

The Neonate with Heart Disease: Initial ED Presentation



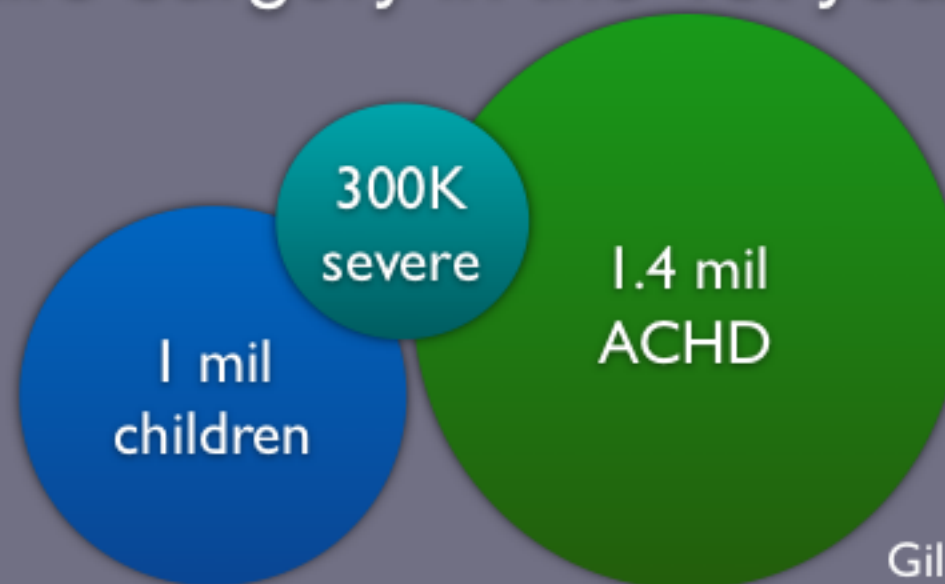
Robert Elder, MD

September 12, 2019



Epidemiology

- CHD occurs in 8/1000 live births
- The most common birth defect
- About 1/4 of infants with CHD will require surgery in the 1st year of life



Fetal echo

- Detect >60% CHD
- Indications
 - CHD
 - Maternal DM
 - Certain maternal exposures
 - Metabolic (PKU)
 - Lupus
 - Rhythm

CCHD screen

- Sept 2011
- Aug 2015: 46 states +
- Done at >24 hrs age but prior to DC
- RA and leg sat

TABLE 1 Conditions Detected Via Screening for CCHD V

Core conditions (CCHD)

- Coarctation of the aorta
- Double-outlet right ventricle
- Ebstein's anomaly
- Hypoplastic left heart syndrome
- Interrupted aortic arch
- Pulmonary atresia
- Single ventricle (not otherwise specified)
- Tetralogy of Fallot
- Total anomalous pulmonary venous return
- D-transposition of the great arteries
- Tricuspid atresia
- Truncus arteriosus
- Other critical cyanotic lesions not otherwise specified

Secondary conditions (non-CCHD)

- Hemoglobinopathy
- Hypothermia
- Infection, including sepsis
- Lung disease (congenital or acquired)
- Noncritical congenital heart defect
- Persistent pulmonary hypertension
- Other hypoxemic condition not otherwise specified

Evaluation of a Screening Program to Detect Critical Congenital Heart Defects in Newborns

Rachel Klausner, BS, Eugene D. Shapiro, MD, Robert W. Elder, MD, Eve Colson, MD, MHPE, Jaspreet Loyal, MD, MS

OBJECTIVES: To report the results of and to identify problems with implementing a screening program to detect critical congenital heart defects (CCHDs) in newborns by using differential pulse oximetry (POx).

ABSTRACT

METHODS: Charts of all live-born infants from 4 Yale–New Haven health system hospitals in Connecticut between January 1 and December 31, 2014, were reviewed.

