APPROACH TO CONGENITAL HEART DISEASE: ACYANOTIC CONGENITAL HEART DISEASE

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OUTLINE

ABBREVIATION	MEANING
CHD	Congenital Heart Disease
BAV	Bicuspid Aortic Valve
ASD	Atrial Septal Defect
VSD	Ventricular Septal Defect
CAVSD	Complete Atrio-Ventricular Septal Defect
PDA	Patent Ductus Arteriosus
PS	Pulmonary Stenosis
AS	Aortic Stenosis
CoA	Coarctation of the Aorta
ECG	Electrocardiogram
SBP	Systolic blood pressure
DBP	Diastolic blood pressure
CXR	Chest X-Ray
RV	Right ventricle
LV	Left ventricle
RVH	Right ventricular hypertrophy
LVH	Left ventricular hypertrophy
RAD	Right axis deviation
LAD	Left axis deviation
MPA	Main pulmonary artery
IRBBB	Incomplete right bundle branch block
PHTN	Pulmonary hypertension
LUSB	Left upper sternal border
TOC	Treatment of choice
PFO	Patent Foramen Ovale
PVR	Pulmonary Vascular Resistance
SVC	Superior Vena Cava

I. INTRODUCTION

- Heart disease
  - Congenital – acyanotic or cyanotic
    - TGA is the most common cyanotic CHD
    - TOF is the cyanotic CHD with the highest survival rates
  - Acquired

- Review of Blood Flow to the Heart (Figure 1)
  - Deoxygenated blood flows to the RA through the IVC and SVC
  - Blood flows from the RA to RV through the tricuspid valve
  - Blood flows from the RV to the lungs through pulmonary arteries
  - Blood becomes oxygenated by the lungs
  - Oxygenated blood flows back to the heart in the LA through the pulmonary veins
  - Blood flows from the LA to the LV through the mitral valve
  - Blood is pumped to the systemic circulation from the LV through the aorta

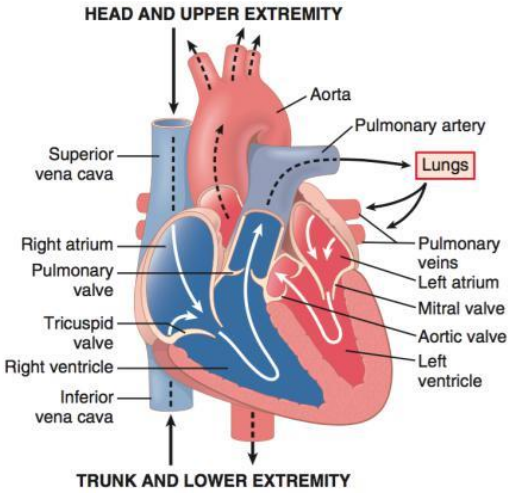

Figure 1. Structure of the heart and course of blood flow through the heart chambers and heart valves. (Hall, 2016)

II. CONGENITAL HEART DISEASE (CHD)

- An anatomic malformation of the heart and/or its vessels
- Occurs during intrauterine development
- Heart is typically formed around week 4
  - A pregnancy test is taken around week 6-8 weeks

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A. Incidence

- 3 to 7 per 10,000 live births
- The following are excluded from overall incidence:
  - **Bicuspid aortic valve (BAV)** – 1 to 2%
    - Most common 'congenital valve disease'
    - The majority are asymptomatic but many will develop into stenosis with age due to calcification. Adverse cardiovascular outcomes in patient with BAV are more common than previously thought
  - **Patent ductus arteriosus (PDA) of preterm infants**
    - In utero, this is normal but is supposed to close upon delivery (**ligamentum arteriosum**)
    - Considered pathologic if it does not close at 6 months
    - Exception: preterm babies who are merely considered to have a "patent ductus arteriosus" – 'semantics'
  - **Patent foramen ovale:** 25%
    - Most common structural abnormality of the heart
    - Opening found between left and right atria
    - Similar to PDA, most patients are asymptomatic until adulthood
    - Only indicated for corrective surgery if the patient undergoes a stroke

B. Indicators

- **Cyanosis**
  - Lack of oxygen
  - Normally oxygen saturation >96%
  - 88-95% is already considered to be oxygen deficient
- **Cardiomegaly** – radiologic or pericardial bulge
- **Pathologic heart murmur**
  - There may be presence of physiologic murmurs; quite difficult to distinguish
  - Absence of murmurs also doesn't mean absence of disease
- **Tachypnea / Overt respiratory distress** (Dyspnea)
  - Fast breathing or breathing with effort
  - Difficult to assess among pediatric patients
- **Sweating during feeding** (Diaphoresis)
  - Breastfeeding is the baby's form of exercise
  - Feeding compounded with CHD makes it even more effortful
- **Intermittency of feeding**
  - Normally, a baby can finish a whole bottle in 5-10 minutes
  - Baby with CHD will keep swallowing until s/he cannot breathe, at which point s/he stops to breathe
  - Swallow → stop → breathe → swallow → stop → breathe
- **Increased or decreased pulses**
  - Peripheral pulses
  - Reflects the heart's pumping power
  - Pulse pressure
    - Difference between diastolic and systolic pressure
    - Wider pulse pressure = more prominent pulse
  - Decreased pulses in the lower extremities = most indicative of CoA
- **Failure to thrive**
  - Growth is stunted
  - Poor weight gain

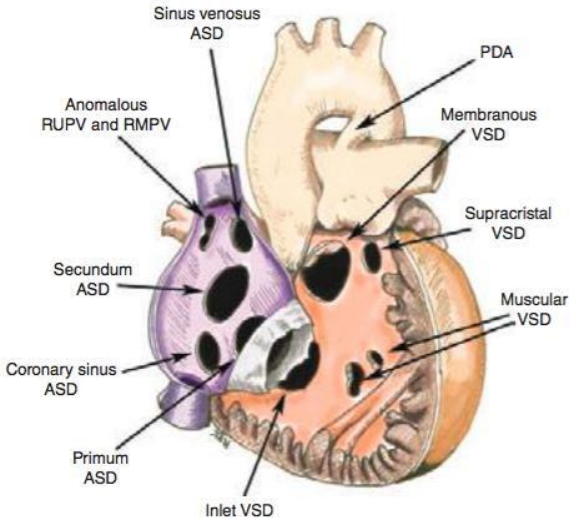

Figure 2. Types and locations of congenital cardiac defects. ASD, atrial septal defect; PDA, patent ductus arteriosus; RMPV, right middle pulmonary vein; RUPV, right upper pulmonary vein; VSD, ventricular septal defect. **(Kasper *et al.*, 2015)**

