Cyanotic Congenital Heart Disease

Iowa ACC- Lecture series
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NO DISCLOSURES

Complex CHD- Learning Objectives
• Types of Cyanotic heart diseases and their pathophysiology
  – Palliated
  – Unpalliated
  – Corrected
• Overview of Treatment and Surgical corrections
• Post operative check lists
• Long term follow up Issues

Cyanotic heart diseases on the hand (5 T’s)

• Truncus Arteriosus
• Transposition of great arteries (Aorta and PA)
• Tricuspid Atresia
• Tetralogy of Fallot
• Total Anomalous Pulmonary Venous return (TAPVR)
• Hypoplastic right and left heart syndromes (HLHS, HRHS)
• Criss cross hearts, Isomerism, Heterotaxy

Relative Frequency of Cyanotic Congenital Heart Disease

<table>
<thead>
<tr>
<th>Lesion</th>
<th>% of All Lesions</th>
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<tr>
<td>Tetralogy of Fallot</td>
<td>5–7</td>
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<tr>
<td>d-Transposition of great arteries</td>
<td>3–5</td>
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<tr>
<td>Truncus arteriosus</td>
<td>1–2</td>
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<tr>
<td>Total anomalous pulmonary venous return</td>
<td>1–2</td>
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Tetralogy of Fallot (TOF)

• One of the most common forms of cyanotic congenital heart disease
• Genetic association- Di-George syndrome
• 1/3rd association with Right Aortic Arch
• Based on severity of pulmonary stenosis it can range from pink Tet to pulmonary atresia
• Coronary artery anomaly- LAD from RCA
• CXR- Boot shaped heart
Anatomic Defects associated with TOF

- Pulmonary Stenosis - infundibular, valvar, supravalvar
- Right ventricular hypertrophy
- Over-riding Aorta
- Ventricular Septal Defect

Bonus Question?
What causes the heart defects of TOF?
Answer?
Anterior-Superior deviation of the conal (infundibular) septum

Tetralogy of Fallot

- Generally rare to establish a primary diagnosis in Adulthood
- Usually repaired within 1st year of life
- Mostly encountered are post operative patients
- Could have had palliative Blalock- Taussig Shunt
- Complete repair is the rule

Palliative Surgery for Tetralogy of Fallot

Classic Blalock-Taussig Shunt
Subclavian artery divided and anastomosed to the branch pulmonary artery

Central shunts
- Waterston shunt: Ascending aorta to RPA
- Potts shunt: Descending aorta to LPA

Systemic-pulmonary artery shunt (modified Blalock-Taussig Shunt)
- Gortex shunt directing blood from the base of innominate artery to the right/left pulmonary artery.

Surgical Options for Complete repair of TOF with a normal Pulmonary Valve
- Reconstruction of main pulmonary artery
- Sub-pulmonary valve patch
- VSD closure
Surgical Options for Complete repair of TOF with a Dysplastic Pulmonary Valve

- Trans-annular right ventricular outflow tract patch
- +/- Pulmonary valvectomy
- VSD closure

What is the sequelae of this surgery?

Pulmonary regurgitation → RV volume overload → RV dilatation

TOF- Questions to be answered Post Operative

- Residual VSD
  - Qp:Qs
- Pulmonary regurgitation
  - Regurgitation fraction
- RV size and Volumes and function
  - RV end diastolic volumes (> 150 ml/m2 surgical indication for Pul. Valve placement)
- Branch pulmonary artery and differential flows
- Myocardial Delayed enhancement- Arrhythmia
- Aorto pulmonary collaterals
- Coronary artery anatomy
- Aortic root and Ascending aorta size

TOF-Post operative- late sequela

- RV dysfunction
- RVOT obstruction/ RVOT aneurysm
- Residual VSD
- Severe PR
- Arrhythmia- SVT, QRS duration more than 180 msec- predisposition to VT
- Aortic regurgitation
- Aortic dilation

Tetralogy of Fallot- Showing right sided enlargement

- After transannular patch, there is Free PR
- Over time leads to RV dilation
- Also can see VSD patch

Tetralogy of Fallot- Unrepaired

- Increased PBF can lead to damage of pulmonary vasculature
- Increases PVR
- Shunt reverses and Eisenmenger syndrome can set in
- If severe PS, can lead to R - Left shunt and cause the hypoxia
D-Transposition of the great arteries (D- TGA)

Transposition of great arteries

- D- TGA (Classical TGA)
- L- TGA (CCTGA)

Simple rules

- Normally Aorta is posterior and right of PA
- Any time aorta is anterior to PA- Transposition
  - Aorta to right: D- TGA
  - Aorta to left: L- TGA
- Morphologic RV to right: D-looped ventricle
- Morphologic RV to left: L-looped ventricle