RESULTS: Of 10 589 newborns, 171 (1.6%) underwent an echocardiogram before screening, 10 320 (97.5%) were screened by POx, and 98 (0.9%) were not screened. Thirteen newborns (0.1%) were diagnosed with a CCHD. No infants with CCHDs were identified through POx screening (POxS) alone. Eleven (85%) were already suspected of having a CCHD lesion on the basis of prenatal ultrasound, 1 (8%) was diagnosed because of clinical concern before undergoing screening, and 1 (8%) had a false-negative screening result, but a CCHD was identified after an echocardiogram was performed because a murmur was heard. Four infants with a positive POx screen showed noncritical cardiac lesions by echocardiogram. The majority of infants were screened within the recommended 24 to 72 hours of age interval and had POx screens that were interpreted and documented correctly. Of 10 316 infants with negative POx screens, 52.1% were still in the Yale–New Haven Hospital health system at 1 year of age and no CCHD lesions were listed in their charts.

What might you see...

- I. Left to right shunts
- II. Left heart obstructive lesions
- III. Cyanotic lesions
- IV. Others

I. Left to Right Shunts

Left to Right Shunting Lesions:

- Ventricular Septal Defects
- Atrial Septal Defects
- Atrioventricular Septal Defects
- Patent Ductus Arteriosus
- Aorto-Pulmonary Window

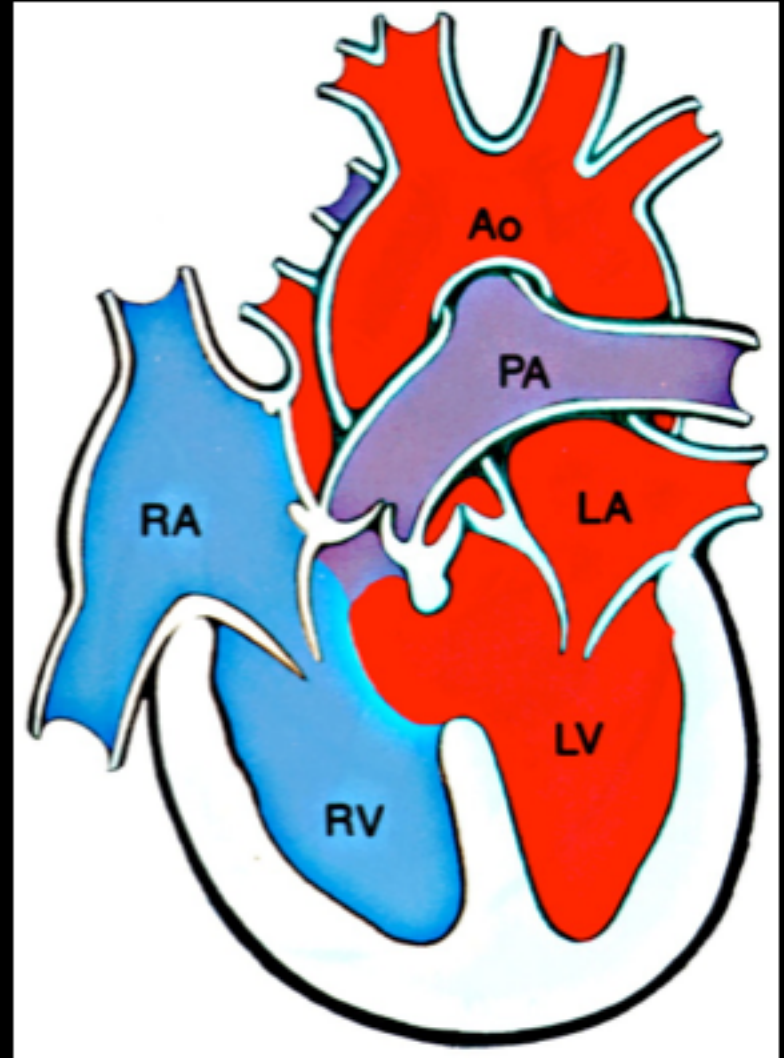
Ventricular Septal Defects

Most common form of CHD

- 20% of all CHD
- Associated with other defects, syndromes, diseases
- May be single or multiple
- May be small or large

