Bridging the Gap: Challenges in Adult Congenital Cardiology

Nebraska Medicine Heart & Vascular Conference 2024
Friday, October 11th

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Objectives

- **Describe the current *population and future growth* of patients with congenital heart disease**
- List current strategies for addressing the ACHD issue
- Explain a few of the *most common congenital heart lesions* and associated long term risks of each
- Be aware of *"high-risk" lesions and situations* for patients with congenital heart disease
- Identify *ACHD resources* within your community

Congenital Heart Disease

CHD Prevalence

- *The* most common birth defect
  - 0.5-2% of all births
  - 40,000 babies/year
- ~60x more common than childhood cancer
- Wide range of severity
  - Simple valve lesions
  - Simple shunts
  - Complex heart disease

Early Experience

"Primum, non nocere"

- CHD known for centuries
- "Don't Touch the Heart"
- Leading cause of congenital associated infant death
- Up to 60% early mortality
- Adulthood survival was rare

Pioneers

- Pediatric Cardiology
  - Helen Taussig
- "Blue Baby" Operation (1944)
  - Alfred Blalock & Vivian Thomas
- CP Bypass (1950s)
- Introduction of CT surgery

Modern Experience

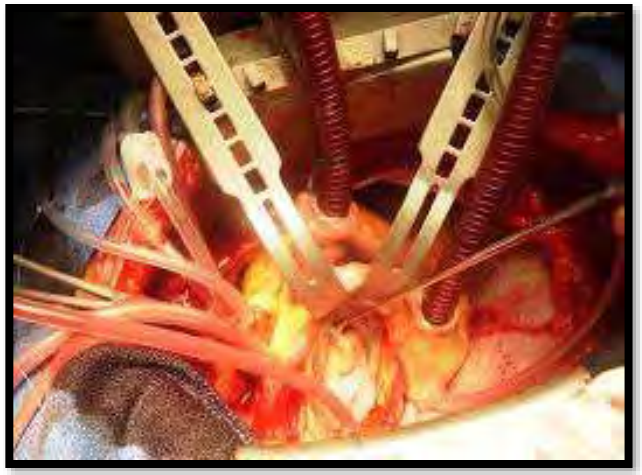
We can get babies to survive

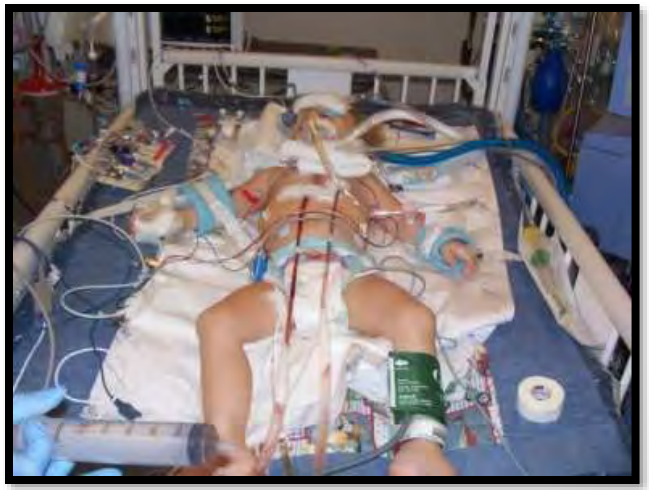
- Surgical mortality is < 5%
  - Moderate defects > 95% survival
- Complex Defects >85%

We can get to adulthood!

- TOF: >95% 25 year survival
- TGA: >95% 15 year survival

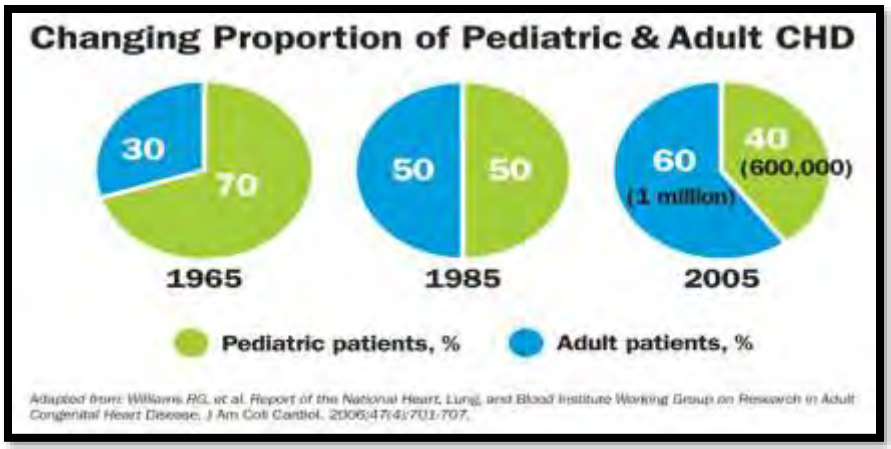
- Advances in medical care
  - Understanding of Natural history
  - Better Intervention timing
  - Non-invasive options
  - Medical advances in care





The ACHD Population

- 1.5-2 million adults with CHD in U.S. alone
  - More adults with CHD than children
- Adults with CHD increasing 5% yearly
  - Estimated growth >10,000 patients per year
- Young adults in the next decade
  - 1:150 adults will have CHD



The ACHD Population

Circulation

American Heart Association®

Lifetime Prevalence of Congenital Heart Disease in the General Population from 2000 to 2010
Ariane J. Marelli, Raluca Ionescu-Ittu, Andrew S. Mackie, Liming Guo, Nandini Dendukuri and Mohammed Kaouache

CHD prevalence (2000-2010)

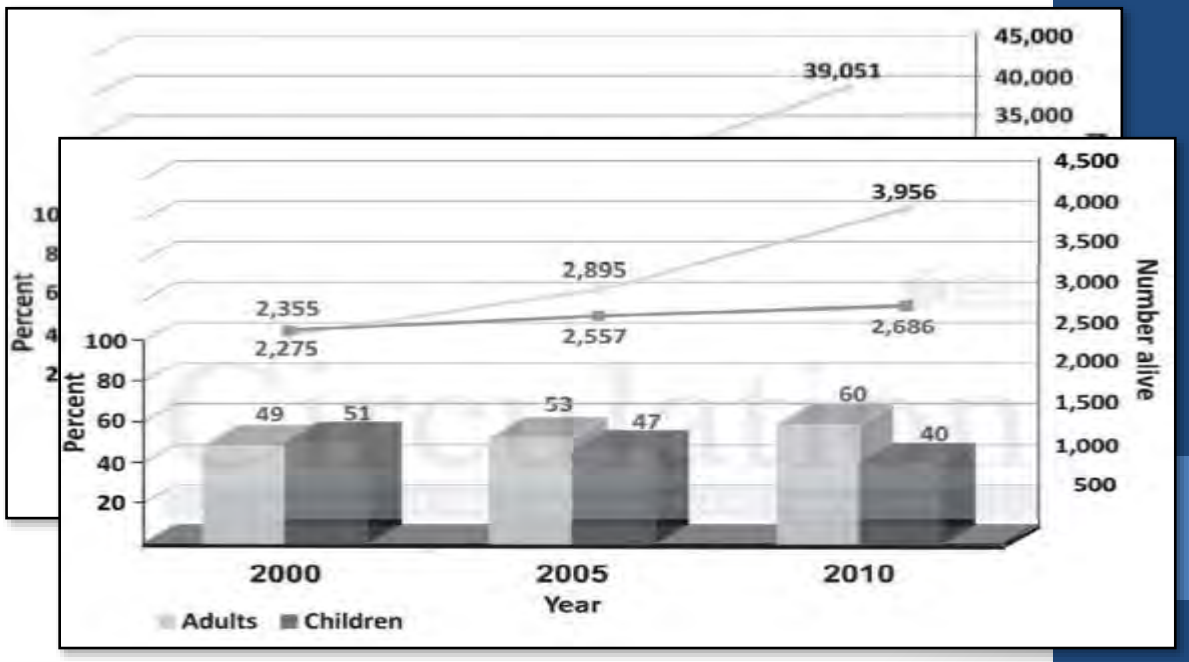
- 57% growth in adults

Severe CHD subgroup

- 55% growth in adults
- 2/3 of the CHD population

Perspective:

- 6.12/1000 Adults
  - 1.5-2 million adults in the US
  - 8000 adults in Nebraska



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The ACHD Population

- How do we care for these patients?
  - **Recognize** the problem
  - **Allocate** Medical Resources
  - **Educate** patients & providers
  - Make resources **accessible**

Recognizing the ACHD Issue

Bethesda Task Force (JACC 2001)

- Rising complexity of adults with CHD
  - 60% had prior surgery; 55% will require additional surgery
- Recommended Region Centers of Excellence

ACHD Care Guidelines (2008 ACC/AHA; Updated 2018)

- Outlines care standards for common ACHD lesions
- Reinforced “centers of experience” working with providers

ACC/AHA Vision 2020 for ACHD

- Board Certification (2015) via ABIM (ABP)
- Center Accreditation (2017)

ACHA
Adult Congenital Heart Association

Allocate Resources

Physician & Teams

Physician Training

- Education integrated in Medical Curriculum
- Fellow (Peds & Adult) educational standards
- Formal training pathway
  - 2yrs of dedicated ACHD fellowship following cardiology

Board certification of ACHD Physicians 2015 (ABIM)

- Roughly 500 board certified ACHD physicians 2022

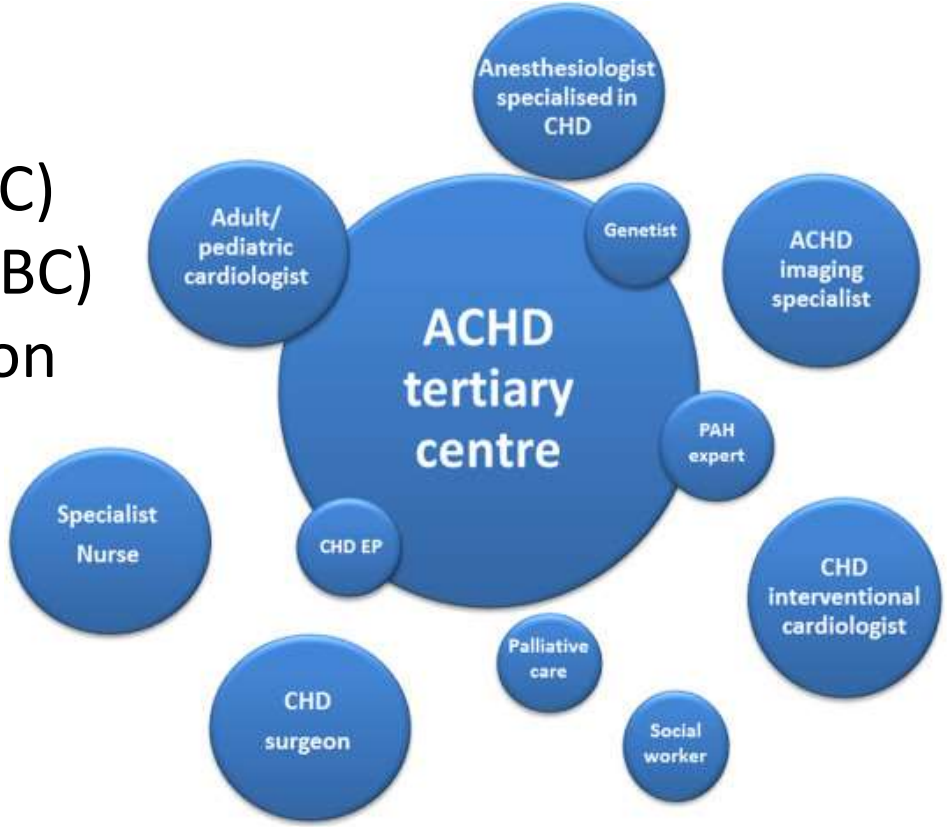
Standards for Centers of Excellence for support teams

- APPs, nursing, social work, etc

ACHD Centers

<u>Allocating Resources</u>:
Institutions

- ACHD cardiologists (BC)
- Congenital surgeons (BC)
- Congenital Intervention
- Cardiac anesthesia
- Congenital EP
- Advanced imaging
- Heart Failure/Tx
- High risk OB/GYN

