Cyanotic Heart Disease
**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
<table>
<thead>
<tr>
<th>Tetralogy of Fallot General</th>
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- 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
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 “coeur 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

- Right aortic arch
- Increased flow
- Cyanosis
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%
Truncus-Type I

Most common (75%)

PA arises on left via short common stem

Shunt vessels

Convex pulmonary artery segment
Truncus Type 1
Truncus Type 1
Pulmonary arteries arise posteriorly from aorta

Truncus-Type II

Uncommon (25%)

Shunt vessels

Concave main pulmonary artery
Truncus Type II
Right and left pulmonary arteries arise laterally

Rare (5%)

Concave main pulmonary artery

Shunt vessels

Truncus-Type III
Truncus Type III
Truncus-Type IV

No pulmonary arteries

Bronchial circulation

TOF with pulmonary atresia

Concave main pulmonary artery

Rare to non-existent
Truncus-Type IV
(TOF with pulmonary atresia)
Bronchial Circulation

Increased flow
## Truncus Arteriosus

<table>
<thead>
<tr>
<th></th>
<th>Pulmonary artery</th>
<th>Shunt vessels</th>
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<tbody>
<tr>
<td>I</td>
<td>Convex</td>
<td>Yes</td>
</tr>
<tr>
<td>II</td>
<td>Concave</td>
<td>Yes</td>
</tr>
<tr>
<td>III</td>
<td>Concave</td>
<td>Yes</td>
</tr>
<tr>
<td>IV</td>
<td>Concave</td>
<td>Bronchial circulation</td>
</tr>
</tbody>
</table>
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
Tricuspid atresia—no transposition

- Systemic blood cannot enter RV
- Blood returns to right heart and then lungs either through VSD or PDA
- Need R → L shunt through patent foramen ovale
- Some unsaturated blood exits aorta

Tricuspid atresia—no transposition
Tricuspid atresia—with transposition

Systemic blood cannot enter RV

Need R → L shunt through patent foramen ovale

Oxygenated blood returns to LA

Un-oxygenated blood flows to lung via transposed PA

Tricuspid atresia—with transposition

Need L → R shunt to get blood into body
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
Normal heart

Anatomic Right ventricle is trabeculated

Anatomic Left ventricle is smooth

Pulmonic infundibulum always stays with the anatomic R ventricle
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

Posterior
Medial
Inferior
To the aortic valve

Normal  Corrected Transposition
Corrected Transposition
Inversion of the Ventrices With Transposition

- Physiologically flow is normal
- Consistent with normal life, except
- Frequently associated with
  - VSD
  - Tricuspid insufficiency
  - Subpulmonic stenosis
  - Complete heart block
Corrected Transposition (L-Trans) Inversion of the Ventricles with Transposition of the Great Vessels

Normal vasculature; or increased with VSD

PA arises from anatomic left ventricle

Aorta arises from pulmonic infundibulum

Acyanotic

RA

LV

LA

RV

PA
Corrected Transposition

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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 ¼ to ½ have patent ductus
- Some have ASD
- Other major finding is obstruction to blood entering pulmonary artery
  - Usually subpulmonic stenosis
Transposition of the Great Vessels (D-Trans)

Cyanotic with increased vasculature

Obligatory shunt since there are 2 separate circulations

Aorta arises from pulmonic infundibulum

PA arises from anatomic left ventricle

ASD
VSD
PDA

RV  LA

RA  LV

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
LVgram with VSD but normal Ao and PA relationships

Corrected Transposition With VSD
Inversion of the Ventricles
Without Transposition

Cyanotic
Ebstein’s Anomaly
Ebstein’s Anomaly
General

- Rare
- Posterior and septal cusps of tricuspid valve displaced into right ventricle
  - Right ventricle smaller or “atrialized”
- Tricuspid insufficiency $\rightarrow$ ↑ right atrial pressure $\rightarrow$ a R $\rightarrow$ L shunt through foramen ovale (or ASD)
  - Cyanosis is present in neonate
Ebstein’s Anomaly
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

- Marked Cardiomegaly
- Decreased flow
- Cyanosis
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
# Single Ventricle Imaging Findings

- No characteristic appearance
- Concave pulmonary artery segment
- Shunt vessels
Single ventricle
The End