Transposition of the great arteries before repair

- Anterior and right ward aorta from RV
- Left ward and posterior PA from LV
- Parallel Circulation
- Primary diagnosis- very rare in adulthood

Initial palliation: Patients with d-TGA

Procedure- Rashkind Balloon Atrial Septostomy

Surgical Repair of d-TGA

- Atrial switch operation
  - Mustard procedure
  - Senning procedure
- Rastelli Procedure
  - In case of LVOT obstruction
  - LV to Aorta baffle via VSD
  - RV to PA conduit

Surgical Repair of d-TGA- Corrective Surgery: Jatene Arterial switch operation

Arterial switch
Coronary artery translocation
Le-Compte maneuver

Serial circulation is achieved
**d- TGA: Questions to be answered**

**Post Operative- Atrial Switch Operation**

- Systemic Venous Baffle obstruction
- Pulmonary venous baffle obstruction
- Baffle leaks
- Systemic Ventricle (RV) function/ dysfunction
- Tricuspid Regurgitation
- Left Ventricular outflow tract obstruction
- Arrhythmias- Atrial/ Ventricular arrhythmias
- Residual VSD if present

**d- TGA: Questions to be answered**

**Post Operative- Arterial switch operation**

- Arterial anastomosis site
  - Supravalvar PS
  - Supravalvar AS
- Branch Pulmonary arteries secondary to
  - Le-Compte Procedure
- Coronary artery kink/ stenosis
- Aortic root dilation
- Aortic regurgitation
- Left Ventricular dysfunction
- Arrhythmia
- Residual VSD

**Mustard/ Senning Operation for d-TGA**

**D- TGA s/p Senning operation**

Systemic Venous Baffle

Pulmonary venous Baffle

**D- TGA s/p Senning operation- RV dysfunction and TR**

Axial stack

Short axis stack

**D- TGA s/p Senning operation- Systemic venous Baffle obstruction**
**D- TGA s/p Senning operation- Systemic venous Baffle leak**

Baffle leak closed with Amplatzer device

**Transposition- Arterial switch Lecompte Manuever**

After the switching of the arteries, the pulmonary arteries end up straddling around the Aorta leading to stretching and narrowing of the pulmonary arteries

**s/p Arterial switch for Transposition- Coronary artery Imaging**

Normally after repair:
- Left from anterior sinus.
- Right from posterior sinus.

Here:
- Right and LAD from anterior sinus
- Left circumflex from posterior sinus and takes retroaortic course

**d-TGA: Arterial switch Operation Aortic root dilation**

**Congenitally Corrected TGA- (CCTGA) (L-TGA)**

- Also known as
  - Physiologically Corrected transposition
  - L-loop TGA
### Congenitally Corrected transposition (L-TGA)

- Associated lesions – VSD, Pulmonary stenosis
- Complete heart block
- Ebstein's Anomaly- WPW
- Dextrocardia

### Congenitally Corrected transposition (L-TGA)

- They can be diagnosed as adults
- Progressive TR
- Systemic RV: progressive dysfunction
- Arrhythmia and heart block
- Symptoms will be related to these issues of systemic RV failure and TR

### Congenitally Corrected transposition (L-TGA)

- With progressive TR, TV replacement is recommended before they are symptomatic and RV failure ensues
- Double switch operation can be done by training morphologic LV
- Conduit replacement for sub pulmonary PS
- Rhythm issues need to be dealt with including pacemaker placement for heart block
- CMR is an excellent diagnostic modality

### Congenitally Corrected transposition (L-TGA)

**Senning- Rastelli Procedure**

- If there is PS/SubPS and VSD is large
- Senning/Mustard –Rastelli
- Senning/Mustard: Atrial switch
- Rastelli procedure: Morphologic LV baffled to Aorta via VSD

### Congenitally Corrected transposition (L-TGA)

**Double Switch Procedure**

**Anatomical repair**

- The LV is the systemic ventricle
- Double switch operation
  - Atrial switch (Mustard or Senning)
  - Arterial switch

### Congenitally Corrected transposition (L-TGA)

**Shows Posterior RV, Tricuspid regurgitation**

![Evan illustrations](image)
Congenitally Corrected transposition (L-TGA)

- Short axis series
- Septal configuration
- Trabeculated dilated posteriorly placed systemic RV
- TR

Congenitally Corrected transposition (I-TGA)

LVOT  RVOT

Truncus Arteriosus

- A single artery arising from the base of the heart which gives origin to the coronary, pulmonary and systemic arteries
- Almost always with a VSD
- The truncal valve is usually abnormal and has stenosis and/or regurgitation
- Can be associated with DiGeorge syndrome.

