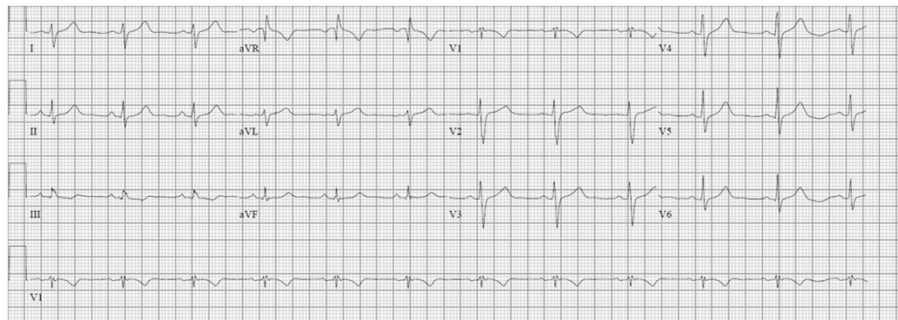


Murmurs in ASD

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

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

EKG



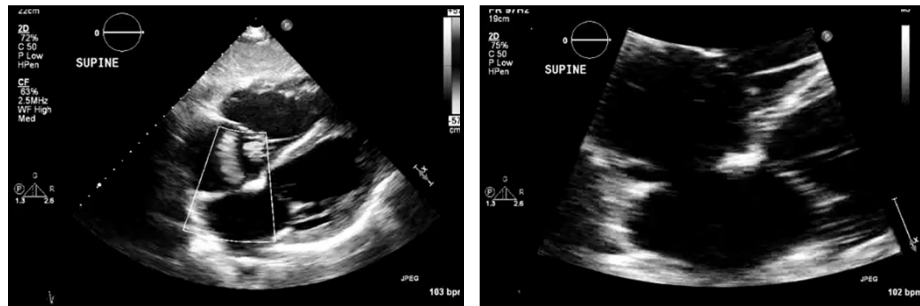
Secundum ASD

**Incomplete RBBB
Right Axis Deviation**

Primum ASD

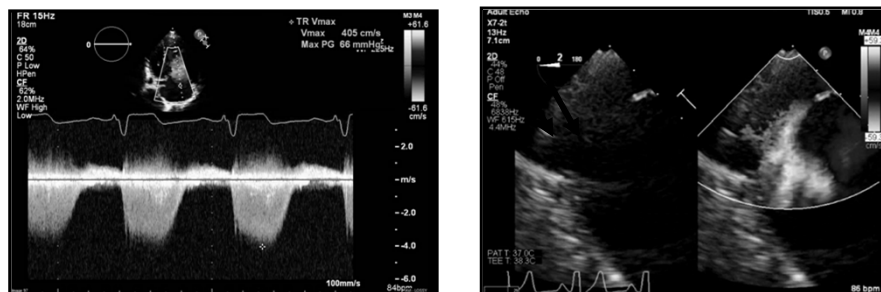
**Incomplete RBBB
Left Axis deviation**

ASD – Paradoxical Embolism



Myocardial Infarction and PE after IVDU

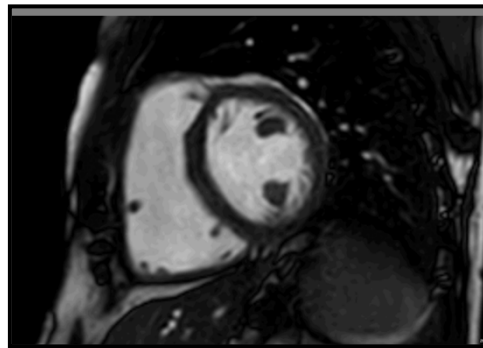
ASD and Pulmonary Hypertension



Associated Anomalies

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

RV 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

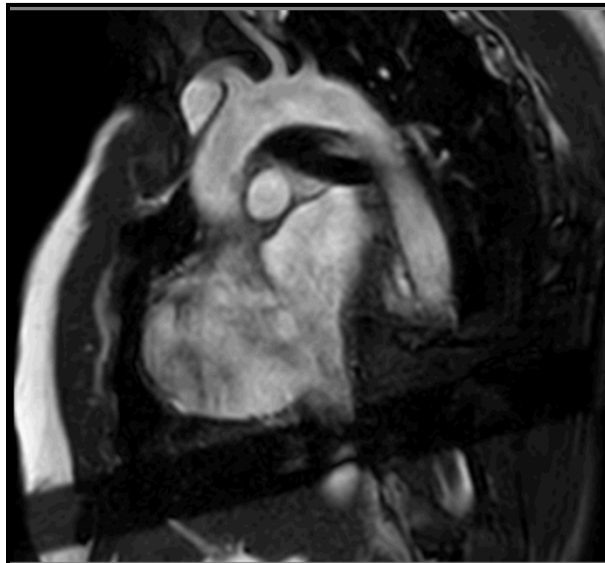