VSD - Physiology

- shunt from high to low pressure ventricle
- Fall of PVR, nadir around 6 weeks



II. Obstructive Lesions

Left Heart Obstruction:

- Mitral Stenosis
- Subaortic Stenosis
- Aortic Stenosis
- Coarctation of the Aorta
- Hypoplastic Left Heart Syndrome

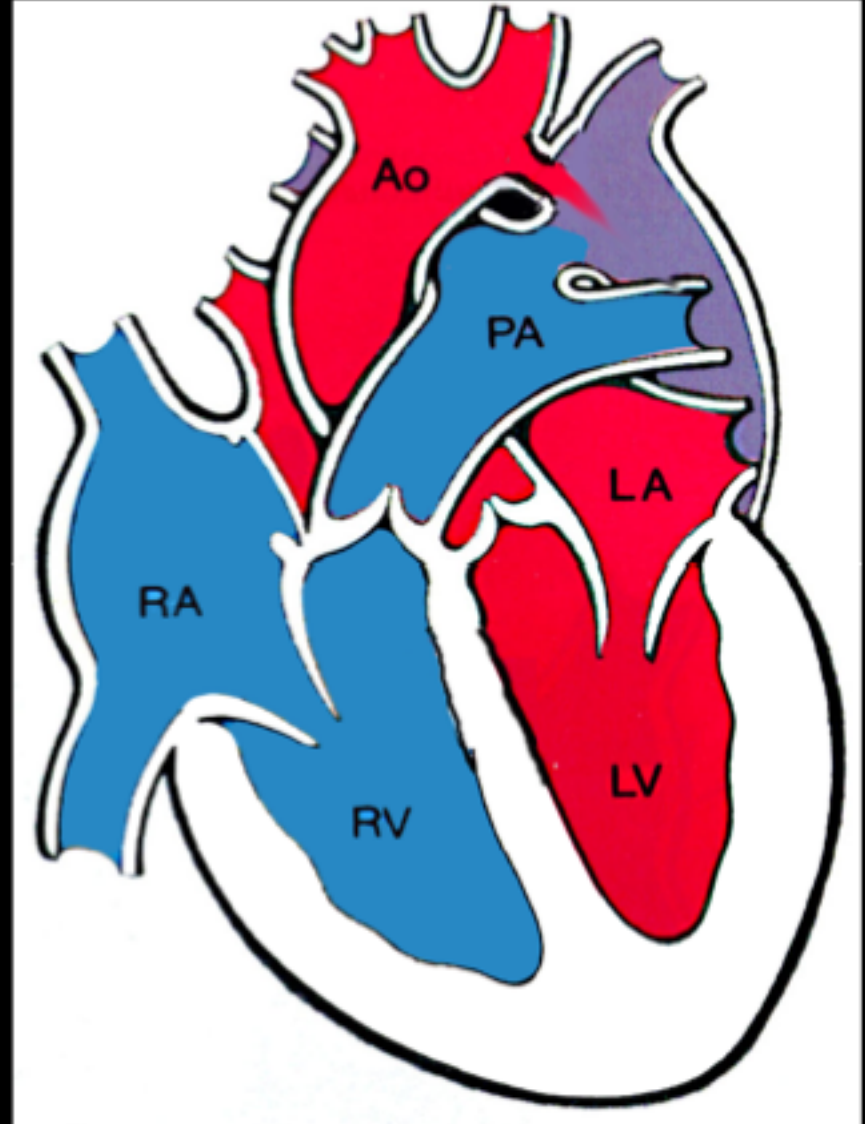
Coarctation of the Aorta

Coarctation of the Aorta

- 6th most common CHD
- 15-25% of Turners (45 XO) patients
- Poor Isthmus Flow in Utero?
- Discrete Ring - Juxtaductal
- Aortic Arch Hypoplasia
- Associated With:
 - Bicuspid aortic valve (> 50%)
 - Ventricular Septal Defects

