BOARD REVIEW

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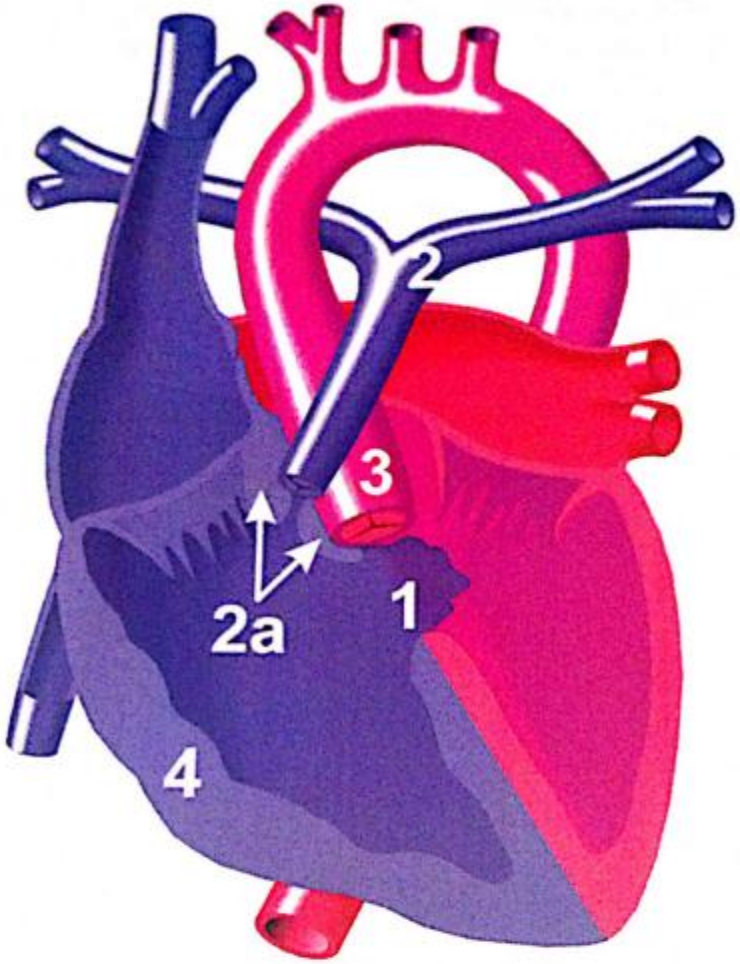
Tom Tarro

Erica Anson

Tetralogy of Fallot

Tetralogy of Fallot (TOF)

- Conotruncal Heart Lesion
- What is the **Primary** issue that developmentally leads to TOF?
  - Anterior displacement/deviation of the infundibular septum (septum that seperates the aortic and pulmonary outflow tracts)
    1. **Large, malaligned ventricular septal defect (VSD)**
    2. **Stenosis of right ventricular outflow tract (RVOT)**
       1. **Stenontic pulmonary artery +/- pulmonary valve**
       2. **Infundibular stenosis**
    3. **Enlarged aorta overiding the VSD (displaced to right)**
    4. **Right ventricular hypertrophy**



Above:

1. Ventricular septal defect (VSD)
2. Stenotic pulmonary valve and artery

2a. Infundibular stenosis

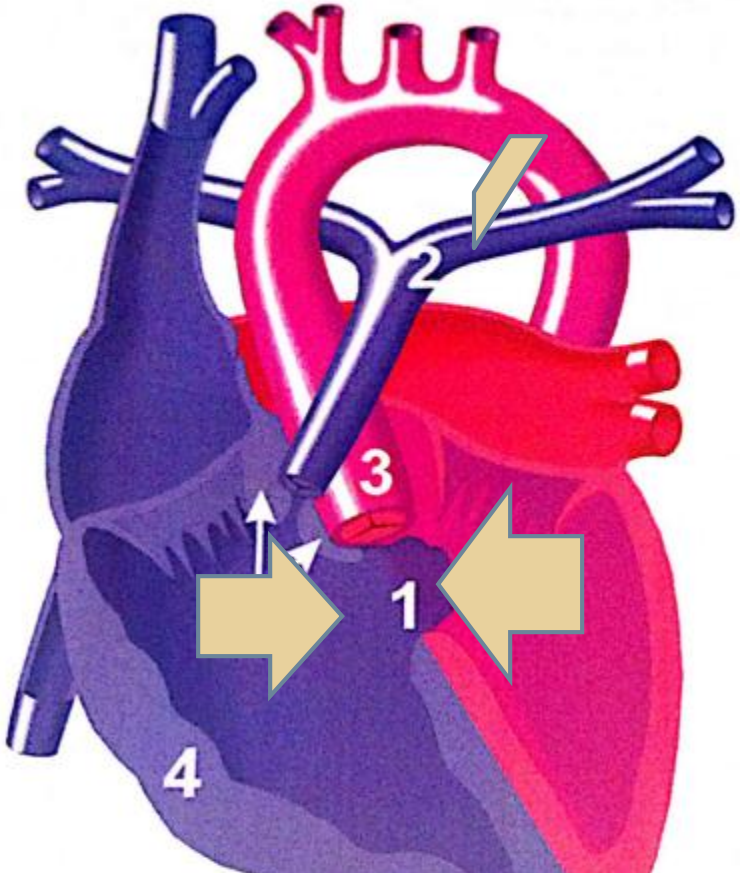
3. Enlarged aorta overriding VSD
4. Right ventricular hypertrophy

TOF- Epidemiology

- Accounts for ~10% of congenital heart disease
- **Most common CYANOTIC congenital heart condition**
  - ~2,700 affected newborns a year
- Affects both sexes equally
- Most common CYANOTIC heart defect beyond infancy

