Cardiac Board-type Case Review

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What’s the DDX?

Cyanotic newborn
Cyanosis With Decreased Vascularity

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

* Also appears on DDx of cyanosis with increased vascularity
What’s the diagnosis?

Ebstein’s Anomaly
What’s the DDX?

8 year-old cyanotic male
## Cyanosis With Increased Vascularity

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

* Also appears on DDx of cyanosis with decreased vascularity
What's the diagnosis?

TAPVR-Supracardiac type 1
What’s the DDX?

Acyanotic newborn
Cardiomegaly with Normal Vasculature

- Viral myocarditis
- Endocardial fibroelastosis
- Aberrant left coronary artery
- Cystic medial necrosis
- Diabetic mother
What’s the diagnosis?

Viral myocarditis
What’s the DDX?

Acyanotic newborn
### Causes of CHF In Newborn

**Impede Return of Flow to Left Heart**

- Infantile coarctation
- Congenital aortic stenosis
- Hypoplastic left heart syndrome
- Congenital mitral stenosis
- Cor triatriatum
- Obstruction to venous return from lungs
  - TAPVR from below diaphragm
What’s the diagnosis?

Hypoplastic Left Heart Syndrome
What’s the diagnosis?

7 yo acyanotic female
Atrial septal defect
Another example

34 yo acyanotic female
ASD (primum) with PAH

34 yo acyanotic female
Ostium Secundum
ASD-MRI
• Discontinuity in the atrial septum with systolic signal void consistent with L to R shunt at atrial level
• Right atrium is slightly dilated; RV, LV and LA size are normal
What’s the diagnosis?

1 yo acyanotic female
Ventricular Septal Defect
Another example-VSD

VSD
Membranous VSD-MRI
What’s the diagnosis?

8 mos old acyanotic female
Patent Ductus Arteriosus
Patent Ductus Arteriosus-MRI

- Jet of signal loss showing continuous flow from the aorta to the MPA consistent with sizeable PDA; MPA is severely dilated at level of PDA
What’s the diagnosis?

9 mos old cyanotic female
TAPVR-supracardiac type
TAPVR
Supracardiac Type 1

Angiographic Appearance
TAPVR-cardiac type-MRI
TAPVR-infracardiac type-MRI
What’s the diagnosis?

10 yo cyanotic male
Tetralogy of Fallot
Tetralogy of Fallot
Tetralogy of Fallot-MRI
Overriding aorta, VSD
Axial spin-echo MR image shows severe infundibular pulmonic stenosis (arrow).

Tetralogy of Fallot
What’s the diagnosis?

12 yo cyanotic male
Truncus arteriosus-Type 1
Truncus Type 1

Radiology Resource and Review
Single large artery (T) arising from the heart.
Pulmonary artery (arrow) originates from the left side of the truncus.
There is a right aortic arch.

Truncus Arteriosus Type 1-MRI
Truncus Type II
ECG-gated spin-echo transaxial image demonstrates a bar of muscle and fat (blue arrow) (tricuspid atresia) separating the right atrium (yellow arrow) from the hypoplastic right ventricle (red arrow)
Tricuspid atresia-MRI

Enlarged right atrium

Small right ventricle

Tricuspid atresia-MRI
What’s the diagnosis?

3 mos old cyanotic male
Transposition of the Great Vessels
In Transposition, pulmonic valve is

Posterior
Medial
Inferior
To the aortic valve

Normal  Corrected Transposition
Anatomic Ventricles

Trabeculated ventricle-
Anatomic Right

Smooth ventricle-
Anatomic Left
Transposition of the Great Vessels - RVgram
Transposition of the Great Vessels - LVgram
Oblique axial spin-echo image shows displaced attachment (thick arrow) of the posterior leaflet (thin arrows)
What’s the diagnosis?

A cyanotic adult
Coarctation of the aorta
Coarctation of the aorta

Arrow points to indentation representing area of coarctation with dilated aorta (or LSCA) above and post-stenotic dilatation below coarct
Coarctation of the Aorta

Convexity above aortic knob due to dilated LSCA or Aorta proximal to coarct

Ascending Ao may be dilated, normal or small

Yellow arrows point to rib-notching

Coarctation of the Aorta
Contrast enhanced MRA shows long segment coarctation of the aorta
Oblique sagittal spin-echo-Coarctation of the Aorta
Axial spin-echo MRI-Coarctation of the Aorta
What’s the diagnosis?

Acyanotic adult
Aortic Stenosis
Prominence of ascending aorta from post-stenotic dilatation

Aortic stenosis
Aortic Stenosis
Coronal cine MRI image demonstrates a systolic signal void originating at the stenotic aortic valve. Ascending aorta is dilated
Hypoplastic Left Heart Syndrome
Hypoplastic Left Heart Syndrome

Atretic aorta

Neufeld, HN, Circulation, 1962
Hypoplastic Left heart Syndrome

Gated spin echo at base of heart shows hypoplastic aorta (arrow) posterior and right of main pulmonary artery
Cor triatriatum

Frontal radiograph demonstrates CHF
Cor Triatriatum - angiography

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Cor Triatriatum - angiography

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Cor Triatriatum
Aortic Regurgitation

Cine MR image during diastole shows signal void emanating from the aortic valve
What’s the diagnosis?