Coarctation - Physiology

- Increased afterload on LV
- Hypertension of LV and upper body
- Left ventricular hypertrophy
- Decreased perfusion of splanchnic and lower body somatic circulation



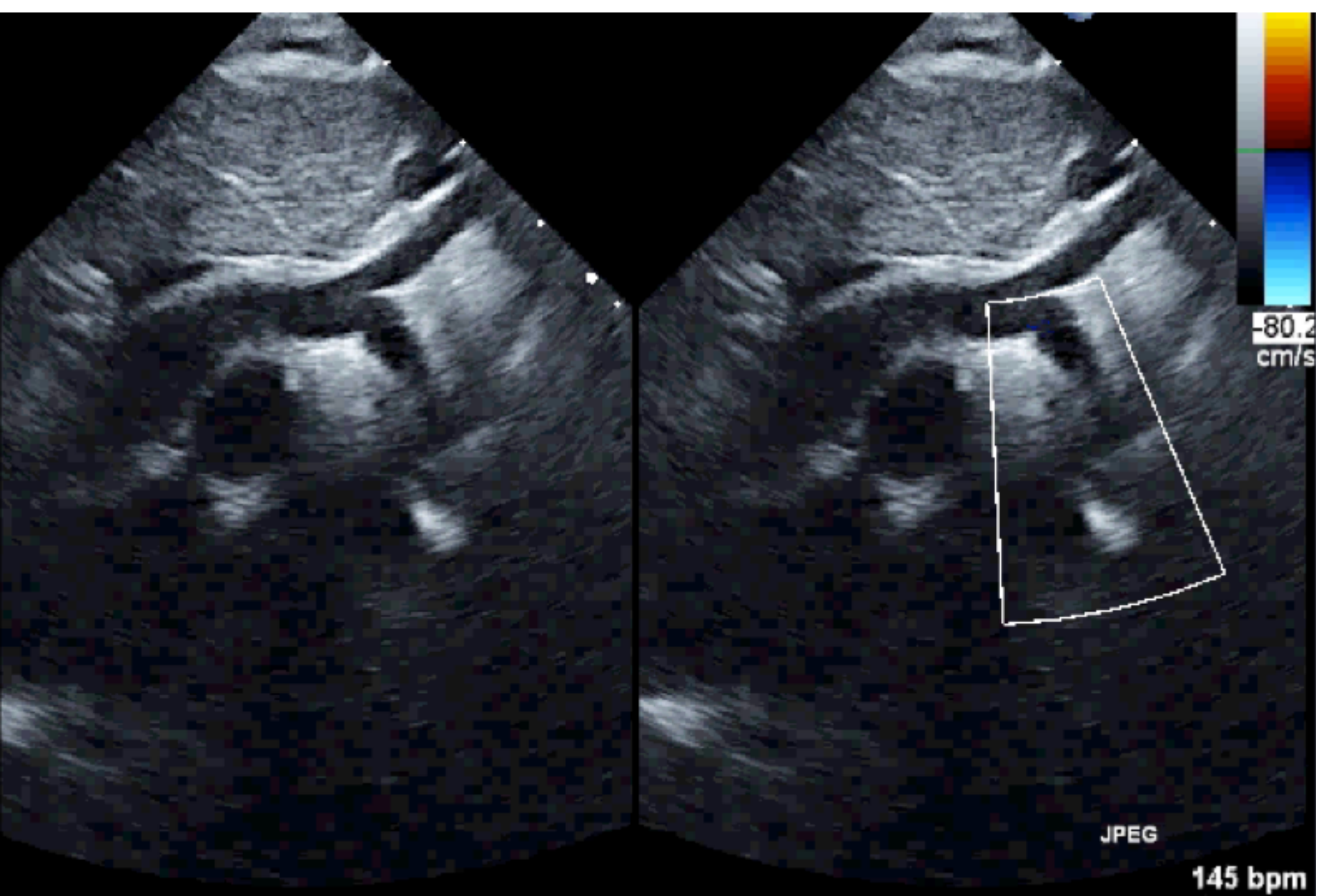
Coarctation - Presentation

- Discrepant pulse
 - radial - femoral impulse quality
 - radial - femoral delay
- Murmur over the back
- CHF
- Shock - cardiovascular collapse