TOF- Pathophysiology

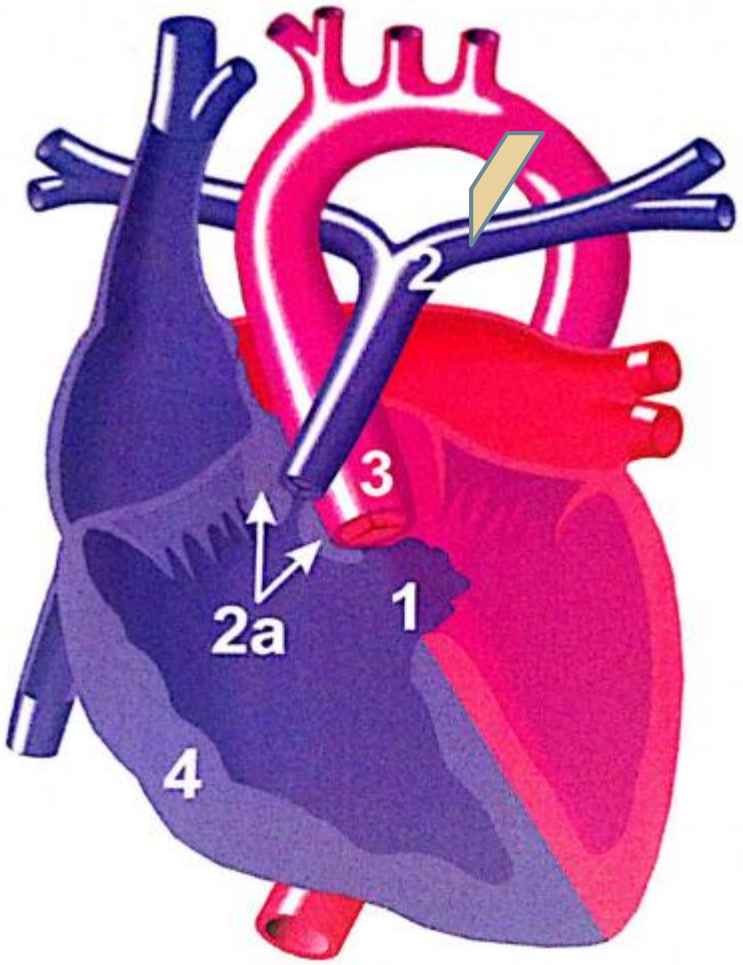
- Cyanosis depends on:
  - Degree of pulmonary stenosis



- Minimal pulmonary stenosis→
  - Large L→R shunt across VSD = (minimal cyanosis)
- Severe pulmonary stenosis→
  - Large R→L shunt across VSD = CYANOSIS!
  - *ductal-dependent pulmonary blood flow!

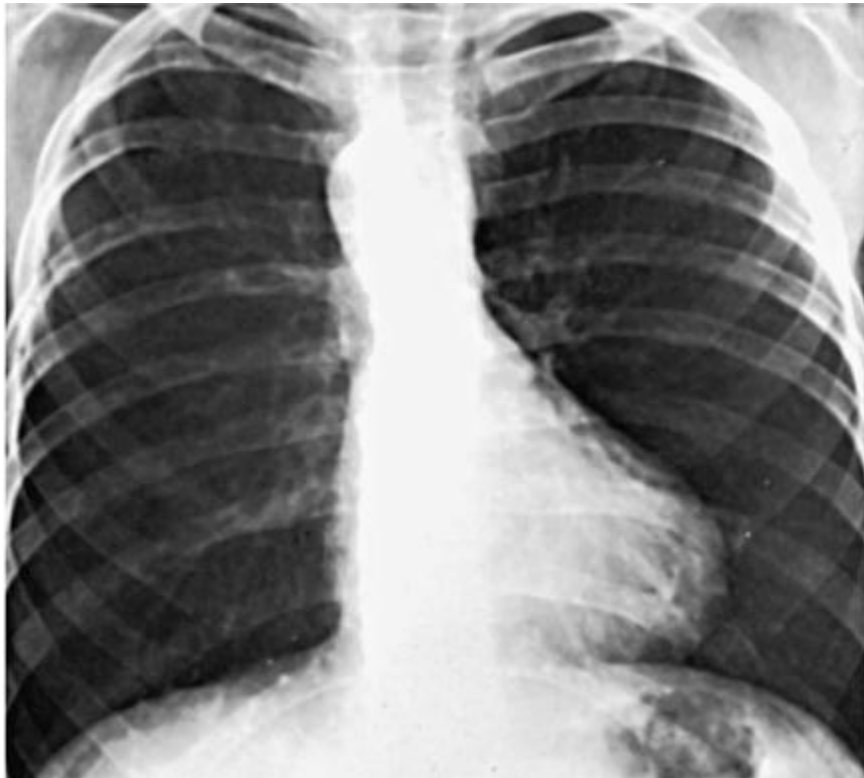
TOF Presentation

- ***VARIABLE!!!***
- The degree of pulmonary outflow obstruction determines degree of cyanosis and age at presentation!!
  - MIGHT BE ASYMPTOMATIC AT BIRTH!!
- If severely cyanotic at birth, need prostaglandin E1 infusion to maintain PDA and pulmonary perfusion
  - Will worsen with PDA closure if not!
- Cyanosis may not appear until months later ("pink TOF")
  - Heart failure common if lots of L→R shunting across VSD!



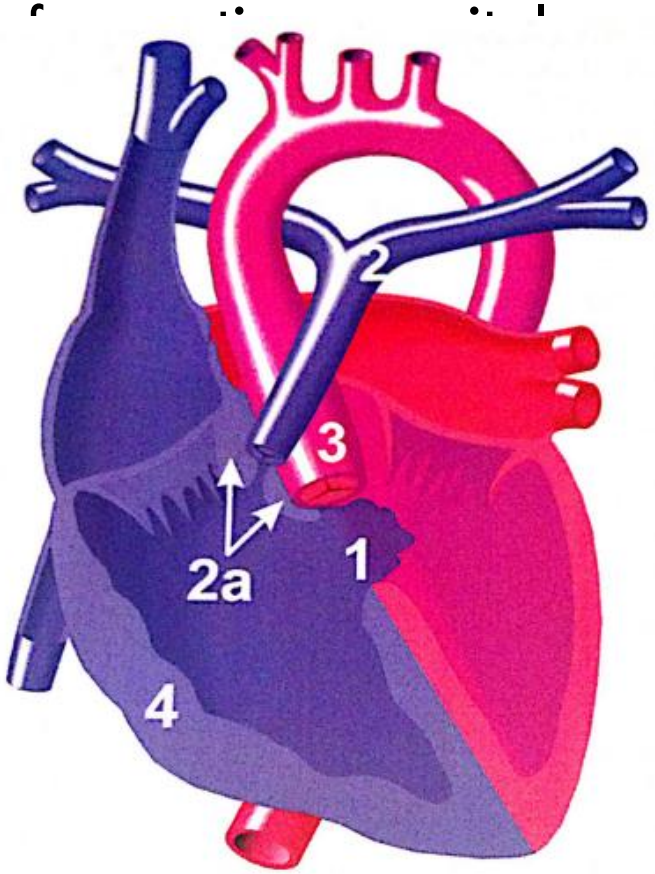
TOF Presentation

- Systolic Murmur transmitted to lungs
  - Louder with worsening RVOT outflow obstruction
  - May disappear with severe obstruction
- Palpable RV impulse
- Single S2
- Normal pressures/pulses
- Boot-shaped heart (x-ray)
- R-axis deviation with RVH
- Cath lab
  - Elevated systolic RV pressure
  - Decreased PA pressures