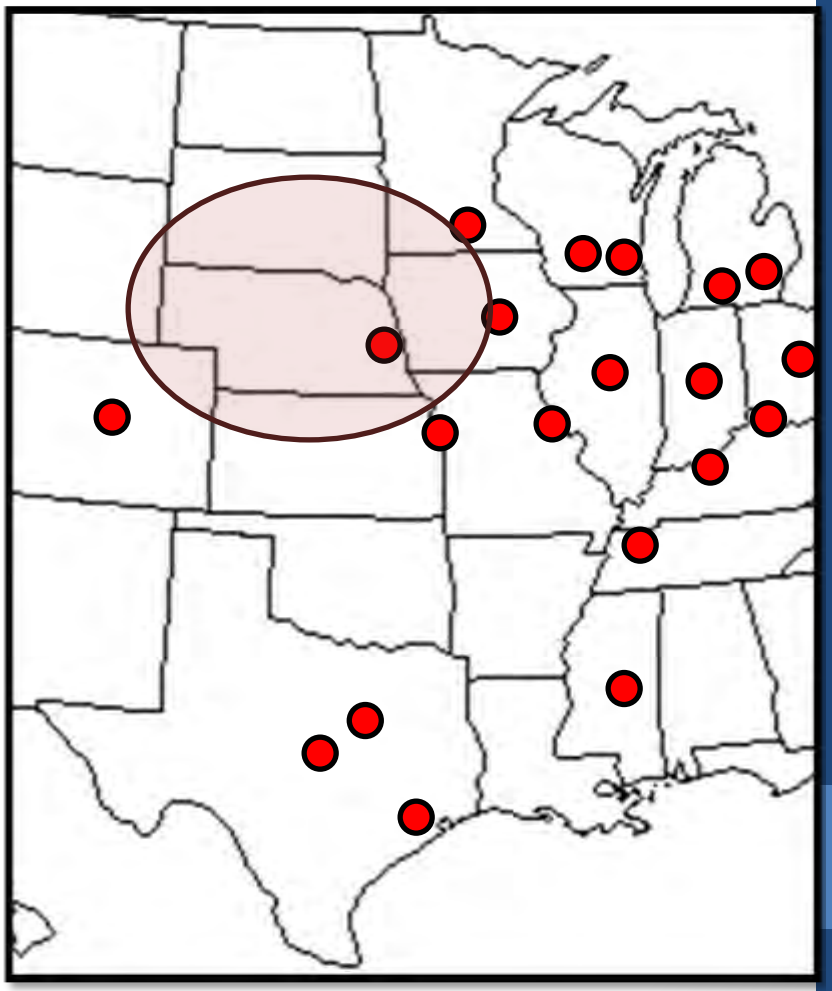


MEDICAL HOME

ACHD Centers

ACHA
Adult Congenital Heart Association

Adult Congenital Heart Association
ACHD ACCREDITED CARE CENTER



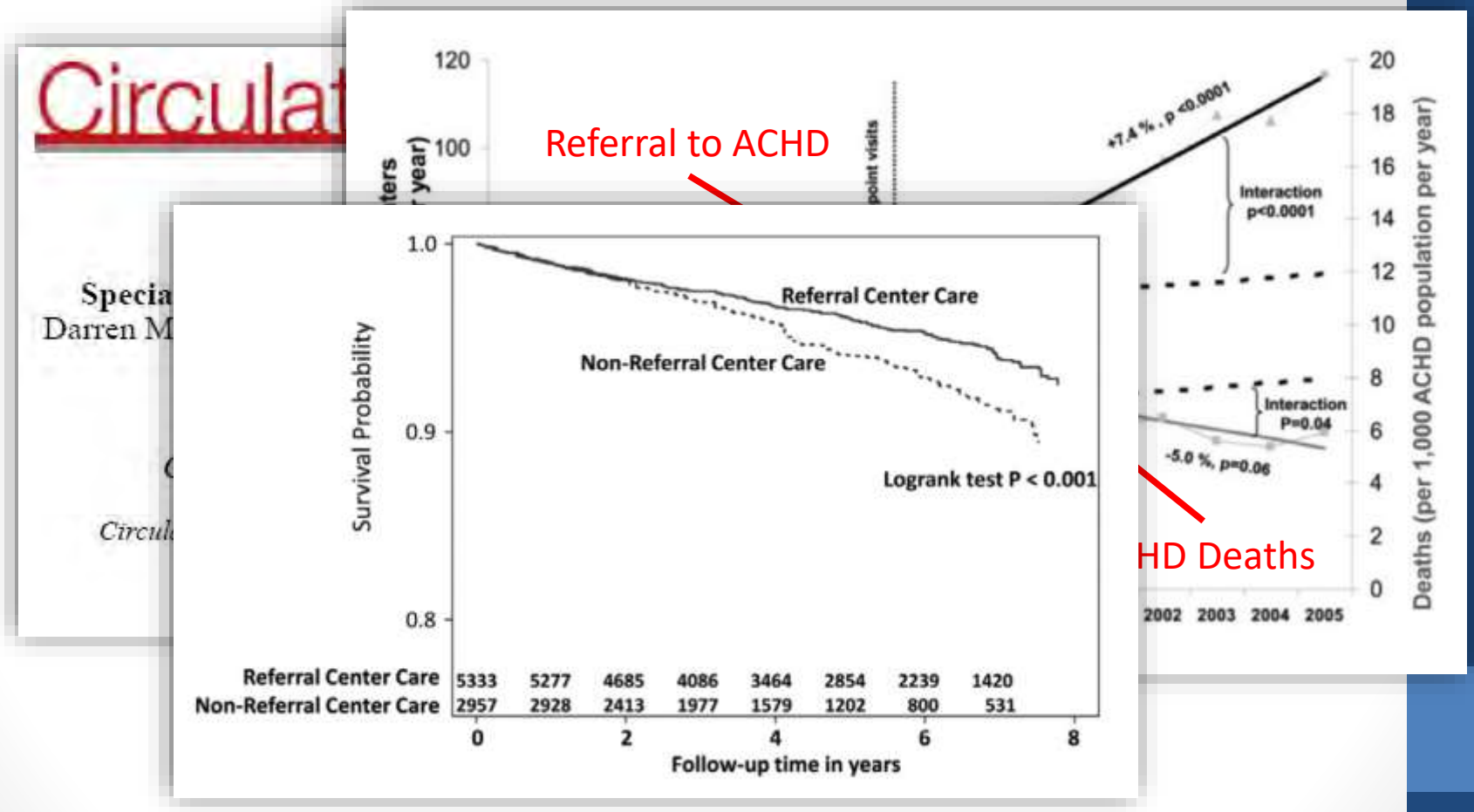
2017 Accreditation (Initial Cycle)

- 15 Accredited Centers in the US

2024 Accreditation (Most Recent Cycle)

- 54 Accredited Centers in the US

Results for Patients?



ACHD Education

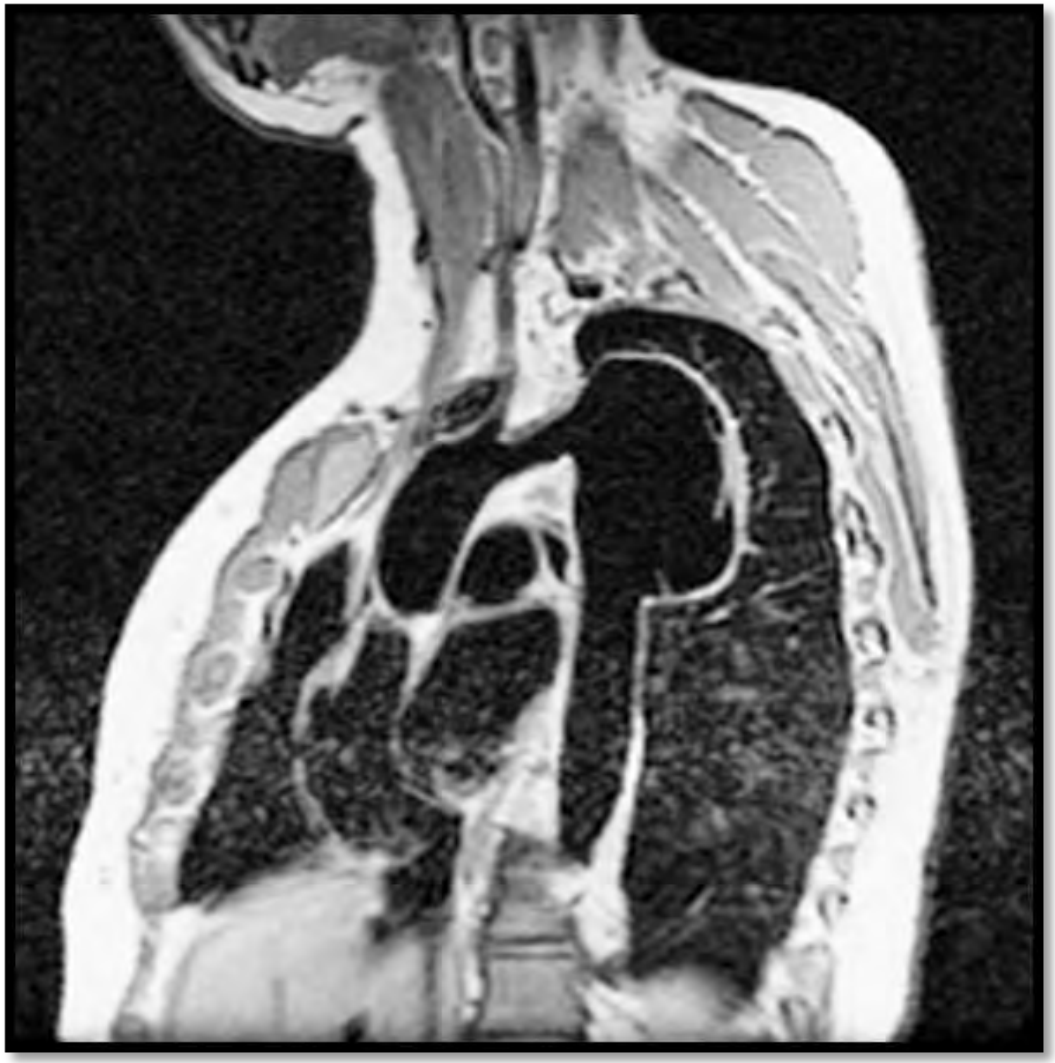
Patients and families

- Empowering patients to know their heart disease
- Understand importance of care & prevention
- Maintaining emotional health & allowing for maturity
- Transition; keeping patients in the system!
  - 45-60% transferred successfully

Learners, Providers, and Allied Health Professionals

- Formal Medical Education
- Learning opportunities across the region

Consequences of ACHD Failure



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ACHD Classification

2018 ACC/AHA ACHD Care Guidelines

- CHD Anatomy (I-III)
  - I or Simple (Isolated or Repaired ASD, VSD, PDA, PVS)
  - II or Moderate (Vast majority of anatomic abnormalities)
  - III or Complex (Single ventricles, TGA, DORV, cyanosis, IAA, etc)
- Physiologic Stage (A-D)
  - A (Asymptotic, no arrhythmia risk, normal exercise, normal anatomy and physiology, no other related organ issues)
  - B (Mild), C (Moderate), D (Severe) in ANY one issues
- Patients with IB-D, IIA-D, and IIIA-D should be managed in collaboration with an ACHD care team (LEVEL I; B-NR)

Common ACHD Lesions

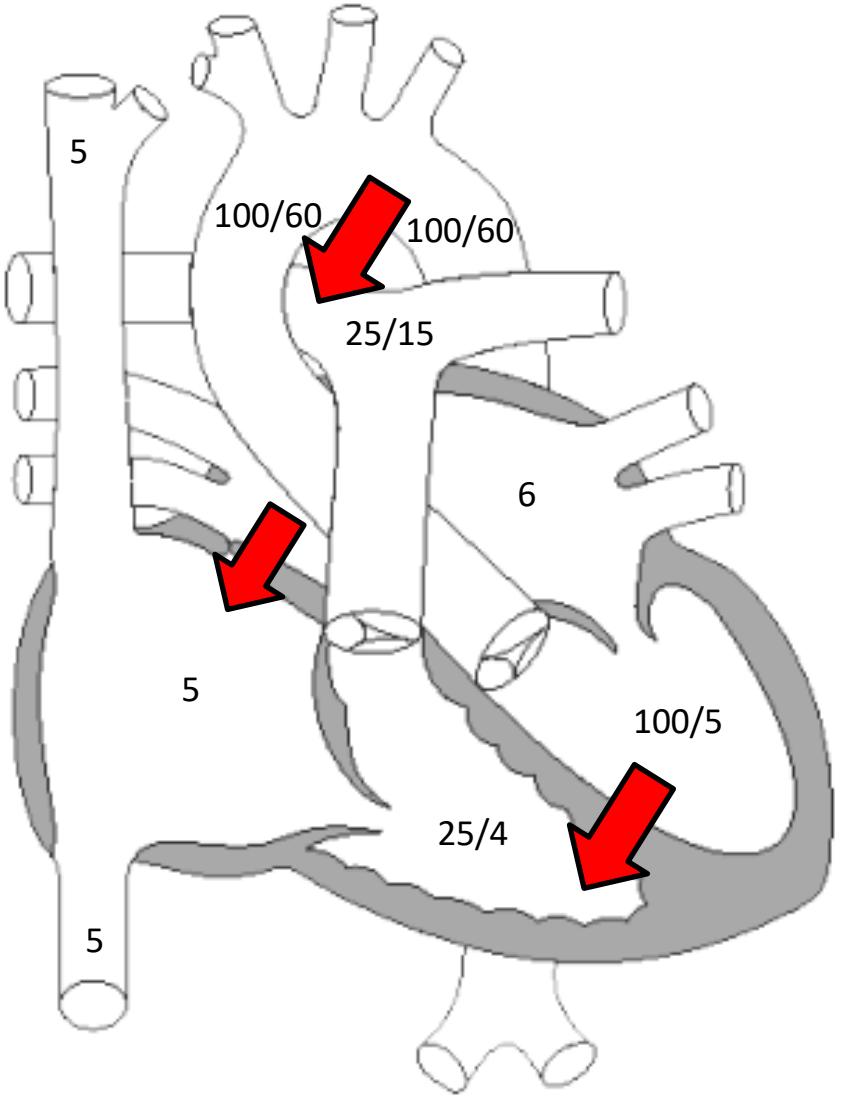
- General Conditions
  - Left to Right Shunts (ASDs, VSDs, PDAs, AVCs, PAPVR)
    - Atrial Septal Defects (ASDs)
  - Right to Left Shunts
    - Tetralogy of Fallot (TOF)
    - Transposition of the Great Arteries (TGA)
  - Obstructive Lesions
    - Bicuspid Aortic Valve
    - Coarctation of the Aorta (CoA)
  - Complex conditions

Physiology

L -> R shunts result in **excessive** pulmonary blood flow (Qp)

Lung to systemic flow (Qp > Qs)

