# Introduction to congenital heart disease

Fifth Year Medical Student Course
Department of Pediatrics
The University of Jordan

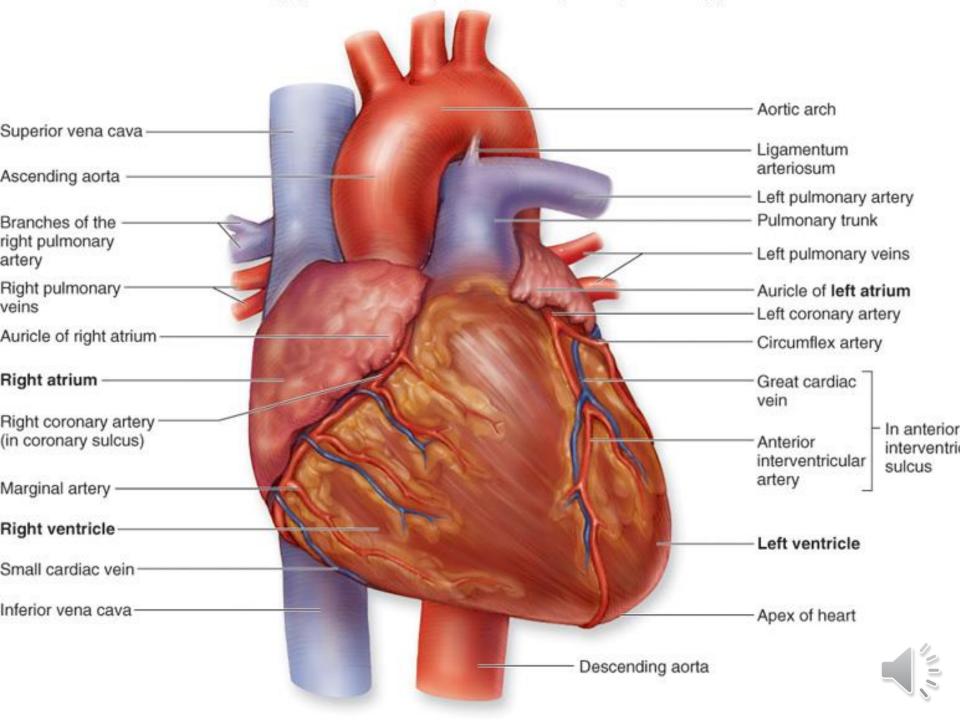
Iyad AL-Ammouri 2020



## Outline

- Review of Anatomy of the Normal heart
- The Cardiac Output
- The Cardiac Cycle
- The Fetal circulation and postnatal changes
- Possibilities and classification of congenital heart disease
- Presentations of CHD
- Mechanisms of Heart Failure in CHD
- Mechanisms of Cyanosis in CHD





Copyright © The McGraw-Hill Companies, Inc. Permission required for reproduction or display. Aortic arch Ligamentum arteriosum Ascending aorta Left pulmonary artery Superior vena cava-Pulmonary trunk Left pulmonary veins Right pulmonary artery Right pulmonary veins Left atrium Aortic semilunar valve Right auricle Left atrioventricular valve Interatrial septum Pulmonary semilunar Right atriumvalve Opening for inferiorvena cava Trabeculae carneae Right atrioventricular valve-Interventricular septum Chordae tendineae-Left ventricle Papillary muscle Right ventricle Inferior vena cava Descending aorta

