

William Herring, M.D. © 2003

Cyanotic Heart Disease

In Slide Show mode, to advance slides, press spacebar
or click left mouse button

Cyanosis With Decreased Vascularity

- Tetralogy
- Truncus-type IV
- Tricuspid atresia*
- Transposition*
- Ebstein's

* Also appears on DDx of Cyanosis with ↑ Vascularity

Cyanosis With Increased Vascularity

- Truncus types I, II, III
- TAPVR
- Tricuspid atresia*
- Transposition*
- Single ventricle

* Also appears on DDx of Cyanosis with ↓ Vascularity

Tetralogy of Fallot

Tetralogy of Fallot

General

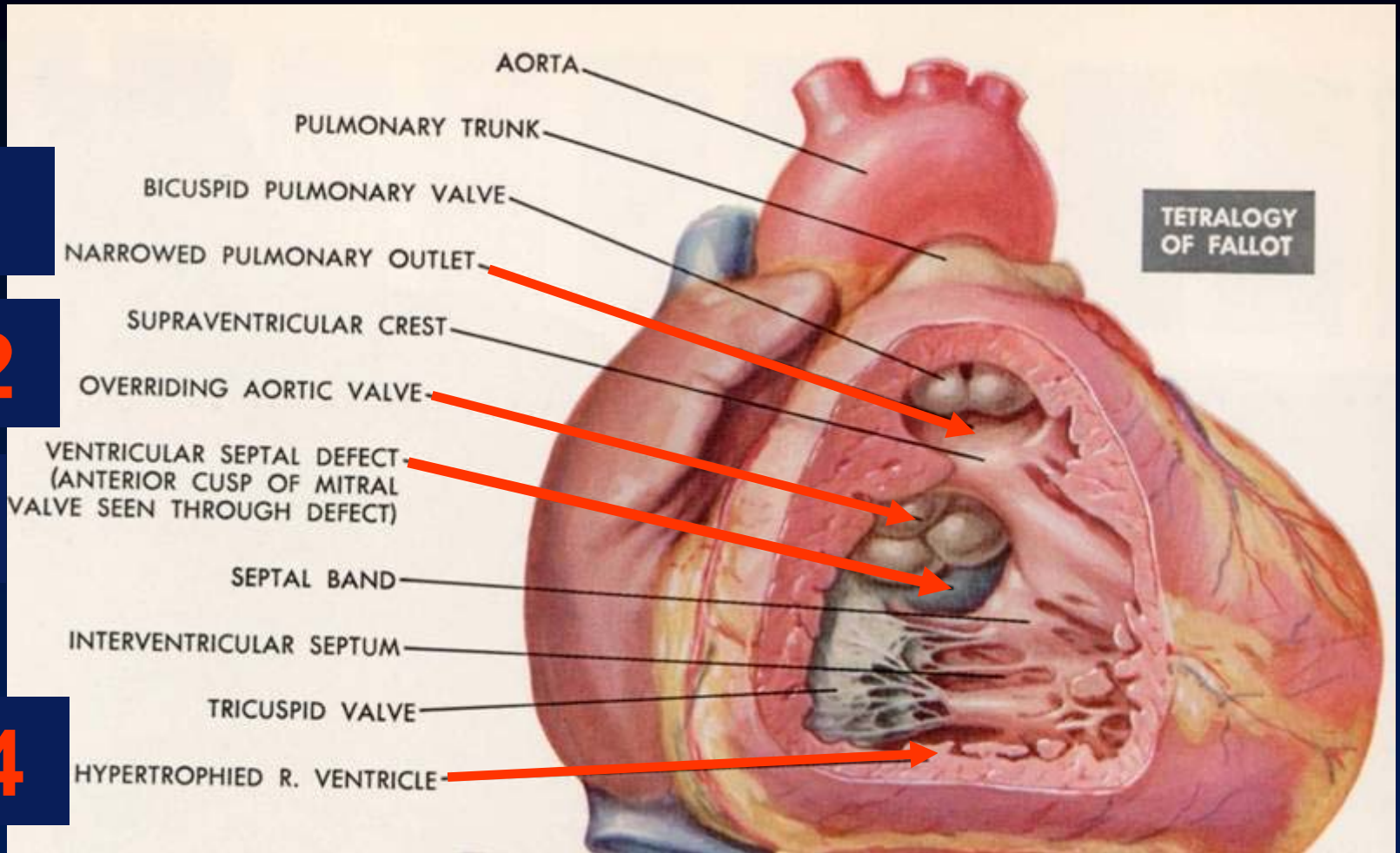
- **About 10% of all congenital heart lesions**
- **Most common cause of cyanotic heart disease beyond neonatal period**

Tetralogy of Fallot

Components

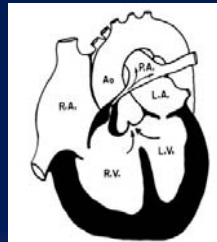
- High VSD
- Pulmonic stenosis, i.e. right ventricular outflow obstruction
 - Usually infundibular, sometimes valvular
- Overriding of the aorta
- Right ventricular hypertrophy





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Tetralogy of Fallot



Tetralogy of Fallot

Other anomalies

- Right aortic arch in 25%
 - Mirror image type
- Left superior vena cava
- ASD
- Tricuspid valve abnormalities
- Anomalies of coronary arteries
 - Aberrant left anterior descending coronary artery arising from right coronary artery



Tetralogy of Fallot

Other anomalies

- **Abnormalities of the pulmonary artery and its branches**
 - **Peripheral PA coarctations, unilateral**
 - **Absence or hypoplasia of pulmonary artery**
 - **Usually left**
 - **Absence of pulmonic valve**
 - **Bicuspid pulmonic valve**



Tetralogy of Fallot

Critical Component

- Degree of pulmonic stenosis
 - Regulates degree of R → L shunt
 - Regulates overriding of aorta
 - Greater the stenosis, the greater the aortic overriding



Tetralogy of Fallot

Clinical findings

- Squatting
- Dyspnea
- Failure to thrive
- Cyanosis-usually
- Severe cases ↔ at birth ↔ severe PS
- Mild cases ↔ much later ↔ mild PS
- “Pink tets” (acyanotic) and “Blue tets” (cyanotic)

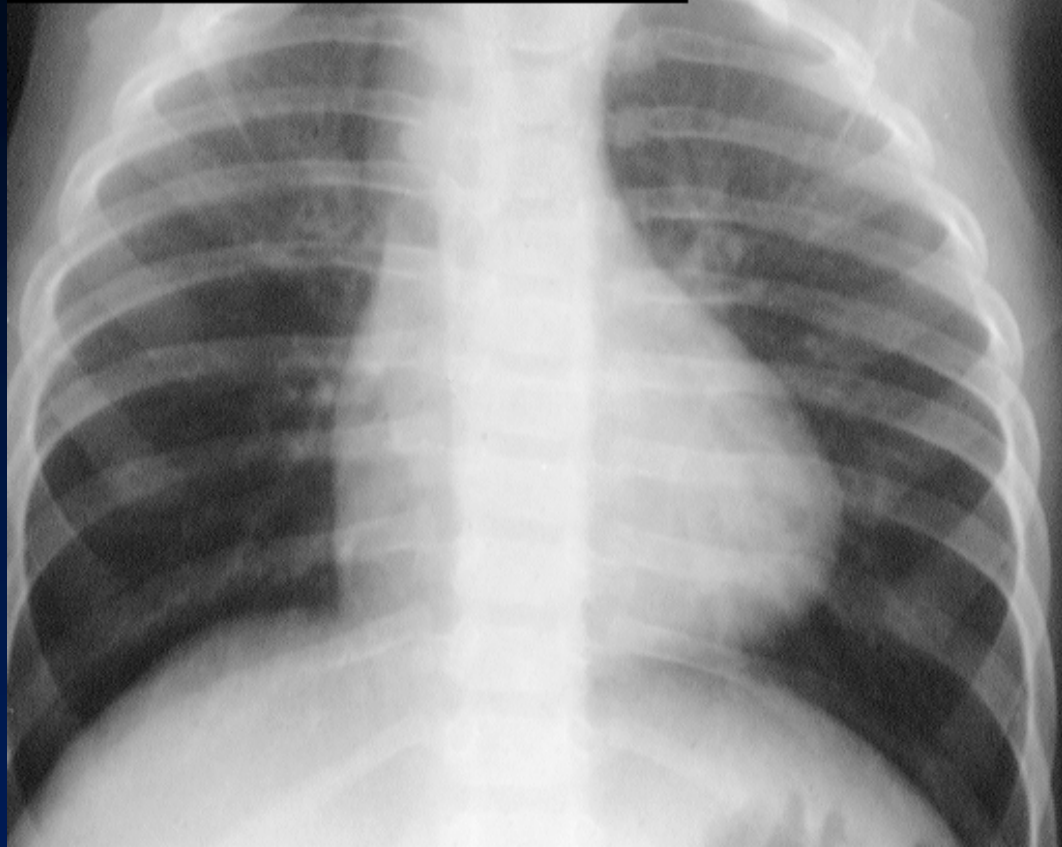


Tetralogy of Fallot

Imaging Findings

- Heart size normal
 - Rarely enlarged
- Cardiac apex displaced upward “coer en sabot”
- PA segment concave
- Decreased vasculature
- R aortic arch in 25%

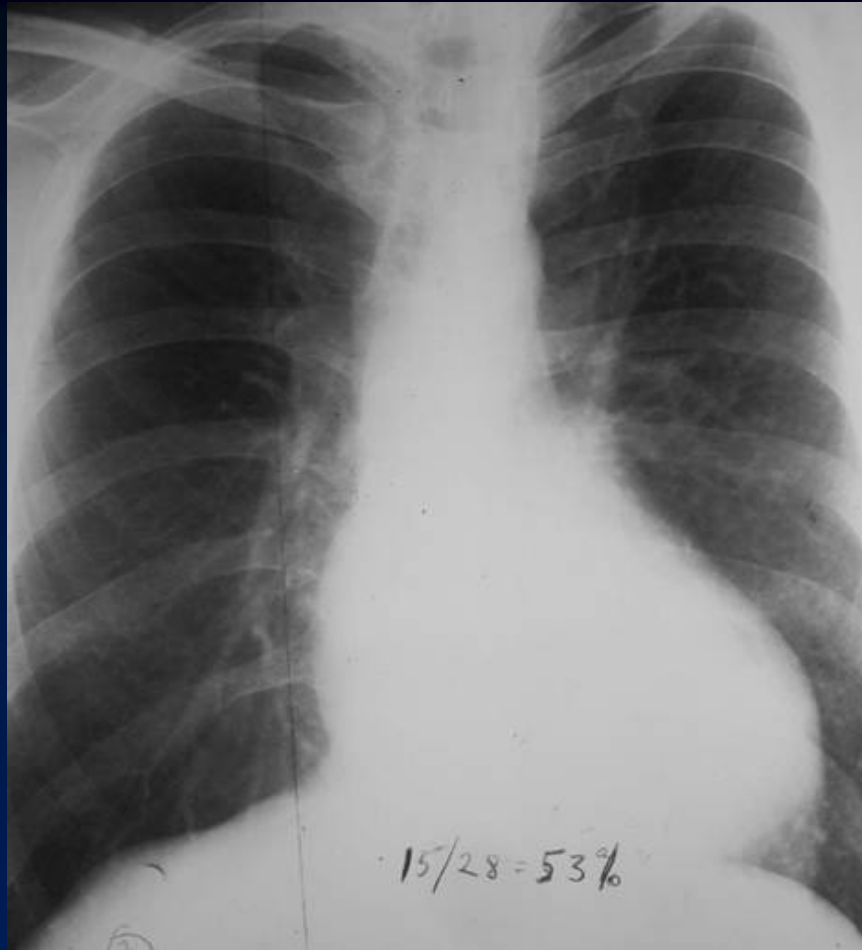




Tetralogy of Fallot



Tetralogy of Fallot



Tetralogy of Fallot

Trilogy of Fallot

- Pulmonic valvular stenosis
- ASD
- Right ventricular hypertrophy

Truncus Arteriosus

Truncus Arteriosus

Embryology

- **Uncommon anomaly 2° failure of primitive common truncus arteriosus to divide into aorta and pulmonary artery**