Degree of shunt determines symptoms

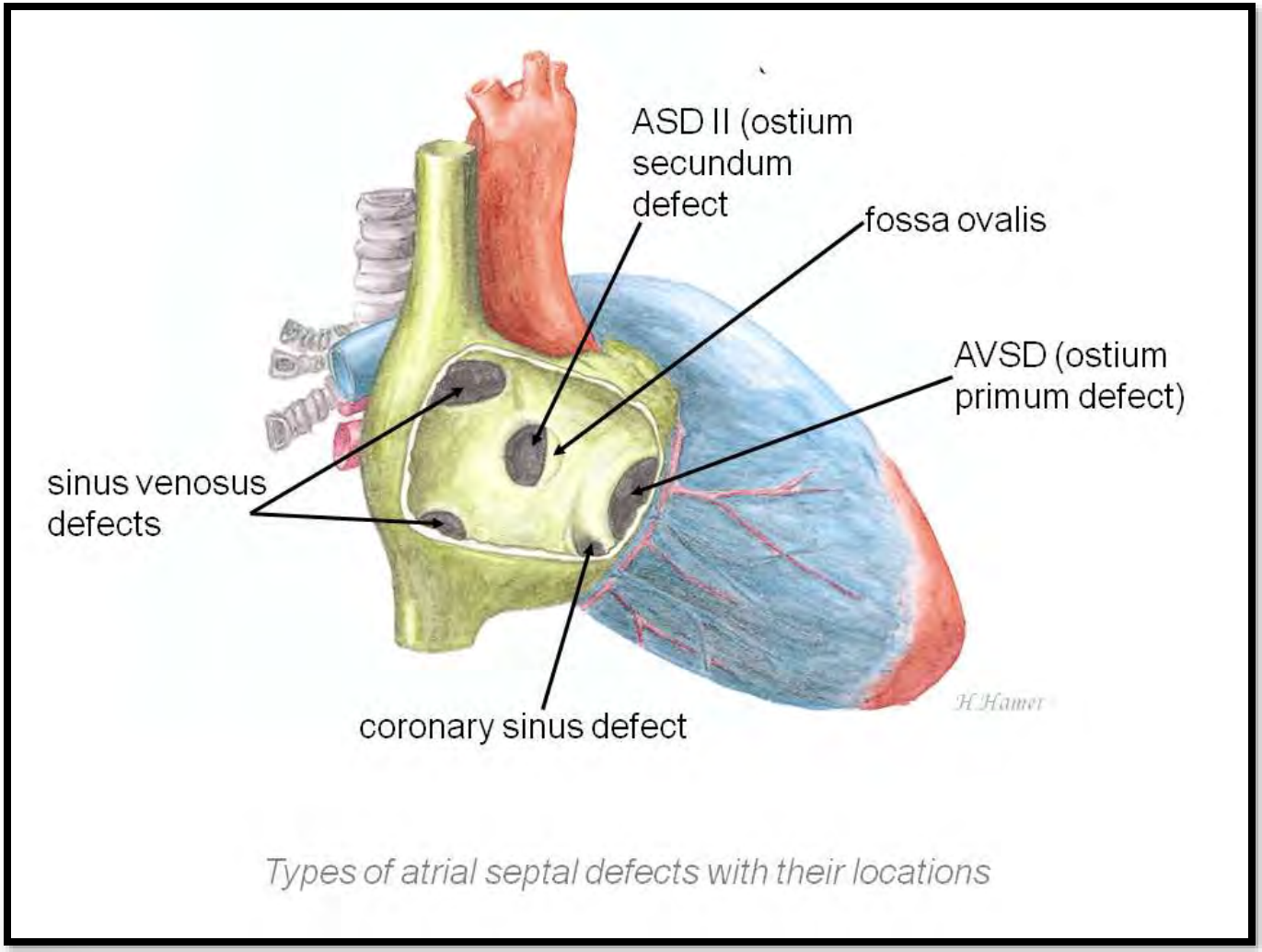


Left to Right Shunt

Natural history of left to right shunts

- As lung resistance drops in neonates, shunt worsens (Qp>Qs)
  - Pulmonary over circulation and congestive heart failure
- Continued volume and pressure load on the lungs results in pulmonary vascular changes (usually >2yrs of age)
- Progressive changes leads to lung resistance > systemic resistance and Eisenmenger physiology sets in (Qp<Qs)
  - Onset of CHD related pulmonary vascular changes results from a combination of shunt (volume) AND pressure
  - ASD and PAPVR are usually volume alone (no PHTN usually)
  - VSD, AVC, PDAs are usually volume AND PRESSURE (PHTN)

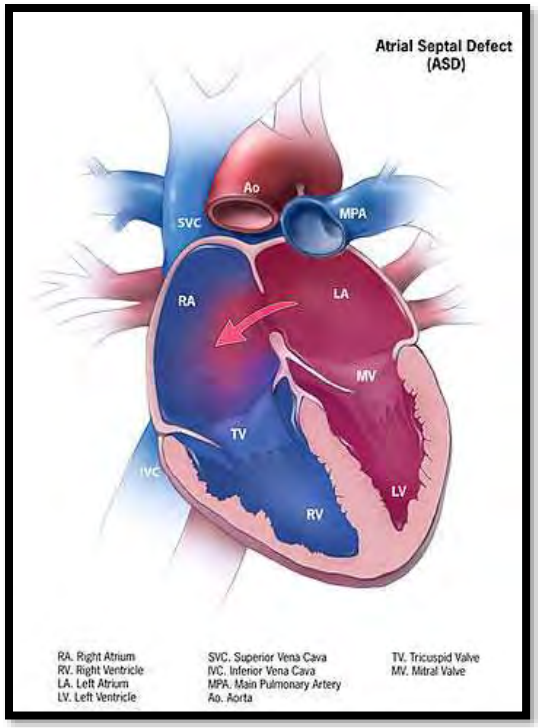
Atrial Septal Defects



Types of atrial septal defects with their locations

Atrial Septal Defects

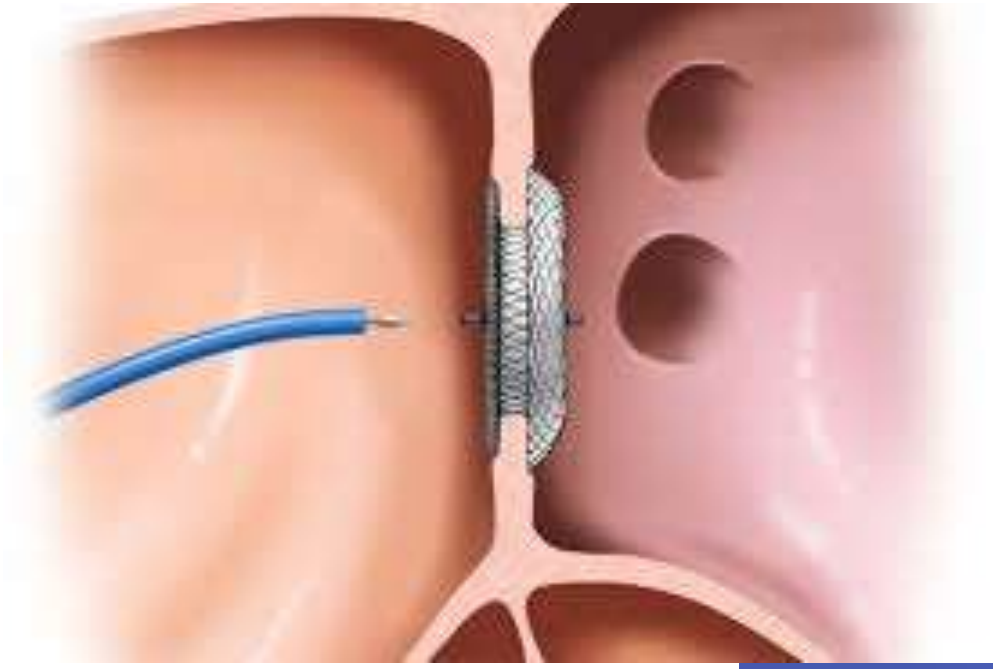
- Low volume left to right shunt
- Moderate sized defects can remain mildly symptomatic
- Long term outcomes change at age 25yr and 40yrs
  - Right heart failure risk rises
  - Atrial arrhythmias may become commonplace
- PFOs and small ASDs; hemodynamically insignificant
  - 10-20% of population may have PFO
  - Closure can be indicated in appropriate circumstances

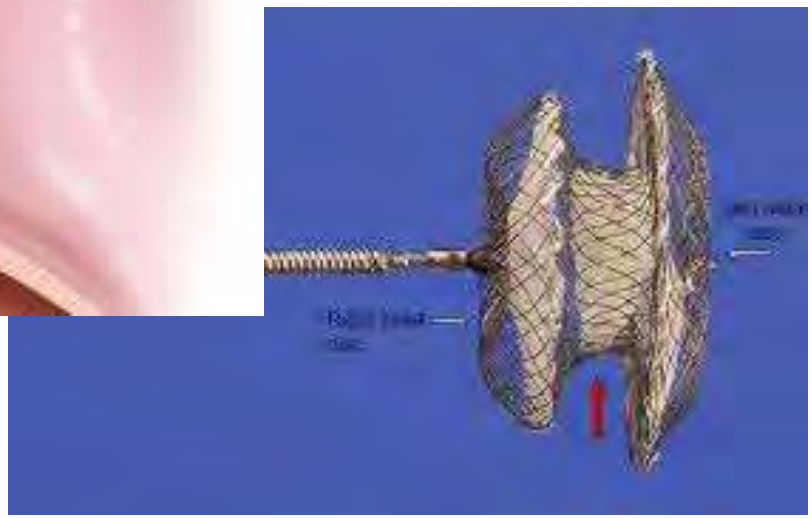


Atrial Septal Defects

- Finding Adult ASDs
  - Right sided enlargement of unknown cause
  - Occasionally murmur, fatigue, CXR
- Indications for Closure
  - ASD anatomy (type, location, rims, pulm. veins)
  - Qp:Qs > 1.5:1 (hemodynamically significant shunt)
  - Absence of pulmonary HTN (PVR rule of thirds)
- Reasons to refer to a ACHD cardiologist
  - Newly diagnosed adult ASDs with any question above
  - ASDs with other cardiac lesion

ASD Closure

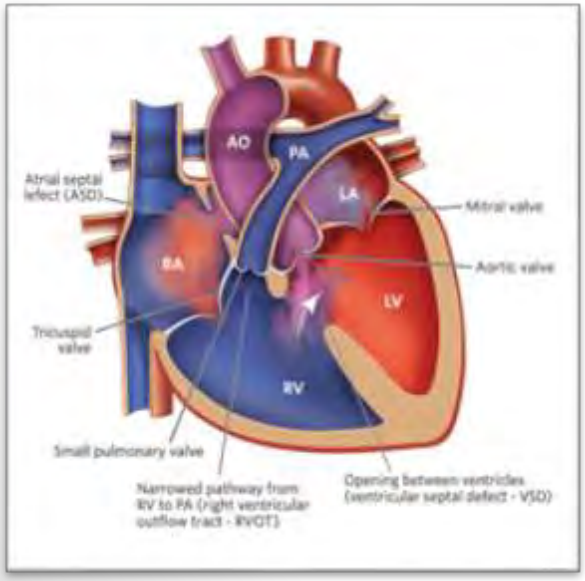


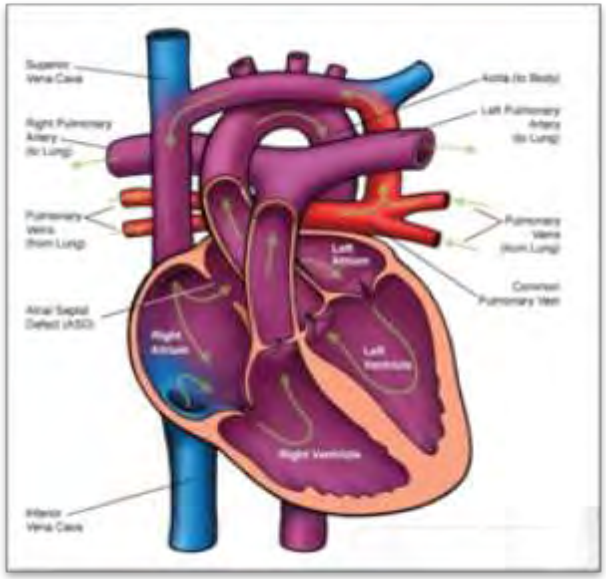


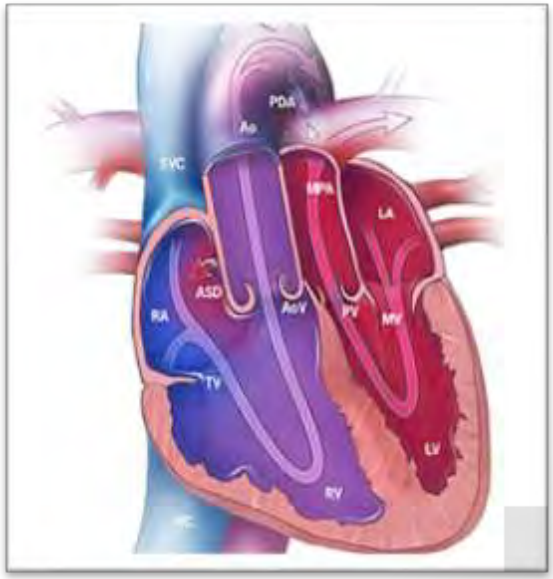
Right to Left Shunts

Cyanotic Congenital Heart Disease

- Complex and heterogeneous group of defects but all have blue, deoxygenated blood entering the aorta
  - TOF has decreased pulmonary blood flow (Qp)
  - TAPVR has normal or high Qp with total mixing of Qs
  - TGA has ineffective Qp with admixing