Coarctation - Diagnosis

Clinical: with your fingers
and your ears

Confirm with the echo



Coarctation: Management

- Infant:
 - PGE until surgery
 - Supportive care
 - End to end anastomosis

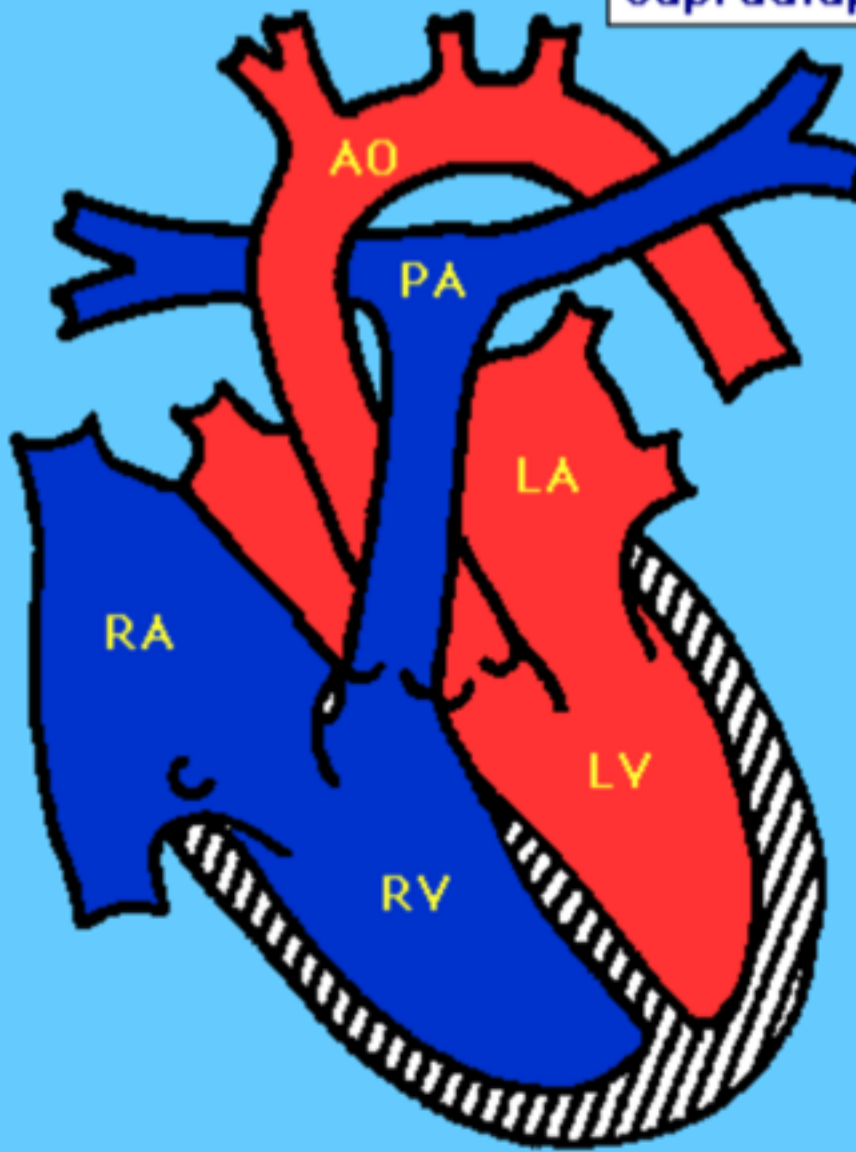


III. Cyanotic Heart Disease

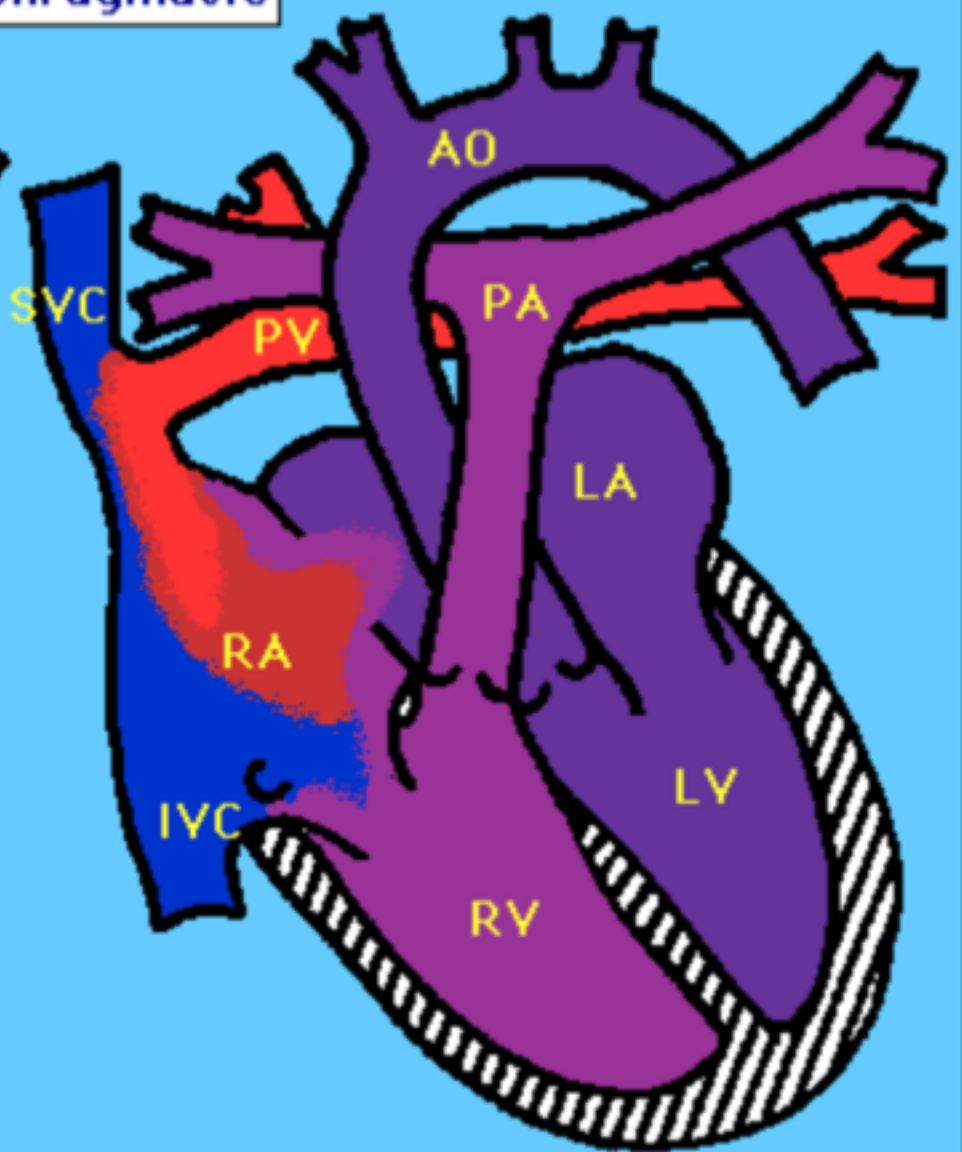
5 T's of Cyanotic Heart Disease:

- Tetralogy of Fallot (TOF)
- Transposition of the Great Arteries (TGA)
- Tricuspid Atresia
- Truncus Arteriosus
- Total Anomalous Pulmonary Venous Return (TAPVR)

Supradiaphragmatic



Normal



TAPVR

IV. Other

- Arrhythmia
- Myocarditis
- Cardiomyopathy (metabolic disease)

Summary

- Good quality fetal echo detects the majority of CHD
- CCHD screening detects most others
- Be on the look out for special cases (coarctation, TAPVR)
- New onset conditions (arrhythmia, myopathies) can be seen in infants

Questions?