C. Heart Disease Considerations

- **Shunt anomalies**
  - Left to right
  - Right to left
- **Valve defects**
  - **Atresia**: complete block
  - Insufficiency / Regurgitation
    - Failure to close
    - Causes volume overload
  - Stenosis
    - Failure to open
    - Causes pressure overload

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- **Obstruction to flow of the great vessels**: obstruction to the aorta and pulmonary vessels
- **Abnormal connections of the great vessels**
  - Normal: 4 pulmonary veins normally drain the left atrium
  - Causes anomalous pulmonary venous return when they are connected wrongly
  - e.g. TGA
- **Congenital coronary abnormalities**: very rare
- **A combination of the above**

D. Approach to Congenital Heart Disease

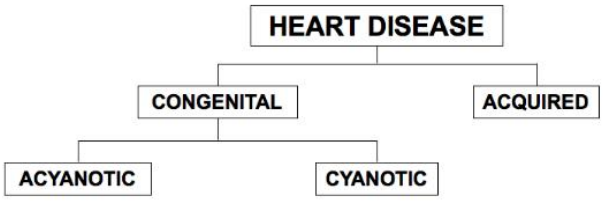

Figure 3. General algorithm for the two types of heart disease

- Determine which type of CHD it is:
  - Acquired heart disease
  - Congenital heart disease
- History and PE remains to be the best way to diagnose CHD

History

- Maternal and family history
- Age of onset of symptoms
  - History of the heart disease
    - Disease manifests when the patient is young
      - Nature of the heart disease is most likely **congenital**
    - If the disease manifests when the patient is a teenager or older
      - Nature of the heart disease is most likely **acquired**
    - Exceptions:
      - CHDs with late onset
      - Acquired heart disease with early onset

Physical Examination

- Chromosomal syndromes
- Cyanosis, murmur, heart failure – probably congenital
- Complete cardiac PE

E. Types of Congenital Heart Disease

Table 1. Types of CHD according to type of shunt and lesion

Left-To-Right Shunts (40%)	Atrial Septal Defect (ASD) Ventricular Septal Defect (VSD) Common Atrioventricular Valve Defect (CAVSD) Patent Ductus Arteriosus (PDA)
Obstructive Lesions (25%)	Pulmonary Stenosis (PS) Aortic Stenosis (AS) Coarctation of the Aorta (CoA)
Right-To-Left Shunts (20%)	Tetralogy of Fallot (TOF) Transposition of the Great Arteries (TGA) Tricuspid Valve Atresia / Pulmonary Atresia
Others (15%)	

III. ACYANOTIC CHD: Left-to-Right Shunts

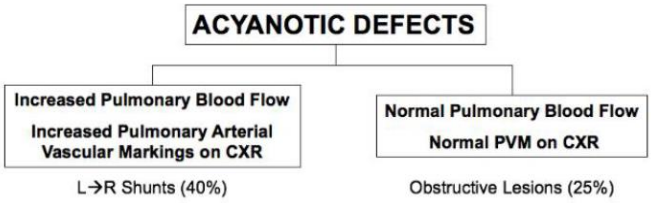

Figure 4. Acyanotic CHD Categories.

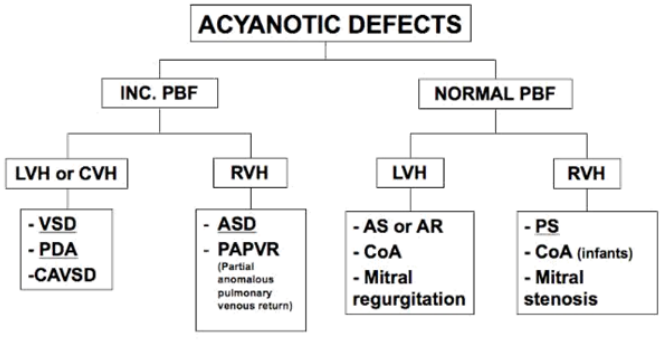

Figure 5. CHDs associated with pulmonic blood flow

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A. Atrial Septal Defect

- Abnormal, fixed openings in the atrial septum that allow communication of blood between the left and right atria
- Types:
  - Primum – worst prognosis
  - Secundum – most common
  - Sinus venosus
- Hypertrophy progresses slower because of the lower pressures in each chamber (See Figure 7 for pressure values)
- Symptoms appear later in life, ≥ 30 years old

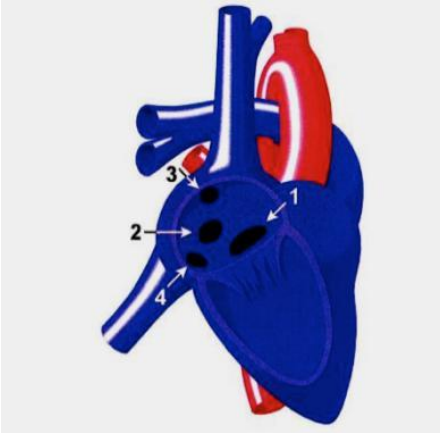

Figure 6. Atrial Septal Defect.
1: primum, 2: secundum, 3: superior sinus venosus, 4: inferior sinus venosus

Pathophysiology

- 100 mL of blood travels the normal path until it reaches the LA (Figure 7)
- A small amount of blood is redirected from the LA back to the RA via the defect in the atrial septum
  - 10 mL goes back to the RA
  - 90 mL goes to the aorta and systemic circulation
  - Because blood flows to the path of least resistance, it goes back to the RA where "there is less pressure [than the LA]"