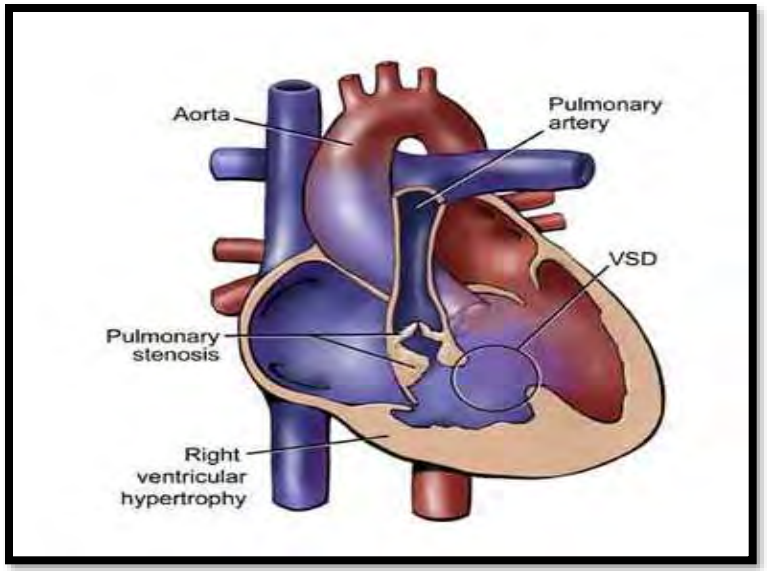


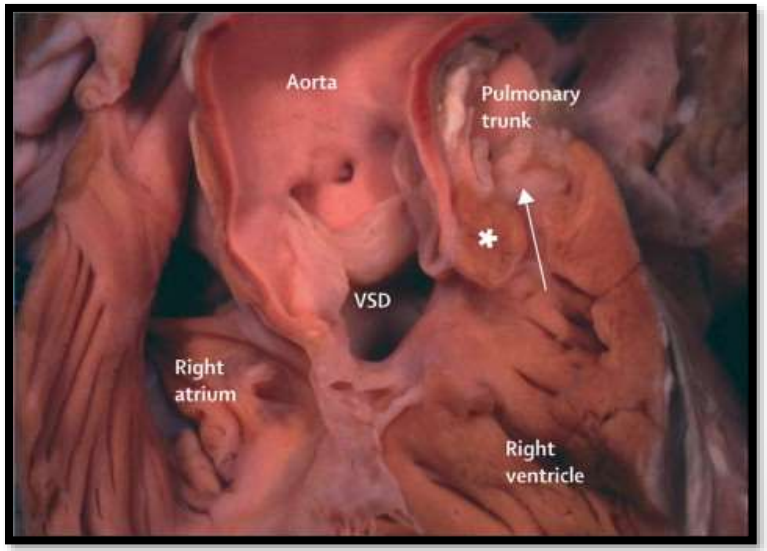




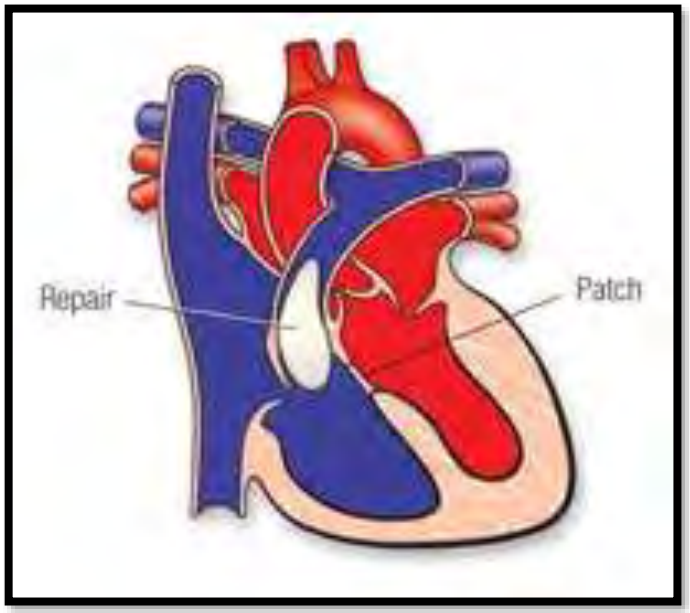
Tetralogy of Fallot

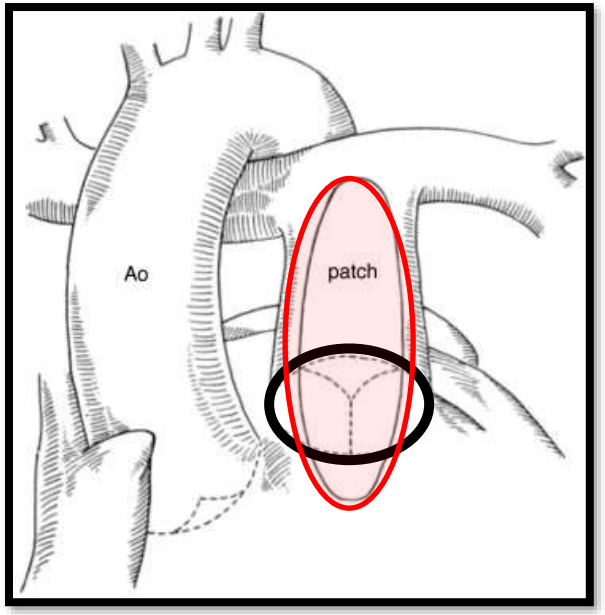
- Most common cyanotic heart defect
- 3.3/10,000 live births
  - 5th most common heart defect
- 7-10% of congenital heart disease
- Typically repaired in the first year of life in single stage

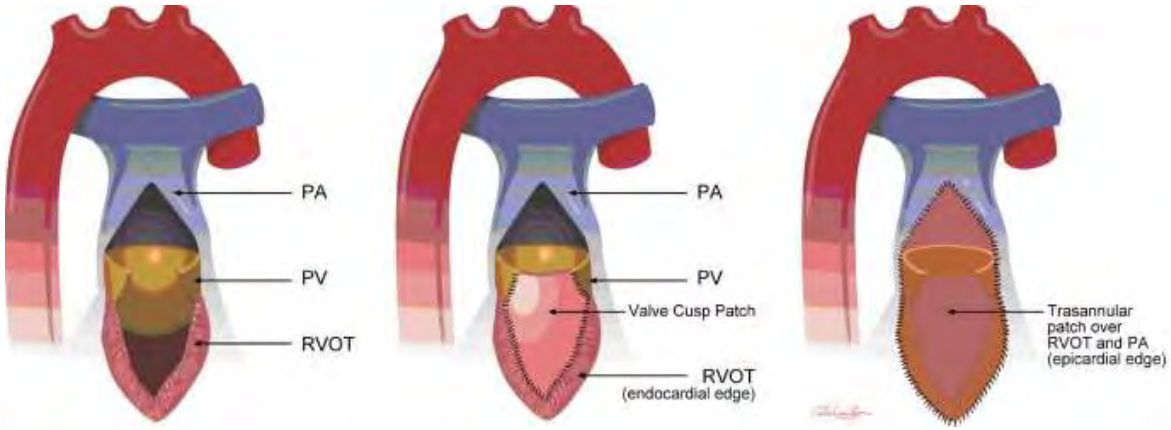




Tetralogy of Fallot

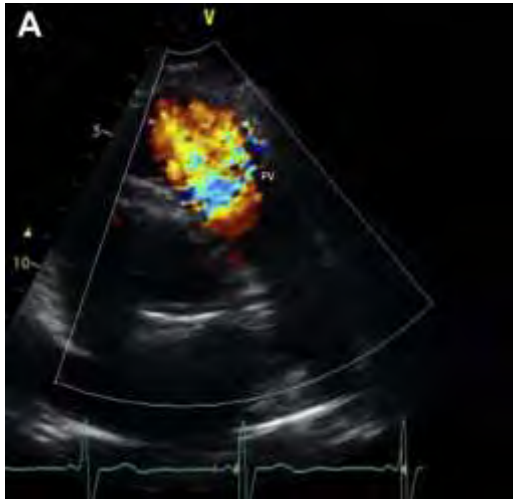






Tetralogy of Fallot

- Early Complications
  - Uncomplicated TOF: Very low mortality rate
- Long term (3rd decade of life)
  - Pulmonary valve regurgitation
  - RV enlargement and failure
  - Residual hemodynamic issues
    - VSD, pulmonary stenosis, etc
  - Electrical abnormalities
    - RBBB seen on EKG in 90% patients
    - Intra-atrial reentry tachycardia
    - Ventricular tachycardia



Tetralogy of Fallot

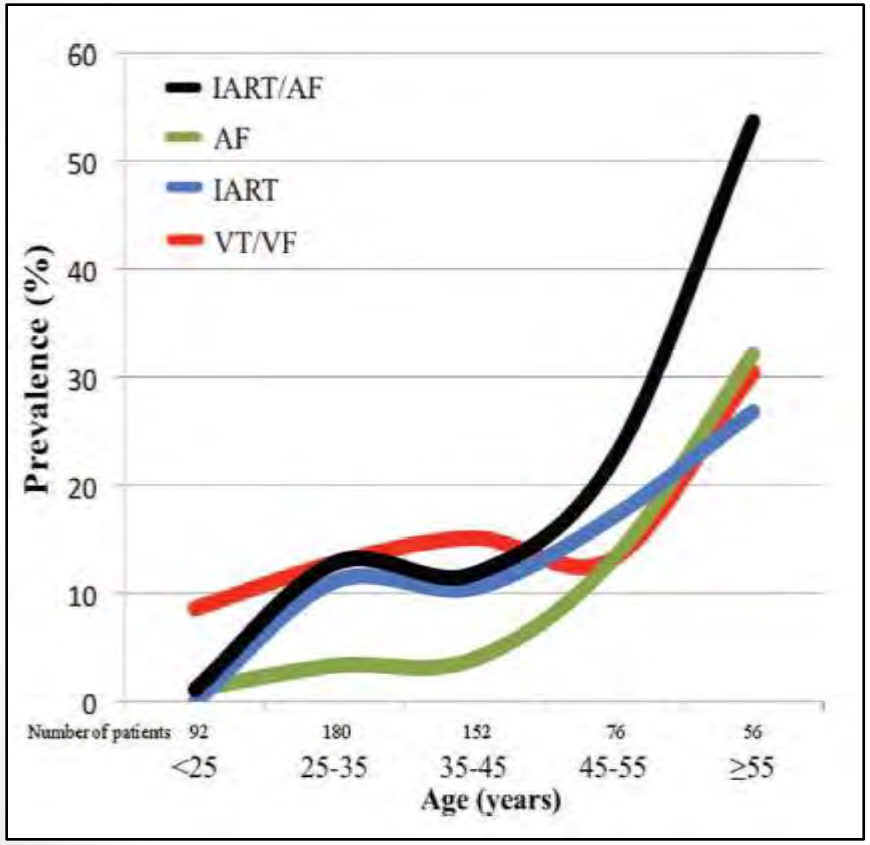


Table 2. Arrhythmia Burden in Adults With Surgically Repaired Tetralogy of Fallot

Characteristic	Prevalence, %	95% CI
Sustained tachyarrhythmia	29.9	26.2–33.7
Atrial tachyarrhythmia	20.1	17.0–23.6
IART	11.5	9.0–14.3
AF	7.4	5.4–9.7
Other	6.7	4.8–8.9
Ventricular tachyarrhythmia	14.6	11.8–17.7
VT	14.2	11.5–17.3
VF	0.5	0.1–1.4
At least 1 arrhythmia intervention	21.4	18.1–24.9
Transcatheter ablation	7.2	5.2–9.5
Implanted cardiac arrhythmia device	18.3	15.3–21.7
Pacemaker	7.9	6.0–10.5
ICD	10.4	8.1–13.1
Sustained tachyarrhythmia and/or intervention	43.3	39.3–47.5

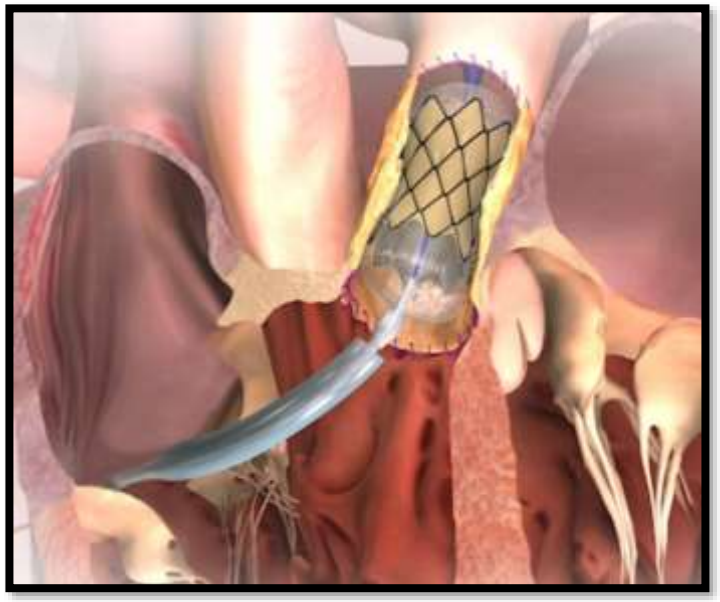
Tetralogy of Fallot

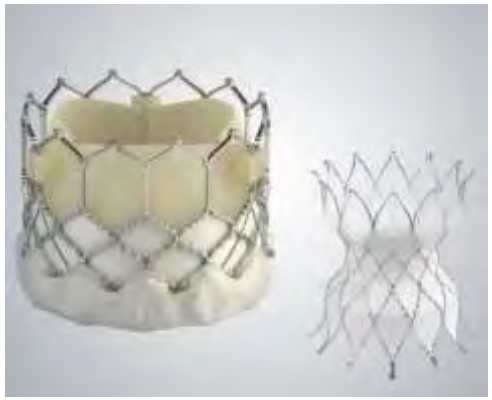
- Recommendation for patients with repaired TOF
  - Yearly follow-up with ACHD
  - Surveillance Imaging
    - Yearly echo to assess PVR, RVE, etc
    - cMRI to assess PR and RV volumes
    - Periodic Holter monitors and ECG
    - Assessment for PVR
  - PVR indicated for right heart dilation/dysfunction
  - Interventions should be done at ACHD Center