Truncus Arteriosus

General

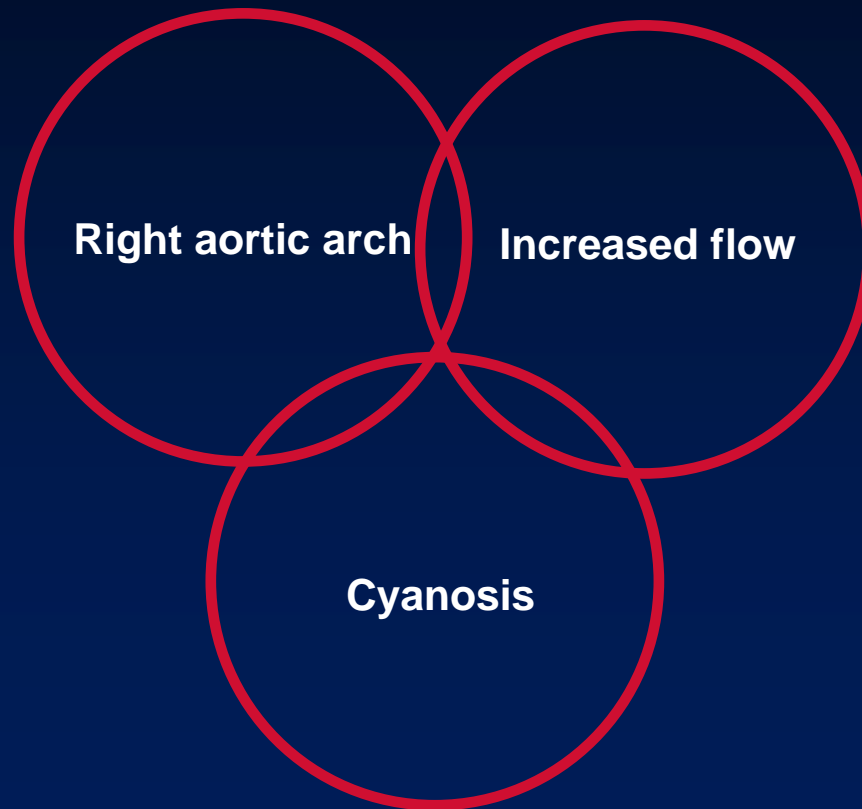
- **The truncal valve is usually tricuspid**
- **Main pulmonary artery segment is concave in types II, III, and IV**
- **Pulmonary vasculature is shunt type in types I, II and III**

Truncus Arteriosus

Right sided aortic arch

- **Right-sided arch in about 33%**
 - **Usually mirror image type**
- **But because truncus is so rare, it accounts for only 6% of all right arches**

Truncus Arteriosus Triad



Truncus Arteriosus

Associations

- **VSD**
 - **Always**
- **Anomalies of the coronary arteries**

Truncus Arteriosus

Clinical Findings

- Infants and small children demonstrate L → R shunt
 - Minimal cyanosis
 - CHF
 - Respiratory infections
 - Growth disturbances
- Majority are dead by 6 mos

Truncus Arteriosus

Clinical Findings

- **Cyanosis is worse in II and III**
 - Can't tell them apart clinically
- **Associated anomalies**
 - Bony
 - Renal
 - Lung
 - Cleft palate

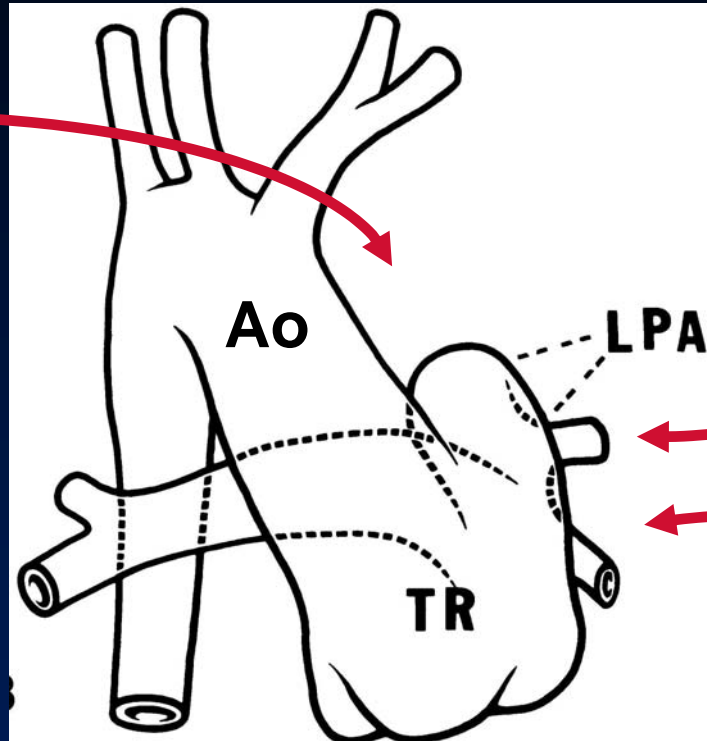
Truncus Arteriosus

X-ray Findings

- **Cardiomegaly**
- **Right aortic arch (33%)**
- **Concave pulmonary artery segment**
- **Enlarged left atrium in 50%**
- **Displacement of hilum**
 - **Elevated right hilum in 20%**
 - **Left hilum in 10%**

Most common (75%)

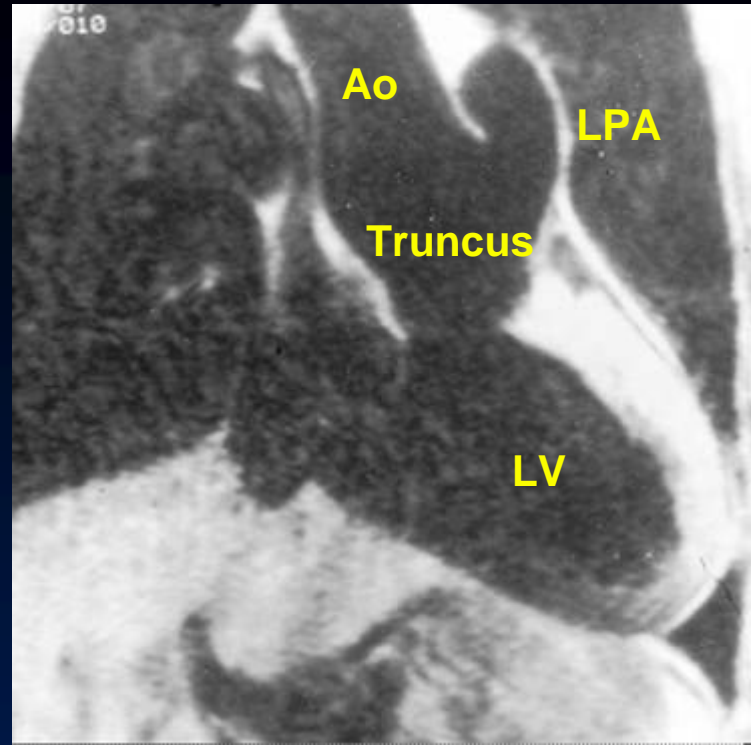
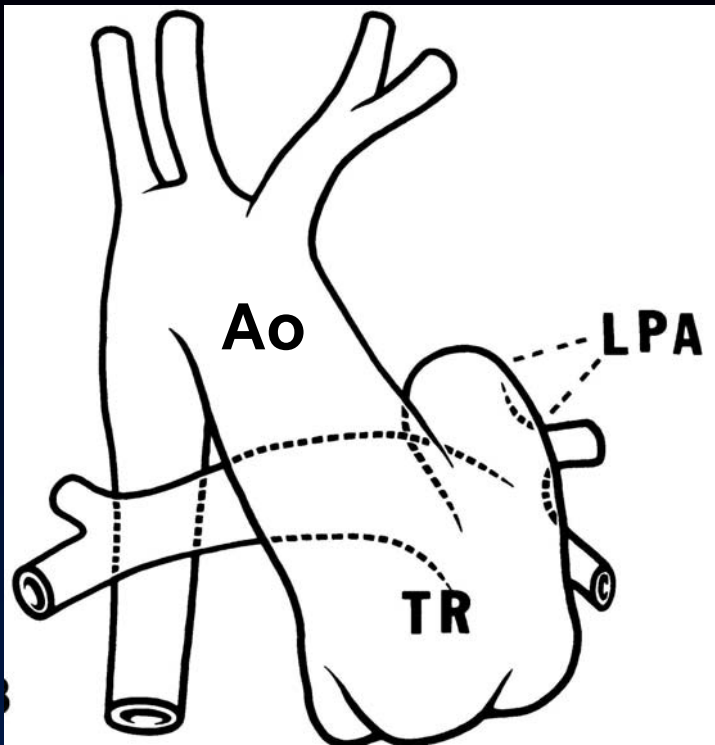
Shunt vessels



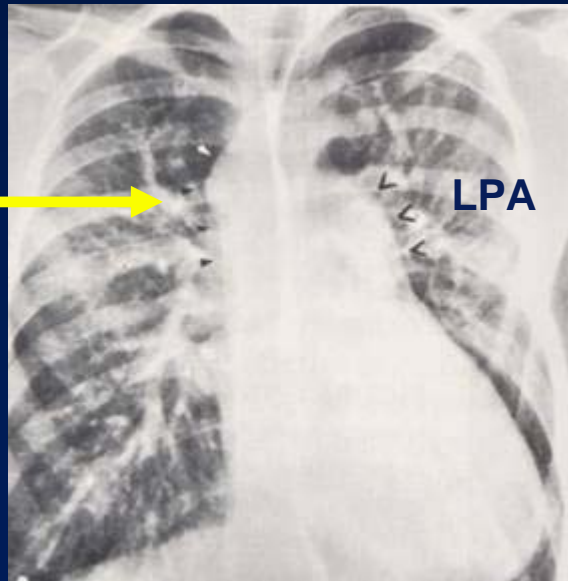
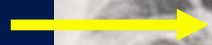
PA arises on left via short common stem

Convex pulmonary artery segment

Truncus-Type I



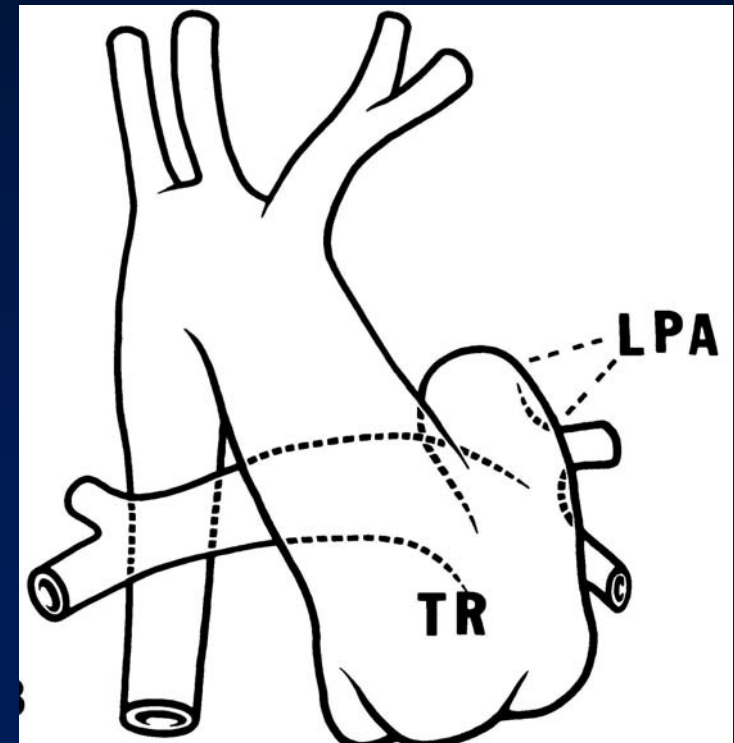
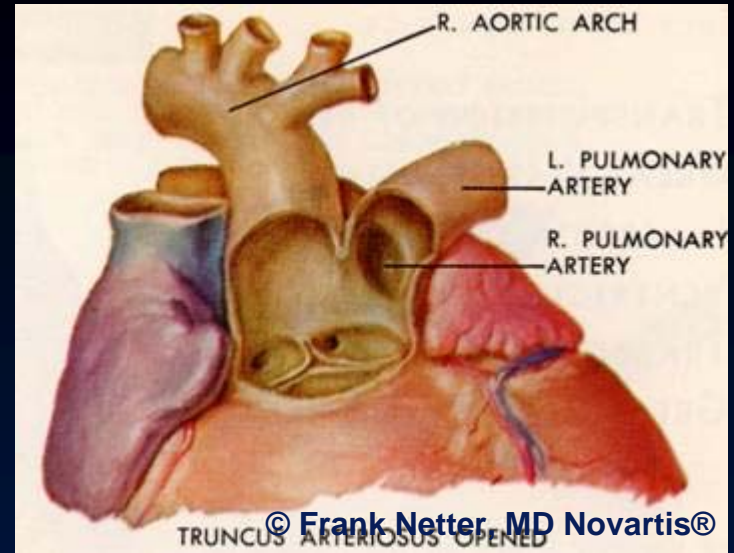
Right Ao Arch



Truncus Type 1

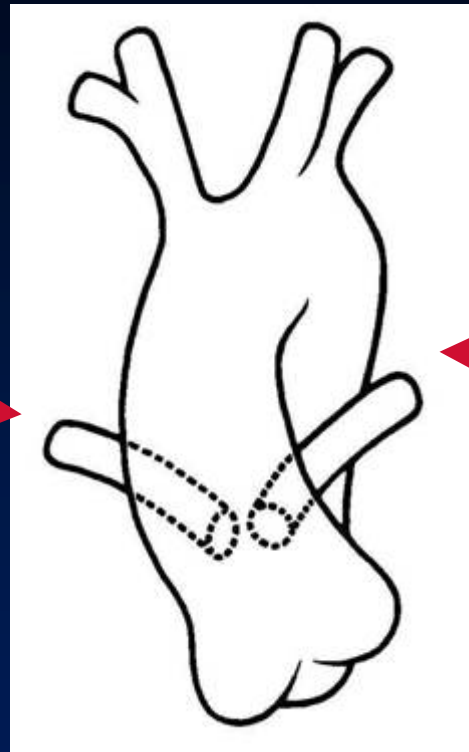


Truncus Type 1



Uncommon (25%)

**Pulmonary
arteries arise
posteriorly
from aorta**



Shunt vessels

**Concave main
pulmonary
artery**

Truncus-Type II



Truncus Type II

Rare (5%)