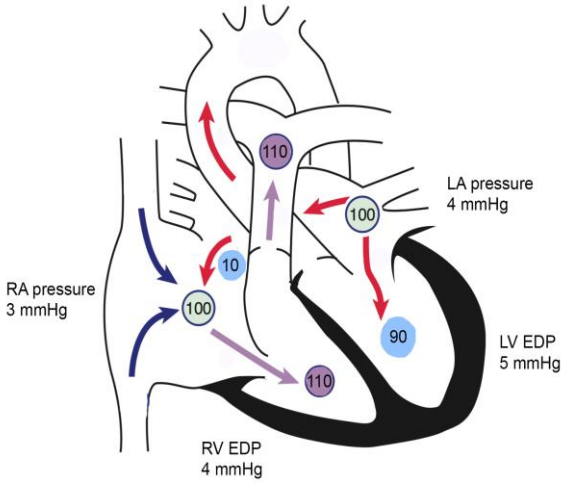

Figure 7. Atrial Septal Defect Pathophysiology. The numbers in green circles show the initial blood flow. The numbers in blue circles show the redirected blood flow through the ASD. The numbers in purple circles show the new blood flow. See the accompanying text above. **(Kliegman *et* al., 2015)**

Hemodynamic Consequences

- Diastolic/ Volume overload of RV
- RA, RV dilation
- Lung congestion

Physical Examination

- Usually asymptomatic, with late presentation
  - "Like a drop filling up a bucket"
  - Pressure gradient between atria is small, hence volume shunt is small as well
- Distended neck veins – seen in adults only
- Fixed split S2
- Soft systolic murmur across the pulmonary valve
- Diastolic rumble across the pulmonary valve

Diagnostic Modalities

- ECG
  - Normal to right axis-deviated
  - Incomplete right bundle branch block (IRBBB) pattern, RVH
- CXR
  - Mild RV cardiomegaly
  - RVH with hypervascular markings – automatically ASD
  - Dilated main PA

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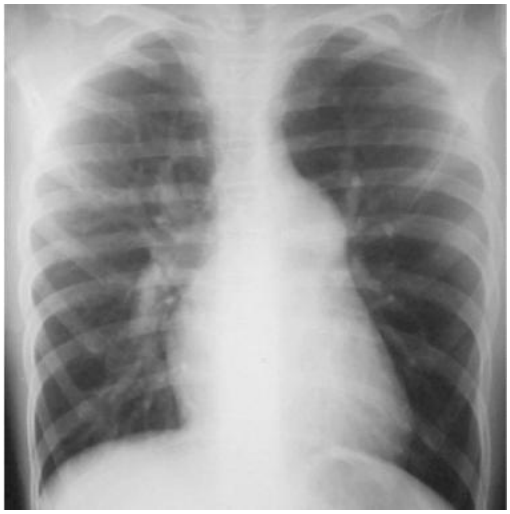

Figure 8. CXR of atrial septal defect.

Treatment

- Surgery
- Non-surgical closure

B. Ventricular Septal Defect

- Incomplete closure of the ventricular septum
- Most common cardiac malformation
  - Accounts for 25% of CHD (Kliegman et al., 2016)
  - Accounts for 42% of CHD (Kumar, Abbas, and Aster, 2015)
    - Take note: This number is based on a 2002 study (see Robbins page 531)

Pathophysiology

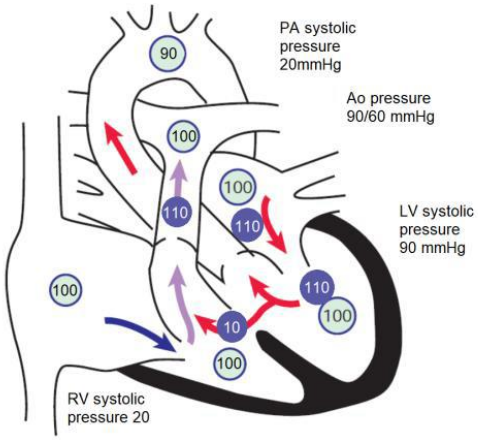

Figure 9. Ventricular Septal Defect Pathophysiology. The black numbers in green circles represent the initial blood flow. The white numbers in blue circles follow the small amount of blood that passes through the VSD. See the accompanying text below. **(Kliegman *et al.*, 2015)**

- An original volume of deoxygenated blood (e.g. 100 mL, green circles) enters the RA, and thus the same volume is expected to enter the RV
- As the RV undergoes ventricular systole to pump deoxygenated blood towards the lungs, the RA will be undergoing atrial diastole in order to refill with blood
- Oxygenated blood flows from the lungs into the LA through pulmonary veins, then to the LV through the mitral valve
- **During left ventricular systole:**
  - Majority of the original blood volume (e.g. 90 mL) flows to systemic circulation through the aorta
  - A small portion of this blood (e.g. 10 mL, blue circle) flows back to the pulmonary arteries
  - Does not "go" to the RV because both ventricles are contracting, then squeezing the blood directly to the PA, which has less resistance / pressure than the RV
    - The PA is the "path of least resistance" because the pressure inside during systole is 20 mmHg
- Since the blood pumped to the systemic circulation is <100 mL, this triggers hematopoiesis to restore the volume back to normal
- Thus, 100mL keeps returning to the right atrium but more and more blood is stuck in the lungs → pulmonary congestion / edema
- The right ventricle undergoes progressive right ventricular hypertrophy in response to increased pulmonary vascular resistance
  - When pulmonary vascular resistance approaches systemic levels, the original left-to-right shunt becomes a right-to-left shunt (**Eisenmenger syndrome**) (Kumar *et al.*, 2015)

Hemodynamic Consequences

- Total blood volume is increased in these patients, producing the following hemodynamic consequences
- **Small-sized hole** (Less than ⅓ of the diameter of the aorta OR 1-2mm OR less than 5mm (Kliegman et al., 2016)
  - No pressure or volume overload
  - Too small to be significant
- **Moderately-sized hole** (Less than ½ of the diameter of the aorta)
  - Volume overload of LV

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- Large-sized hole (Greater than ½ of the diameter of the aorta or greater than 10 mm (Kliegman et al., 2016)
  - Volume overload of LV
  - Pressure overload of RV

Natural History

- **Location of defect**
  - Muscular and perimembranous have high incidence of spontaneous closure
- **Size of defect**
  - Small
    - Tend to be asymptomatic
    - Normal growth and development
    - No CHF
    - More common compared to other sizes (Kliegman et al., 2016)
  - Moderate to Large
    - Easy fatigability, intermittent feeding
    - Delayed growth and development
    - Repeated respiratory tract infection
    - More likely to develop CHF sooner
  - The larger the hole, the more symptoms there will be and the younger the onset will be
- **Pulmonary vascular resistance**
  - Pulmonary HPN or high resistance will limit total shunt flow

