Complex Congenital Heart Disease

Iowa ACC- Echo Lecture series
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NO DISCLOSURES

Complex CHD-
Learning Objectives

• Types of Cyanotic heart diseases and their pathophysiology
  – Not palliated/ Not repaired
  – Palliated
  – Corrected
• Treatment and Surgical corrections
• Overview of Echo Findings
• 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)

Miscellaneous
- Ebsteins' Anomaly
- CCTGA
- Criss cross hearts, Isomerism, Heterotaxy

Relative Frequency of Cyanotic Congenital Heart Disease

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

Commonest cyanotic/complex congenital heart disease- Tetralogy of Fallot
Syndromes and CHD

- Di-George/ 22.q 11 deletion: Cono-trunical lesions (TOF, TA)
- Downs Syndrome: VSD and AV canal
- Marfan Syndrome: Aortic dilation and dissection, MVP
- Loeys-Dietz Syndrome: Aortopathy with dissection
- Noonan Syndrome: PS, HCMP
- Turners syndrome: BAV, COA and PAPVR
- Williams Syndrome: Supravalvar AS, Peripheral PS
- Holt Oram Syndrome: ASD and radial bone abnormalities
- Trisomy 13: VSD, PDA
- Trisomy 18: VSD, PDA

Surgeries in CHD

- Blalock Taussig Shunt (B-T shunt): Systemic to Pulmonary shunt (In TOF)
- Potts Shunt: Descending aorta to LPA shunt
- Waterston shunt: Ascending aorta to RPA shunt
- Norwood Operation: reconstruction to make single outflow Aorta and PA combined (Single Ventricle- first stage)
  - Sano shunt: RV to PA conduit (Single ventricle )
  - B-T shunt
- Bidirectional Glenn shunt: SVC to RPA (Single ventricle 2nd stage)
- Fontan Operation: IVC to PA: (Single ventricle- 3rd stage)
### Surgeries in CHD

- **Rashkind Procedure**: Balloon atrial septostomy (d-TGA)
- **Jatene Operation**: Arterial switch operation (d-TGA)
  - Le-Compte Maneuver: PA's wrapping over the aorta
- **Mustard/ Senning**: Atrial switch operation (d-TGA)
- **Rastelli Procedure**: RV to PA conduit and generally VSD closure
- **Warden Operation**: Rerouting anomalous PAPVR to LA and SVC anastomosis to Rt. atrial appendage
- **Brock Operation**: Pulmonary valvotomy

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### Tetralogy of Fallot (TOF)
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

Tetralogy of Fallot – PLAX View
Tetralogy of Fallot – PLAX Sweep

Tetralogy of Fallot – SAX view
Tetralogy of Fallot – Apical Sweep

Tetralogy of Fallot – Pulmonary Atresia
PLAX and SAX views
Tetralogy of Fallot – status Post Repair
Apical view

Tetralogy of Fallot – RVOT Stent
Tetralogy of Fallot –s/p repair with RV to PA Conduit

Tetralogy of Fallot –s/p repair with residual VSD
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
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

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 great arteries- PLAX
Transposition of great arteries-
SAX

Transposition of great arteries-
Subcostal sweep
Transposition of great arteries-
Apical view

Transposition of great arteries-
SAX Branch PA’s
Transposition of great arteries- s/p Senning Procedure

Transposition of great arteries- s/p Senning: Baffle Leak
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

Normal atrio-ventricular concordance
Normal ventriculo-arterial concordance
ASD closure
** 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
D- TGA s/p Senning operation

Systemic Venous Baffle  Pulmonary venous Baffle

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

- Also know 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) PLAX view
Congenitally Corrected transposition (L-TGA)- PSAX View

Congenitally Corrected transposition (L-TGA)- Apical View
**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

From DiBardino DJ, et al.

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

**Shows Posterior RV, Tricuspid regurgitation**
Congenitally Corrected transposition (L-TGA)

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

Truncus Arteriosus
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.

Truncus Arteriosus - Subcostal view
Truncus Arteriosus-
Subcostal view

Truncus Arteriosus
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
Total Anomalous Pulmonary Venous Return (TAPVR)

Classification of TAPVR

- **Supra-cardiac type**
  - TAPVC drainage to the innominate/SVC/RA

- **Cardiac type**
  - TAPVC drainage to the coronary sinus

- **Infra-cardiac type**
  - TAPVC drainage to the ductus venosus/IVC/Hepatics
TAPVR- Echo

Supra-cardiac type
Showing the confluence behind the LA

Supra-cardiac type
TAPVC drainage to the innominate/SVC/RA

Supra-cardiac type
Showing obligatory Right to left shunt

TAPVC: Pulmonary veins in relationship to other trachea, aorta and PAs
TAPVR- Correction

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

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
Ebsteins Anomaly

Ebsteins Anomaly - 4 Ch and PLAX
Ebsteins Anomaly- TR

Ebsteins’ Anomaly- Inflow outflow and PSAX view
Single Ventricle

Single Ventricle- DILV
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
Hypoplastic left Heart Syndrome

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
Atriopulmonary Fontan
Third and final Staged operation: Glenn with Intracardiac Lateral tunnel 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
- Glenn anastomosis
- Fontan circuit patency
- Branch pulmonary arteries
- Pulmonary venous issues
- Aortopulmonary collaterals
- Veno-Venous Collaterals
- Arteriovenous malformations
Thank you for your attention !!!!
Questions ???