Right and left pulmonary arteries arise laterally



Shunt vessels

Concave main pulmonary artery

Truncus-Type III

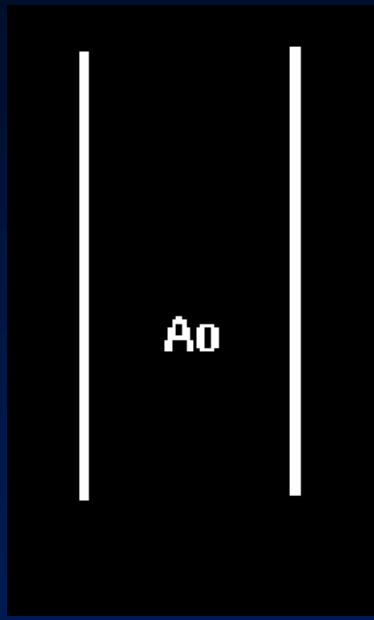


Truncus Type III

Rare to non-existent

No pulmonary arteries

Bronchial circulation



TOF with pulmonary atresia

Concave main pulmonary artery

Truncus-Type IV



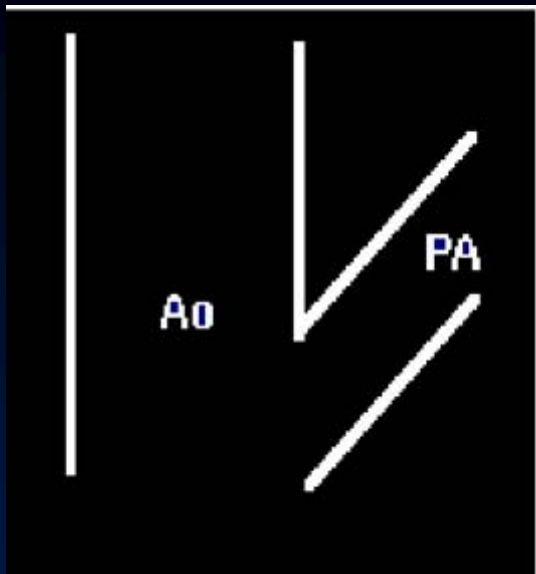
**Truncus-Type IV
(TOF with pulmonary atresia)**



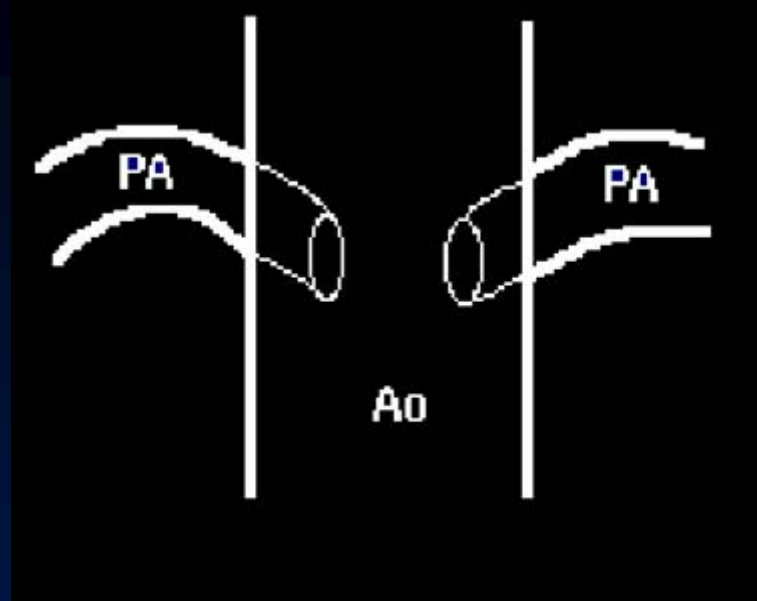
Bronchial Circulation



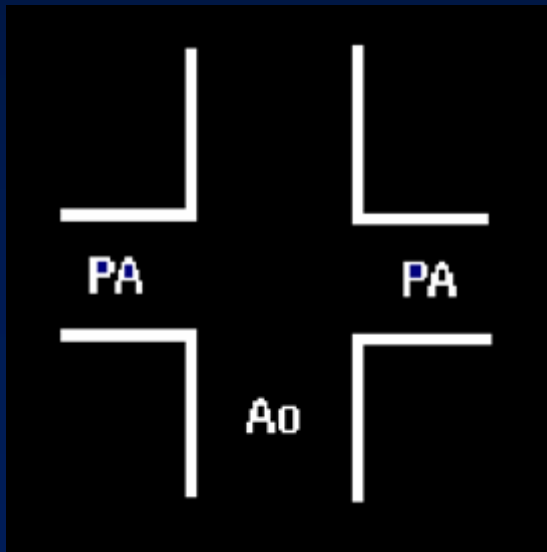
Increased flow



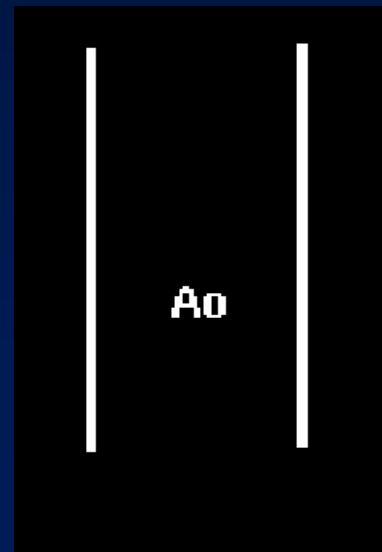
I



II



III



IV

Truncus Arteriosus

| | Pulmonary artery | Shunt vessels |
|------------|------------------|-----------------------|
| I | Convex | Yes |
| II | Concave | Yes |
| III | Concave | Yes |
| IV | Concave | Bronchial circulation |

Truncus Arteriosus

Angiographic Findings

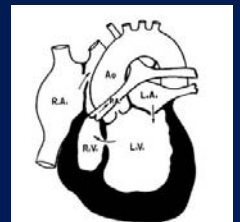
- On angiogram, truncal valve (common valve) may have 2-6 cusps
 - Most often tricuspid
- In lateral projection, plane of truncal valve is distinctive
 - Tilts anteriorly toward patient's toes
 - Helpful to distinguish truncus from aorticopulmonary window or TOF with pulmonic atresia



Tricuspid Atresia

Tricuspid Atresia

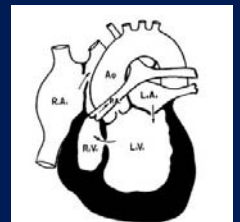
- Fusion of dorsal and ventral endocardial cushions occurs too far to the right → obliteration of tricuspid valve, and
- Hypoplasia of right heart
 - Tricuspid valve, Right ventricle and pulmonary artery



Tricuspid Atresia

Shunts needed

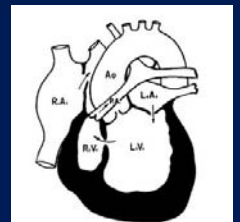
- **Complete obstruction to outflow from RA**
 - **Need R → L shunt: Patent foramen ovale or ASD**
 - **Small ASD → elevated RA pressures and enlarged RA**
 - **Large ASD → lower RA pressures and minimal enlargement of RA**
- **Blood in L heart must get back to lungs**
 - **Also have associated VSD or PDA**



Tricuspid Atresia

Transposition of Great Vessels

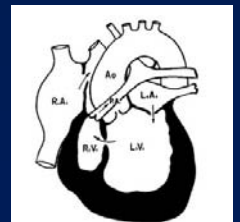
- 70% have normal relationships of great vessels
- 30% have transposition of great arteries

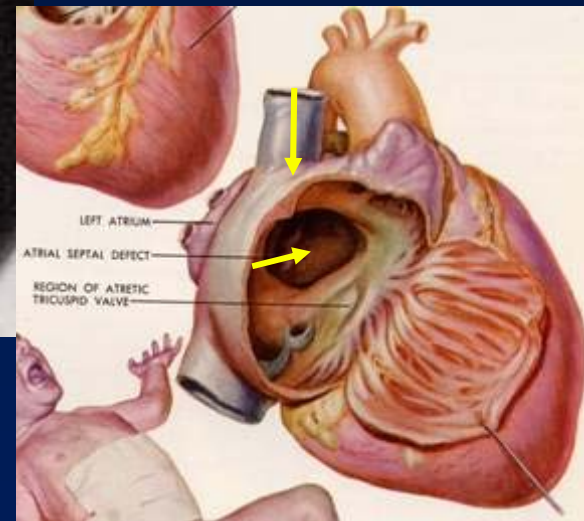
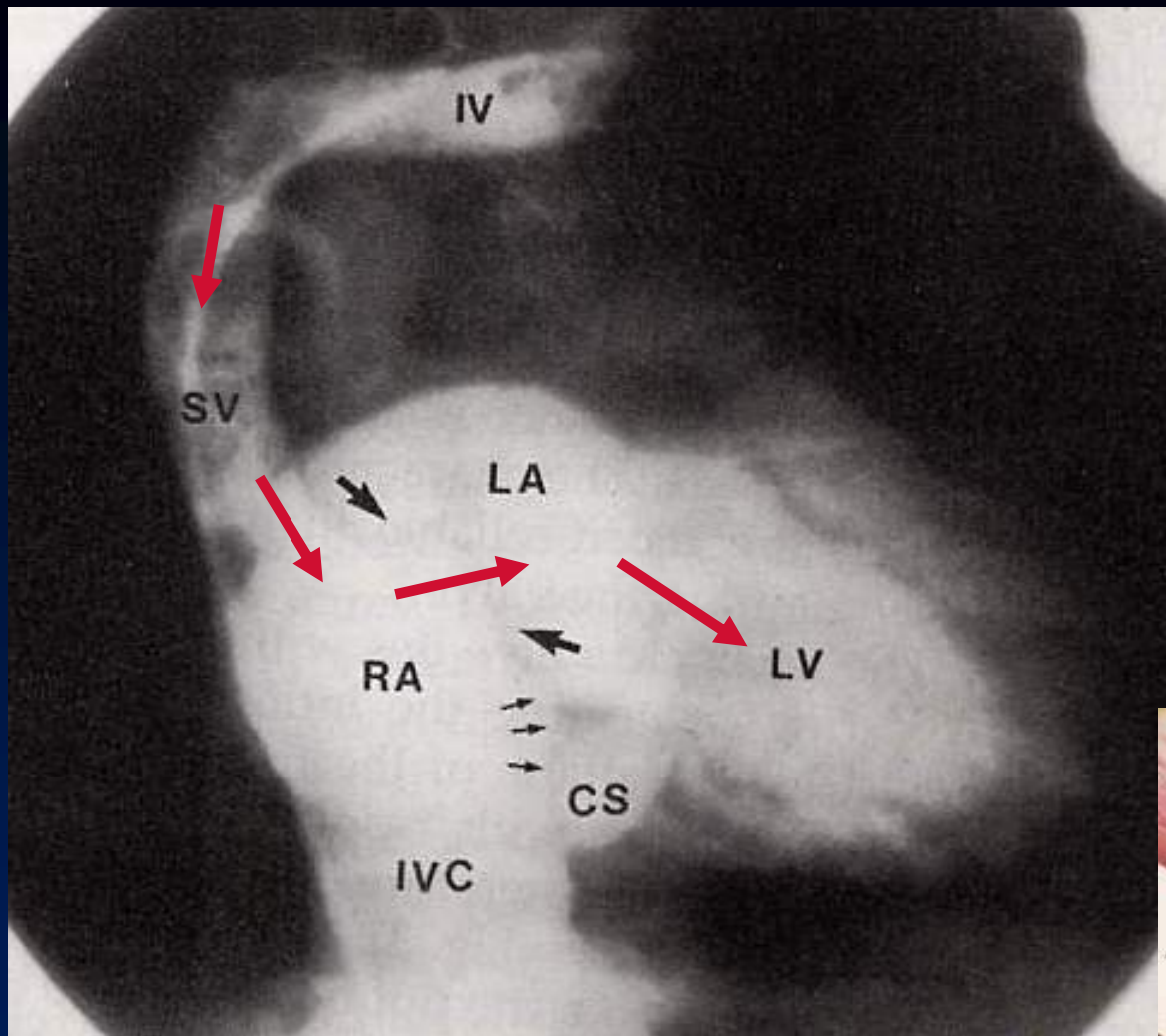


Tricuspid Atresia

Two main types

- **No Transposition of the Great Arteries**
 - Some degree of PS
 - Majority (70%)
- **Transposition of the Great Arteries**
 - No pulmonic stenosis
 - Minority (30%)