Physical Examination

- Pansystolic or holosystolic murmur at left lower parasternal border
  - Characteristic differs depending on the defect size
- Small VSD
  - Cardiac chambers are not appreciably enlarged
  - Pulmonary vascular bed is probably normal
  - Loud, harsh, or blowing holosystolic murmur accompanied by a thrill
- Moderate to large defects
  - LV, LA, and lungs are enlarged
    - Dynamic and bulging left precordium
  - Lung findings
    - Increased pulmonary vascular markings
    - Congestion
  - Signs of CHF
    - Cyanosis is usually absent, but duskiness is sometimes noted during infections or crying
  - Palpable parasternal lift
  - Laterally displaced apical impulse and apical thrust
  - Systolic thrill
  - Less harsh holosystolic murmur compared to small defects
  - Normal to increased P2 because of pulmonary HPN

Diagnostic Modalities

- **ECG**
  - Small: normal
  - Moderate to large: LVH to CVH
  - Severe PHTN: RVH
- **CXR**
  - Normal to cardiomegaly (LV, RV)
  - Dilated MPA; hypervascular markings
- **Treatment** (Kliegman et al., 2016)
  - Small
    - 30-50% close spontaneously during the first two years of life
    - Up to 80% of small muscular VSDs close and up to 35% of membranous VSDs close
    - Small, hemodynamically insignificant VSD (Qp:Qs <1.5) is not an indication for surgery
  - Large
    - Catheter or surgical closure indicated for patients at any age and if with clinical symptoms and failure to thrive that cannot be controlled medically

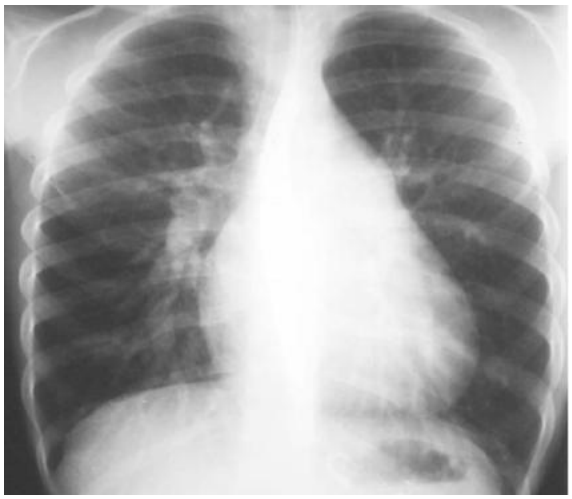

Figure 10. CXR of a patient with small VSD

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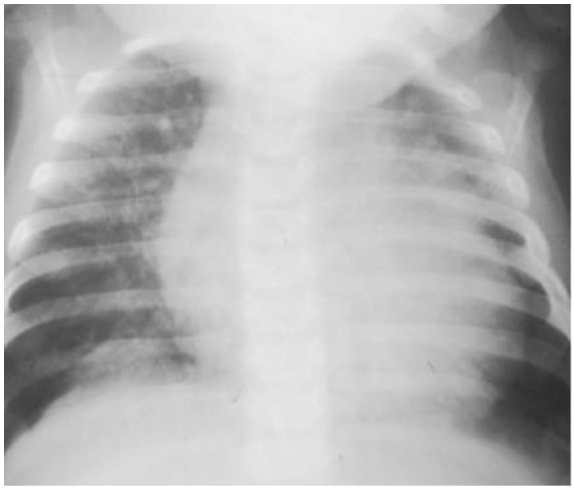

Figure 11. CXR of a patient with large VSD.

Table 2. Summary of VSD

	SMALL	MODERATE	LARGE
Size compared to aortic diameter (Infant to Adult)	Less than ⅓ (OR 1-2mm OR less than 5mm)	Less than ½	Greater than ½ (or usually greater than 10mm)
Hemodynamic Consequences	No pressure or volume overload Too small to be significant	Volume overload of LV	Volume overload of LV Pressure overload of RV
Natural History	Tend to be asymptomatic Normal growth and development No CHF	The larger the defect, the more quickly symptoms and CHF will develop Easy fatigability, intermittent feeding Delayed growth and development Repeated respiratory tract infection With CHF	
Physical Examination	Cardiac chamber not appreciably enlarged	LV and LA are enlarged Lungs are congested and have increased pulmonary vascular bed is probably normal Holosystolic murmur loud and harsh	pulmonary vascular markings There are signs of CHF (but no cyanosis) Holosystolic murmur is less harsh, there is a normal or increased P2 because of pulmonary HTN
ECG	Normal		LVH to CVH If with severe PHTN: RVH

C. Complete Atrio-Ventricular Septal Defect (CAVSD)

- a.k.a. **Endocardial cushion defect, Canal defect**
- Common in children with Down Syndrome (25%)
  - 50% of individuals with Down Syndrome have congenital heart disease
- 50% of these individuals have CAVSD
- Atrial and ventricular septa do not meet, which prevents septation of the common AV valve
- Basically a combination of VSD and a primum ASD
- Age of presentation is infancy

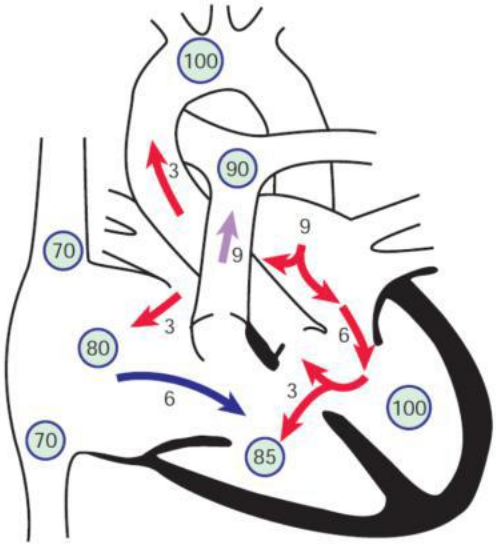

Figure 12. Blood flow in a hypothetical patient with Complete Atrioventricular Septal Defect. The encircled numbers represent OXYGEN SATURATION. The arrows indicate the direction of blood flow and the small numbers beside these arrows indicate the amount of blood in liters. **(Kliegman *et al.*, 2015)**

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Pathophysiology

- Blood is shunted from the left chambers to the right chambers
- Right ventricular pressure is equal to left ventricular pressure due to an unrestricted septal defect

Hemodynamic Consequences

- All chambers are enlarged
  - LA, LV and RV enlargement/hypertrophy – from the VSD aspect
  - RA and RV enlargement – from the ASD aspect
- Lungs
  - The pulmonary vascular markings are increased/ congested

D. Patent Ductus Arteriosus (PDA)

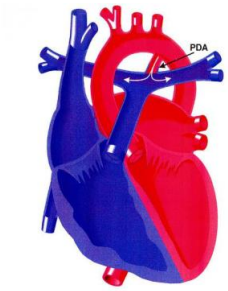

Figure 13. Patent ductus arteriosus.