TOF

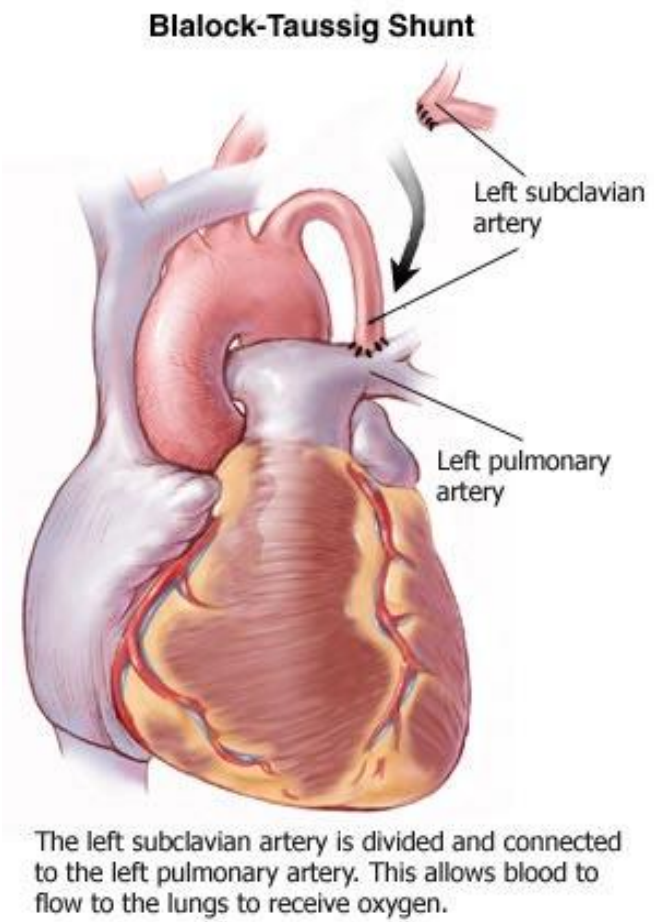
- If "Pink TOF"
  - Develop long-standing manifestation [illegible] heart disease
    - Heart failure
    - Clubbing of digits
    - Duskiness
    - Poor growth
    - Delayed puberty
    - Polycythemia
      - Rare complication: Cerebral thrombose
    - Iron-deficiency anemia
    - Dyspnea on exertion
      - → resolves with squatting position

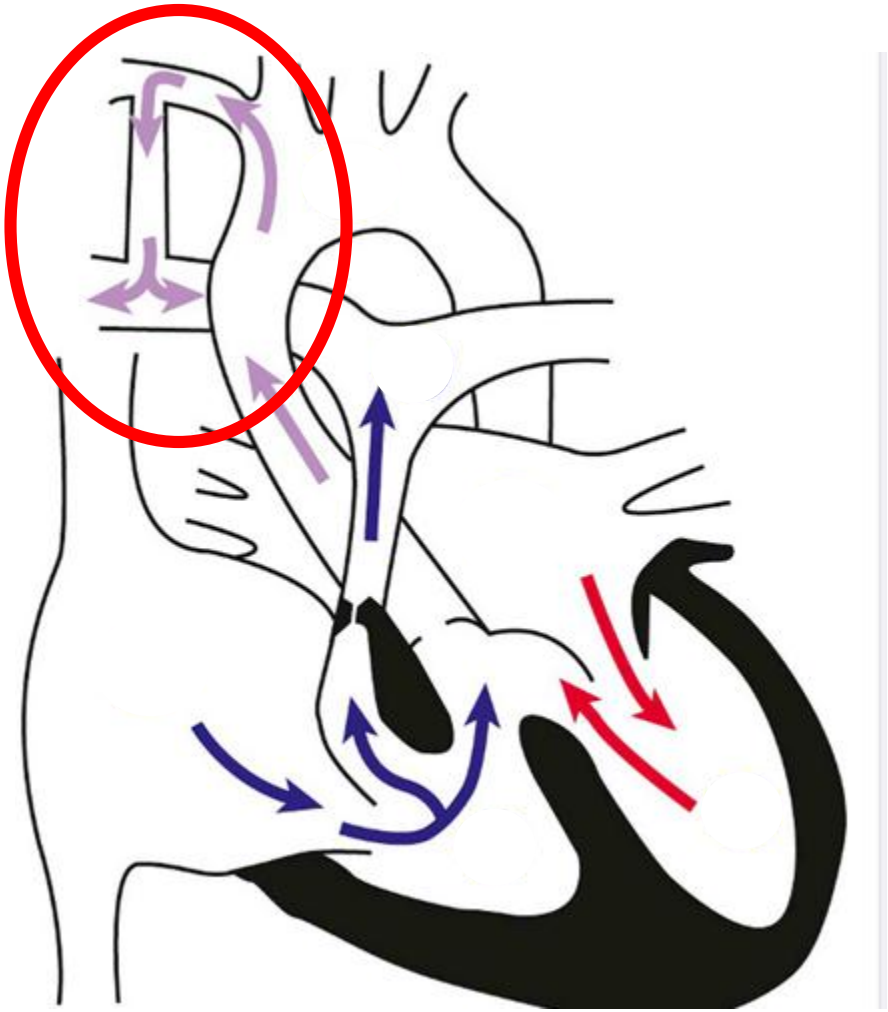


TOF-Treatment

- If severe cyanosis at birth
  - Prostin! → need the PDA to provide pulmonary blood flow
  - Neonatal repair
- If "pink Tet"
  - Prompt treatment of dehydration
  - Surgical treatment once spells begin

TOF-Treatment

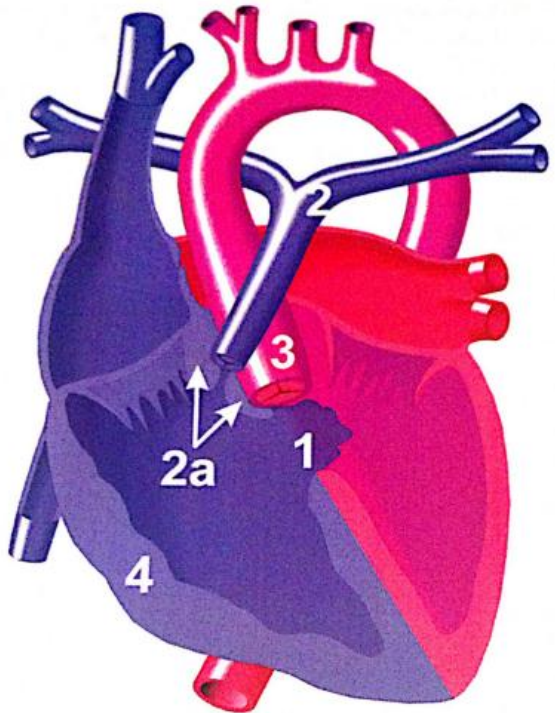




TOF-Treatment

- Total Corrective surgical repair
  - Resect obstructive portion of infundibulum
  - Patch closure of VSD
  - Valve correction (valvotomy vs. valvectomy)

- Post-operatively
  - Chance for arryhthmias and syncope



TET Spells

- Spells of extreme cyanosis "hypercyanotic spells"
- Increased infundibular/pulmonary stenosis