Pulmonary artery hypertension

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**

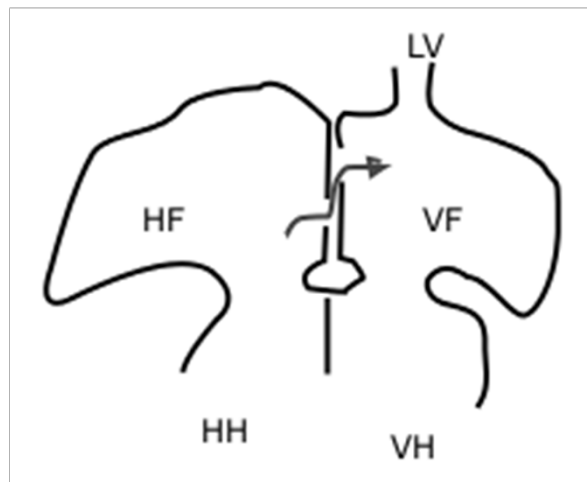
ASD Closure

- **Surgical**
- **Transcatheter**

Other Issues with ASDs

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

Patent Foramen Ovale

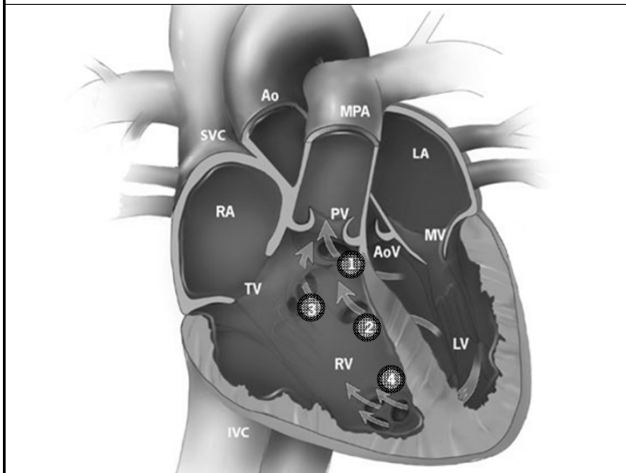


Drawn by: Kjetil Lenes

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



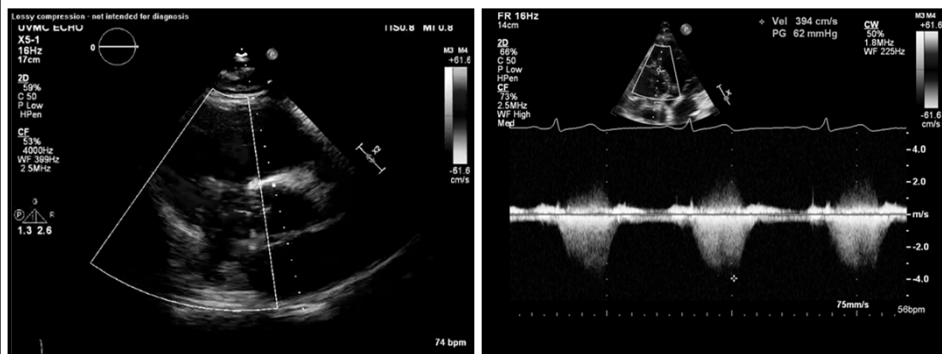
1. Conoventricular
2. Membranous
3. Inlet
4. Muscular

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**

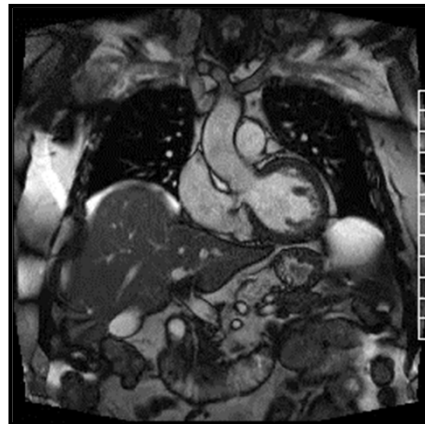
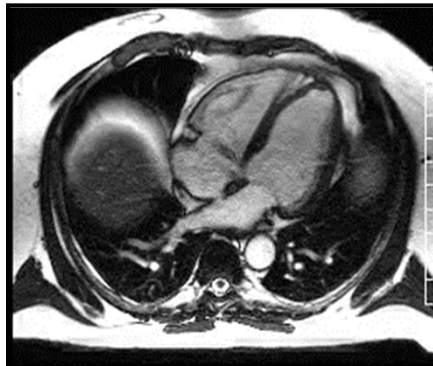
VSD Closure in Adults