- In fetal life, the ductus arteriosus shunts blood from the pulmonary artery to the aorta
- At birth, an increase in arterial oxygen concentration causes a decrease in the pulmonary vascular resistance and the beginning of the constriction of the ductus arteriosus
  - Functional closure occurs within the first 24 hours of life
  - Normal anatomic closure is complete by 2 weeks in 90%
- Persistence beyond 4-6 months is pathologic
- Two things to consider
  - The **size** of the PDA (diameter and length), which affects
  - The pressure gradient between the aorta and the pulmonary arteries

Pathophysiology

- In a theoretical patient, 100 mL of blood travels normally across the heart and lungs, and finally into the aorta (Figure 14)
- Some amount of blood (e.g. 10 mL) is shunted from the aorta back to the PA via the DA, while the remaining blood (e.g. 90 mL) continues its systemic circulation
  - The amount shunted in actuality depends on the size of the PDA
- 110 mL of blood enters the pulmonary arteries, lungs, LA, LV, aorta, and so forth

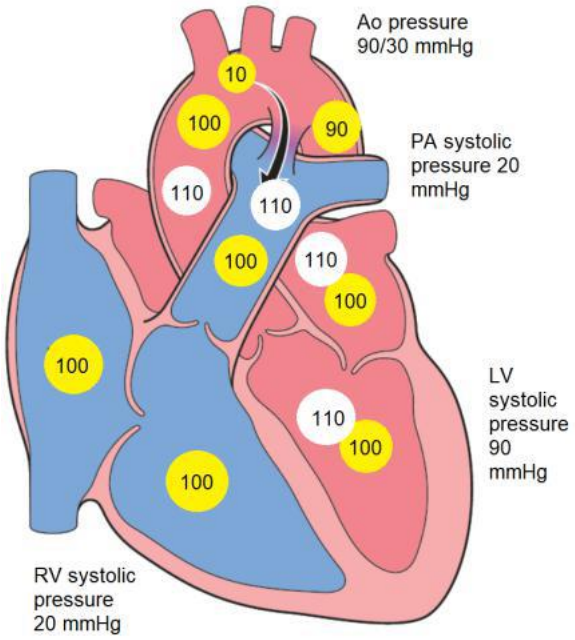

Figure 14. Patent ductus arteriosus in a hypothetical patient. The yellow circles represent blood entering the heart from the great veins in mL. The white circles represent the blood (mL) entering the pulmonary arteries via the PDA and its mixing with the next set of blood flowing in from the great veins.

Hemodynamic Consequences

- Moderate size
  - Volume overload of LV
- Large size
  - Volume overload of LV
  - Pressure overload of RV
- Consequences are similar to VSD

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Physical Examination

- **Small PDAs**
  - Associated with normal peripheral pulses
  - Normal size
- The following bullets refer to large PDAs
- **Wide pulse pressure**
  - Large SBP-DBP difference
  - Occurs due to runoff of blood into the pulmonary artery during diastole (Kliegman et al., 2016)
  - There is a difference between how much blood is pumped out by the LV and how much blood actually enters the systemic circulation
- **Bounding pulse**
  - Recall: larger pulse pressure, more prominent pulse
- **Cardiomegaly**
  - Apical impulse is prominent and is heaving
  - Thrill that is maximal in the 2nd left interspace, down the left sternal border, or toward the apex
  - Dynamic precordium
- **Soft to increased P2**
- **Continuous, machinery-like murmur**
  - **Pathognomonic for PDA**
  - May be localized to the 2nd intercostal space or radiate down the left sternal border or to the left clavicle
  - Occurs all throughout systole and diastole since aortic pressure is consistently higher than the PA, leading to continuous shunting

Table 3. Comparison of PDA and VSD

	PDA	VSD
Murmur	Continuous	Holosystolic
Location	Base	Apex
Pulse	Bounding	Normal
Pulse pressure	Wide	Normal

Diagnostic Modalities

- **ECG**
  - Normal
  - LVH, CVH, or RVH
- **CXR**
  - Normal
  - Cardiomegaly (LV, CV, or RV)
  - Dilated MPA; hypervascular markings
  - Dilated aortic arch

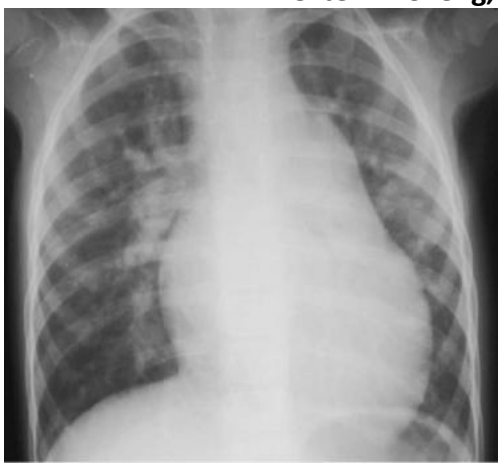

Figure 15. CXR of PDA.