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Right ventricle

Left atrium

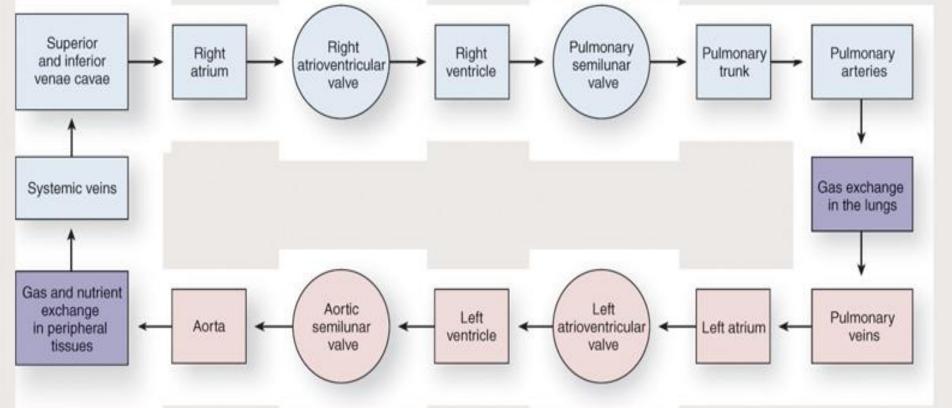
Left ventricle

#### **Blood Flow Through the Heart**

Right atrium

Left atrium

Pulmonary veins



Gas and nutrient exchange in peripheral tissues	Aorta Aortic semilunar valve	Left ventricle Left atrioventricular valve	Gas exchange in the lungs  Pulmonary veins
Chamber of the Heart	Receives Blood From	Sends Blood To	Valves Through Which Blood Flows
Right atrium	Superior vena cava, inferior vena cava, coronary sinus	Right ventricle	Right AV valve

Pulmonary trunk (blood enters pulmonary circuit of vessels)

Aorta (blood enters systemic circuit

Left ventricle

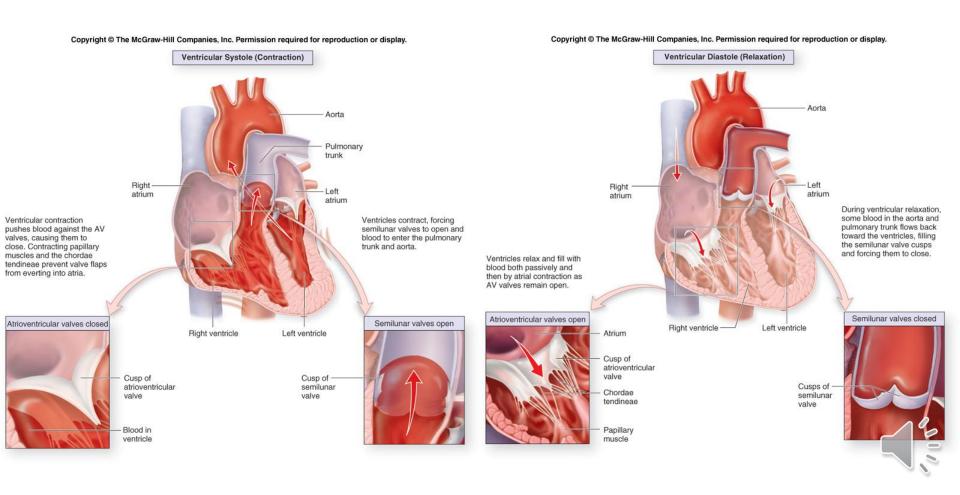
of vessels)

Pulmonary semilunar valve

Aortic semilunar valve

Left AV valve

## Ventricular systole and diastole



## Cardiac function

- Pumping of oxygenated blood to the systemic organs through systemic circulation, and pumping of de-oxygenated blood to the pulmonary circulation
- Blood flow to the systemic circulation is measured as liters/minute and is called (Cardiac output)
- Cardiac output is regulated by tissue demand for oxygen
- Cardiac output is a result of : stroke volume X heart rate per minute
- Stroke volume is the volume of blood ejected to the systemic circulation in one beat
- In normal heart

stroke volume = end diastolic volume - end systolic volume



## Cardiac Output

Heart rate

X

Stroke Volume

Regulated by sympathetic and Parasympathetic nervous system

Symp→ ↑
Parasymp→ ↓

Determined by:

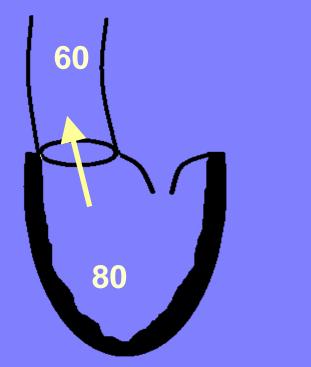
- •Preload (volume)
- •Afterload (resistance)
- Contractility



## Stroke volume

LVEDV=80 (preload)
LVESV=20

Stroke volume= LVEDV-LVESV=60



Preload: volume of blood in LV at end diastole

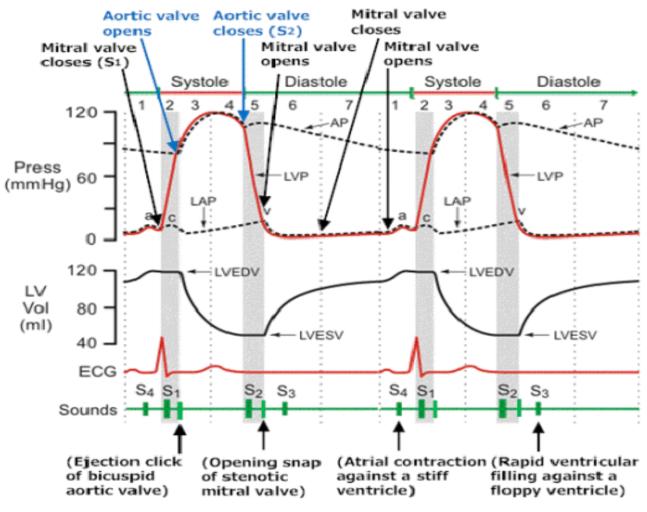
(venous return)

Afterload: resistence against which the LV is pumping

Contractility: degree of shortening of the muscle when it contracts (strength of muscle)



#### The Cardiac Cycle



- 1 -> Atrial Contraction --> Mitral Valve Opens
- 2 -> Isovolumic Ventricular Contraction --> Mitral Valve Closes ---> Aortic Valve Opens
- 3 -> Rapid Ejection
- 4 -> Reduced Ejection
- 5 -> Isovolumic Ventricular Relaxation --> Aortic Valve Closes ---> Mitral Valve Opens
- 6 -> Rapid Ventricular Filling
- 7 -> Diastasis ---> Mitral Valve Closes
- 1 -> Atrial Contraction --> Mitral Valve Opens

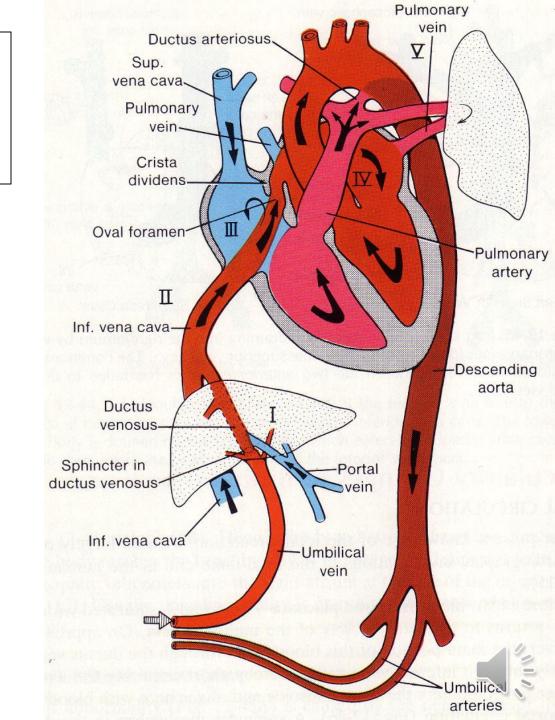


## Circulation in the Fetus



Principle differences in fetal circulation compared to post-natal circulation:

- Three critical anatomic communications
  - PFO (from RA to LA)
  - PDA (from PA to AO)
  - Ductus venosus (from UV to IVC)
- Organ responsible for oxygenation is Placenta
- Pulmonary vascular resistance is high due to constricted pulmonary vessels



## PFO and PDA during fetal life

- PFO: directs blood coming from inferior vena cava (from umbilical vein) to the left atrium
- PDA: directs blood from the pulmonary artery to the descending aorta because the lungs are not being used for oxygenation in the fetus
- Both PFO and PDA are important for fetal survival



## PFO and PDA after birth

- PFO: when blood flow to the left atrium increased because of more blood flow to the lung after the baby is born, the flap of foramen ovale closes
- PDA: Increased oxygenation and absence of placental prostaglandins once the cord is clamped causes constriction and closure of the PDA within few days after birth

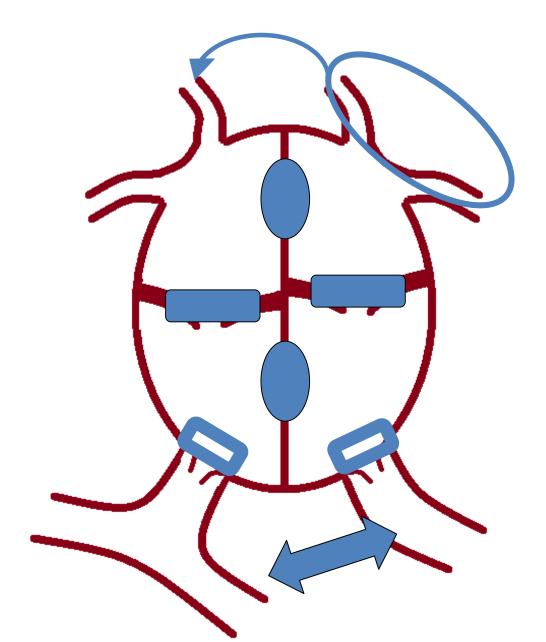




# Congenital heart disease



### Congenital heart disease – Many possibilities





#### Structural heart disease

# **Acyanotic** with shunt

- •ASD
- •VSD
- •PDA

#### Cyanotic

- •TOF
- •TGA
- •Truncus
- •Tricuspid Atresia
- •TAPVR

### **Non Shunt lesions**

#### **Obstruction**

- Aortic stenosis AS
- •Supravalvar AS
- •Subaortic S
- •Coarctation
- •Mitral Stenosis
- •Pulmonary Stenosis

#### Regurgitation

- •Aortic regurgitation
- •Mitral regurgitation
- •Pulmonary reg.



# Symptoms of congenital heart disease





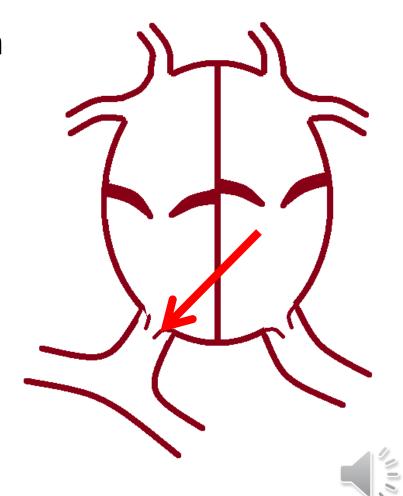
Cyanosis Lesions with shunts **HLHS** TOF **VSD** Aortic atresia **TGA Tricuspid Truncus PDA** atresia artersiosus **Pulmonary TAPVR** atresia

Many asymptomatic patients



# Mechanisms of heart failure in CHD 1) Overcirculation

- Pulmonary overcirculation with high pressure
  - VSD
  - PDA
  - AVCanal
  - Truncus arteriosus
  - Single ventricle without pulmonary stenosis



# Mechanisms of heart failure in CHD 2) Obstruction to blood flow

- Critical obstruction to systemic blood flow causing metabolic acidosis and shock
- Presents when PDA starts to constrict
  - Critical aortic stenosis
  - Critical coarctation of aorta
  - Hypoplastic left heart syndrome