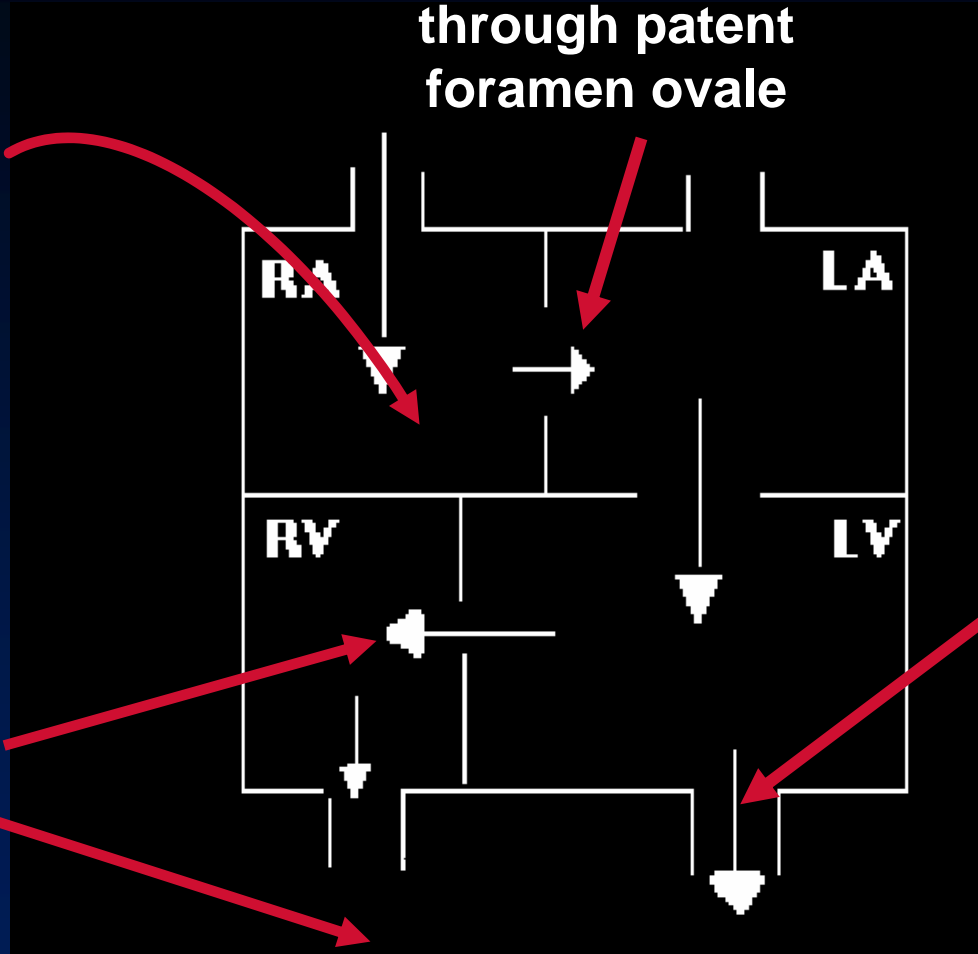
Tricuspid atresia—no transposition

Systemic blood can not enter RV

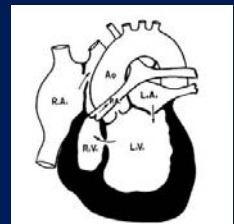
Need R → L shunt through patent foramen ovale

Blood returns to right heart and then lungs either through VSD or PDA

Some unsaturated blood exits aorta



Tricuspid atresia—no transposition

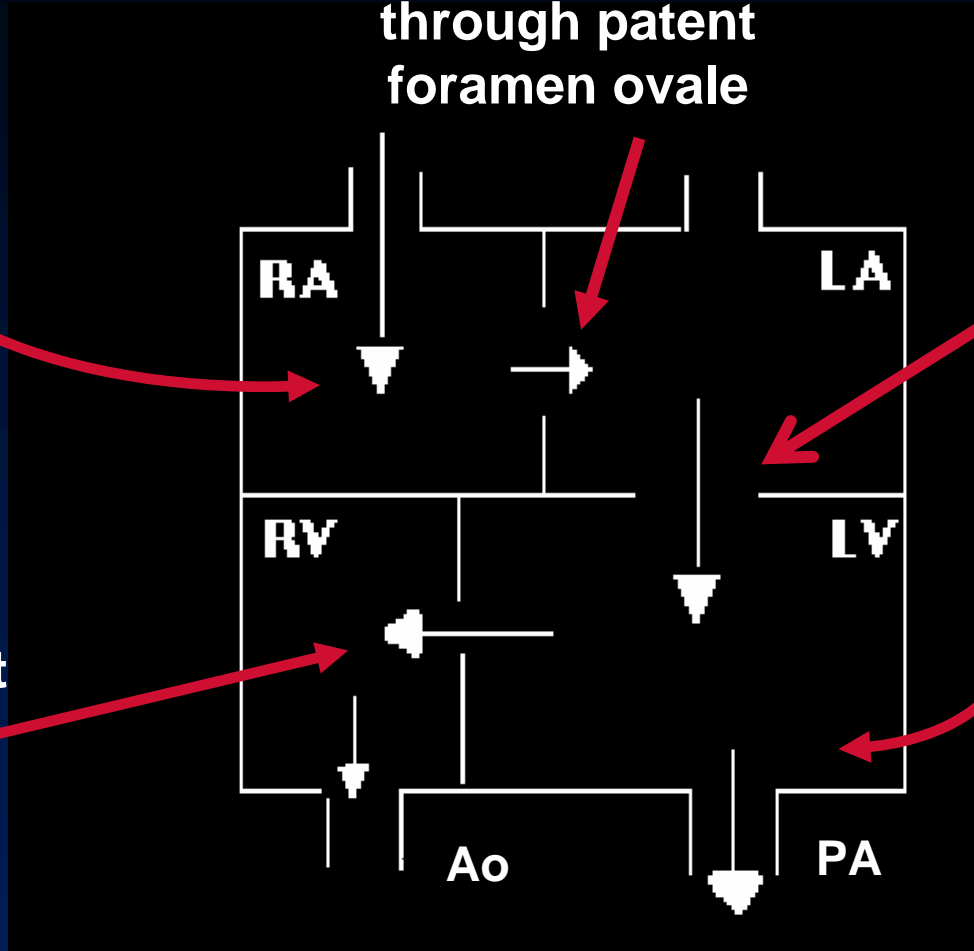


Systemic blood can not enter RV

Need R → L shunt through patent foramen ovale

Oxygenated blood returns to LA

Need L → R shunt to get blood into body



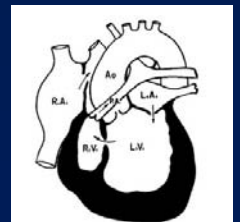
Un-oxygenated blood flows to lung via transposed PA → ↑ flow

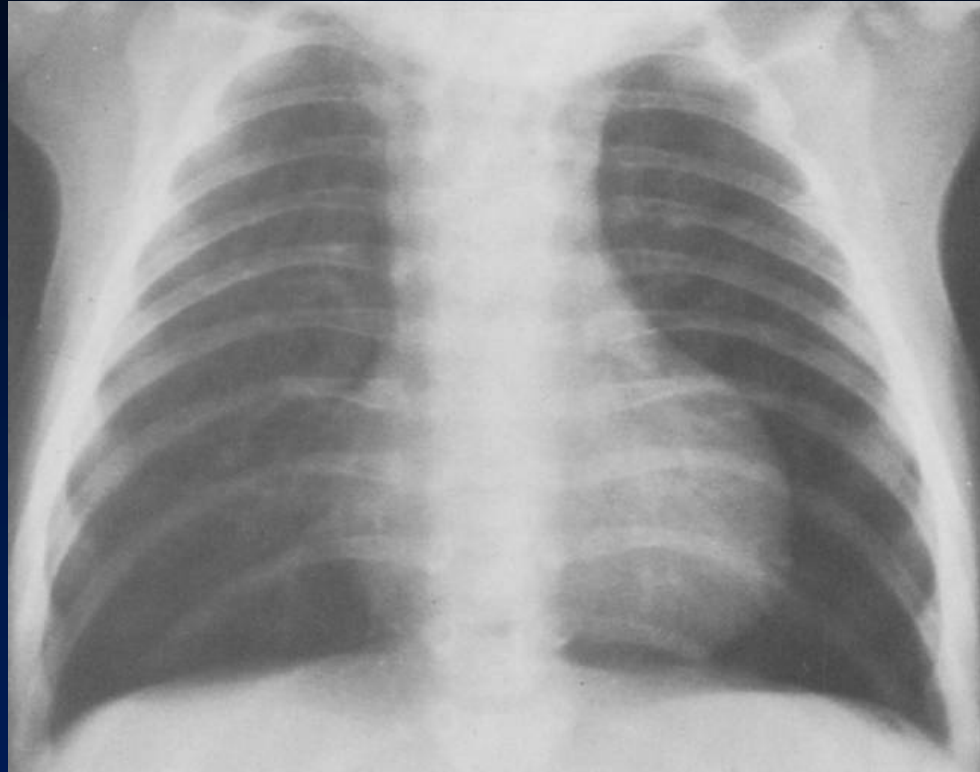
Tricuspid atresia—with transposition

Tricuspid atresia

X-ray Findings - No transposition

- Normal-sized heart
- Decreased pulmonary vessels (60-70%)
- Flat/concave pulmonary artery
- Small ASD → enlarged RA
- Large ASD → normal or slightly enlarged RA



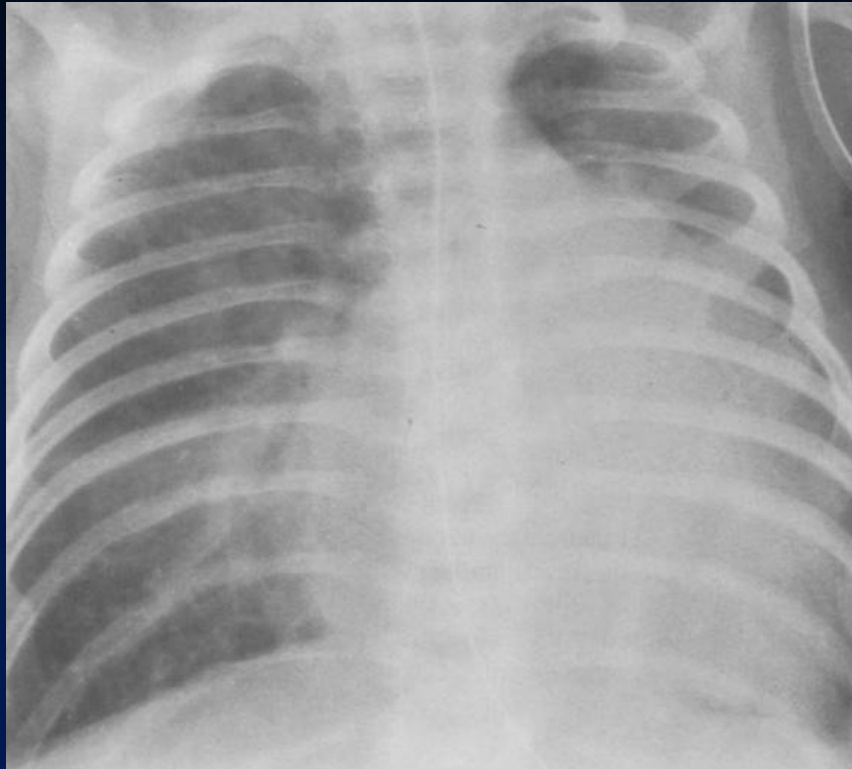


Tricuspid atresia—some PS, no transposition

Tricuspid atresia

X-ray Findings - Transposition

- **Mild cardiomegaly**
- **Engorged pulmonary vessels**
- **No characteristic appearance to heart**



Tricuspid atresia—no PS, shunt vessels

Transposition of The Great Vessels

The “TR” Lesions

Cyanosis with ↑ or ↓ vasculature

- **Tricuspid atresia** ↑ or ↓
- **Transposition** ↑ or ↓
- **Truncus arteriosus**
 - Type I, II, III ↑
 - Type IV ↓
- **Tetralogy** ↓
- **TAPVR** ↑
- **TrEbstein’s** ↓

The Rules

- **Since anatomic side (i.e. “left” or “right”) in complex lesions is frequently reversed or indeterminate**
- **Naming conventions for**
 - **Atria**
 - **AV valves**
 - **Ventricles**
 - **Ventricular outflow tracts**

The Rules

How the atria are named

- **Anatomic right atrium is on the side of the trilobed lung and liver**
 - **Trilobed lung=upper, middle and lower**
- **The anatomic left atrium is on the side of the bilobed lung and spleen**
 - **Bilobed lung=upper and lower**

The Rules

Mitral and tricuspid valves

- **The tricuspid valve belongs to the anatomic right ventricle**
 - **Not right atrium**
- **The mitral valve belongs to the anatomic left ventricle**
 - **Not left atrium**

The Rules

How the ventricles are named

- **The anatomic right ventricle is the trabeculated ventricle**
- **The anatomic left ventricle is the smooth-walled ventricle**

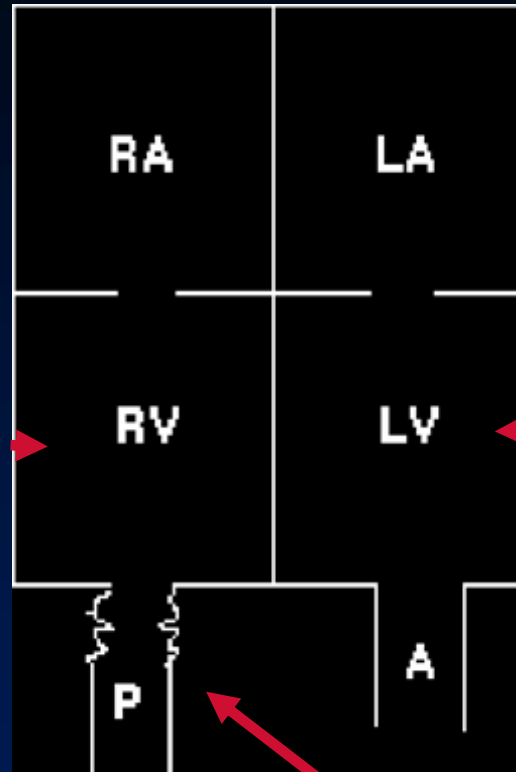
The Rules

Aortic and pulmonic valves

- **The pulmonic valve is part of pulmonary artery**
 - **Not anatomic right ventricle**
- **The aortic valve is part of aorta**
 - **Not anatomic left ventricle**
- **The pulmonic infundibulum is part of anatomic right ventricle**