  (May be secondary to anemia, stress, crying, dehydration, fever)
  - Worsening of R→ L shunting across VSD = CYANOSIS
    - Hyperpneic
    - Restless
    - Gasping
    - Syncope
  - Can be fatal
  - Often first thing in the morning or with vigorous crying and ACUTE onset
  - Murmur disappears as flow across restricted RVOT diminishes
  - Often followed by weakness/sleep (similar to post-ictal period!)
  - If untreated, hypoxia and metabolic acidosis insue and worsen spell

**** Identify the clinical characteristics of a Tet spell**

Tet Spell

- Treatment:
  - Squatting
  - Knee-chest position
  - Oxygen!
  - Morphine
  - Volume expansion
  - Calming
  - Sodium bicarbonate
    - Recovery often insues with normalization of pH
  - Last resort- intubation and sedation

    Phenylephrine, Propranolol

TOF-Associated Defects

- Main pulmonary artery (PA) may be small with varying degrees of branched PA stenosis
- Often with significant pulmonary valve hypoplasia
- Infundibular muscle can be severely hypertrophied
- Major aortopulmonary collateral arteries (MAPCAs) may be providing pulmonary blood flow
- Atrial septal defects (ASD) also common
- Right-sided aortic arch occurs in 25-33% of patients with TOF
- Coronary anomalies
- Pulmonary anomalies/hypoplasia
- Can be associated with DiGeorge (22q11 defect)

TOF- Prognosis **

- Do very well long-term generally
- Exercise intolerance
- Persistent pulmonary valve problems
- Residual pulmonary stenosis
- Still at risk for arrythmias and syncope

Question 5

- You are treating a 4-month-old infant who was born with Tetralogy of Fallot. Her mother brings her to clinic because she has had diarrhea and fever since the previous evening. On physical exam, the infant is irritable and has cyanosis and HR of 180. Of the following, the finding that is MOST consistent with a "TET spell" is:
  a. Clubbing of the digits
  b. Hepatomegaly
  c. Inability to hear murmur
  d. Normal chest x-ray
  e. S3 gallop rhythm

Question 14

During a routine health supervision visit, the mother of one of your patients informs you that she is 28-weeks pregnant and that fetal echocardiogram has revealed Tetralogy of Fallot in an otherwise normal fetus. She asks you about the prognosis for children who have this condition. Of the following, the MOST appropriate response is that:

a. Cardiac catheterization techniques may help avoid surgery
b. Complete repair is associated with an excellent result with <5% mortality
c. Surgery most likely will be undertaken before the child is discharged after birth
d. Tetralogy of Fallot rarely is associated with chromosomal abnormalities
e. The usual surgical approach is palliative and requires three stages

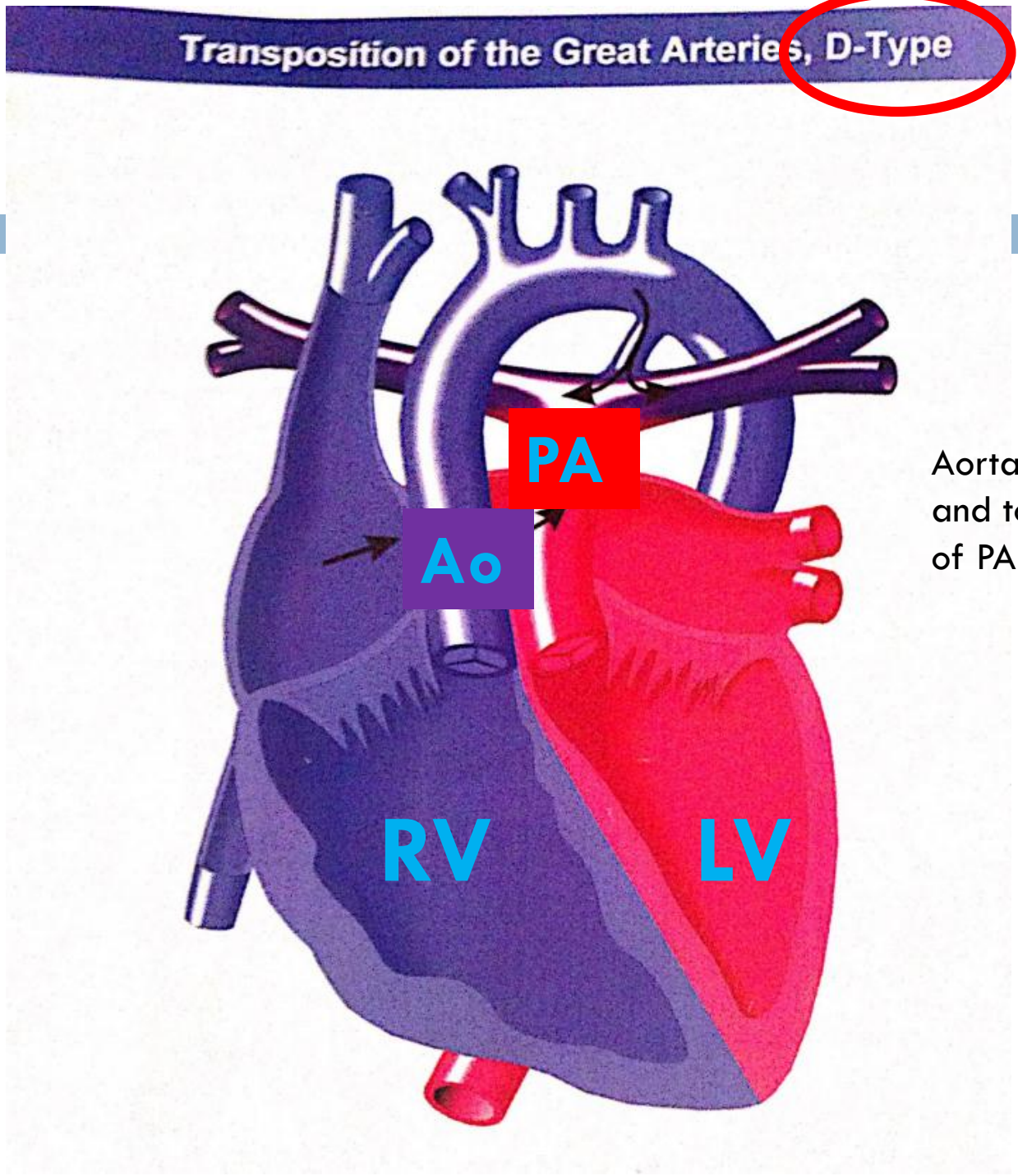
Question 15

- The primary developmental defect in Tetralogy of Fallot that is responsible for the characteristic four cardiac defects is what?
  a. Anomaly in arch development
  b. Anterior displacement of the infundibular septum
  c. Downward displacement of the tricuspid valve
  d. Abnormal cardiac conduction development
  e. Failure of the atrial septum secundum to fuse with the septum primum