# Mechanisms of heart failure in CHD 3) Muscle dysfunction

- Left ventricular dysfunction, takes long time to happen
- Due to:
  - Long standing pressure overload (eg, AS, COA)
  - Long standing volume overload (eg, VSD, mitral or aortic regurgitation
  - Rarely due to myocardial ischemia (severe cyanosis, severe diastolic hypotension, relative ischemia due to severe hypertrophy, coronary anomalies)



### **Mechanisms of Cardiac Cyanosis**

Right to left shunt

Mixing

Recirculation

**TOF** 

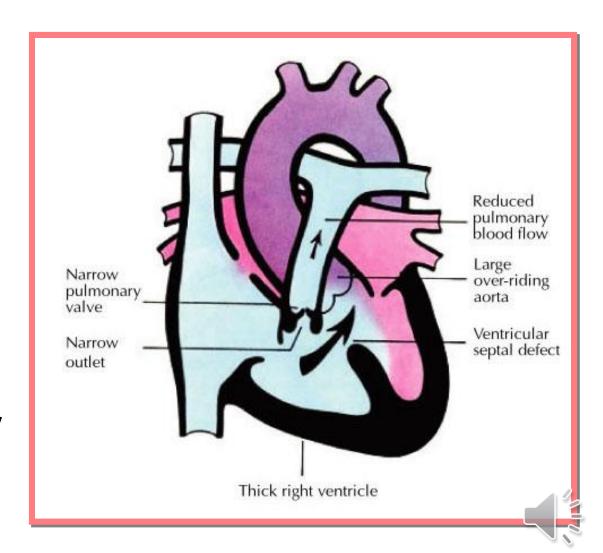
Tricuspid atresia
Truncus arteriosus
Single ventricle

**TGA** 



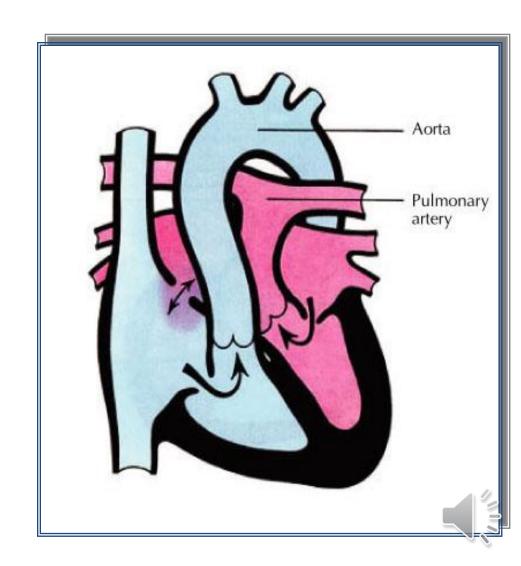
## Tetralogy of Fallot (TOF)

- RVOT obstruction
- 2. VSD
- 3. Overriding aorta
- 4. RV hypertrophy



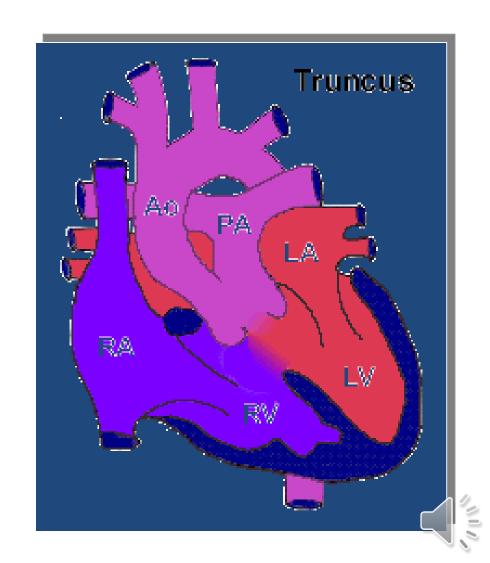
## Transposition of Great Areries (TGA)

