Corrected Congenital Heart Disease for the Thoracic Radiologist
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GOALS AND OUTLINE
- To highlight some of the more commonly used surgical procedures for CHD in the adult
  - Review the radiologic impact
    - On protocols
    - On imaging findings
  - Review the more common complications
    - With emphasis on those encountered on cross-sectional imaging
- I have no relevant disclosures

GROWN UPS WITH CHD (GUCH)
- Minor CHD presenting as an adult
- Definitive surgical correction as an adult
- Adults presenting for expected reoperations
- Repair of residual defects after correction
- Heart failure after correction or palliation
- Acquired heart disease in addition to CHD

As Cardiac Radiologists, we are familiar with these indications

CONGENITAL HEART DISEASE
- Most common birth defect (moderate - severe disease 4-6 per 1,000 live births)
  - More recent estimates: 20 per 1,000
- > 85% of neonates will survive to adulthood
- 2004: 0.8 to 1.3 million adults with CHD in US
- Expected to increase 5% per year

In next decade:
- 1/150 young adults will have some form of CHD
- Higher percentage of adults with CHD will consist of more complicated cases
- “The dramatic improvement in survival should lead to a larger number of GUCH patients than children with CHD”

CONGENITAL HEART DISEASE
- US GUCH population on the rise
- The CHD surgeries can confound a Chest CT
  - Need to be aware of some well-known complications
  - May affect a protocol
  - The normal post-op appearance can simulate disease
  - A diseased repair can simulate normal
- Higher incidence of pulmonary hypertension

BUT WHY CARE AS A THORACIC RADIOLOGIST

I have no relevant disclosures
CONDITIONS THAT MAY PRESENT IN ADULTHOOD

- Ebstein Anomaly
- Bicuspid aortic valve
- ASD (secundum) and other septal defects
  - VSDs most common but >85% heal spontaneously
  - Anomalous pulmonary venous return
- Coronary artery anomalies
- Aortic coarctation
- Silent Ductus (PDA)

*For sake of time, let us skip these lesions.*

ADULTS WITH REPAIRED CHD

- Tetralogy of Fallot
  - Blalock Taussig
- Univentricular physiology
  - Glenn
  - Fontan and Modifications
- Congenital valvular
  - Ross
- Repaired TGA
  - Baffles
  - Jatene

TETRALOGY OF FALLOT (TOF)

- Long-term survival is great (>89% at 30 yrs)
- Definitive repair with homograft and VSD closure is treatment of choice

TETRALOGY OF FALLOT (TOF)

- Radiologic Impact
  - Dilated aortic root (unknown, ?TGF-Beta)
  - Peripheral pulmonic stenoses
  - Coronaries have a clockwise orientation
Patients may undergo palliation with Blalock-Taussig (BT) shunt
- Subclavian Artery to PA
- Contralateral side of the Aortic Arch

Classic uses proximal SCA; now use graft material

TOF Impact:
- Unilateral, ipsilateral rib notching (Classic)
- Vessel in paratracheal region

Complications of BT:
- Thrombosis
- Seroma adjacent to graft
- Occluded ipsilateral PA or stenotic PA
- Enlarged bronchial arteries (MAPCAS)
The goal is to let the lungs (pulmonary arteries) fill passively and convert the heart to a systemic ventricle.

- **Glenn (SVC to PA)**
- **Fontan (RA to PA)**
- **Modified Fontan is the standard**

**UNIVENTRICLE HEART**

**Glenn**

- Radiologic Impact
  - Altered flow dynamics: Side with shunt opacifies early with arm injection
- Complications
  - Pulmonary arteriovenous malformations

**Fontan**

- Radiologic Impact
  - Altered flow dynamics: Depends on the type but classically, left lung opacifies late (unless leg injection)
- Complications
  - Pulmonary arteriovenous malformations
  - Big right atrium (arrythmias)
  - Thrombus
  - Hepatomegaly (cirrhosis and possible HCC)
  - Protein losing enteropathy
CAVOPULMONARY SHUNT

- Sometimes called modified Fontan
- Inferior tunnel may be
  - Intracardiac or extracardiac
  - Fenestrated or not
- Often times imaging is performed to address the question: is the RV failing or is it a conduit failure?