Anatomic
Right ventricle
is trabeculated

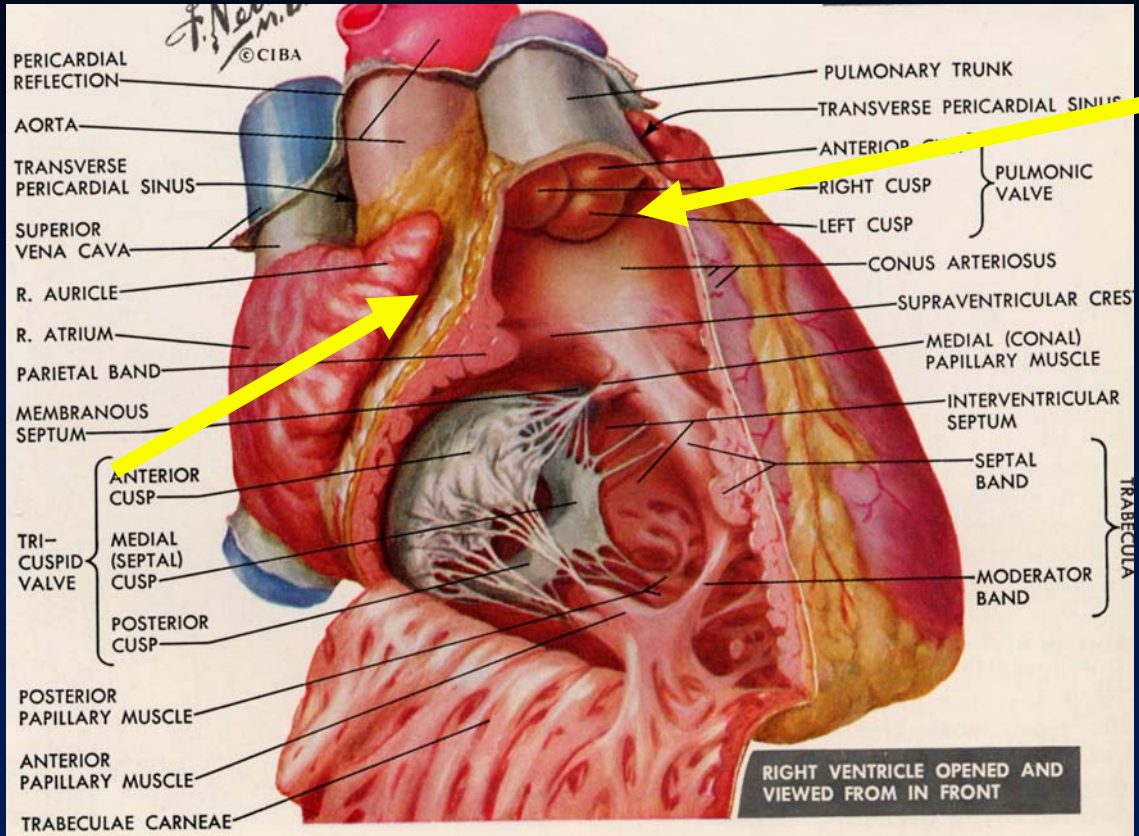
Anatomic
Left ventricle is
smooth



Pulmonic
infundibulum
always stays with
the anatomic R
ventricle

Normal heart

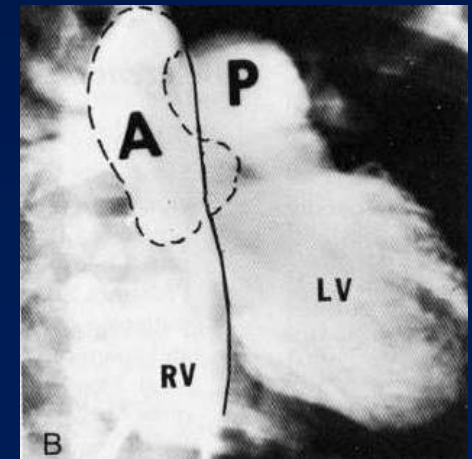
Normal relationship of aortic to pulmonic valves



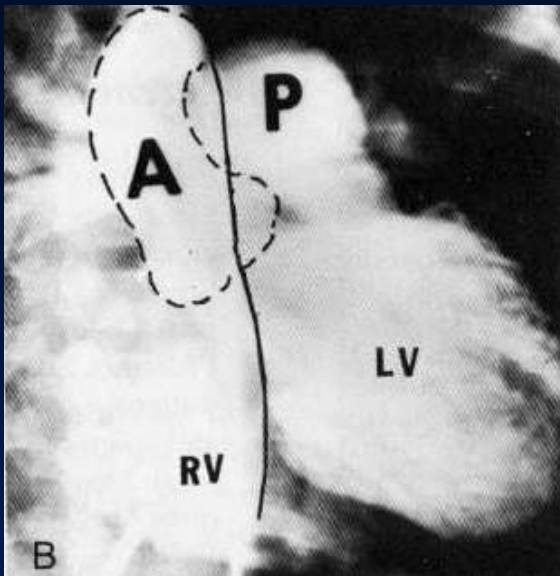
Pulmonic valve is
Anterior
Lateral
Superior
To the aortic valve

PALS

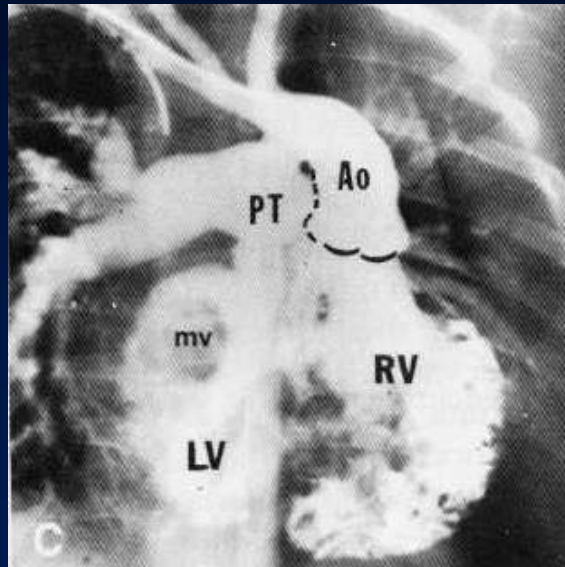
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In Transposition, pulmonic valve is



Normal



Corrected Transposition

Posterior

Medial

Inferior

To the aortic valve

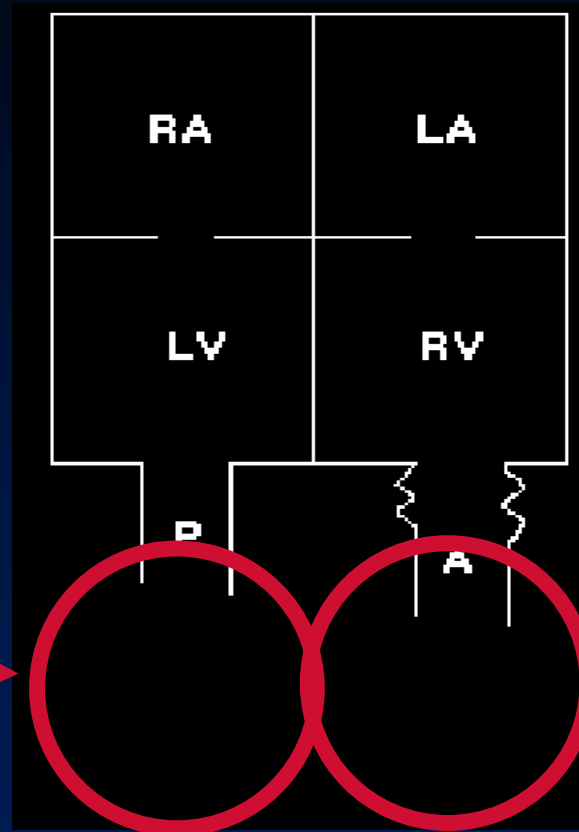
Corrected Transposition

Inversion of the Ventricles With Transposition

- Physiologically flow is normal
- Consistent with normal life, except
- Frequently associated with
 - VSD
 - Tricuspid insufficiency
 - Subpulmonic stenosis
 - Complete heart block