Transposition of the Great Arteries

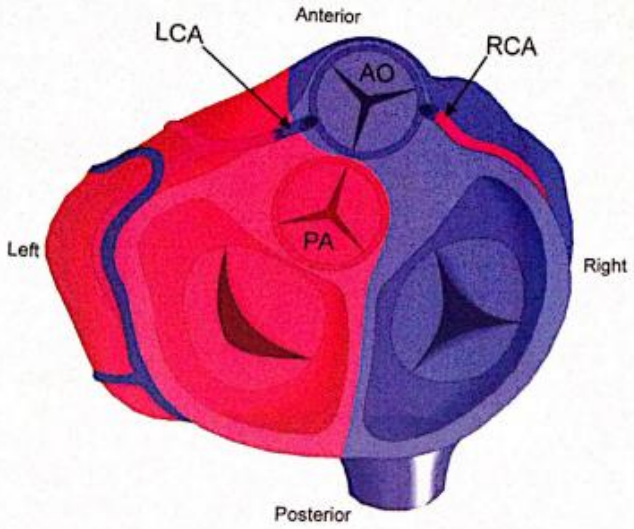
Transposition of the Great Arteries (TGA)

- Ventricular-arterial relationship discordance

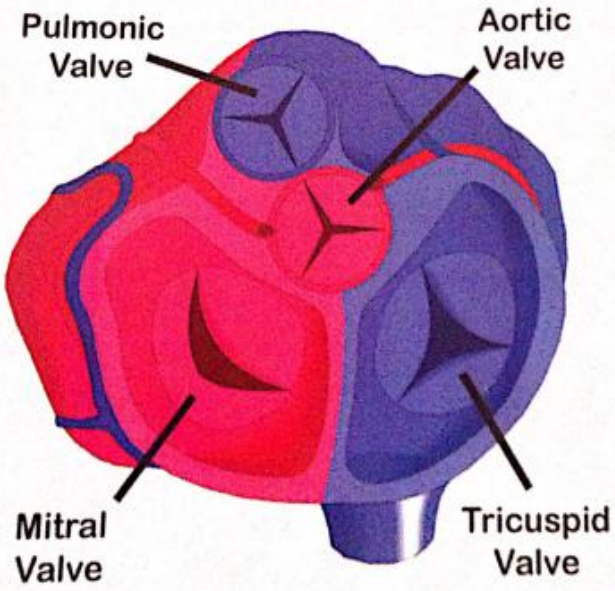


Aorta is anterior and to the right of PA

Transposition of the Great Arteries, D-Type - cont.



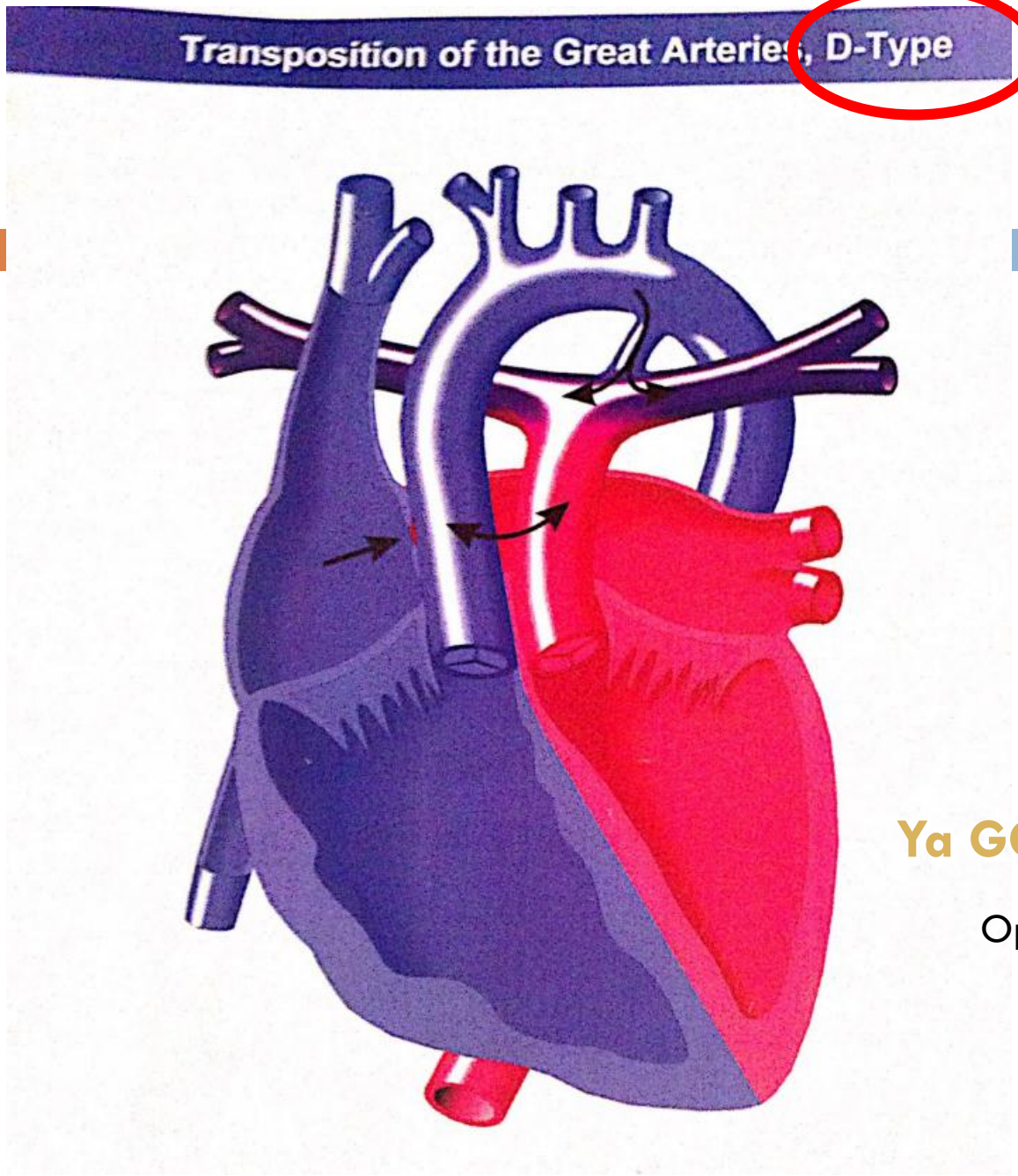
TGA - D



Normal Heart

TGA-Epidemiology

- Accounts of ~ 5% of congenital heart disease
- The most common cause of CYANOTIC heart disease IDENTIFIED in newborns
- More common in infants of diabetic mothers
- More common in males than females (3:1)
- More common in patients with DiGeorge Syndrome (chromosome 22q11)



Aorta: Carries DEOXYGENATED blood to systemic circulation

Pulmonary Artery: Carries OXYGENATED blood back to pulmonary circulation

Ya GOTTA MIX SOMEWHERE!!!