- **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**

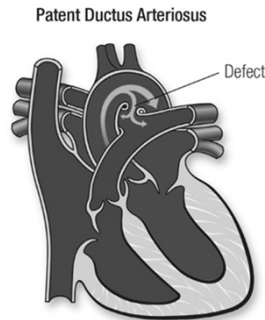
Lifelong Follow Up -VSD

- **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

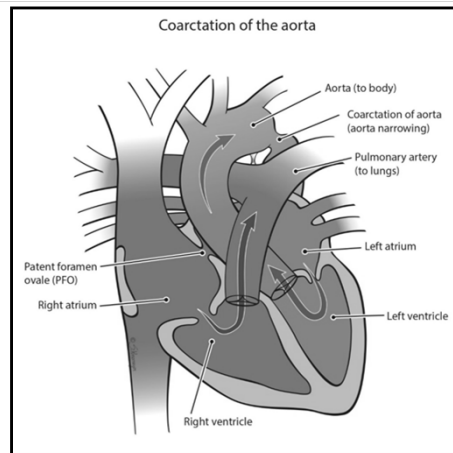
- Symptoms
- LA and LV enlargement

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

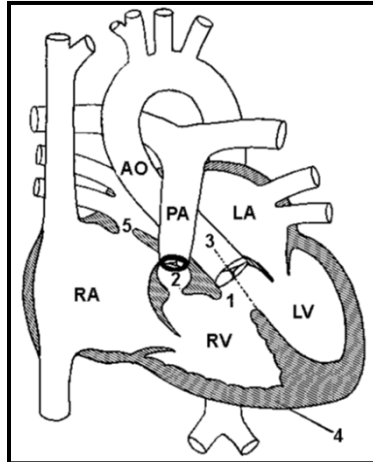
- **Familial risk**
- **Turner syndrome**
- **Associated anomalies**
 - ASD
 - VSD
 - Bicuspid aortic valve
- **Hypertension**
- **Brachial-femoral delay**
- **Premature CAD, Stroke**
- **Intracranial aneurysms**
- **Surgery, Balloon Angioplasty, Stent**



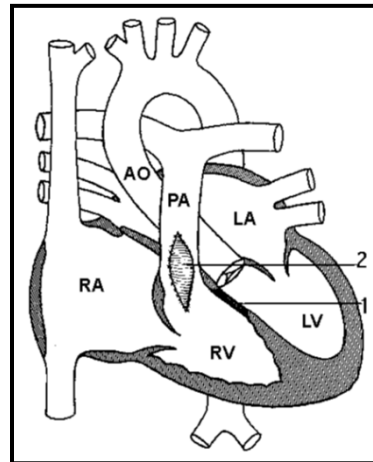
Courtesy: Ohio Fetal Medicine Collaborative

Tetralogy Of Fallot (TOF)