**Normal
vasculature;
or increased
with VSD**

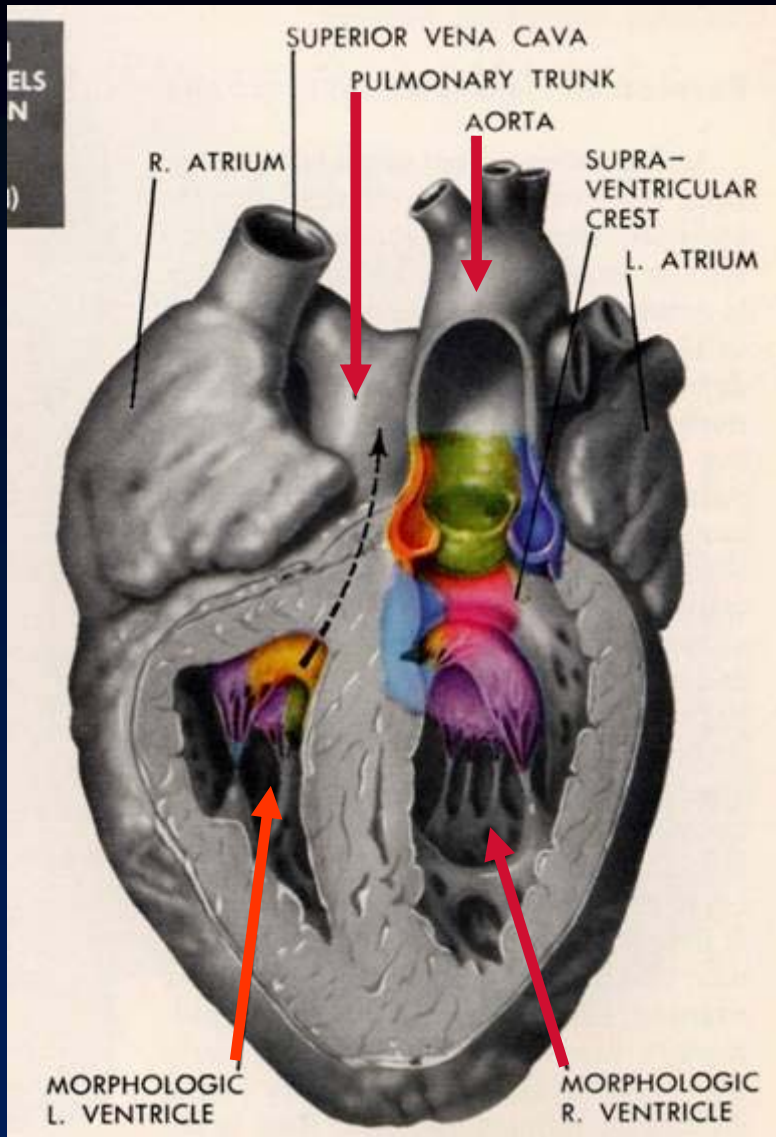
PA arises from
anatomic left
ventricle



Acyanotic

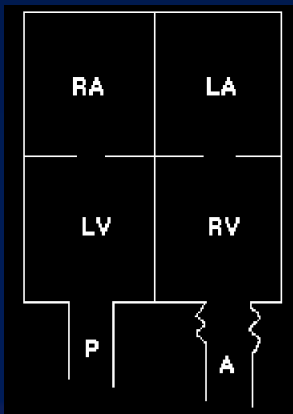
Aorta arises
from pulmonic
infundibulum

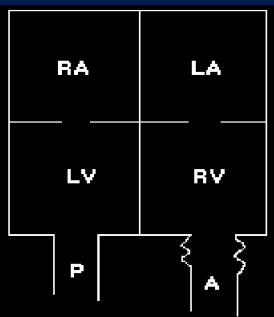
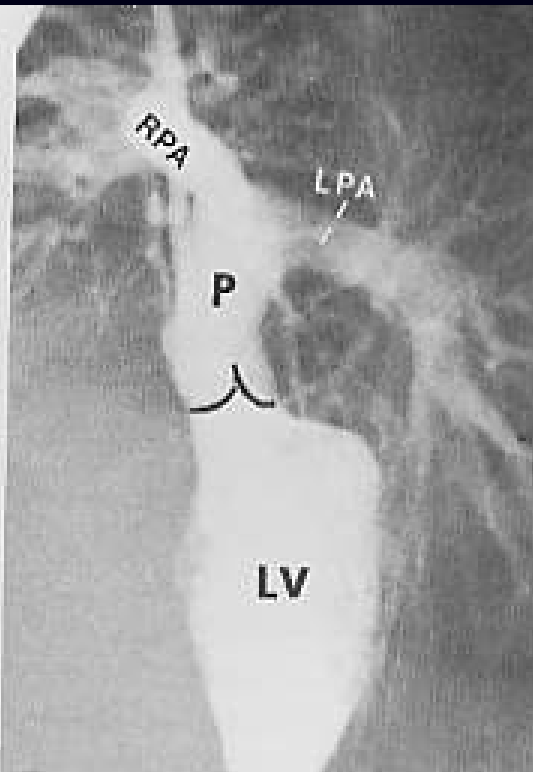
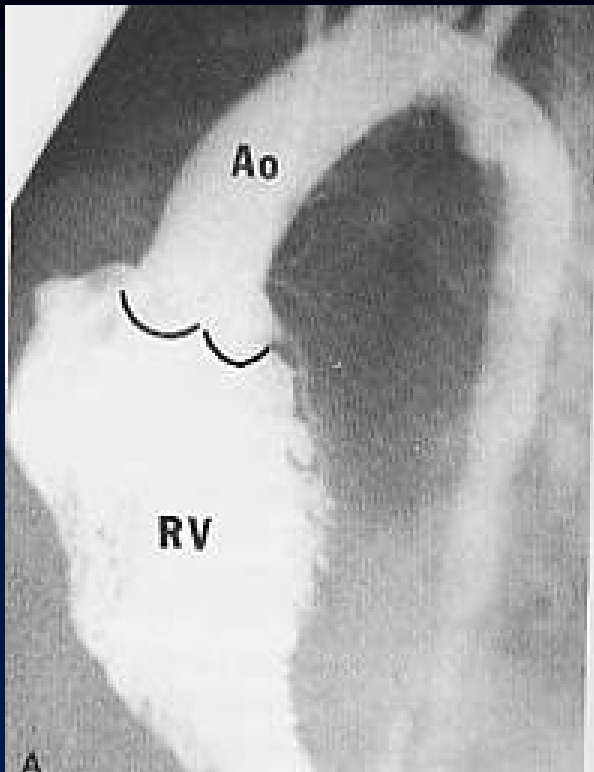
**Corrected Transposition (L-Trans)
Inversion of the Ventricles with Transposition of the
Great Vessels**



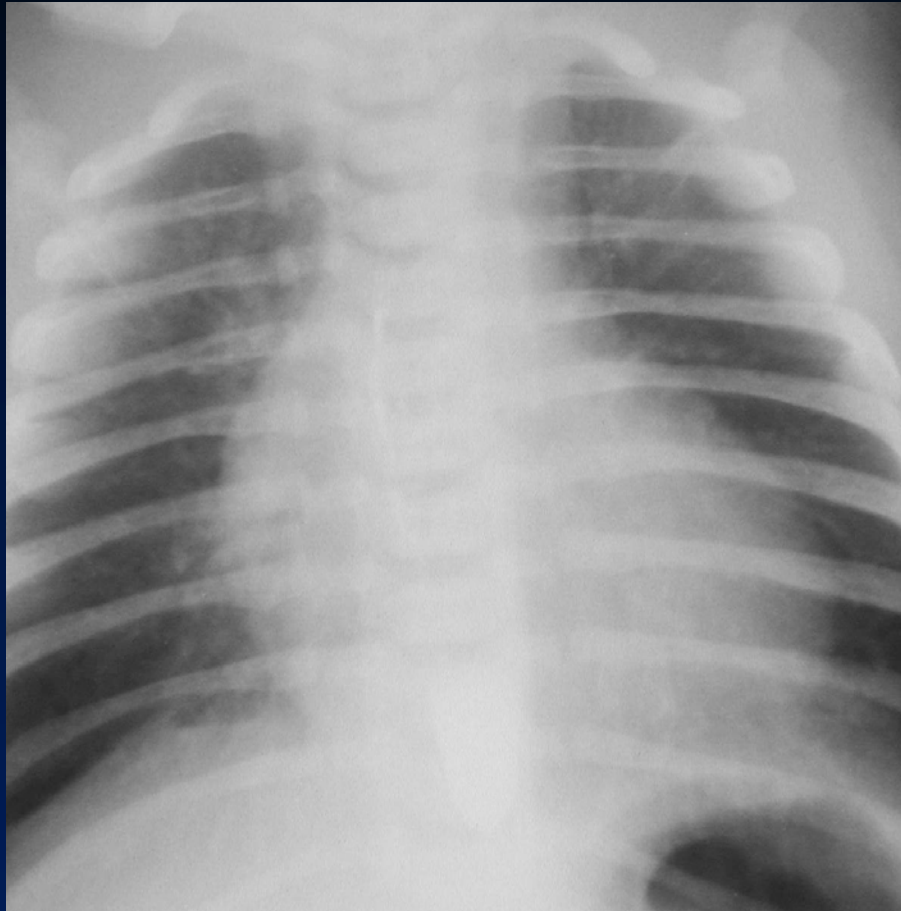
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Corrected Transposition





Corrected Transposition

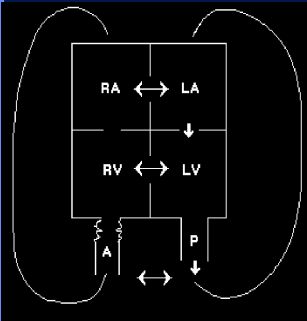


Corrected Transposition

Transposition of the Great Arteries

General

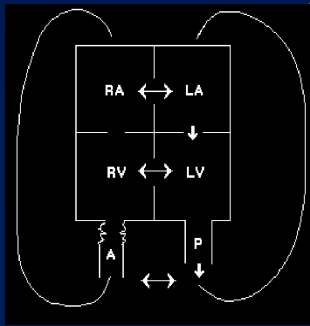
- Second most common cause of cyanosis in infancy
- Pulmonary and systemic circulations form two separate circuits
- Must be mixing between two circuits for life



Transposition of the Great Arteries

Associated abnormalities

- About 1/3 have VSD
 - Larger the shunt, more likely CHF
- About 1/4 to 1/2 have patent ductus
- Some have ASD
- Other major finding is obstruction to blood entering pulmonary artery
 - Usually subpulmonic stenosis

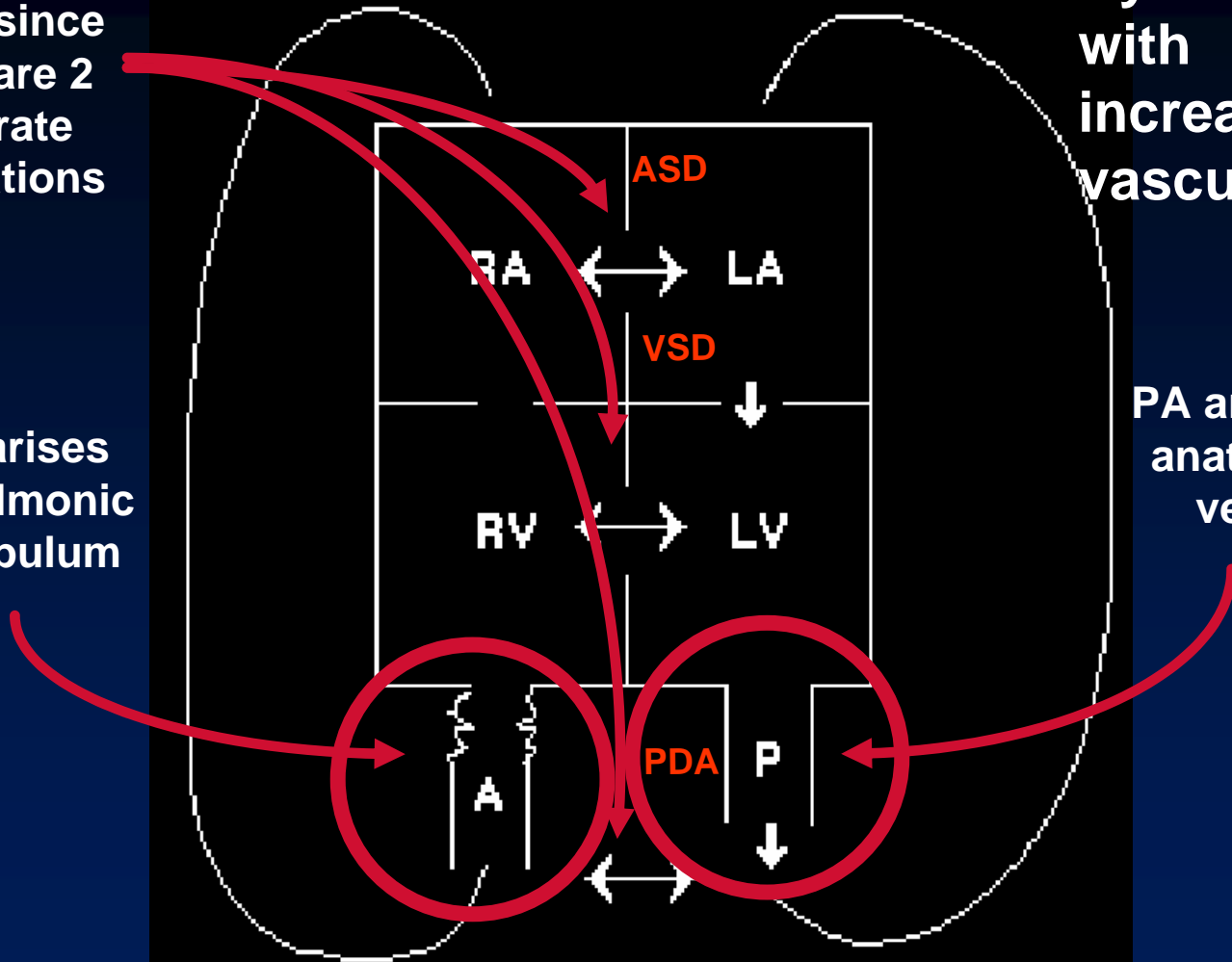


Obligatory shunt since there are 2 separate circulations

Cyanotic with increased vasculature

Aorta arises from pulmonic infundibulum

PA arises from anatomic left ventricle

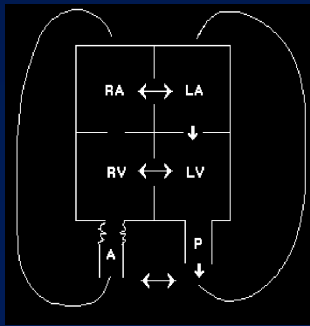


Transposition of the Great Vessels (D-Trans)

Transposition of the Great Arteries

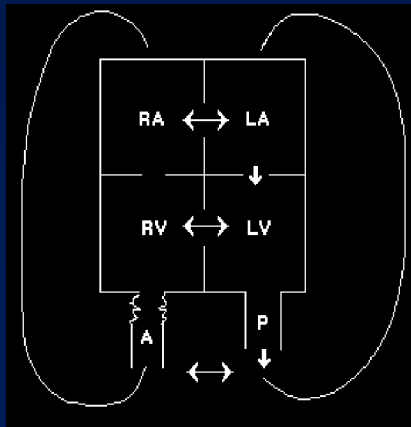
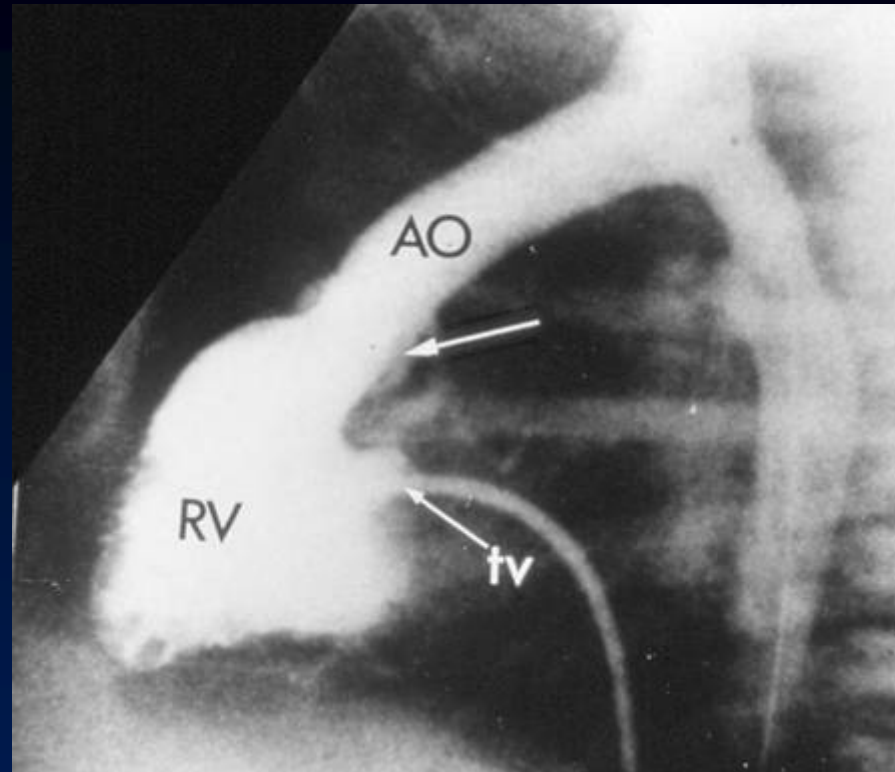
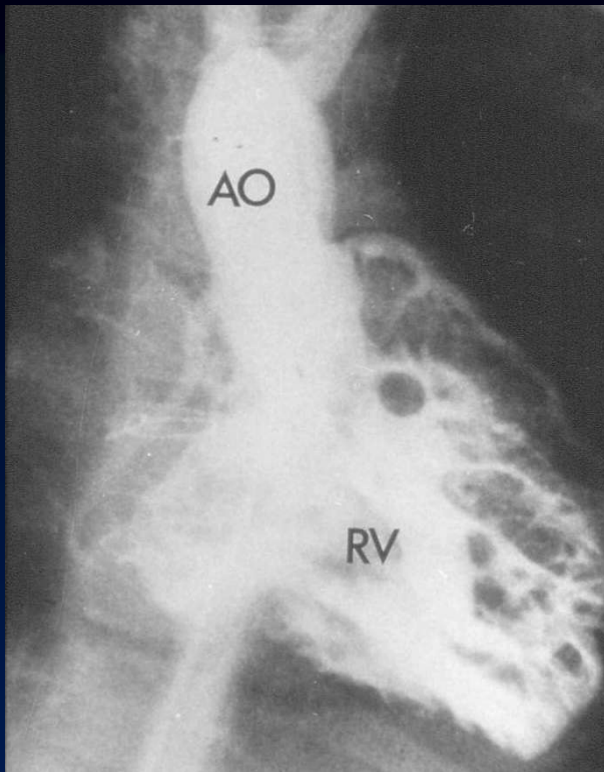
X-ray findings