CAVOPULMONARY SHUNT

- Radiologic Impact:
  + Altered flow dynamics: right lung opacifies early and left lung opacifies late
- Complications:
  + Thrombus
  + Other complications are less common than with Glenn or Fontan alone
  + Collateral arteries may be seen with Fontan, Glenn or Cavopulmonary Shunts

CAVOPULMONARY SHUNT

- When there is a fear of subaortic obstruction, the cavopulmonary may be combined with an outlet procedure: Damus-Kaye-Stansel
  - The DKS is an anastomosis of the proximal PA and the Aorta
  - Tricuspid Atresia + TGA
- Radiology Impact:
  - The outflow ill have an odd appearance and may simulate an AA aneurysm
**RETIRED CLASSICS**

- Waterston-Cooley
  - Formerly used for TOF, tricuspid atresia, P Atresia
  - Ascending Aorta to R PA
  - Complications: stenosis of R PA and PH
- Potts
  - Formerly used for TOF, tricuspid atresia, P Atresia
  - Descending Aorta to L PA
  - Complications: stenosis of LPA and PH

**VALVULAR HEART DISEASE**

- Bicuspid aortic valve is one of the more common CHDs
- Congenital AS may undergo Ross Procedure
- In a Ross, the native P Valve is used for the A Valve and a cadaveric P valve is used
  - Great idea!
  - Bad idea- 1 valve disease → 2 valve disease
- The native aorta becomes dilated and the main PA is stenotic (does not grow)
**TRANSPOSITION**

- The definition of transposition:
  - RV gives rise to Aorta
  - AV valves follow the ventricles
  - The coronary arteries follow the ventricles
  - In D-TGA, the RV communicates with the RA ${\text{RA} \rightarrow \text{RV} \rightarrow \text{Aorta}}$; $\text{LA} \rightarrow \text{LV} \rightarrow \text{PA}$
  - In L-TGA, the RV communicates with the LA $\text{RA} \rightarrow \text{LV} \rightarrow \text{PA}$; $\text{LA} \rightarrow \text{RV} \rightarrow \text{Aorta}$

**D-TGA**

- Arterial switch (Jatene) is now the preferred correction
  - Radiologic Impact: Classic shape of the great vessels (akin to a Dutch hat)
  - Complications [Few]:
    - Coronary artery kinking in the immediate post-op period
    - Branch pulmonary artery stenoses
    - Neoaortic root dilatation
  - Inflow switch (Baffles) were used up until the late 1980’s. Mustard (Pericardium); Senning (Atrial tissue)
  - Radiologic Impact: SVC, IVC are directed to the left ventricle and the pulmonary veins to the right ventricle
  - Complications:
    - Baffle leaks
    - Baffle stenoses
    - Arrhythmias requiring pacemakers
GUCH patients are on the rise
- No longer a pediatric condition
- No longer only a Cardiac Imager’s Issue
- Many of these conditions will affect our protocols (Glenn, Fontan, Cavopulmonary)
- My result in anatomic changes
- Associated with extracardiac complications
    + AVMs
    + Ascending aortic aneurysms
    + Cirrhosis

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<td>Pulmonary stenoses related coronary dilation ascending aorta calcified aneurysm of RVOT</td>
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<td>Blalock-Taussig (SDA to PA)</td>
<td>Occlusion</td>
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<td>Glenn (SVC to PA)</td>
<td>Pulmonary AVMs</td>
<td>Opacifies ipsilateral lung early</td>
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<td>Fontan (RA to PA)</td>
<td>Large RA (thrombus) Cirrhosis</td>
<td>Altered flow (may require delayed imaging) May simulate aneurysm or mediastinal mass</td>
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<td>Damus-Kaye-Stansel (aortic obstruction)</td>
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<td>Congenital Aortic Stenosis</td>
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