Acyanotic adult
Mitral Stenosis
Mitral Stenosis

Convexity from enlarged left atrial appendage
Mitral Stenosis

Convexity from enlarged left atrial appendage
Upper lobe vessels equal to or larger than size of lower lobe vessels = Cephalization
Mitral Stenosis

Cine MR image in axial plane demonstrates a diastolic signal void emanating from the mitral valve.
Left Atrial Myxoma
Contrast-enhanced CT shows large filling defect in lumen of LA
Left Atrial Myxoma

Cine MRI shows soft tissue mass arising from wall of left atrium and projecting into lumen
What’s the diagnosis?

Acyanotic adult
Mitral regurgitation
Mitral Regurgitation

Cine MR image in axial plane during systole depicts a signal void emanating from the mitral valve
Difference in heart size – MS and MR

Mitral Stenosis

Mitral Regurgitation
What’s the diagnosis?

Acyanotic adult
Pulmonic stenosis
Pulmonic Stenosis

- Normal-sized heart
- Prominent main pulmonary artery segment
- Enlargement of left pulmonary artery

Pulmonic Stenosis
What’s the diagnosis?

Acyanotic adult
Right Arch with Aberrant Left SCA

Trachea is bowed forward by aberrant left subclavian artery (arrow)
Right Aortic Arch with Aberrant Left Subclavian (Arrows)
What’s the diagnosis?

36 yo cyanotic female
Mirror image Right aortic arch with TOF

Trachea is not bowed forward
Mirror Image Right Aortic Arch
Left Aortic Arch with Aberrant R SCA
Review-
Name the abnormalities. Are they the “good” or “bad” variety?
Left Aortic Arch with Aberrant R SCA

Right Aortic Arch with Aberrant Left Subclavian

Mirror Image
Right Aortic Arch
What’s the diagnosis?

2 month old with stridor
Double aortic arch

Right arch is larger and higher
Left arch is smaller and lower
Double Aortic Arch
Double Aortic Arch-angiographic appearance
Double Aortic Arch
Impressions on Trachea and Esophagus
What’s the diagnosis?

4 month old with stridor
Pulmonary Sling
Pulmonary Sling
Tracheal/esophageal impressions

- Aberrant SCA
- Double Ao Arch
- Isolated Anomalies (Rare)
- Pulmonary Sling

Dahnert
If you see cases like these, you passed...
Myxoma in Left Atrium
Thrombus in left atrial appendage
Dilated Cardiomyopathy
Dilated Cardiomyopathy

Cine MR images in the short axis plane show little change in size between end diastole and end systole
Arrhythmogenic Right Ventricular Dysplasia

Left-thickening and replacement of RV anterior wall by fatty tissue.
Fat suppression (right) - loss of signal in RV anterior wall, confirming fatty nature of these changes
Restrictive cardiomyopathy

ECG-gated spin-echo image - enlargement of both atria and normal size of ventricles with thickened walls
Hypertrophic Cardiomyopathy

ECG-gated spin-echo image in coronal plane - severe symmetrical hypertrophy of LV
Hypertrophic Cardiomyopathy

Asymmetric septal hypertrophy

Thickened apex

Hypertrophic Cardiomyopathy
Hypertrophic Cardiomyopathy

- Marked wall thickening
- Mitral Regurgitation From SAM
- Almost complete emptying of LV
- LV
- LA

Hypertrophic Cardiomyopathy
Congenital Defect in the Pericardium
Cardiac Malpositions
Cardiac Malpositions
Types

- Situs solitus with dextrocardia
- Situs inversus with levocardia
- Situs inversus with dextrocardia
Rule of Thumb

- If aortic arch/apex of heart are on opposite sides from stomach bubble, high incidence of CHD
Asplenia
Bilateral Right-sidedness

- Male
- Cyanotic
- High risk of infection
- Severe cardiac abnormalities
  - Transposition
  - TAPVR
Polysplenia
Bilateral left-sidedness

- Female
- Abnormalities are more benign
  - Azygous continuation of IVC
  - Bilateral superior vena cava
  - PAPVR
  - ASD
Asplenia/Polysplenia

- Asplenia – bad boy
- Polysplenia – good girl
Situs Ambiguous-polysplenia
**CARDIAC MALPOSITIONS**

**Situs solitus**
- Aortic arch and LA are always on the same side except with isolated R arch
- 0.6-0.8% CHD

**Situs inversus with dextrocardia**
- 3.5% CHD
- Most common is corrected transposition (Kartegener's)

**Situs inversus with levocardia**
- Rare, but 95-100% CHD; no specific prevalence. If asplenia, 100% have common ventricle. Interruption of IVC common.

**Situs solitus with malposition of the stomach**
- R/O asplenia
- Most have CIID (L→R shunt)
- Most with polysplenia andazygous continuation of IVC

**Situs inversus with malposition of the stomach**
- 95% CHD of which 80% are corrected transposition. If cyanosis with shunt vessels, then tricuspid atresia. If cyanosis and Jаve, then corrected transposition. If asplenia, 100% have common ventricle. Interrupted IVC common.
Good Luck