- Mild cardiomegaly
- Concave pulmonary artery segment
- Narrow mediastinum (Egg-on-string)
- Shunt vessels
 - Depends on size of shunt and degree of PS

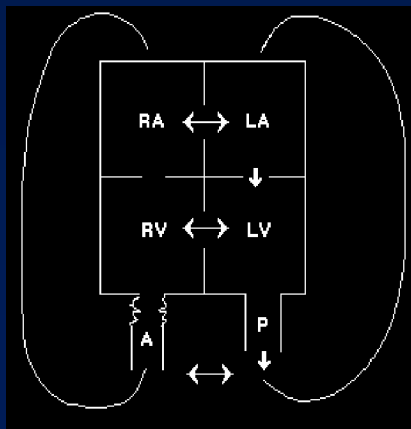
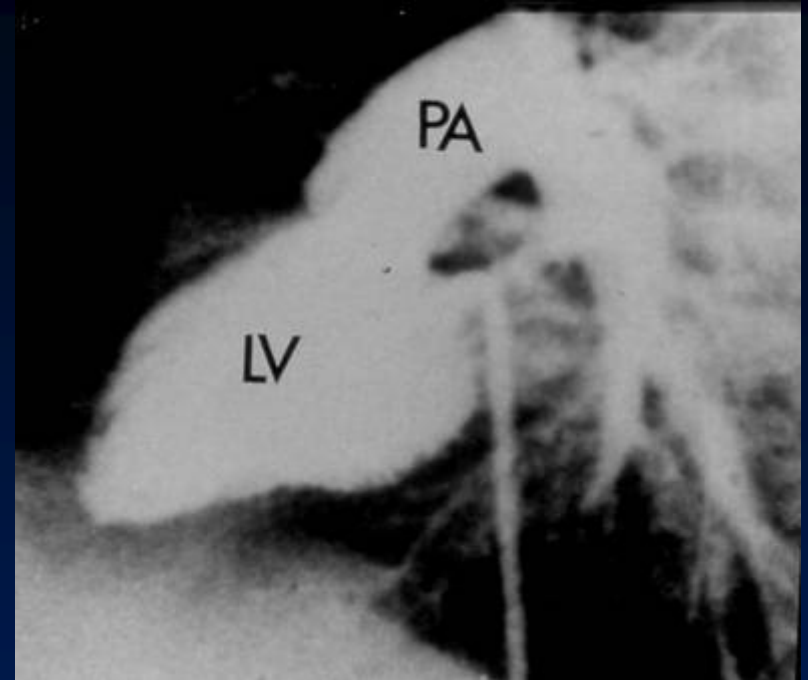
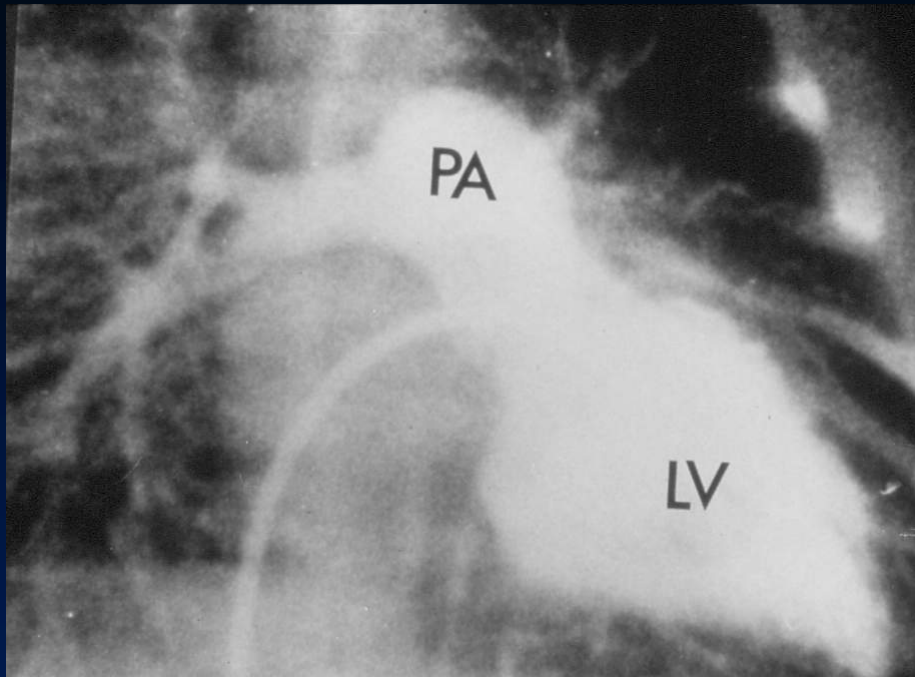




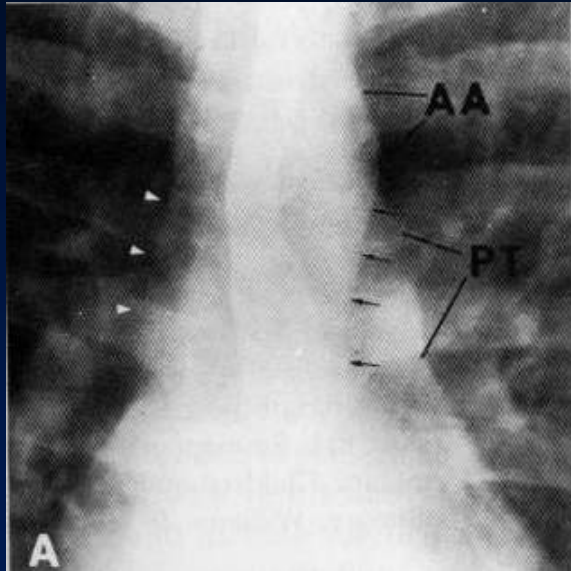
**Transposition of the Great Vessels
Cyanotic with - vasculature**



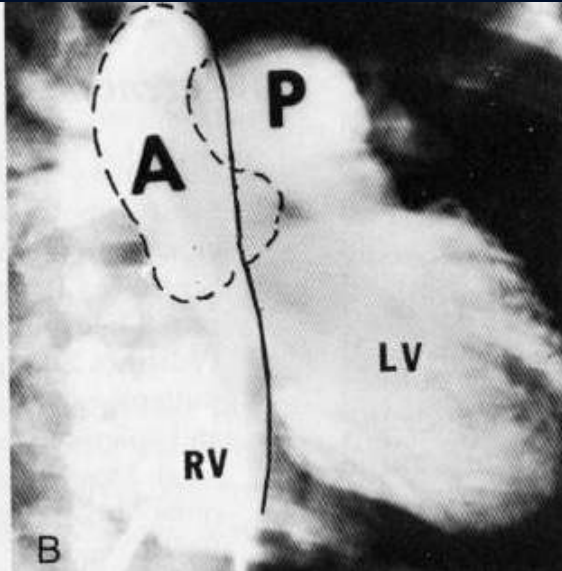
Transposition of the Great Vessels - RVgram



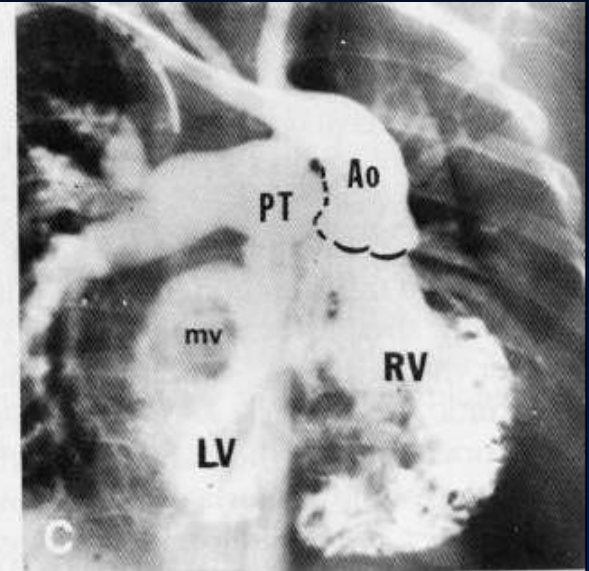
Transposition of the Great Vessels - LVgram



**Narrow waist in
Transposition**

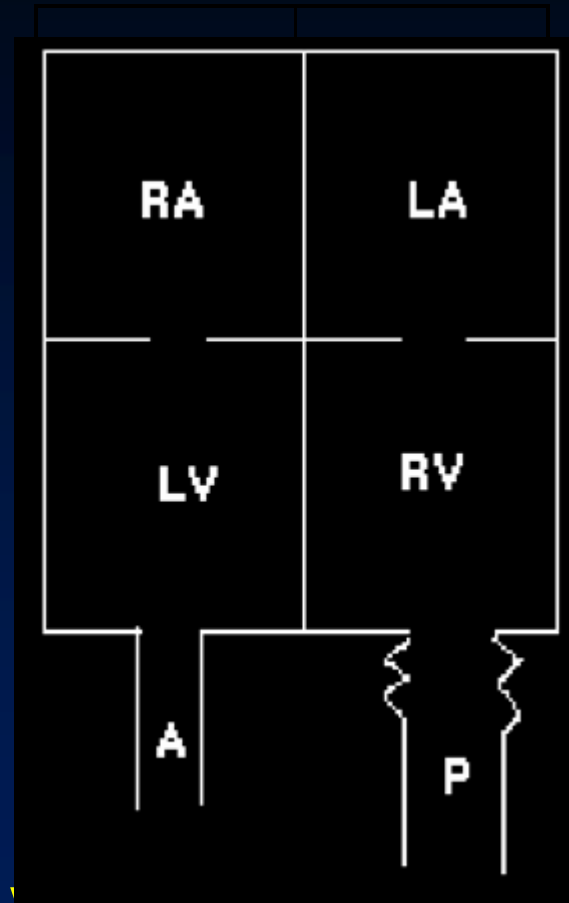


**LVgram with VSD but
normal Ao and PA
relationships**



**Corrected Transposition
With VSD**

Cyanotic



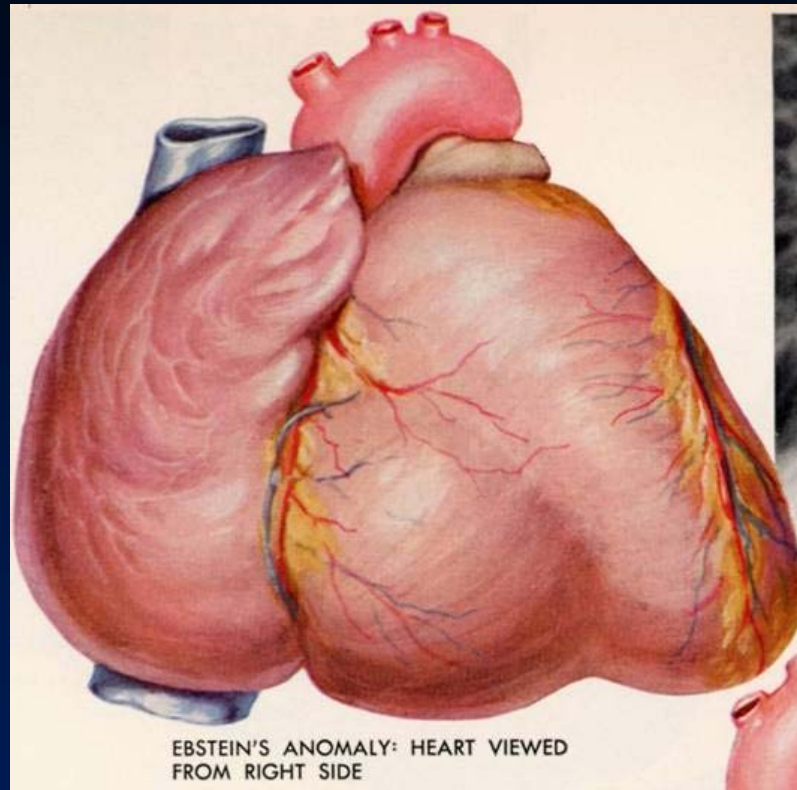
**Inversion of the ventricles
Without Transposition**