Management

- Closure of the PDA via surgery or transcatheter
- Treatment should be done irrespective of age
- In patients with small PDA, closure is done to prevent bacterial endarteritis or other late complications
- In patients with moderate to large PDA, closure is done to prevent the development of pulmonary vascular disease, to treat heart failure, or both
  - Once the diagnosis of a moderate to large PDA is made, treatment should not be unduly postponed after adequate medical therapy for cardiac failure has been instituted - Kliegman et al., 2016

IV. ACYANOTIC CHD: Obstructive Lesions

A. Pulmonary Stenosis (PS)

- Malformation leading to obstruction at the level of the pulmonary valve
- 5-8% of congenital heart disease
- Associated with several diseases
  - Congenital rubella
    - Pulmonary stenosis
    - Also leads to sensorineural deafness, eye abnormalities, CNS defects (e.g. mental retardation), etc.
  - Noonan and William Syndrome

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- Manifestations are typically asymptomatic unless severe
  - Severe manifestations may produce right-sided heart failure in later years

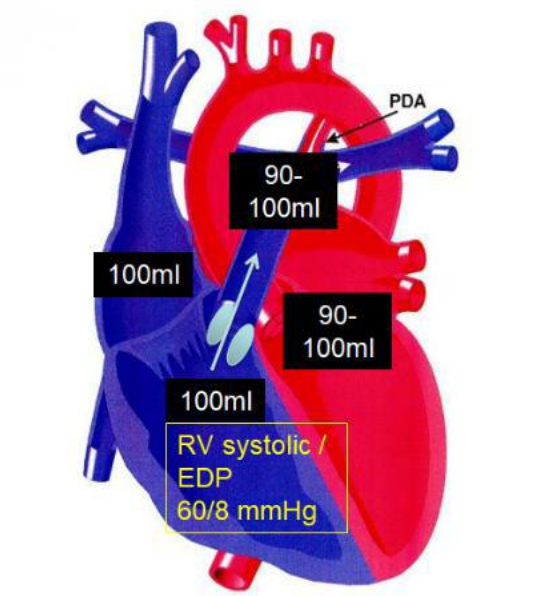

Figure 16. A hypothetical patient with pulmonary stenosis. The right ventricular systolic/end diastolic pressure (EDP) is increased.

Types of Pulmonary Stenosis

- Valvular
  - Best prognosis since it can just be ballooned open
  - Most common
  - Accounts for 7-10% of all congenital heart defects
- Sub-valvular (Infundibular)
- Supra-valvular
- Branch

Hemodynamic Consequences

- **Pressure overload of RV**
  - Because of the outflow obstruction, there is increased right ventricular systolic pressure and wall stress, producing hypertrophy

Physical Examination

- RV heave
  - Enlarged heart with a conspicuous parasternal right ventricular lift that frequently extends to the left midclavicular line in cases with severe stenosis (Kliegman et al., 2016)
- Systolic thrill (valve PS)
- Systolic ejection murmur
  - LUSB (left upper sternal border) with radiation to the back (interscapular area and bilateral axilla)
  - In severe stenosis, there is a loud, long, and harsh systolic ejection murmur that is maximally audible at the pulmonic area and may radiate over the entire precordium (Kliegman et al., 2016)
- Soft P2

Diagnostic Modalities

- ECG
  - R-axis deviation
  - IRBBB if mild
  - RVH
- CXR
  - Normal or RV cardiomegaly
  - Normal or dilated MPA (post-stenotic dilation)
  - Intrapulmonary vascularity is normal to decreased

B. Aortic Stenosis (AS)

- Congenital narrowing and obstruction of aortic valve
- The most frequent cause of aortic stenosis is calcification and sclerosis of anatomically normal or congenitally bicuspid aortic valves (Kumar et al., 2015)
  - Congenital bicuspid aortic valves are not considered as a CHD because it is not debilitating nor is it life-threatening
  - As we get older, our valves become more calcified

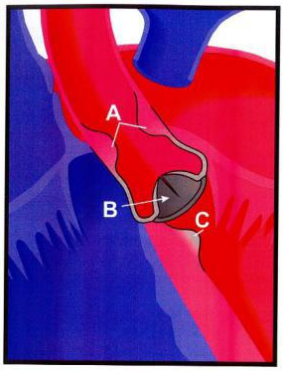

Figure 17. Aortic stenosis.

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Hemodynamic Consequences

- Pressure overload of LV
- LVH, normal vascular markings

Types of Aortic Stenosis

- **Valvular aortic stenosis**
  - Most common type
  - Leaflets are thickened
  - Commissures are fused to varying degrees
  - Left ventricular systolic pressure is increased as a result of the obstruction to outflow
    - Left ventricular wall hypertrophies in compensation
- **Subvalvular (subaortic) stenosis** with a discrete fibromuscular shelf below the aortic valve
  - Lesions are frequently associated with other forms of CHD
    - Mitral stenosis
    - Coarctation of the aorta (Shone syndrome)
  - May progress rapidly in severity
- **Supravalvular aortic stenosis**
  - Least common type

Aortic Stenosis vs. Pulmonary Stenosis

- To distinguish one from the other, listen to the radiation of the murmur
  - The aorta points towards the head and neck
  - The pulmonary valve and artery points towards the back, the interscapular area, then branches left and right
- Aortic Stenosis
  - Sound goes up the ascending aorta, towards the head and neck, in the carotid arteries
  - Bruits will be heard in the left and right side of the neck (carotid arteries), and in the suprasternal notch
  - Thrills will be felt on the suprasternal notch
- Pulmonary Stenosis
  - From the front, the sound radiates to the back (interscapular area), and follows the pulmonary arteries to the left and right
  - Bruits will be heard in the left and right axillary area

C. Coarctation of the Aorta (CoA)

- Narrowing or constriction of the lumen of the aorta may occur:
  - Anywhere along its length
  - **Most commonly located distal to the origin of the left subclavian artery**
    - Near the insertion of the ligamentum arteriosum
  - More proximal CoAs occur in Caucasians

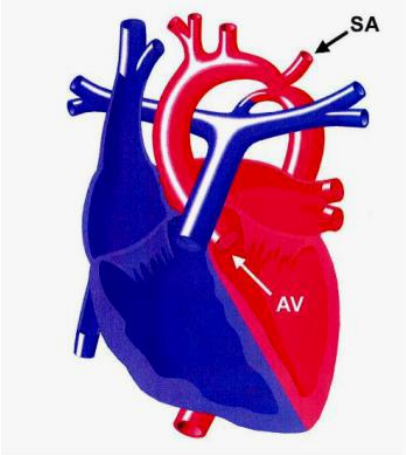

Figure 18. Coarctation of the aorta.