Surgical repair of Truncus Arteriosus

- Repair includes VSD closure
- Separation of Pulmonary artery from aorta
- RV to PA conduit
**Truncus Arteriosus**

**Check list**

- Residual VSD
- Truncal valve stenosis
- Truncal valve regurgitation
- RV to PA conduit stenosis
- RV to PA conduit regurgitation
- Branch pulmonary artery stenosis

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**Total Anomalous Pulmonary Venous Return (TAPVR)**

**Classification of TAPVR**

<table>
<thead>
<tr>
<th>Type</th>
<th>Drainage to</th>
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<tbody>
<tr>
<td>Supra-cardiac</td>
<td>innominate/SVC/RA</td>
</tr>
<tr>
<td>Cardiac</td>
<td>coronary sinus</td>
</tr>
<tr>
<td>Infra-cardiac</td>
<td>ductus venosus/IVC/Hepatics</td>
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**TAPVR- Correction**

- The confluence is anastomosed to the back of the left atrium
- ASD is closed
- The vertical vein is generally ligated

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**TAPVR- Follow up**

- Generally they do very well
- Follow up is still recommended - no set guidelines
  - Check anastomosis
  - Check pulmonary veins - for stenosis
  - Arrhythmia

Echo can be primary modality
MRI will be the ideal modality
Cath if need intervention for PV stenosis

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**Single Ventricle**
Tricuspid Atresia/Hypoplastic right heart syndrome

- Complete agenesis of the tricuspid valve
- No direct communication of the right atrium and right ventricle.
- Also spectrum of Hypoplastic right heart syndrome with pulmonary atresia

Tricuspid Atresia
Hypoplastic right heart syndrome

Hypoplastic left Heart Syndrome

- Basic concept: The left side cannot handle a full cardiac output
- Mitral valve atresia/stenosis
- Varying degrees of LV hypoplasia
- Aortic atresia/stenosis
- Aortic arch hypoplasia

Stage I, Surgical Palliation Surgery for Single ventricle physiology- Norwood Operation – At birth

- Atrial septectomy
- Concept: Functioning single ventricle connected to one systemic artery (PA and Aorta combined to form neo-aorta)
- Borrow blood from the single systemic artery to supply blood to lungs
- Systemic to Pulmonary artery shunt- modified Blalock-Taussig shunt
- Sano Shunt: direct connection from single ventricle to PA

Stage II, Surgical Palliation for Single Ventricle- At around 4-6 months of age

Bi-directional Glenn Shunt
- Anastomosis of the Superior vena cava and Right pulmonary artery
- Called bidirectional since blood goes to both PA's
- Usually with take down of the A-P shunt/Sano shunt
Stage III. Surgical Palliation for Single Ventricle—around 4 years of age

Fontan Procedure

- Conduit from the IVC and hepatic veins to the RPA.
- Previously intracardiac baffle
- Presently extracardiac baffle

IVC → SVC → RPA → Lungs → Pulmonary veins → LA → LV → Aorta

Single Ventricle Pathway—Summary

- One functioning ventricle
- First operation—
  - Norwood and associated shunt
    - B-T shunt or RV to PA (Sano) Shunt
- Second Operation
  - Bidirectional Glenn Operation
- Third and Final Operation
  - Fontan Operation
    - Intracardiac or extracardiac
    - Fenestration or no fenestration

First Stage Operation—Norwood with BT shunt

| Aorta to pulmonary artery shunt - BT Shunt |
| Both arteries are combined to form one outlet - Norwood Procedure |

First Stage Operation—Norwood with RV to PA shunt (Sano Shunt)

| Both arteries are combined to form one outlet plus RV to PA conduit |

Second Stage Operation—Glenn Procedure (BLBDG)

| Here the (SVC) is connected directly to the pulmonary artery |

Intracardiac Fontan Operation

| Atropulmonary Fontan |
Third and final Staged operation: Glenn with Intracardiac Lateral tunnel Fontan

Bidirectional Glenn and Extracardiac Fontan

Check List- Single Ventricle Structural Issues
- Unrestrictive atrial level communication
- Fontan fenestration- YES or NO
- AV Valve regurgitation
- Ventricular function
- Neo-aortic regurgitation
- Aortic arch patency

Check List- Single Ventricle Other Issues
- Arrhythmia-  
  - Due to suture lines and chamber enlargement  
  - Inherent predisposition  
  - Pacemaker/ ICD  
- Thromboembolic events  
  - Due to poor Ventricular Function  
  - Sluggish flow in Fontan circuit  
  - Hypercoagulable state  
- Coronary ischemia  
- Exercise capacity  
- Hepatic dysfunction  
- Plastic Bronchitis  
- Protein Losing Enteropathy (PLE)  
- Suitability for pregnancy  
- Suitability for Transplant