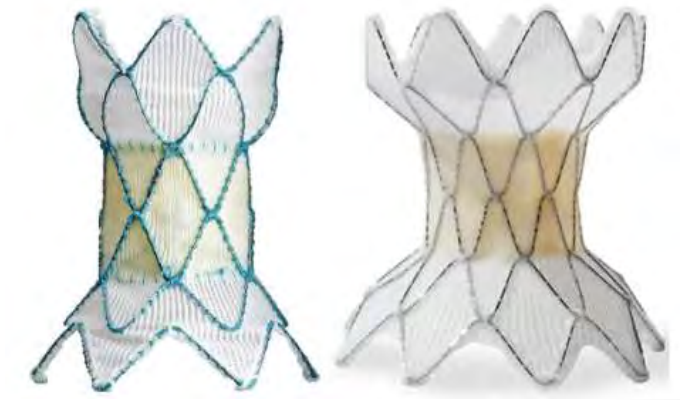
PVR Options in 2024

Options based on
1. Native Outflow vs. Surgical PVR
2. Size and anatomy

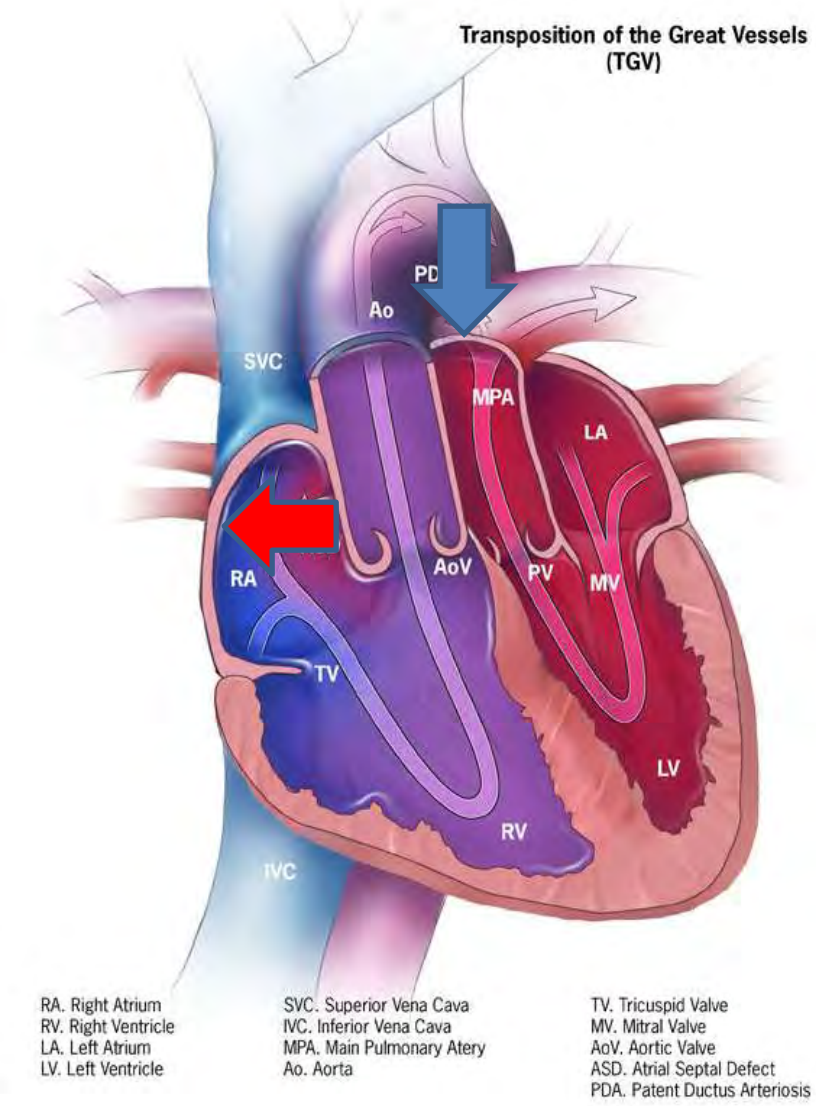
Advanced imaging for sizing







Transposition Great Arteries

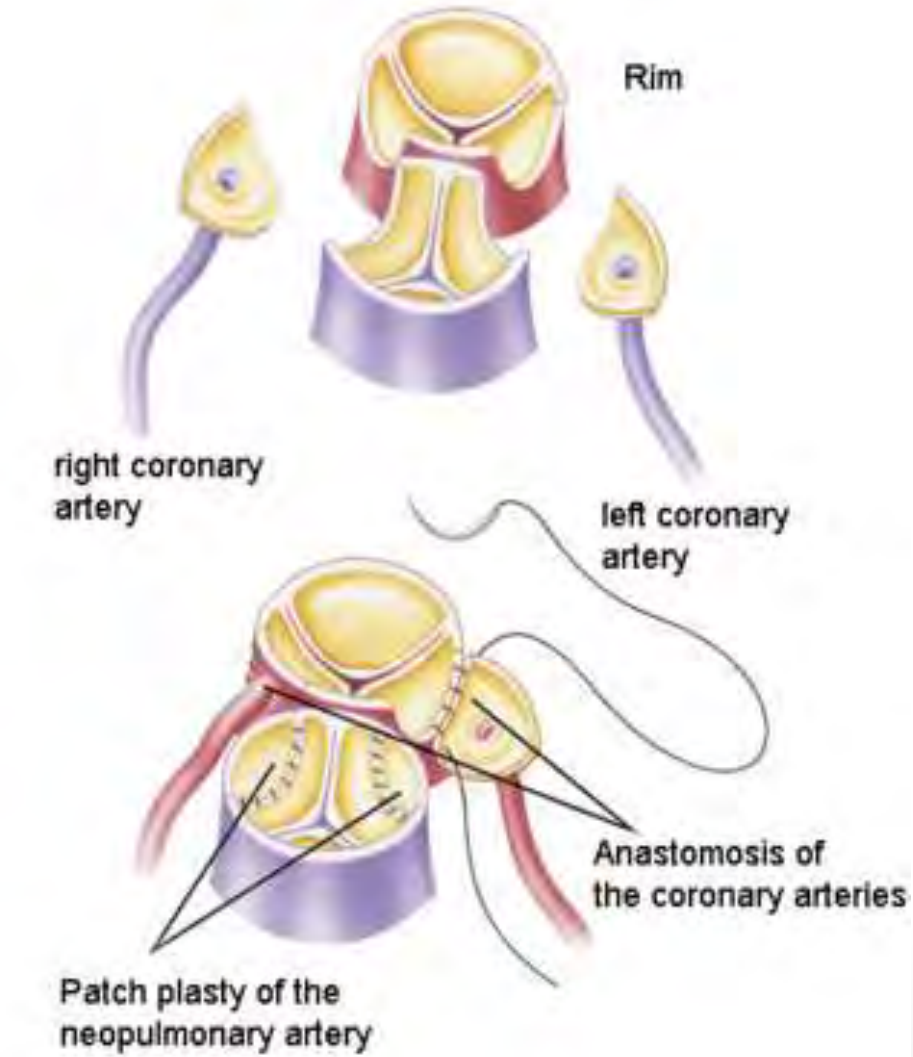


D-TGA

Atrial switch era

- Pre-1980s
- Inability to switch the coronary arteries
- High early death rate

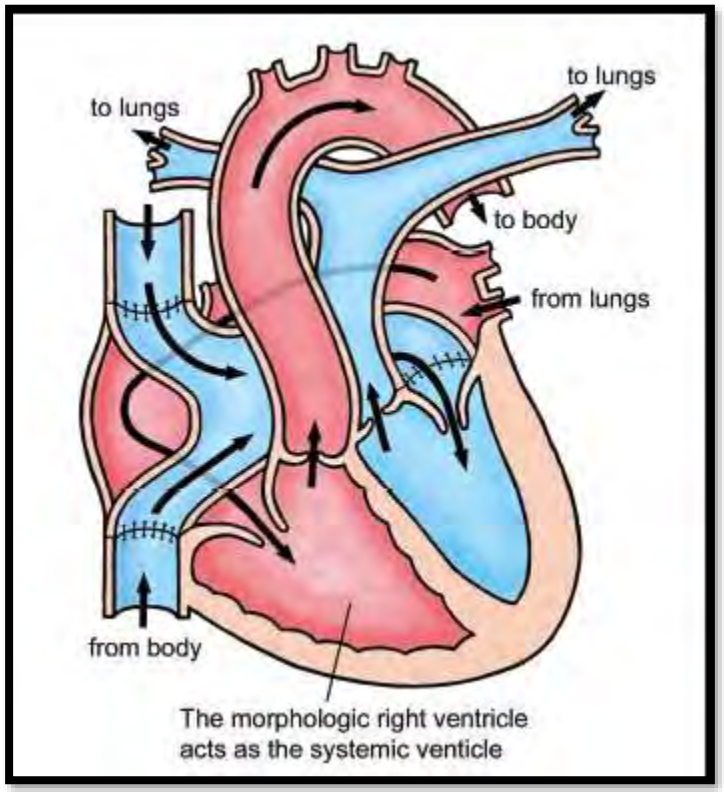
- Abandoned for “atrial switch procedure”



D-TGA

Surgical Repair

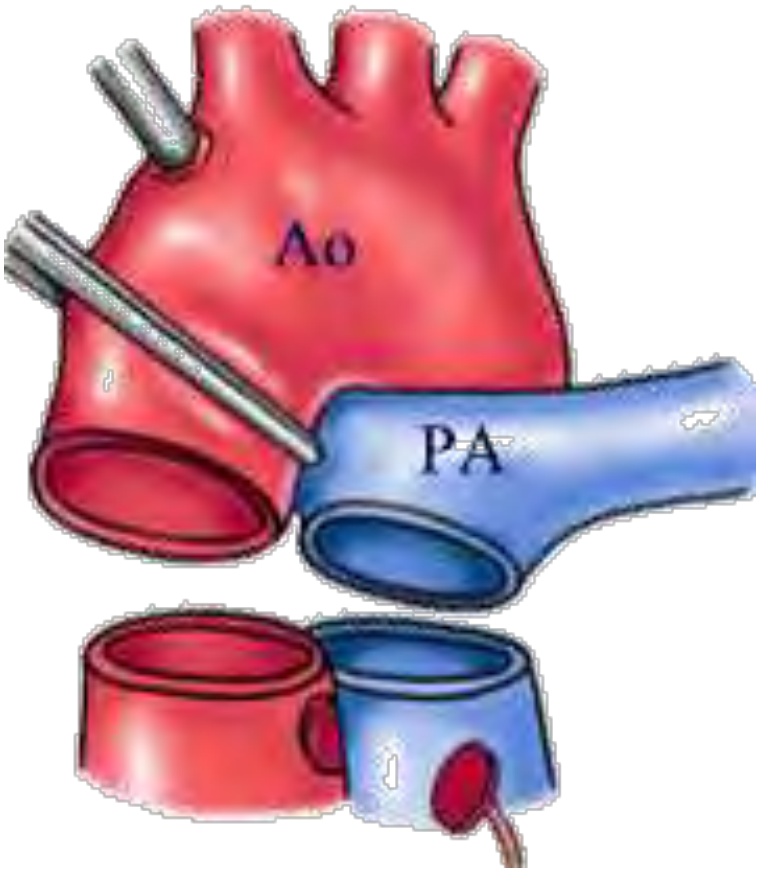
- ***Atrial*** Switch
  - Mustard/Senning
  - Surgery of choice from 1960s – 1980s
- Excellent early survival
- Significant late comorbidities
  - Baffle problems
  - Systemic RV failure
  - Atrial arrhythmias



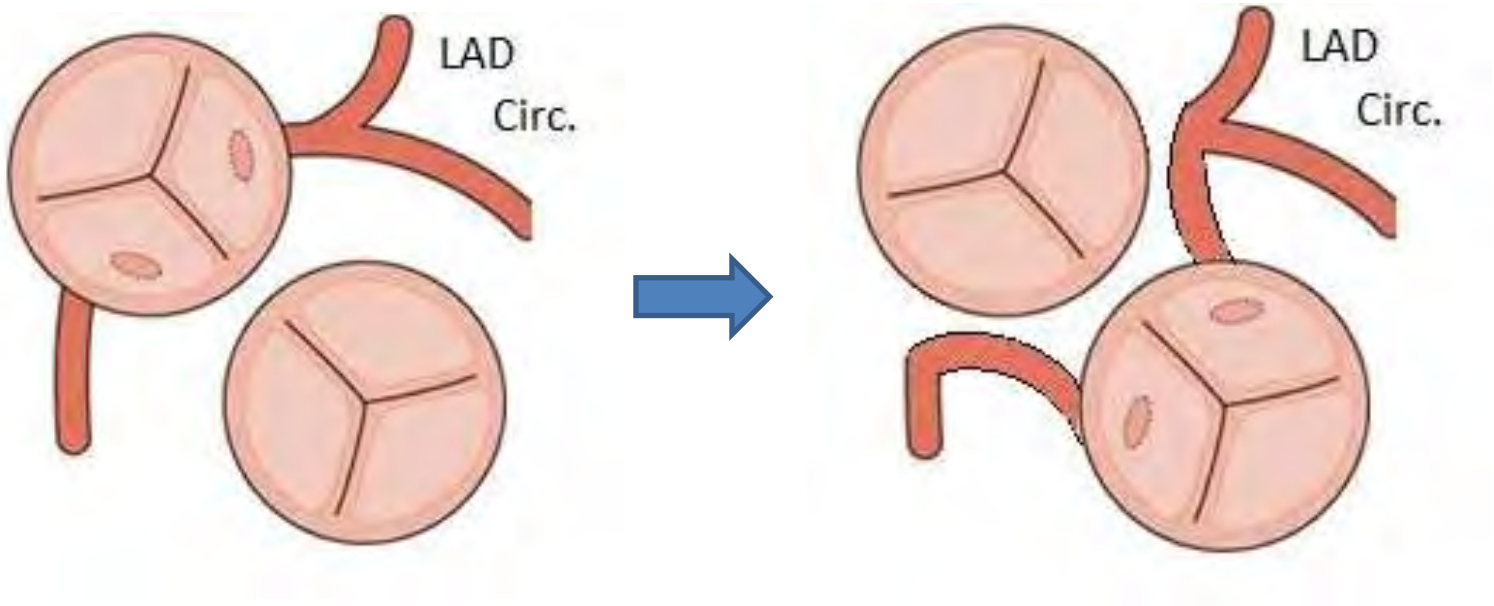
D-TGA

Arterial Switch

- Surgery of choice from 1980s-present
  - LV is systemic pump
  - No arrhythmias
- Excellent early and late survival
- Limited long term complications