Options for mixing:

- Patent Foramen ovale
- Patent ductus arteriosus

TGA-Presentation **

- Should be your first thought in a cyanotic newborn!
- Low saturations *without* respiratory distress
- Simple D-TGA (no VSD)
  - Often with worsening cyanosis/tachypnea/CV collapse in first few days of life as PDA closes and mixing is drastically reduced
  - Loud and single 2nd heart sound
  - +/- parasternal heave
  - Often no murmur!
  - "Egg-shaped heart" with narrow mediastinum
  - Increased pulmonary markings
  - No improvement with hyperoxia test

**Recognize that the absence of improvement in Pa02 with 100% oxygen in comparion with RA is compatible with cyanotic congenital heart disease

Therapy

- At birth, patient needs prostaglandin E1 to maintain ductal patency**
- May consider balloon atrial septostomy to enlarge PFO/ASD

- Corrective surgery undertaken in newborn period
  - Arterial Switch operation
    - Can't wait too long or LV won't be able to generate systemic pressures
    - Don't forget the coronaries have to be moved as well!

TGA- Associated Defects

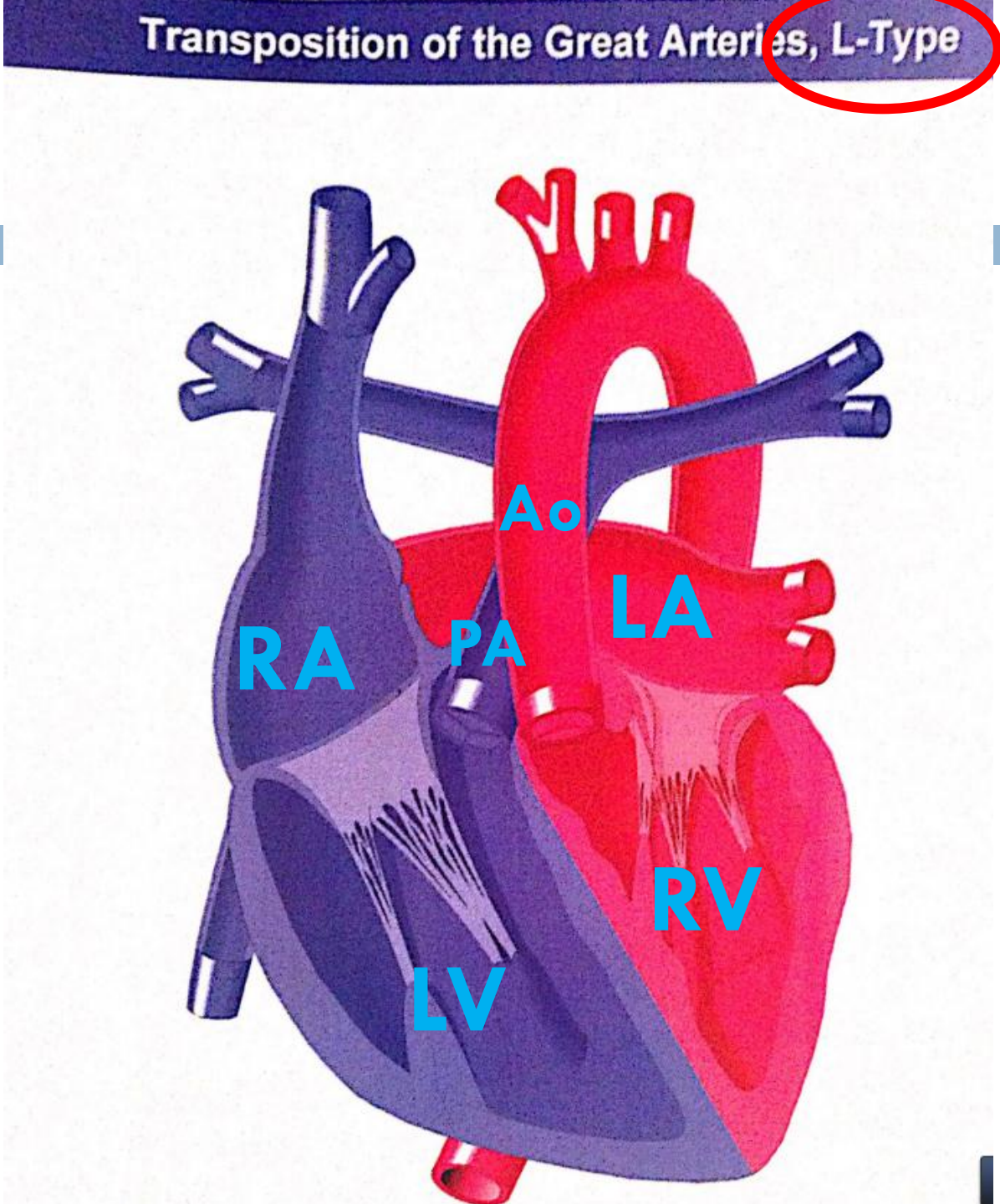
- Ventricular septal defects (50%- allows for mixing!)
- Coronary artery anomalies (10-15%)

D-TGA with VSD

- Now mixing can occur→ cyanosis less severe
- Now a harsh systolic murmur is present
- Now heart failure symptoms are noted
- Now cardiomegaly is more severe
- Still need surgery in newborn period
  - Arterial Switch with VSD closure

Discordant atrio-ventricular relationship & ventricular arterial relationship

But…
"corrected" physiology



Question 6

You discover a heart murmur in a term, 3.1kg male infant immediately after birth. The infant demonstrates cyanosis and his oxygen saturation is 65%. His breathing is unlabored. There is no hepatomegaly or splenomegaly, and the distal pulses are excellent. A CXR demonstrates clear lung fields and a normal cardiac silhouette but a narrowed mediastinal shadow. You order an echocardiogram. Of the following the most likely diagnosis is:

a. Coarctation of the aorta
b. Complete AV canal defect
c. Hypertrophic cardiomyopathy
d. Large VSD
e. Transposition of the great arteries

Question 21

You are evaluating a 6-hour-old male infant who was born after a term pregnancy and normal delivery and weighs 4 kg. The infant is comfortable but exhibits mild tachypnea with a RR: 50 and HR: 150. Oxygen saturation is 60% in all extremities and does not increase significantly with the administration of oxygen via facemask. His lungs are clear, and there are no murmurs, gallops, or rubs. You suspect transposition of the great arteries. Of the following, the BEST management strategy is:

a. Diuretic therapy intravenously for pulmonary edema
b. Increasing left to right (or aorta-pulmonary artery) shunt at the ductus arteriosus
c. Increasing the right to left shunt at the foramen ovale
d. Increase the right to left (or pulmonary-aorta artery) shunt at the ductus arteriosus
e. Intubation and mechanical ventilation with an FiO_2 of 1.0