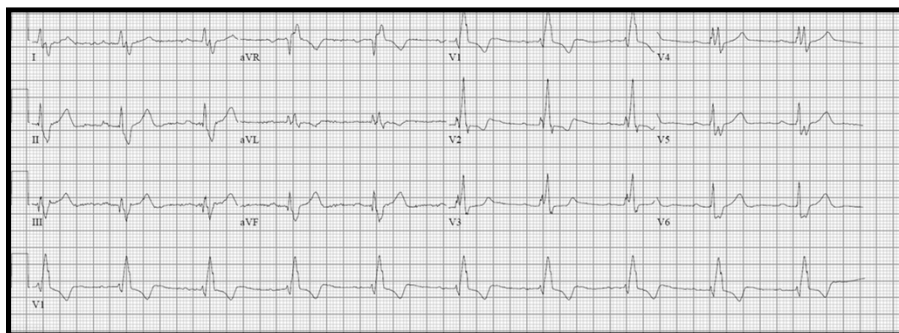
Unrepaired



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

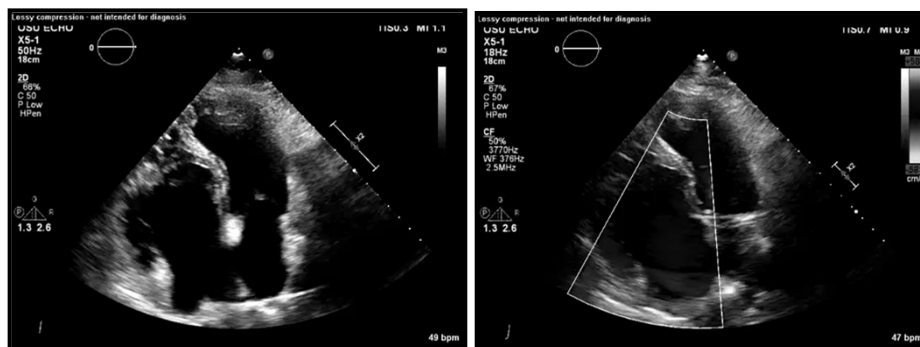
Tetralogy Of Fallot



Adults with Repaired TOF

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

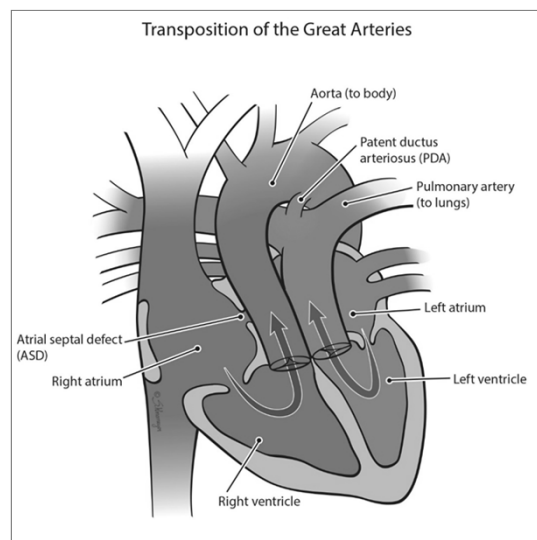
Ebstein Anomaly



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

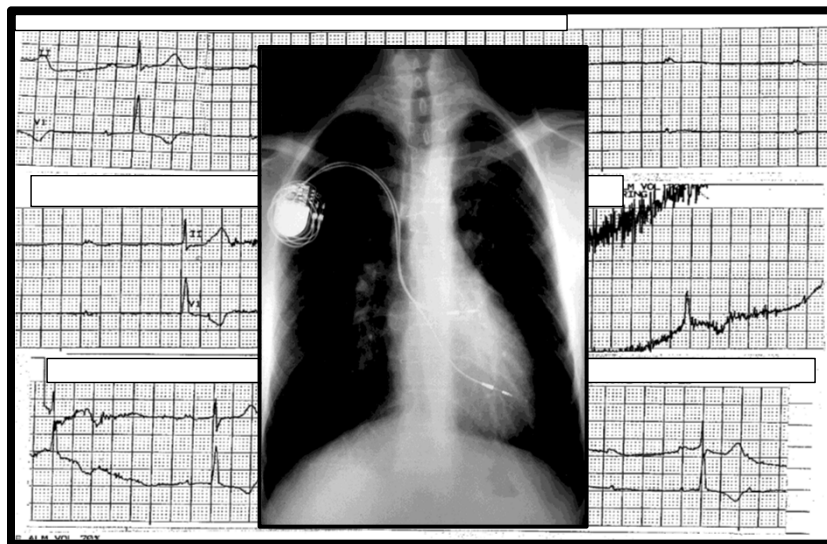


Courtesy: Ohio Fetal Medicine Collaborative

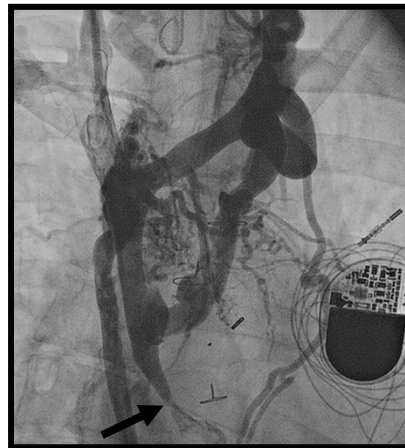
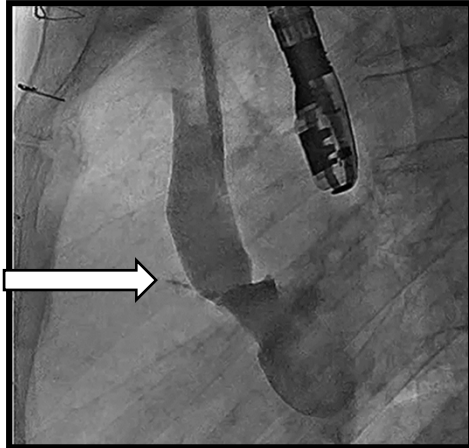
Transposition of Great Arteries