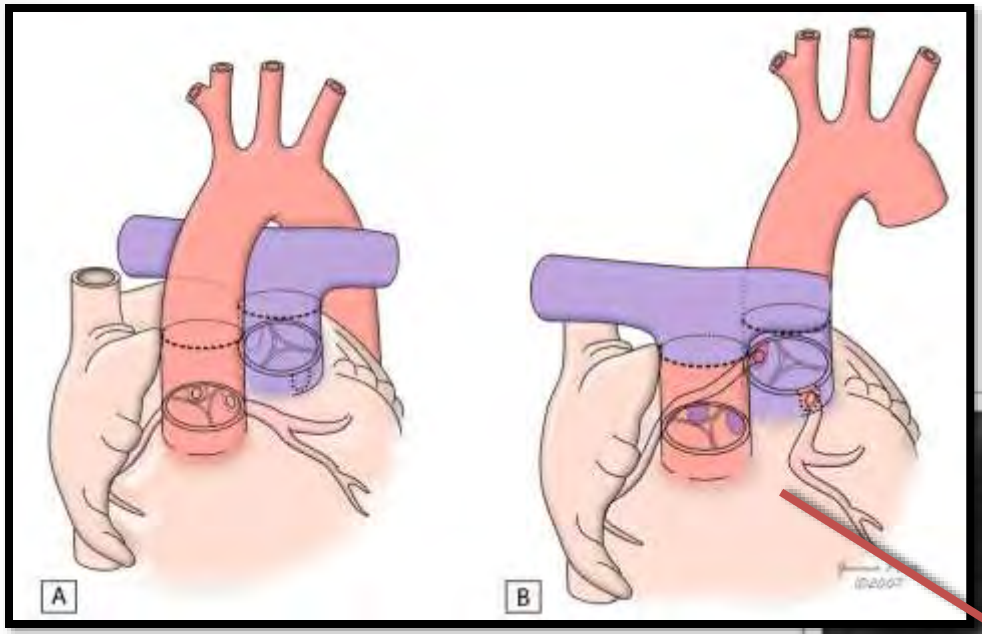
D-TGA



Coronary Switch Issues

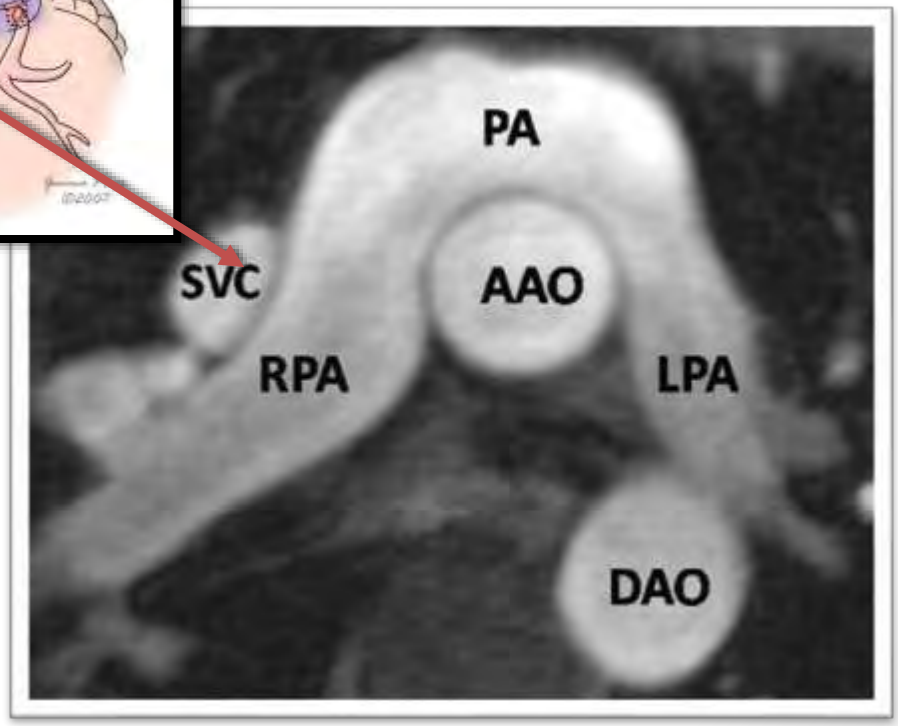
- Second most common long-term complication (10%)
  - Likely less in recent era
- Can frequently be subclinical
- Unknown impact on CAD later in life

D-TGA

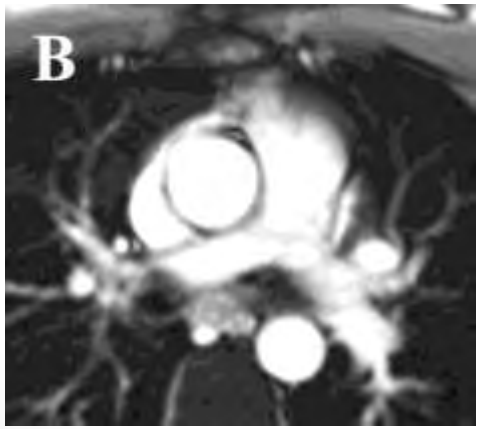


LeCompte Maneuver

- RPA anterior to Aorta
- Lead to MPA/RPA/LPA stenosis
- Most common long-term issue

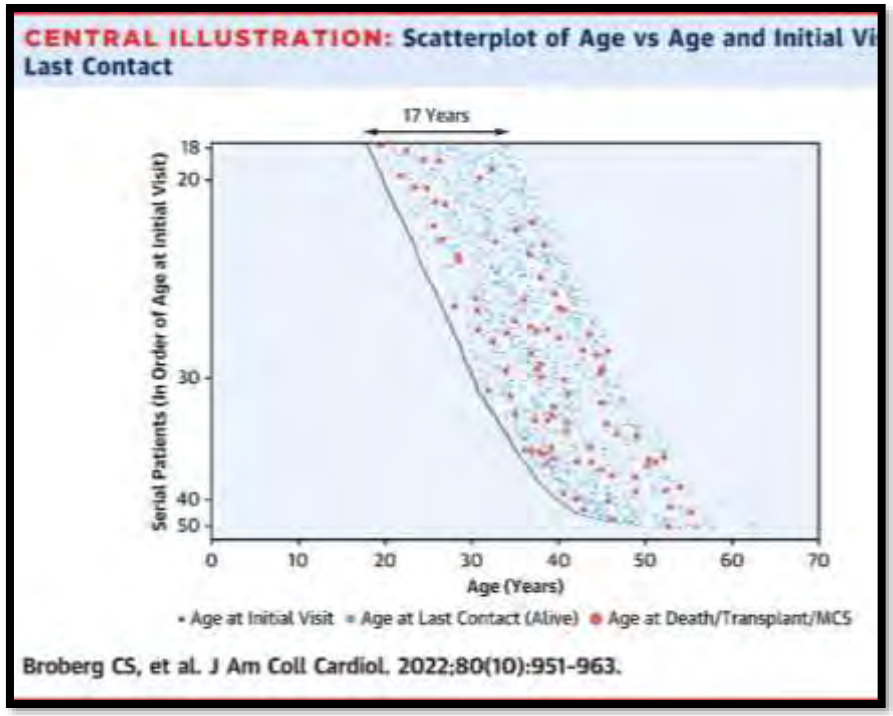


normal



Outcomes for Systemic RVs

- Systemic right heart failure is the rule, with some exceptions
- Arrhythmias risk is significant
- Significant residual lesions
- Initial events occur in in 30-40s



Obstructive Lesions

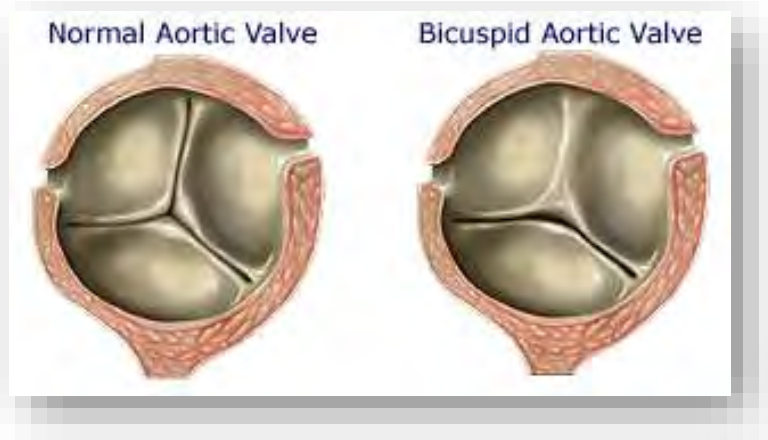
Left sided obstructive lesions

- Most common forms of CHD
  - More often affect males
  - Can run in families
- Frequently can present in series
- Common types
  - Mitral valve: Supravalvar mitral ring, parchute mitral valve, double orifice mitral valve, or congenital mitral stenosis
  - LVOT: subaortic membrane
  - Aortic valve: bicuspid aortic valve, valvar hypoplasia, congenital aortic stenosis
  - Aorta: coarctation of the aorta

Valvar Abnormalities

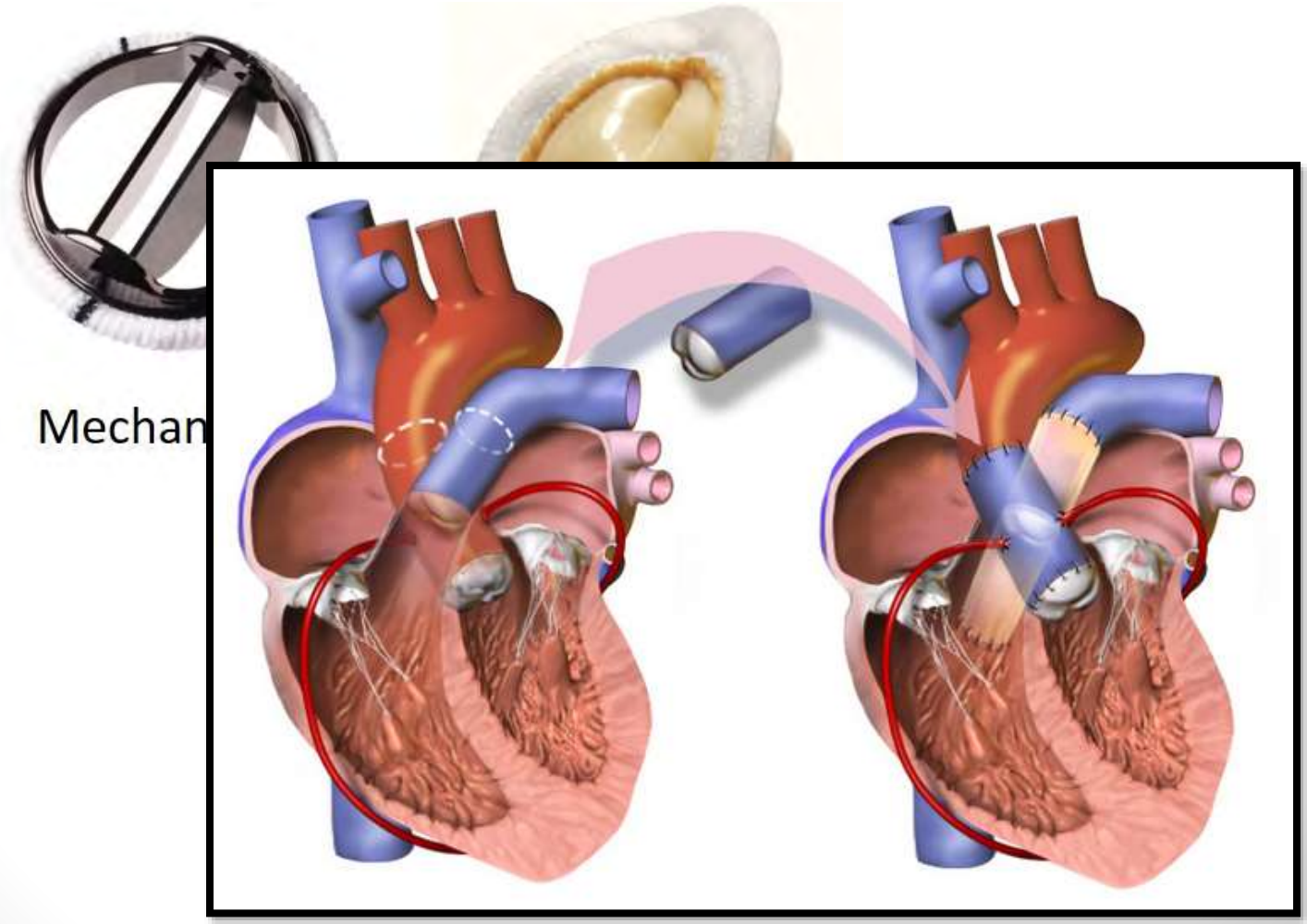
Bicuspid aortic valve

- 1-2% of patients
- Range of Presentation
  - Asymptomatic to severely compromised
- 50% will need intervention by age 50yrs
  - 85% by age 65yrs