Pathophysiology

- Left Ventricular Hypertrophy Pathophysiology
  - The narrowing of the aorta creates a very high resistance to circulation with which the left ventricle must contract against
  - The increased workload for the LV causes LV hypertrophy

Hemodynamic Consequences

- **Pressure overload of LV**

Physical Examination

- LV heave
- Systolic murmur at the back along the aorta on the left of the spine
- Delayed or weak lower extremity pulses versus strong or good upper extremity
  - Classic sign

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- Disparity in pulsation and blood pressure in the arms and legs
  - Weak (or absent) pulses in the femoral, popliteal, posterior tibial, and dorsalis pedis
  - Pulses on the arms and carotid vessels are bounding
  - Check both the radial and femoral pulses for a radial-dorsalis or brachial-femoral delay
- Common chief complaint: upper extremity hypertension or leg pains
  - Lower extremities
    - Poor pulse, often difficult to obtain
  - Upper extremities
    - Strong pulse and hypertension

Diagnostic Modalities

- ECG
  - LVH
  - Left axis deviation
- CXR
  - LVH
  - Cardiac enlargement and pulmonary congestion in severe coarctation
  - **Rib notching** of inferior portion
    - Occurs due to pressure erosion by enlarged collateral vessels
    - Common occurrence by late childhood

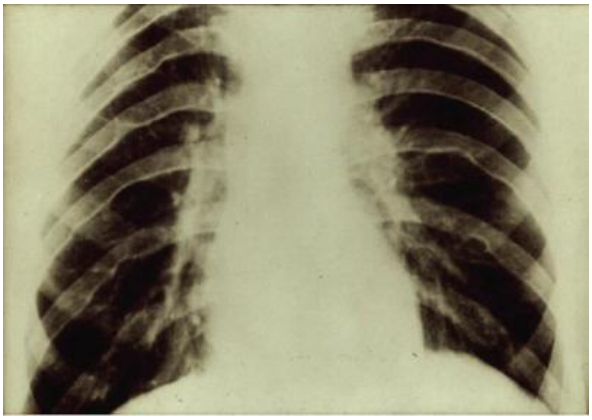

Figure 19. CXR of CoA. Figure shows notched ribs, a classic CXR finding in patients with CoA.

Management

- **Prostaglandin E1**
  - To reopen the ductus and reestablish adequate lower extremity blood flow
  - Not Prostaglandin E2, which is the natural type found in the body
- Surgery
  - Gold standard
- Doing surgery to treat CoA runs the risk of paraplegia due to prolonged clamping of the aorta in order to stitch it
- Non-surgical
  - The gold standard for post-operative re-coarctation
- There is currently a debate on when surgical and non-surgical treatments should be done
  - In younger pediatric patients
    - Surgery is the treatment of choice
  - In older pediatric or adult patients
    - A primary stent placement is recommended
    - Initially, the stretching of the stent results to a tearing of the intima layer.
    - Risk of rupture if over dilated
- Pulses in the lower extremities could still be normal as a result of collateral blood vessels
  - From the ascending aorta, vertebral collaterals could be used to bypass the coarctation and supply the descending aorta to the lower extremities
  - These arteries expand around the ribs, eroding the bones

APPROACH TO CONGENITAL HEART DISEASE: ACYANOTIC CONGENITAL HEART DISEASE

Dexter D. Cheng, MD

Quick Review

A. Acyanotic Defects: Left-To-Right Shunts

Atrial Septal Defect (ASD)

- Abnormality
  - Opening in the atrial septum, allowing blood to flow between the left and right atria
  - Types
    - Primum: worst prognosis
    - Secundum: most common
- Pathophysiology
  - Small amount of blood flows back from the LA to the RA, which has a lower pressure gradient
- Hemodynamic Consequences
  - RA hypertrophy
  - RV hypertrophy
- Physical Examination
  - Usually asymptomatic, has a late presentation
  - Fixed split S2
- Diagnostic Modalities
  - ECG: right-axis deviation
  - CXR: dilated main PA

Ventricular Septal Defect (VSD)

- Abnormality: incomplete closing of the ventricular septum
- Pathophysiology
  - During ventricular systole, a small portion of blood flows back to the pulmonary artery
  - Hematopoiesis is triggered because less blood is returning to systemic circulation
  - More and more blood is directed to the lunges, causing pulmonary congestion
  - 60% muscular, 40% perimembranous
- Hemodynamic Consequences
  - Small-sized hole: <1/3 of aortic diameter
    - Too small to be insignificant
  - Moderately-sized hole: 1/3 of aortic diameter
    - LV volume overload
  - Large-sized hole: ⅓ to > ½ of aortic diameter
    - LV volume overload
    - RV pressure overload
- Physical Examination
  - Dynamic and bulging precordium
  - Pansystolic murmur heard at the end of systole
- Diagnostic Modalities
  - ECG
    - Small: normal
    - Moderate: LVH
    - Large: RVH
  - CXR
    - Small: looks similar to ASD
    - Moderate or Large
      - Cardiomegaly
      - Dilated MPA

Complete Atrio-Ventricular Septal Defect (CAVSD)

- Abnormality
  - Basically a combination of ASD and VSD
  - a.ka. endocardial cushion defect or canal defect
  - Most commonly found in Down Syndrome (25% of patients)
- Pathophysiology
  - ASD component: blood shunted to RA
  - VSD component: blood shunted to RV
- Hemodynamic Consequences
  - ASD component: RV and RA hypertrophy
  - VSD component: LV and LA hypertrophy
  - Early development of pulmonary congestion

Patent Ductus Arteriosus (PDA)

- Abnormality
  - Before birth, the aorta and pulmonary artery are connected by the ductus arteriosus, which is expected to close within 24 hours after birth
  - In PDA, it does not close such that oxygen-rich blood from the aorta is mixed with oxygen-poor blood from the pulmonary artery
- Pathophysiology
  - Blood is shunted from the aorta back to the pulmonary artery through the ductus arteriosus
  - This causes an increased volume of blood entering the LA and LV
- Hemodynamic Consequences
  - Moderately-sized hole
    - LV volume overload
  - Large-sized hole
    - LV volume overload
    - RV pressure overload
    - Dilated ascending aorta
- Physical Examination
  - Continuous, machinery-like murmur
    - Main differentiation between VSD
    - Murmur is heard throughout the entire systole and diastole because aortic pressure is consistently higher than PA
- Diagnostic Modalities
  - ECG
    -
  - CXR
    - Looks similar to VSD
    - Differentiating trait: dilated aorta