Heart Failure

Heart Failure

- Cardiac output cannot meet metabolic demands of the body
  - Decreased CO → metabolite build-up in underserved tissues; ACIDOSIS!!!
- Compensatory mechanisms kick in
  - Renin-aldosterone-angiotensin system
  - Sympathetic nervous system
  - Cytokine-induced inflammation
- Ultimately→ cardiac remodeling
  - Protective mechanisms: ANP/BNP, IGF-1, GH

Heart Failure-Presentation

- Infants **
  - Dyspnea, irritability, increased fatigability, failure to thrive, increased work of breathing, tachypnea, grunting, tachycardia, gallop rhythm, hepatomegaly, decreased feed volumes
- Older kids **
  - Exercise-intolerance, early fatigue, somnolence, anorexia, cough, wheezing, crackles, jugular venous distention, peripheral edema, hepatomegaly, cardiomegaly, gallop rhythm

Heart Failure-Etiology **

- Excessive Preload
- Excessive Afterload
- Abnormal rhythm
- Decreased Contractility

Heart Failure- Etiology

Table 2. Sources of Heart Failure With a Structurally Normal Heart

Primary Cardiac

- Cardiomyopathy
- Myocarditis
- Myocardial infarction
- Acquired valve disorders
- Hypertension
- Kawasaki syndrome
- Arrhythmia (bradycardia or tachycardia)

Noncardiac

- Anemia
- Sepsis
- Hypoglycemia
- Diabetic ketoacidosis
- Hypothyroidism
- Other endocrinopathies
- Arteriovenous fistula
- Renal failure
- Muscular dystrophies

Heart Failure- Excessive Preload

- Most common physiology:
  - L→ R shunting at ventricular level
  - Left heart overload→ backs up into lungs as pulmonary pressures lower in infants **
- VSD/PDA
    - "dependent shunts" = dependent on pressure gradient between systemic and pulmonary circuits
- AVM
  - "obligate shunts" = shunt blood to venous system
- Valvular regurgitation
- Septic shock (High output)

**Understand the association between systemic AVM and CHF in a newborn infant!!

Heart Failure- Excessive Preload

Pericarditis can cause HF with *decreased* preload!!

Heart Failure- Excessive Afterload

- Left heart obstructive lesions
  - Mitral stenosis
  - Aortic stenosis
  - Coarctation of the aorta

→ Increased end-diastolic filling pressures with decreased pressure gradient between the ventricle and the aorta→ sub-endocardial ischemia

Heart Failure- Rhythm abnormalities

- Tachycardia related:
  - Decreased diastolic filling time→ decreased CO
- Bradycardia related:
  - Increased stroke volume→ enlarged LV

Heart Failure-Contractility Disorders

- Cardiomyopathy
  - Dilated→ idiopathic dilated chambers with impaired systolic and diastolic function
  - Restrictive→ infiltrative or storage diseases lead to impaired diastolic function
  - Hypertrophic

Heart Failure- Studies

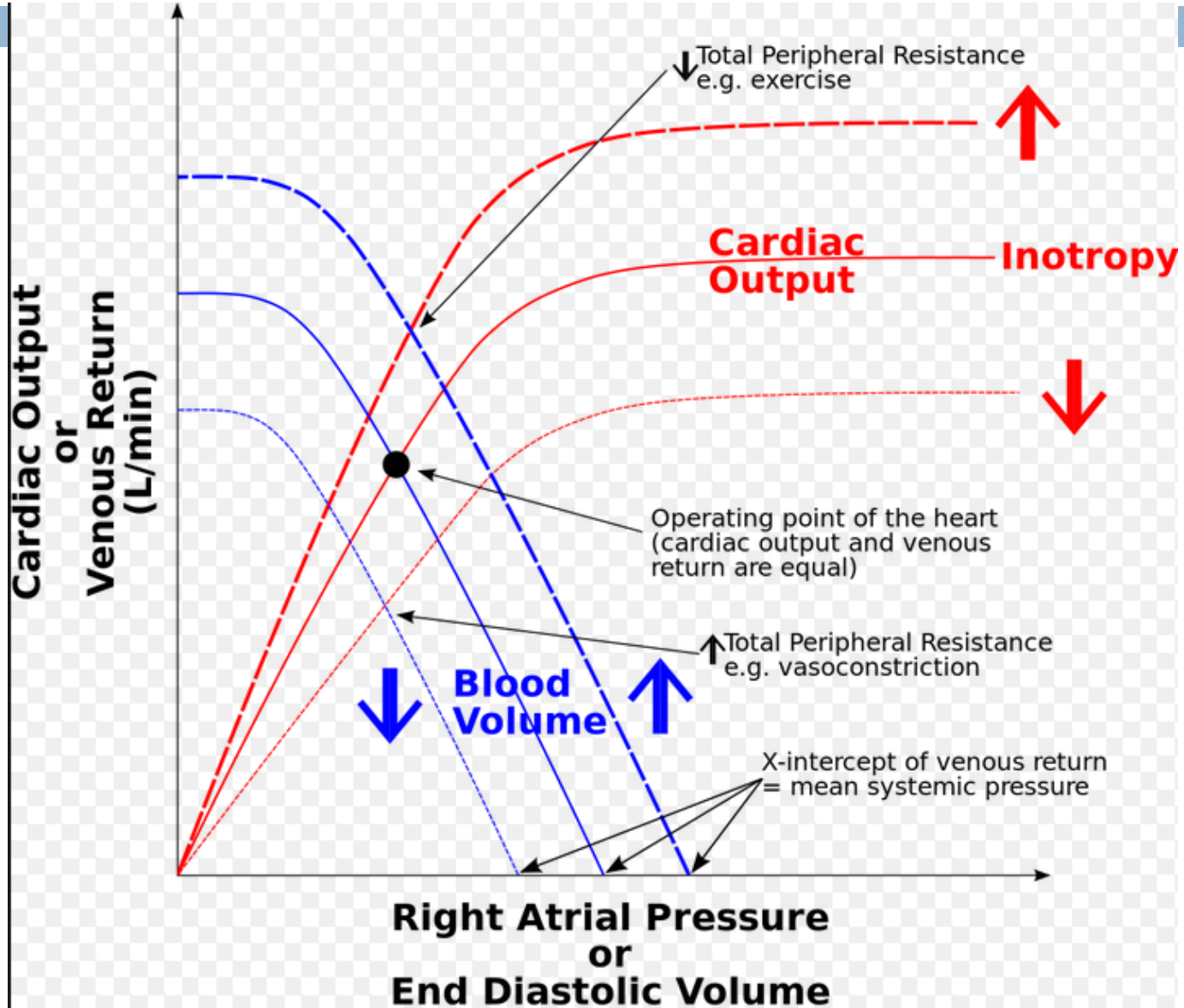
- Pulse oximetry
- 12-lead EKG
- CXR
- Echocardiography
- Labs:
  - BNP
  - CRP
  - TNF-alpha