- Aorta originating from the right ventricle, and pulmonary artery originating from the left ventricle
- Accounts for 5-7% of all congenital heart disease



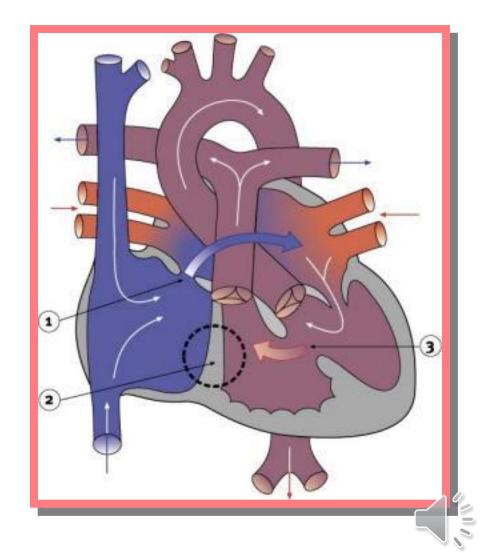
### **Truncus Arteriosus**

- The presence of a common trunk that supply the systemic, pulmonary and coronary circulation
- Almost always associated with VSD
- No PDA
- 1.2-2.5% of all congenital heart disease

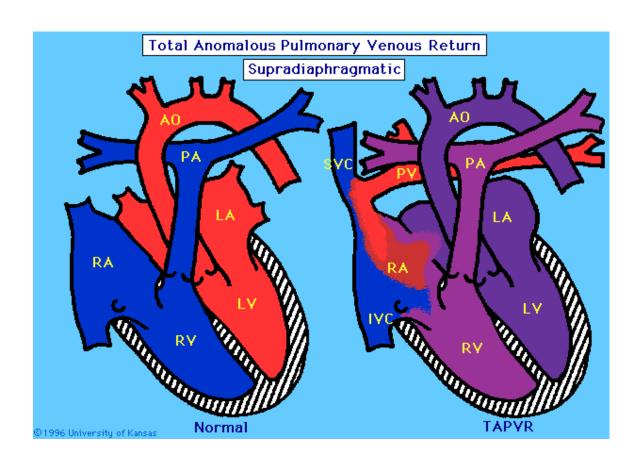


## Trcuspid Atresia

- Complete absence of communication between the right atrium and right ventricle
- About 3 % of congenital heart disease

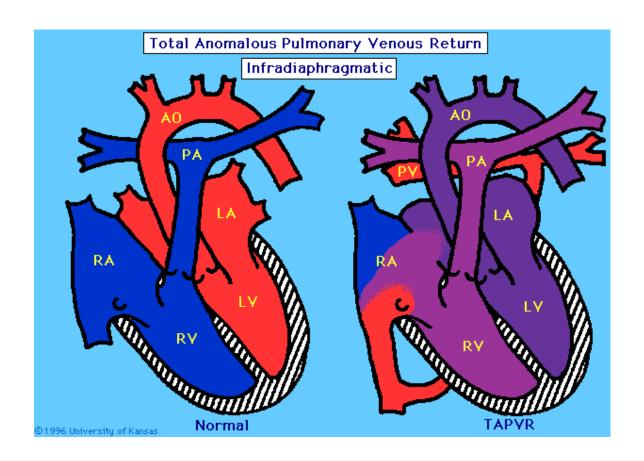


# Total Anomalous Pulmonary Venous Return (TAPVR)





## TAPVR-Infracardiac





### non-shunt Acyanotic heart lesions

#### **Obstruction**

- Aortic stenosis AS
- •Supravalvar AS
- •Subaortic S
- Coarctation
- •Mitral Stenosis
- •Pulmonary Stenosis

Generally cause pressure overload

#### Regurgitation

- •Aortic regurgitation
- •Mitral regurgitation
- •Pulmonary regurgitation

Generally cause Volume overload



# The end