QUESTIONS:

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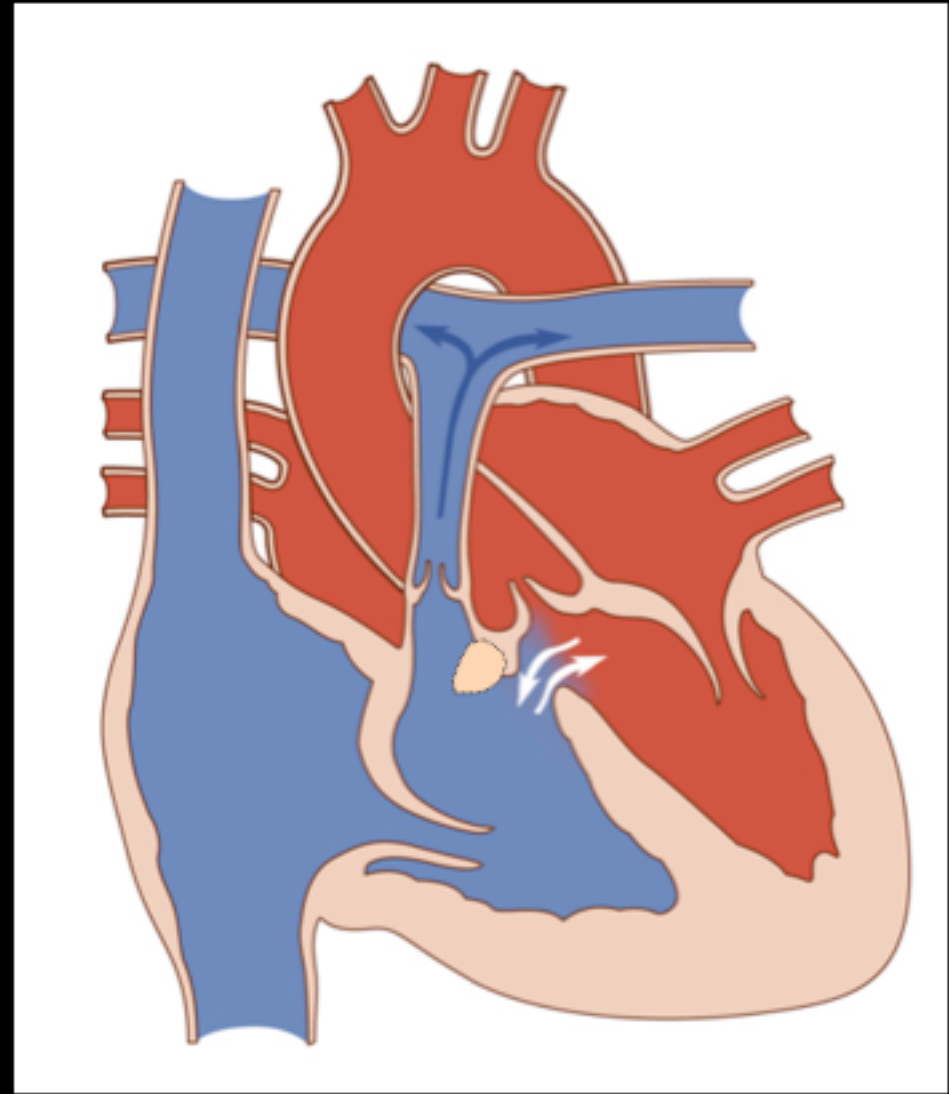
Associate Professor of Pediatrics and
Internal Medicine

203-785-2022

robert.elder@yale.edu

TOF - Physiology

- Degree of cyanosis results from amount of pulmonary blood flow
- Murmur reflects pulmonary blood flow
 - no murmur = no flow
- Spectrum