**Know how an imaging study of the chest may help diagnose CHF!!

Heart Failure- Management **

- Treat the Cause!!
- Goals: maximize CO, maximize tissue perfusion, limit myocardial oxygen consumption
  - Reduce afterload stress (decrease SVR)
    - ACE inhibitors
    - ARB's
    - PDE inhibitors
    - Nitrates
  - Reduce EXCESSIVE preload stress
    - Diuretics
  - Inhibit sympathetic activation
    - Beta-blockers
  - BNP analogs
    - Nesiritide
  - Inotropy
    - Digoxin

Frank Starling



Heart Failure- Management

Table 3. Principles of Managing Heart Failure

Recognition and Treatment of Underlying Systemic Disease

Timely Surgical Repair of Structural Anomalies

Afterload Reduction

- Angiotensin–converting enzyme inhibitors
- Angiotensin receptor blockers
- Milrinone
- Nitrates
- Brain natriuretic peptide (BNP)

Preload Reduction

- Diuretics
- BNP

Sympathetic Inhibition

- Beta blockers
- BNP
- Digoxin

Cardiac Remodeling Prevention

- Mineralocorticoid inhibitors

Inotropy

- Digoxin

Question 9

You are seeing a 4-week-old previously healthy infant in your office because of concern about poor feeding. On questioning, the parents report that the child has developed grunting respirations associated with feedings, diaphoresis, pallor, and prolonged periods of sleep. On physical exam, HR: 160, RR: 55, BP (R arm): 75/48, BP (L leg): 88/55, oxygen sat: 95%. He exhibits tachypnea, rales and retractions, a II/VI low-pitched holosystolic murmur across the precordium, and a palpable liver 2 cm below the right costal margin. Of the following, the MOST likely explanation of the child's findings of congestive heart failure is:

a. Aortic valve stenosis
b. Coarctation of the aorta
c. Tetralogy of Fallot
d. Transposition of the Great Arteries
e. Ventricular septal defect

Question 12

A 6-year-old previously healthy boy presents with a recent development of nocturnal dyspnea. On questioning of his parents you discover that the child has experienced exercise intolerance, two episodes of syncope while running, poor appetite, and a cough without congestion for the past year. Exam reveals HR: 120, RR: 26, a gallop rhythm, a III/VI high pitched blowing systolic murmur at the apex, hepatomegaly, and diminished pulses. Chest radiography documents an enlarged cardiac silhouette with pulmonary vascular congestion. An echo demonstrates a regurgitant mitral valve with a dilated left ventricle and markedly reduced systolic contractility. Of the following, the most likely cause of this child's dilated cardiomyopathy:

a. Congenital mitral valve abnormality
b. Duchenne's muscular dystrophy
c. Freidreich's ataxia
d. Rheumatic heart disease
e. Sickle cell disease

Question 4

The symptom MOST suggestive of early heart failure in a 2-month-old infant is:

a. Apnea
b. Clubbing
c. Pedal Edema
d. Seizure
e. Slowed feeding

Question 16

Included in your rounds today is a 36-hour-old boy who was born at term by normal spontaneous vaginal delivery. His RR 80, HR 168. He has easily palpable bounding pulses in all 4 extremities. His BP 72/30. Precordial examination reveals a lift and a 3/6 systolic ejection murmur at the upper left sternal border. You also note a murmur over the anterior fontanelle. Of the following, the most likely diagnosis is:

a. Aortic coarctation with congestive heart failure
b. Aortic insufficiency
c. Large VSD with congestive heart failure
d. Left-to-right extracardiac shunting with congestive heart failure
e. Right-to-left extracardiac shunting with right heart failure

****Understand the association between systemic AVM and CHF in a newborn infant!!**

Question 19

You are seeing a 6-week-old infant who was born with Trisomy 21 and a large AV septal defect. Over the previous week, she has tired with feeding and has not gained weight. RR: 60, HR 150. Auscultation reveals mild retractions and a II/VI systolic murmur with a gallop rhythm. The liver is palpable at 2-cm below the costal margin, and the perfusion is good. You decide to increase the caloric content of the formula to 24 kcal/oz, and you contact her pediatric cardiologist to discuss referral for surgical repair. Of the following, the best therapeutic option while awaiting surgical repair is:

a. Captopril
b. Furosemide
c. Hydralazine
d. Propranolol
e. Verapamil

Congenital Heart Disease

Question 13

You are caring for a 2-year-old girl who has cardiomyopathy and is awaiting cardiac transplantation. She is receiving a continuous infusion of milrinone at 0.5 mcg/kg/min, IV furosemide three times a day, and 2L/min of oxygen administered via nasal cannula. On physical exam, Temperature: 39°C, HR: 130, RR: 30, BP: 80/40, 02 sat: 92%. An arterial blood gas shows a pH 7.35, $PaCO_2$ 40, $Pa0_2$ 50 with a hemoglobin of 8. Of the following, the treatment that can BEST increase her tissue oxygen delivery is:

a. Administration of 10 mg/kg acetaminophen
b. Increased furosemide administration to four times a day
c. Increased oxygen flow to achieve an oxygen sat of 95%
d. Reduction of the milrinone infusion to 0.25 mcg/kg/min
e. Transfusion with 15 ml/kg of packed red blood cells

Question 22

A 3-year-old girl presents for a health supervision visit. At birth, she was diagnosed with hypoplastic left heart syndrome and underwent uncomplicated, staged surgical palliation. Today, her mother asks you if her daughter's heart disease could affect her development. Of the following, you are MOST likely to advise the mother that:

a. A small percentage of children who undergo neonatal heart surgery may develop transient motor delay
b. Cognitive problems are rare with neonatal heart surgery
c. Evaluation for developmental delay should wait until after kindergarten
d. Speech and behavioral disorders are common among those who undergo neonatal heart surgery
e. The only children who sutter developmental delay after neonatal heart surgery are those who have pre- or post-operative complications

****Understand the prognosis for cognitive development in patients with cyanotic congenital heart disease!!**

Question 10

Yesterday you received a call from the newborn nursery that they were referring to you a term infant who was being discharged at 4 days of age. The female newborn's birth weight was 3 kg, and the delivery was by repeat C-Section. Findings on examination at discharge include heart rate, respiratory, and blood pressures were normal. Her lungs were clear and no murmurs were noted. She was breastfeeding without difficulty. Today, her mother calls to tell you that she is difficult to awaken, pale, and breathing much more rapidly than she was in the hospital nursery. She has had one wet diaper in the past 12 hours. When you meet them in the emergency department, you note that the infant has cool extremities, weak pulses, and lethargy. Of the following, the most likely cause of this newborn's condition is:

a. Aortic coarctation
b. Atroventricular septal defect
c. Tetralogy of Fallot
d. Transposition of the Great Arteries
e. Ventricular septal defect

**** Know that shock maybe the initial finding in a newborn infant with congenital heart disease**