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



D-TGA Atrial Switch

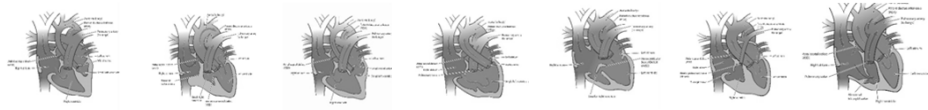


41.1

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

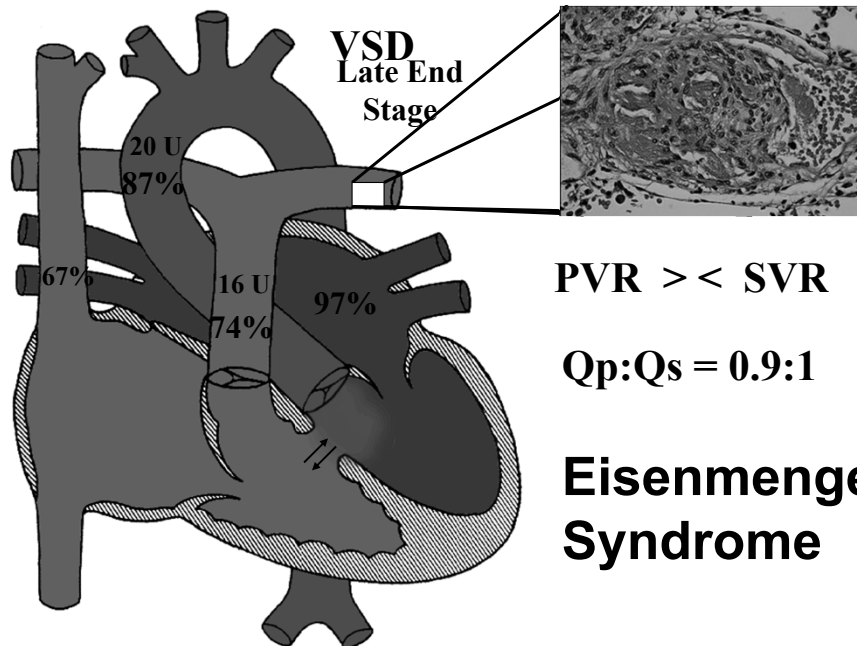
Ebstein

Surgical
Shunts

- BT
- Central
- Waterston
- Potts



Courtesy: Ohio Fetal Medicine Collaborative



$PVR > SVR$

$Qp:Qs = 0.9:1$

**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

- 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**

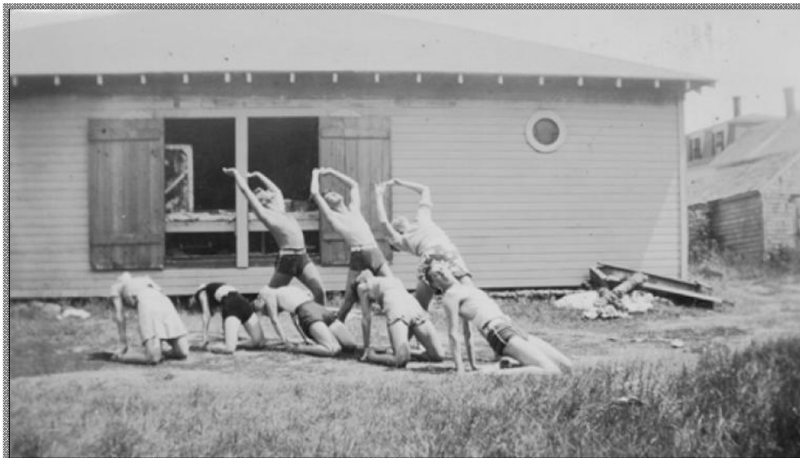
Pregnancy

- **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 ± 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

COACH

Columbus Ohio Adult Congenital Heart Program



COACH

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