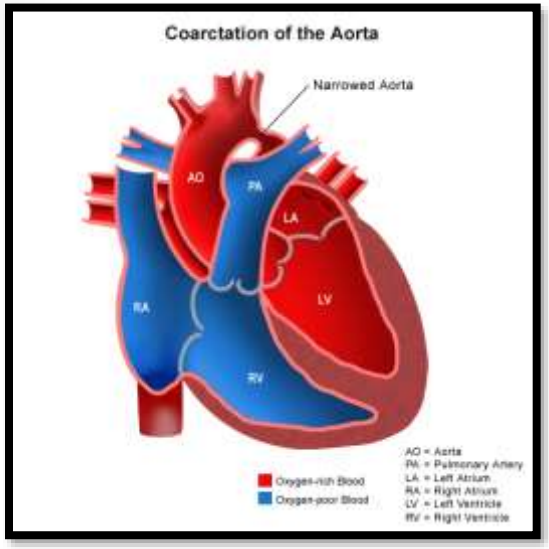


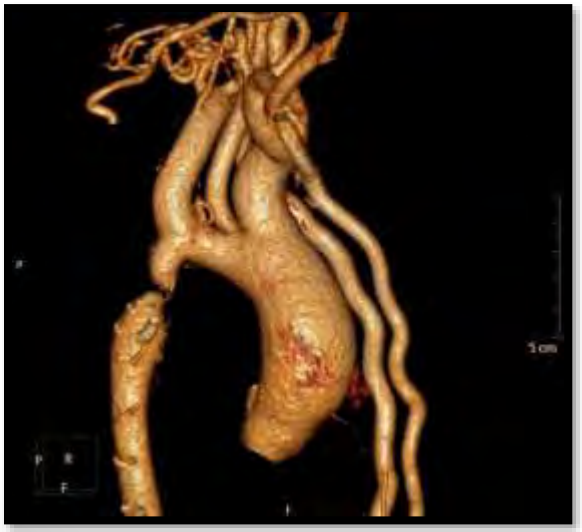
- Aortic root dilation
- Associated abnormalities
- Genetic status

BAV Options



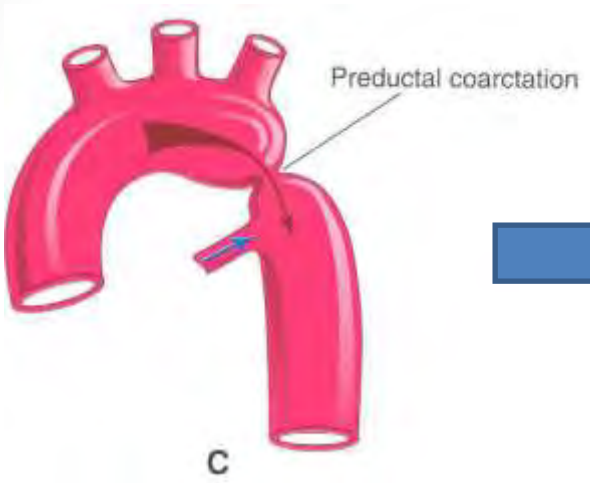
Coarctation

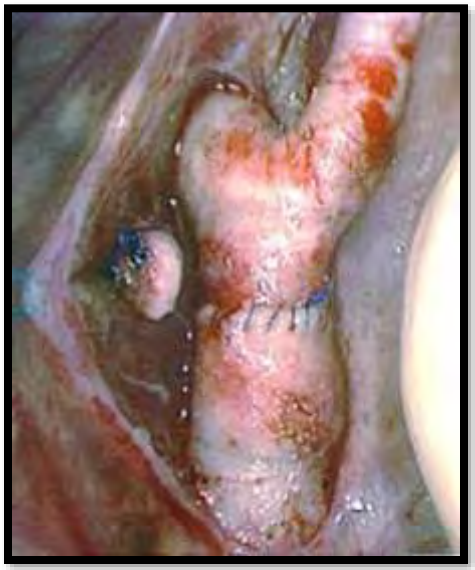


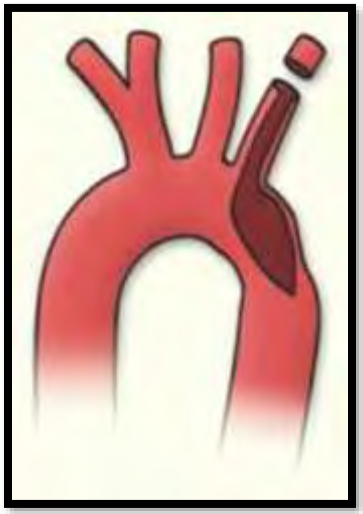


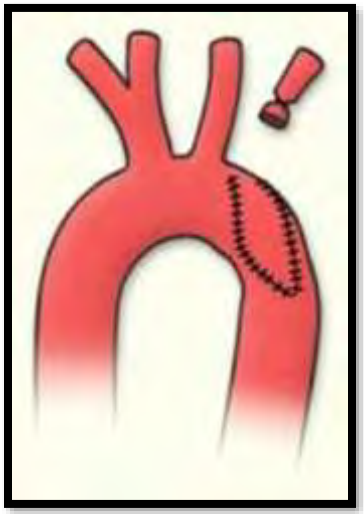
- One of the most common cardiac conditions
  - Commonly repaired in childhood
  - Occasionally diagnosed later in life
- Correct diagnosis of associated lesions

Coarctation



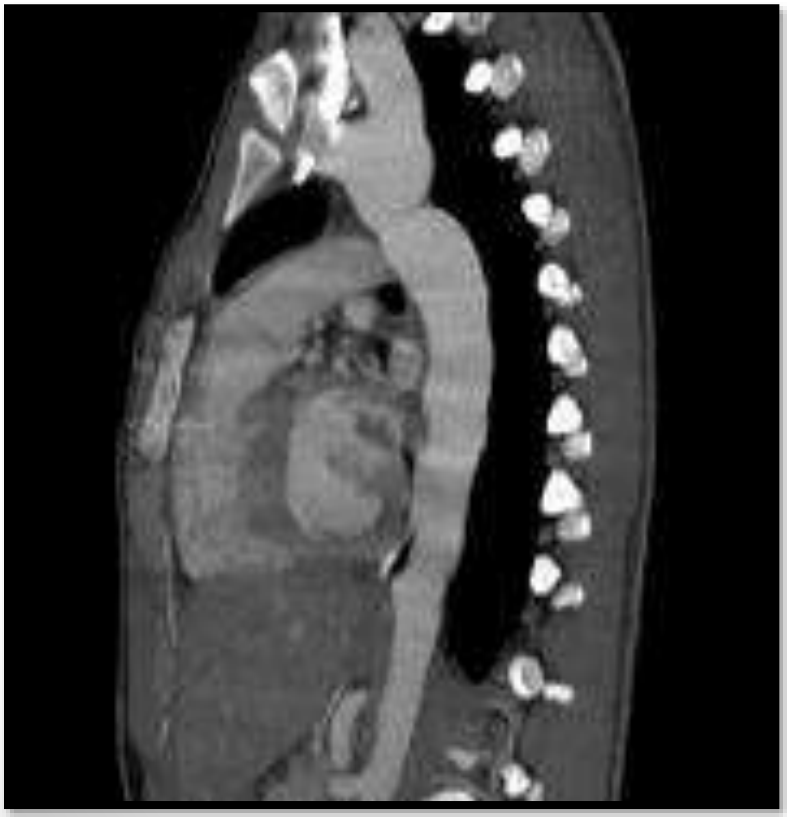




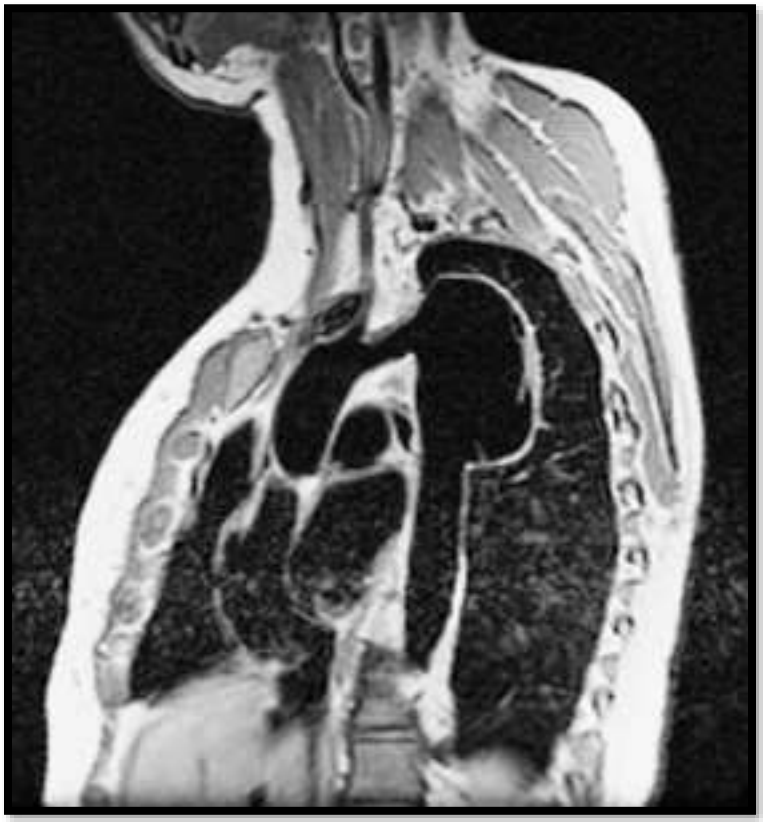


Coarctation

Recoarctation

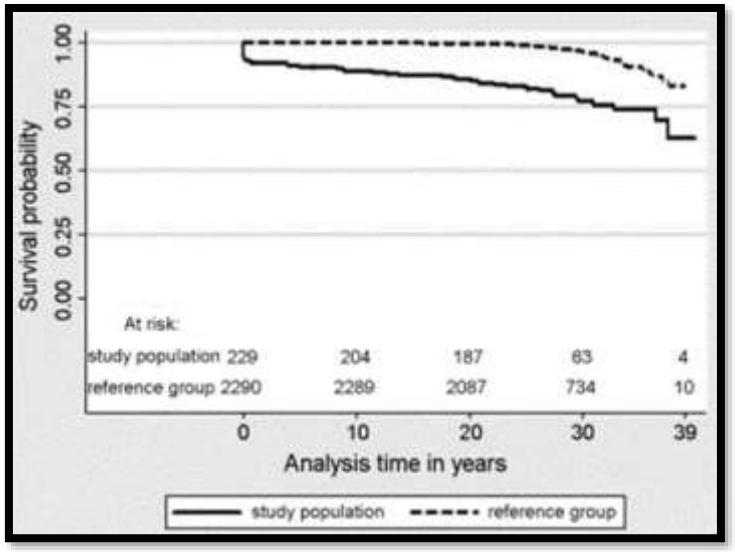


Aneurysm



Coarctation

- One of the most common "*fixed*" lesions
- Long term follow-up is necessary
  - HTN occurs in 30-80% of patients
  - Recoartctation: 10-20% lifetime
  - Aortic aneurysm 10-40% depending on type of repair
  - CAD, endarteritis, etc
- Correct diagnosis of associated lesions is key



Objectives

- Describe the current *population and future growth* of patients with congenital heart disease
- List current strategies for addressing the ACHD issue
- Explain a few of the *most common congenital heart lesions* and associated long term risks of each
- **Be aware of *"high-risk" lesions and situations* for patients with congenital heart disease**
- Identify *ACHD resources* within your community

High Risk ACHD

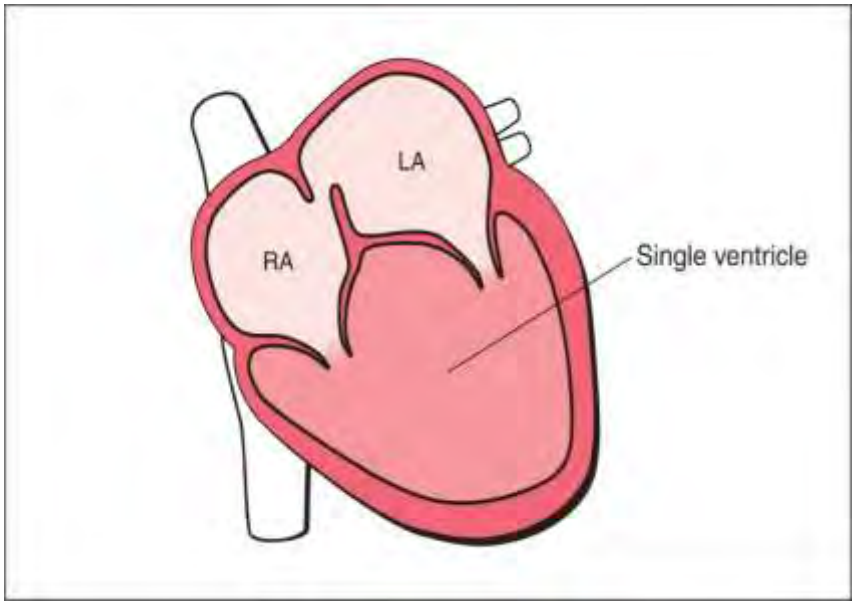
Highest Risk Patients (Grade III or I/II Class C-D)

- Single Ventricles
  - Fontans
- Systemic Right Ventricles
  - Atrial Switches
  - L-TGAs
  - Fontans
- Cyanotic Heart Disease/Eisenmenger
- CHD associated Pulmonary Hypertension
- Complex CHD associated arrhythmias

Single Ventricles

Single Ventricles

- Single Left Ventricles
  - Double inlet LV
  - Tricuspid atresia
- Single Right Ventricles
  - HLHS
  - DORV/MA
- Other Single Ventricles
  - Unbalanced AVC