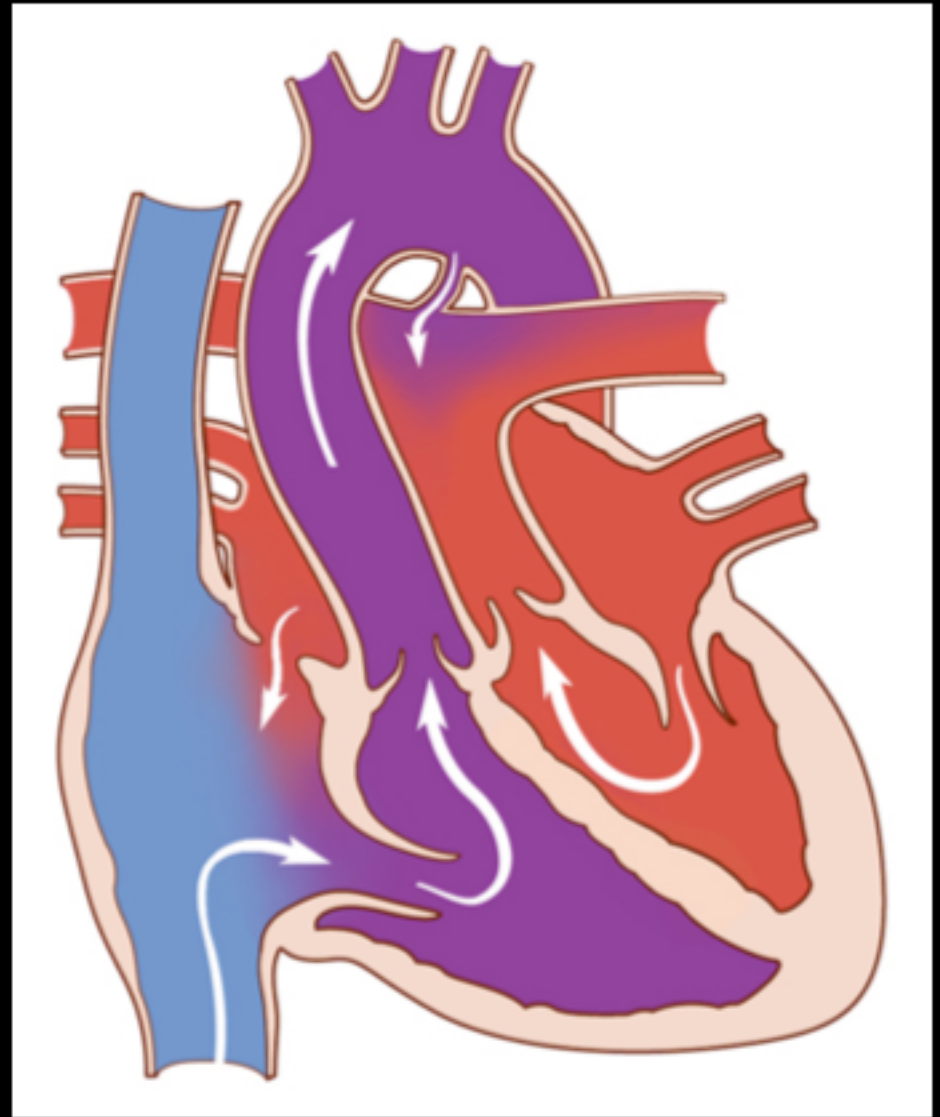
TOF - Presentation

Clinical Manifestations:

- Most patients present with murmur rather than cyanosis
- Progressive cyanosis
- Single S2, loud murmur
- Cyanotic “Tet” spell

TGA - Physiology

- Parallel Circulations
- Shunting
 - Ductus Arteriosus
- Mixing
 - Atrial Septum (PFO, ASD)
 - Ventricular Septum if VSD



TGA - Management

- PGE: maintain shunt at PDA
- Balloon Atrial Septostomy: maintain atrial mixing
- Surgical: Arterial Switch Operation 98% survival