B. Acyanotic Defects: Obstructive Lesions

Pulmonary Stenosis

- Abnormality
  - Malformations that cause pulmonary valve obstruction
  - Associated with congenital rubella or Noonan and William Syndrome
  - The valvular type is the most common and has the best prognosis
- Pathophysiology
  - Stenosis is the failure of valve opening, which essentially causes pressure overload
- Hemodynamic Consequences
  - Asymptomatic unless severe
  - RV pressure overload
    - If chronic, it can result in RVH or RHF
- Physical Examination
  - RV tap and heave
  - Systolic thrill
- Diagnostic Modalities
  - ECG
    - RVH
    - R-axis deviation
  - CXR
    - RVH
    - Less white areas are seen

APPROACH TO CONGENITAL HEART DISEASE: ACYANOTIC CONGENITAL HEART DISEASE

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Aortic Stenosis

- Abnormality
  - Congenital narrowing and obstruction of the aortic valve
  - Not strictly considered as a CHD
  - Types
    - Valvular aortic stenosis: most common type
    - Subvaulvular
    - Supravalvular: lest common type
- Pathophysiology
  - Stenosis is the failure of valve opening, which essentially causes pressure overload
- Hemodynamics
  - LV pressure overload
  - LVH

Coarctation of the Aorta (CoA)

- Abnormality
  - Narrowing or constriction of the lumen of the aorta
  - Most common location: distal to the origin of the left subclavian artery
- Pathophysiology
  - LVH Pathophysiology
    - Aorta coarctation creates a higher circulatory resistance that the LV must contract against in order to efficiently pump blood
    - Due to increased workload, LVH will occur
  - RVH Pathophysiology
    - More common in infants
    - Aorta coarctation prevents blood being pumped to the descending aorta
    - Infants remain asymptomatic due to the presence of PDA. However, increased RV workload will eventually cause RVH
- Hemodynamic Consequences
  - LV pressure overload
    - Eventual LVH
  - The more proximal the location of the coarctation, the less common for Asians
- Physical Examination
  - Classical sign: disparity in the pulsation and blood pressure between the arms and legs
- Diagnostic Modalities
  - ECG
    - LVH
  - CXR
    - Rib notching of inferior portion

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Appendix

Table 5. Acyanotic Congenital Heart Disease: Left-To-Right Shunts

Shunt	Hemodynamic Consequences	Physical Examination	Diagnostic Modalities	Other Notes
Atrial Septal Defect	• Diastolic overload of RV	• Fixed split S_2 • Soft systolic murmur	• **ECG**: normal to RAD, IRBBB pattern, RVH • **CXR**: mild RV cardiomegaly, dilated MPA, hypervascular markings	• Repair ○ Surgery: gold standard ○ Non-surgical: for secundum type only
Ventricular Septal Defect • Most common L to R shunt	• Small size ○ No pressure or volume overload • Moderate size ○ Volume overload of LV • Large size ○ Volume overload of LV ○ Pressure overload of RV	• **Signs of congestive heart failure** (large VSD) • Dynamic and bulging precordium • **Systolic thrill** • **Pansystolic murmur** • Normal to increased P_2	• **ECG**: ○ **Small**: Normal ○ **Moderate to large**: LVH to CVH ○ **Severe PHTN**: RVH • **CXR**: Normal to **cardiomegaly** (LV combined to RV), dilated MPA, hypervascular markings, dilated aorta	• Treatment ○ Predominantly **surgical** • Natural History ○ **Location of defect**: muscular and perimembranous VSD have high incidence of spontaneous closure ○ **Size of defect**: ▪ **Larger defect**: more likely to develop CHF sooner ▪ **Small VSD**: normal growth and development; no CHF ▪ **Moderate to large VSD**: easy fatigability, intermittent feeding, delayed growth and development, repeated RTI, CHF ○ High PVR limits total shunt flow
Complete Atrio-Ventricular Septal Defect • Endocardial Cushion Defect / Canal Defect				
Patent Ductus Arteriosus	• Moderate size ○ Volume overload of LV • Large size ○ Volume overload of LV ○ Pressure overload of RV	• Wide pulse pressure • Bounding pulses • Dynamic precordium, thrill • Soft to increased P_2 • **Continuous murmur (machinery-like)**	• **ECG**: Normal, LVH, CVH, or RVH ○ RVH will only occur if there is PHTN • **CXR**: Normal, cardiomegaly (LV, CV, or RV), dilated MPA, hypervascular markings, dilated aortic knob	• Management: ○ **Surgery**: gold standard for patients less than 4 kg ○ **Transcatheter**

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Table 6. Acyanotic Congenital Heart Disease: Obstructive Lesions

Lesion	Hemodynamic Consequences	Physical Examination	Diagnostic Modalities	Other Notes
Pulmonary Stenosis	• Pressure overload of RV • RVH, normal vascular markings	• RV heave • Systolic thrill (valve PS) • Systolic ejection murmur LUSB with radiation to the back (interscapular area and bilateral axilla) • Soft P_2	• **ECG**: RAD, IRBBB if mild, RVH • **CXR**: Normal or RV cardiomegaly, normal or dilated MPA (post-stenotic dilatation)	• 5-8% of CHD • No pulmonary congestion • May develop to RHF years later • Associated with congenital rubella; Noonan and William syndrome • **Types**: Valvar, subvalvar (infundibular), supravalvar or peripheral • **Manifestations**: asymptomatic unless severe
Aortic Stenosis	• Pressure overload of LV • LVH, normal vascular markings			
Coarctation of the Aorta	• Pressure overload of LV	• LV heave • Systolic murmur at the back • **Delayed or weak lower extremity pulses vs. upper extremity**	• **ECG**: LVH, LAD • **CXR**: LVH, pulmonary congestion, **rib notching**	• Management: ○ **Surgery**: gold standard, TOC for younger pediatric patients ○ **Non-surgical**: gold standard for post-operative re-coarctation ○ **Primary stent placement** in older pediatric patients