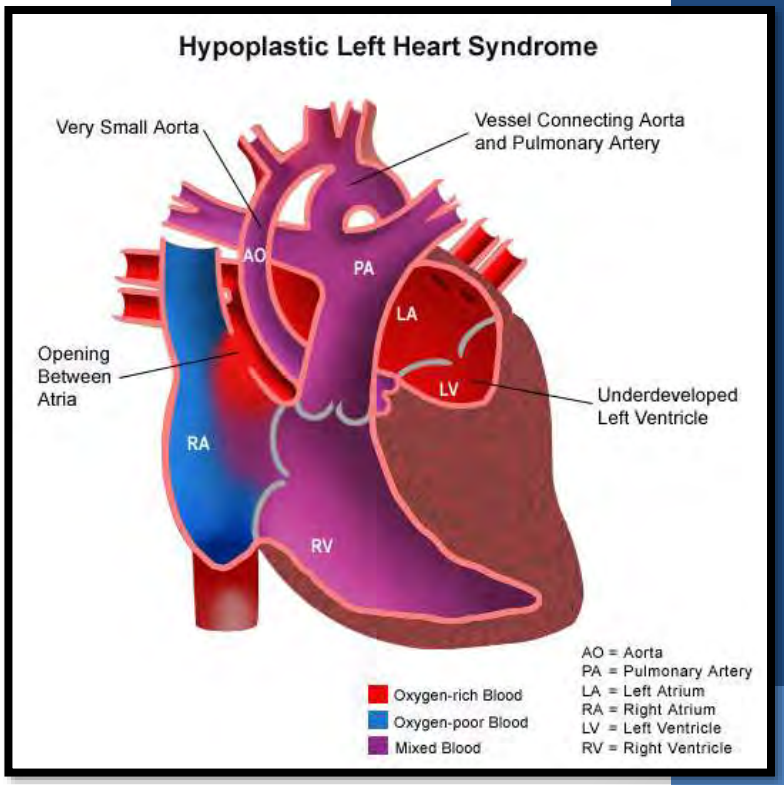


All follow common pathway

Single Ventricle

Hypoplastic Left Heart Syndrome (HLHS)

- Critical left heart underdevelopment
  - Mitral & aortic stenosis/atresia
  - Aortic arch hypoplasia
  - Ductal dependent systemic blood flow



Palliation: **Single Ventricle Pathway**

1. Stage I (Norwood Repair)
2. Stage II (Glenn Operation)
3. Stage III (Fontan Completion)

Fontan pathway is for single ventricles

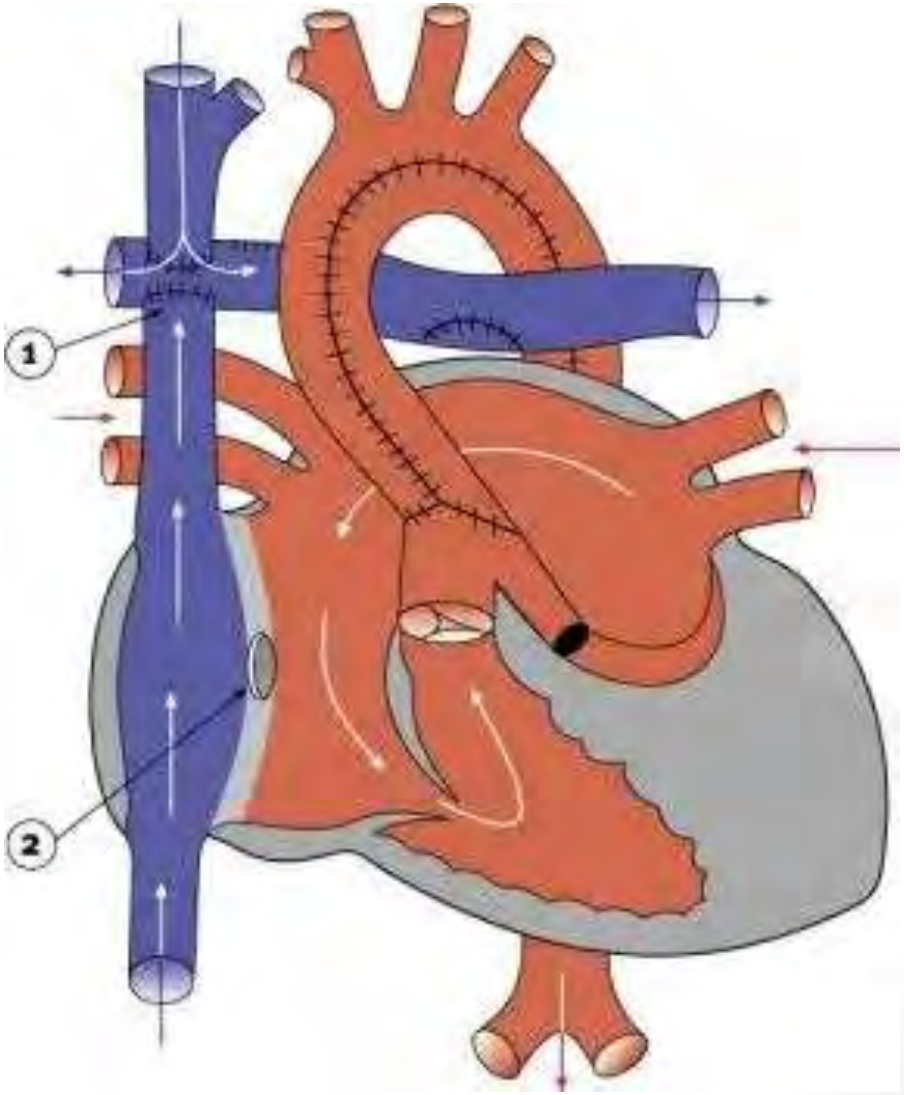
Fontan

IVC to PA connection

With or without fenestration

Starts the clock ticking for Fontan complications

- Heart Failure
- Fontan Failure
- Associated Issues
  - Liver failure
  - Thrombus
  - Protein losing enteropathy
  - Complex arrhythmias



High Risk ACHD

Pregnancy

- Changes in Hemodynamics
  - Substantial increase in cardiac output
    - HR ↑, SVR and PVR ↓, 02 consumption ↑
  - 30-50% rise in plasma volume & relative anemia
- Peaks at 32 weak pregnancy (30-50% +)
  - Cardiac output after 32nd week can be positional

Delivery makes it worse

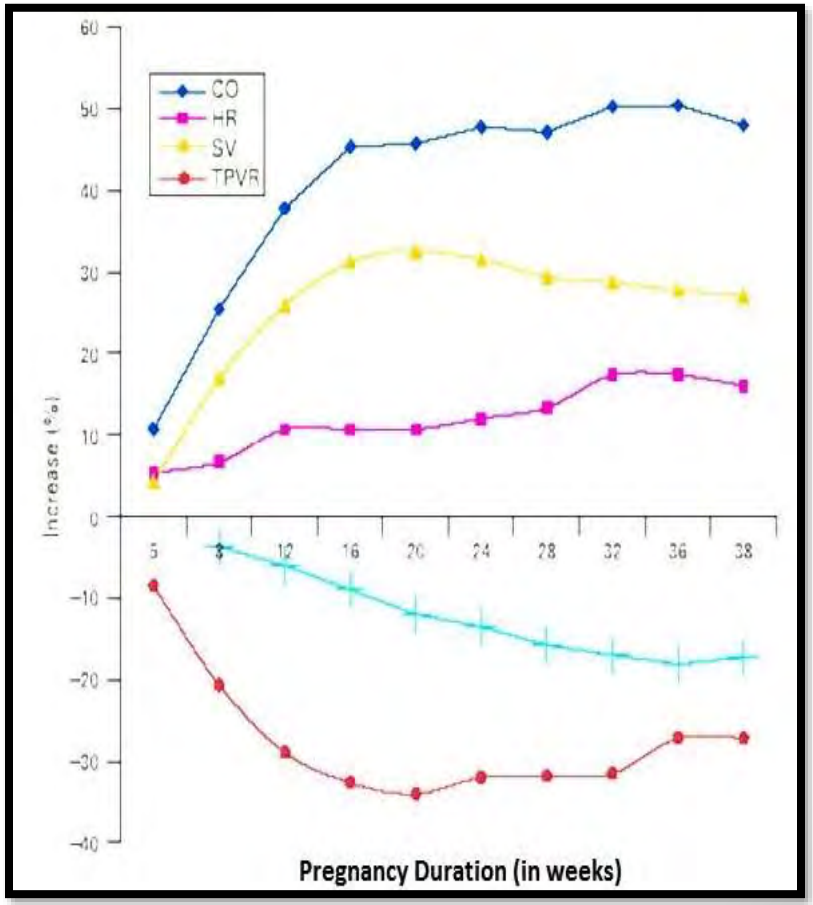
- Contractions release 300-500cc blood
- CO drastically increases (preload & HR)
  - Pain & stress increase SVR & PVR
- Results in <u>80%</u> increase in CO (up to 9L/min)

High Risk ACHD

Who do we worry about?

Those who can't achieve the graph

1. Hemodynamic challenges
2. Medical Challenges
   - Prosthetic valves
3. Poorly defined risks
   - Big aortas
   - Fontans



High Risk ACHD

ACHD Clinic-Risk stratification/Planning

Prior to pregnancy!

- Clinical assessment of exercise ability
- Careful assessment of hemodynamic issues
  - Echocardiography, MRI or CT, Catheter
- Pregnancy/Delivery Planning
  - Monitoring throughout pregnancy
  - Careful delivery plan with OB-GYN
  - Fetal assessment of child for CHD
  - Anesthesia plan with anesthesiologist

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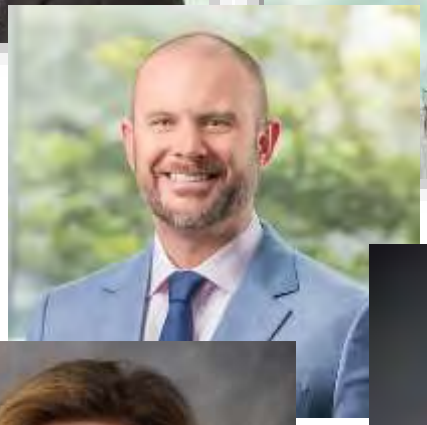
ACHD Physicians



ACHD Cardiologist

- Jonathan Cramer, MD
  ACHD and Imaging
- Shane Tsai, MD
  EP and ACHD
- Anji Yetman, MD
  ACHD & Connective Tissue
- Vincent Gonzalez, MD

Congenital Surgery

- Ali Ibrahimiye
- Camille Hancock-Friesen

Other Team Members

- Cardiac Cath/Intervention
- ACHD EP
- Nurse Practitioners
- Nurse Coordinators

Addressing the ACHD Issue

Accessibility: What are the resources?

General ACHD Clinics

- Children's Nebraska
- Nebraska Medicine
- Outreach (Lincoln, Rapid City, Kearney, Sioux City)

Specialty Clinics

- Transition services
- Eisenmenger's clinic (UNMC)
- Marfan & Aortopathy clinics

Take Home Points

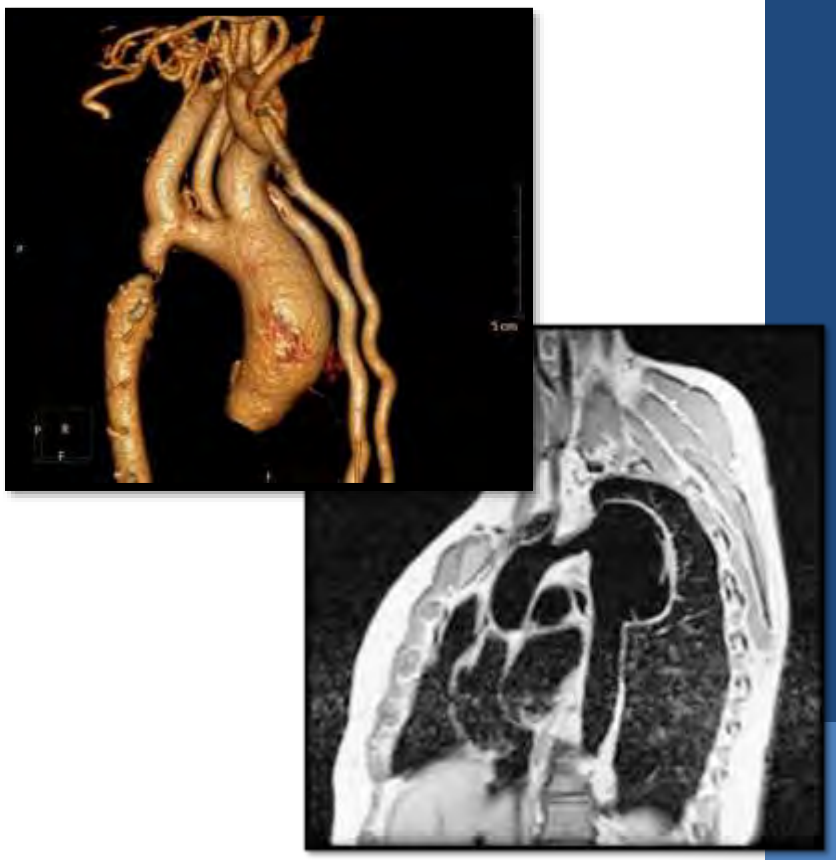
1. Adults with congenital heart disease are here and undergoing serious growth
2. ACC, AHA, & other bodies have identified ACHD as a poorly served population. These bodies have made appropriate CHD care a priority at may levels.
3. Nebraska has 4 board certified/board eligible ACHD physicians; with a regional center of excellence since 2017
4. Know when and how to engage the ACHD team

When to Reach Out

1. All unrepaired CHD of any age
2. Repaired CHD > IB
3. Aortopathies or CTD
4. Pregnant ACHD

<u>KEY Rules of Thumb for ACHD</u>

- Patients don't know their history
- Asymptomatic ≠ Good
- Refer
  - Anyone with a surgical scar
  - Anyone followed by a Ped Cards



Thank-you