Ebstein's Anomaly

Ebstein's Anomaly

General

- **Rare**
- **Posterior and septal cusps of tricuspid valve displaced in to right ventricle**
 - **Right ventricle smaller or “atrialized”**
- **Tricuspid insufficiency → ↑ right atrial pressure → a R → L shunt through foramen ovale (or ASD)**
 - **Cyanosis is present in neonate**



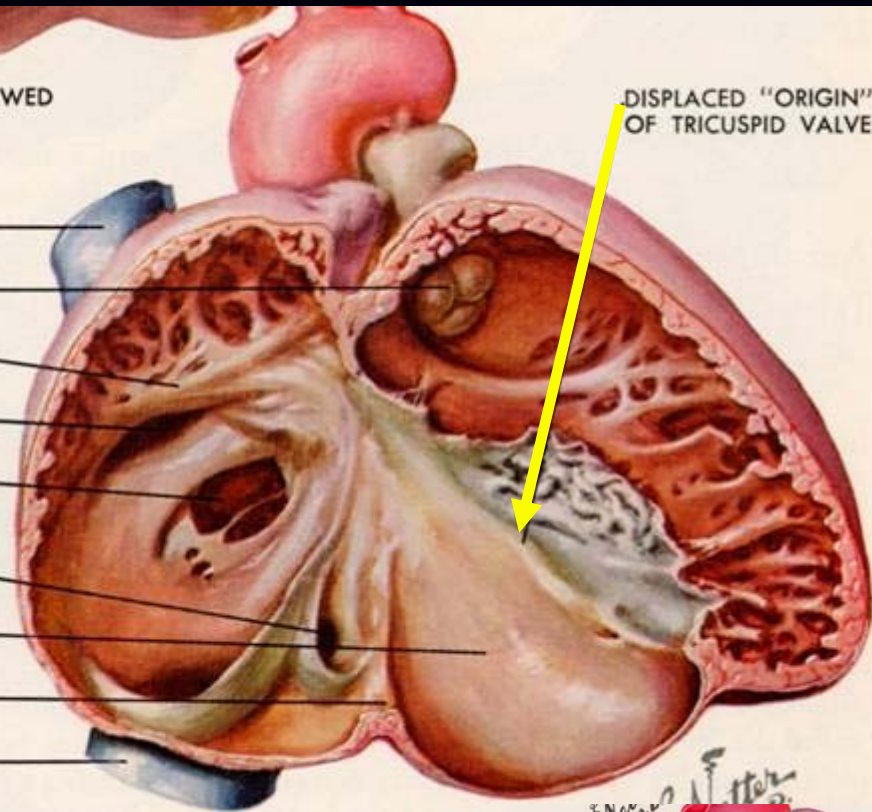
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Ebstein's Anomaly

EBSTEIN'S ANOMALY: HEART VIEWED FROM RIGHT SIDE

DISPLACED "ORIGIN" OF TRICUSPID VALVE

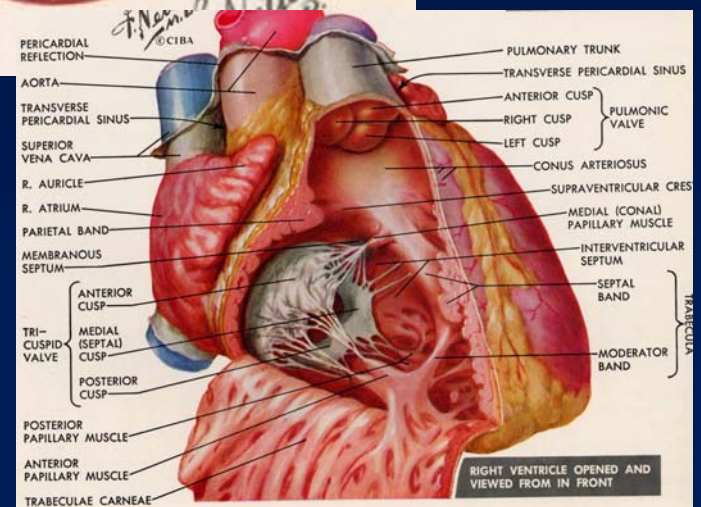
- SUPERIOR VENA CAVA
- PULMONARY VALVE
- CRISTA TERMINALIS
- OSTIUM OF SUPERIOR VENA CAVA
- FORAMEN OVALE (ATRIAL SEPTAL DEFECT)
- ORIFICE OF CORONARY SINUS
- "ATRIALIZED" PORTION OF R. VENTRICLE
- ATRIOVENTRICULAR JUNCTION
- INFERIOR VENA CAVA



Normal

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Ebstein's Anomaly



Ebstein's Anomaly

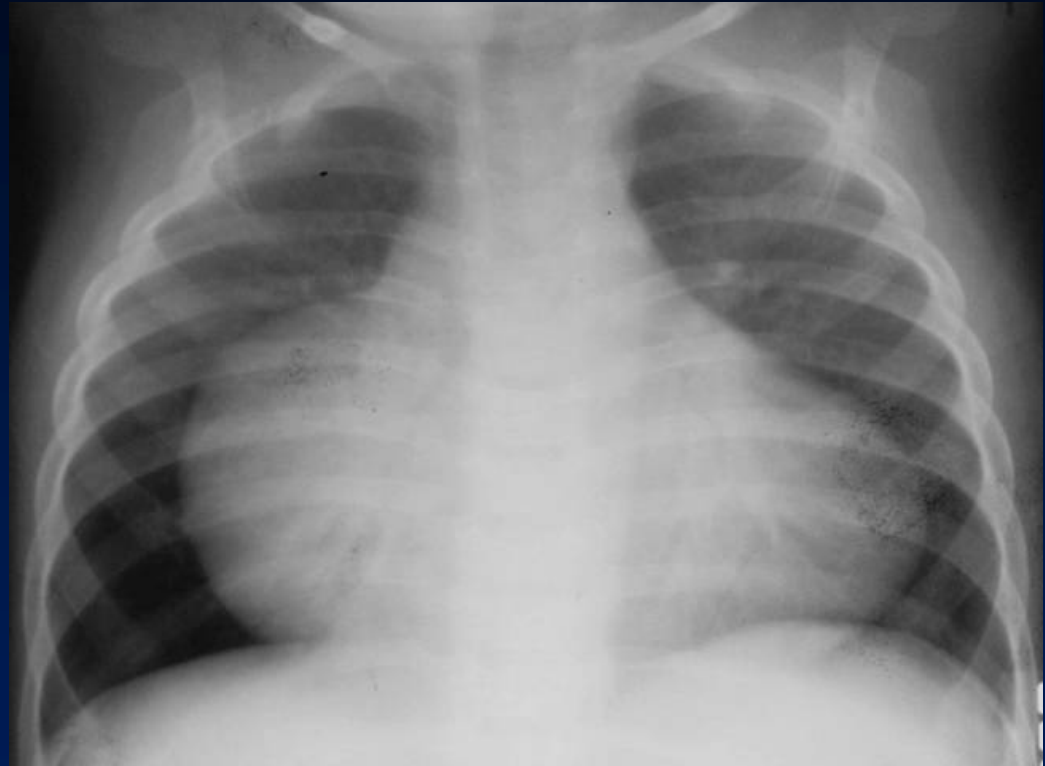
X-ray Findings

- **Cardiomegaly**
 - One of few conditions → cardiomegaly first few days of life
- **Unusual prominence to right heart border**
- **Pulmonary flow is decreased**

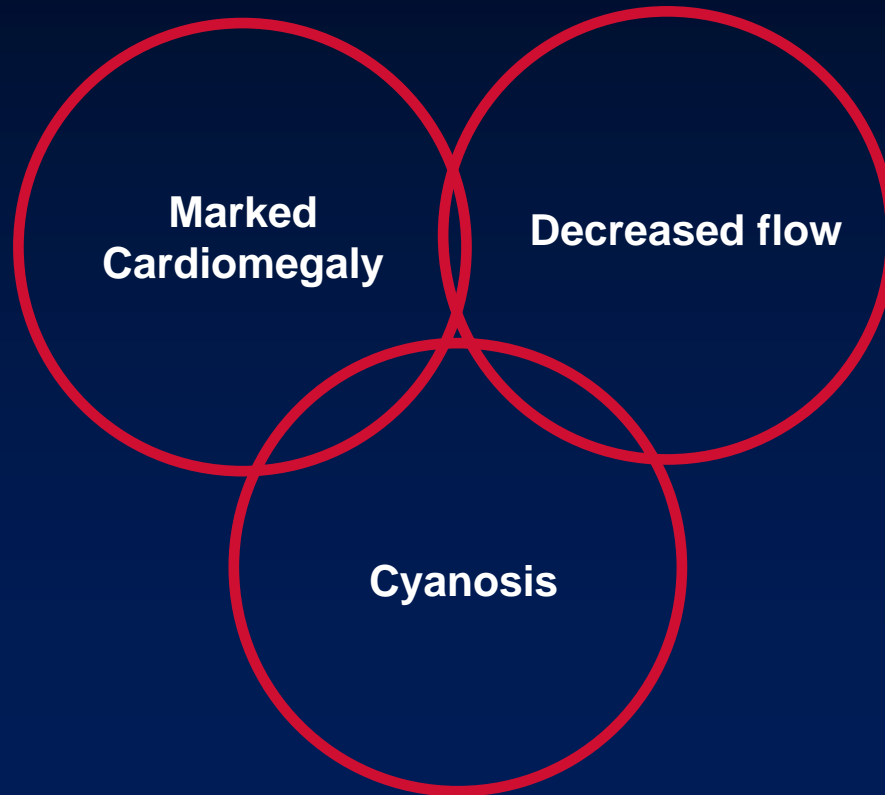
Ebstein's Anomaly



Ebstein's Anomaly



Ebstein's Anomaly Triad



Single Ventricle

Single Ventricle

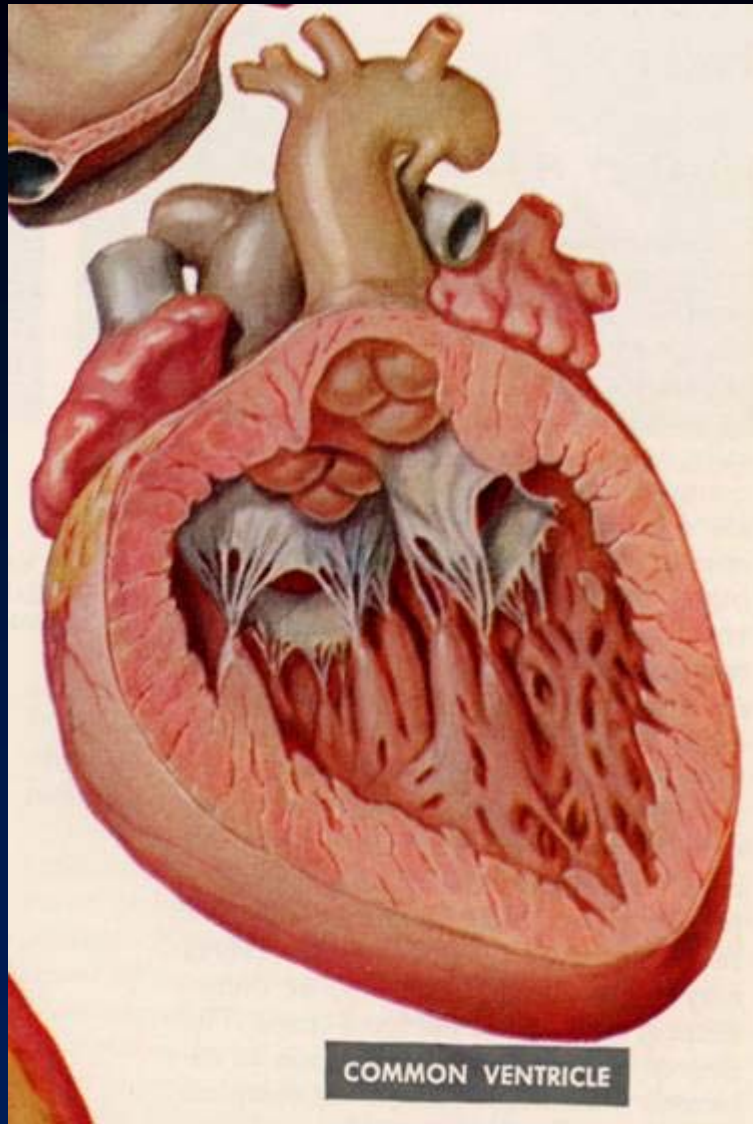
- **Surprise!**
 - There are usually two ventricles in this disease
- **Single ventricle: one ventricle with two atria**
- **Three types of Single Ventricle**
 - Morphologic LV with a rudimentary RV (common)
 - Morphologic RV with a rudimentary LV (rare)
 - Morphologically indeterminate ventricle (rare)

Single Ventricle

- **Most common**
 - Morphologic LV with rudimentary RV
- **Also called**
 - Double-inlet left ventricle
 - Common ventricle
 - Univentricular heart
- **Frequently difficult to determine which anatomic ventricle is present**

Single Ventricle Associated Findings

- **Pulmonic stenosis**
 - Valvular or subvalvular (66%)
- **Pulmonary atresia**
- **PAPVR**
- **PDA**



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Single Ventricle Imaging Findings

- **No characteristic appearance**
- **Concave pulmonary artery segment**
- **Shunt vessels**



Single ventricle

The End