Check List- Single Ventricle Evaluation- Testing
- Echocardiography  
  - TTE  
  - TEE (before cardioversion for sure)  
- CMR- Ideal  
- CT scan  
- Exercise Stress tests  
- Holter/ Event Monitors  
- Labs annually

Check List- Single Ventricle Evaluation- Testing
- New onset tachycardia-  
  - Hemodynamic evaluation by catheterization  
- Image the liver – US/ CMR - Hepatic congestion, Cirrhosis, Hepatocellular carcinoma  
- Evaluation for cardiac transplant (PLE, Plastic bronchitis)
Check List- Single Ventricle
Rhythm Issues

- Sinus node dysfunction 45 % of adults
- Atrial tachycardia- can occur in 60 % of adults with Fontan
  - Rate control
  - Rhythm control
- New onset tachycardia- Hemodynamic evaluation by catheterization
- Catheter ablation especially with a Maze procedure
- AV synchrony has to be achieved
- TEE needs to be done before cardioversion to rule out thrombus

Check List- Single Ventricle
Medications

- Diuretics
- Anticoagulation
  - Suspected thrombus
  - Embolic events
- Usage of pulmonary vasoactive medications
  - Sildenafil, Bosentan
- Aldosterone antagonists
- Sub cutaneous heparin
- Budesonide for PLE and plastic bronchitis

Failing Fontan

- Fontan – obligatory increased CVP
- Decreases cardiac output
  - Protein loosing enteropathy
  - Hepatic dysfunction
  - Lower extremity venous congestion
  - Exercise limitation
  - Plastic Bronchitis

Single Ventricle
Protein Loosing enteropathy (PLE)

- More than 10% of Fontan patients develop PLE
- 5 year survival – 50%
- Increased systemic venous pressure
- Increased retrograde pressure and thoracic duct pressure
- Dilated lymphatic vessels
- Loose protein, lymphocyte and chylomicrons in the gut
- Pleural effusion, ascites, peripheral edema, diarrhea

Diagnosis:
- Decreased serum protein/albumin
- Increased alpha 1 antitrypsin concentration in stool
- r/o urine protein loss

Protein Loosing enteropathy (PLE)

- Average time from Fontan operation to diagnosis of PLE is 3.5 years.
- Severe PLE may manifest hyponatremia (both relative and absolute)
- Increased susceptibility to infections due to lymphopenia (intestinal loss of circulating lymphocytes) and/or hypogammaglobulinemia (intestinal loss of circulating immunoglobulins).
- Rarely, a diffuse coagopathy can occur due to liver congestion and to loss of coagulation factors.
- Gold standard for diagnosis of PLE is a 24-hour stool α-1-antitrypsin (AAT) clearance study.
- If performed, biopsy of the small intestine can reveal dilated lacteals and other pathologic changes that are also hallmarks of PLE

Grading of PLE
  - Mild (Grade I PLE)- Serum albumin between 2.5 and 3.5 mg/dL
  - Moderate (Grade II), between 2.0 and 2.5 mg/dL
  - Severe (Grade III)- less than 2.0 mg/dL

Protein Loosing enteropathy (PLE)- Treatment

- First try to get to the bottom of the reason
  - Echo, Cath, CMR
  - Normalize rhythm

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<tr>
<th>Treatment</th>
<th>Description</th>
<th>Monitoring/ Risk Factors</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dietary</td>
<td>High protein, high medium-chain triglycerides, contained sodium, low fiber</td>
<td>Monitor fluid status, albumin, compliance, electrolytes</td>
</tr>
<tr>
<td>Pharmacologic</td>
<td>1 mg/kg oral increasing x 1 modeled on symptoms</td>
<td>Hypokalemia, other electrolyte problems</td>
</tr>
<tr>
<td>Spironolactone</td>
<td>2-3 mg/kg per day divided bid</td>
<td>Hypokalemia, other electrolyte problems, deconditioning</td>
</tr>
<tr>
<td>Vasopressin</td>
<td>0.5-1 mg/kg three weekly given with 25% D5W</td>
<td>Hypopotassemia, hypoxia, arterial suppression, vasoplegia</td>
</tr>
<tr>
<td>Bosentan</td>
<td>125 mg twice daily</td>
<td>Hypoxia, systemic hypertension, pulmonary hypertension, arterial suppression, vasoplegia</td>
</tr>
<tr>
<td>Budesonide</td>
<td>6-8 mg per day</td>
<td>Renal, hepatic, autoimmune, immunosuppression</td>
</tr>
<tr>
<td>Steroids</td>
<td>Body weight adjusted low molecular weight</td>
<td>Renal failure, heart failure, organ failure, infection, bleeding, steroid related complications</td>
</tr>
</tbody>
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International classification of disease classification (ICD-10)

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