ECHOCARDIOGRAPHY IN ADULT CONGENITAL HEART DISEASE
Adult congenital heart disease have a variety of abnormalities.

- Includes:
  - i. Left sided outflow abnormalities
  - ii. Right sided outflow abnormalities
  - iii. Left to Right shunting lesion
  - iv. Cyanotic congenital defect
  - v. Valve abnormalities
The most common diagnoses are

- VSD
- ASD
- AS
- TOF
- Coartation
- TGA
- AVSD

(Ohio State University Medical Centre 1996 to 1999)
- Untreated congenital heart disease during childhood can become more complex in terms of patient management.

- Pulmonary hypertension is the primary cause a change in the patient’s life.

- Medical therapy, cardiac intervention and also cardiac surgery is needed to treat these patient.
In our institution a lot of complex congenital heart diseases are commonly seen.

Almost all congenital heart diseases require particular attention during the echo assessment. These include:

1. ASD
2. VSD
3. TOF
4. PDA
5. TGA
6. CCTGA
7. Ebstein’s Anomaly
8. Coartation
9. Pulmonary Stenosis
Deficiency of tissue from the interatrial septum
About 7 to 10% of infants with congenital heart disease have ASD
In 30% of ASD other associated lesions are:

1. Partial or total anomalous venous drainage
2. Congenital mitral stenosis
3. Mitral Valve Prolapse
4. Ventricle Septal Defect
5. PDA
6. Pulmonary Stenosis
7. Coarctation Of Aorta
TYPES OF ASD

ASD

OSTIUM SECUNDUM
OSTIUM PRIMUM
SINUS VENOSUS
CORONARY SINUS DEFECT
Primun ASD

Secundum ASD

Sinus Venosus ASD
TYPES OF VSD

VSD

PERIMEMBRANOUS
MUSCULAR
DCSA
INLET
Perimembranous VSD

- Located at the junction of muscular, atrioventricular & outlet portion of the septum

- Immediately adjacent to the aortic and tricuspid valve

Muscular VSD

- Muscular VSDs are surrounded by a complete muscular rim and are most often located in the apical two thirds of septal myocardium.

- Muscular defect can also present in the posterior inlet septum
- **Doubly Committed Subarterial VSD (DCSA)**
  - Located in the outlet septum
  - Defect adjacent to aortic & pulmonary valve annulus
  - RCC frequently prolapsed into VSD causing aortic valve regurgitation

- **Inlet VSD**
  - This VSD usually occur as part of a complete AV canal defect but it may occasionally be seen in isolation.
  - This type VSD occur near the AV valves.
**PMVSD**
- Defect located near the TV from short axis view

**DCSA VSD**
- Near to PV and Aortic valve at outlet portion
Muscular VSD
- Located at 2/3 of apical ventricle septum

Inlet VSD
-Inlet VSD is near to AV valves
PDA

- Ductus arteriosus close in the first 48 hours after birth
- More common in female than male
- Size and origin of the duct varies
- PDA shunt is determined by the resistance of the duct and the level of PVR
Suprasternal View

Parasternal Short Axis

Ductal View
PDA DOPPLER PATTERN

SMALL PDA

MODERATE SIZE PDA
TOF occurs in 10% of all congenital heart diseases.

This is the most common cyanotic heart defect seen in children beyond infancy.
The description of TOF includes 4 abnormalities:

- Large VSD
- Right Ventricle Outflow Obstruction
- Right Ventricle Hypertrophy
- Overriding of Aorta
The most frequent form of RVOT obstruction is infundibular stenosis (45%) and Pulm. Valvular stenosis (10%). A combination of 2 may also occur (30%).

Usually Pulm. Valve annulus and main PA are hypoplastic in most patient.
Overriding Of Aorta

RVOT Obstruction
Doppler of CW in RVOT Obstruction
What do u think?
D- TGA occur in about 5 % of all congenital heart defect. It is most common in males than in females (3 : 1).

In adult population D- TGA (simple TGA) is a rare condition. But TGA will present with other complex lesion in adult such as TGA with VSD and PS.

TGA without ASD or VSD will not present in adult population because usually they either require early surgical intervention or die early.
Pathology

- In D-TGA the aorta arises anteriorly from RV carrying desaturated blood and pulmonary artery arises posteriorly from LV carrying oxygenated blood to lung.
  (in normal heart aorta arises from LV chamber (post) and pulmonary arises from RV chamber (ant))

- VSD is present in 30% to 40% with D-TGA heart.
Pulmonary is arising from LV

PA Branches

In Parasternal Long Axis Aorta And Pulmonary is in Parallel position
Aorta is Ant. while Pulm. is posterior
(in normal heart pulm should be ant. while aorta is post.)

Parallel Aorta and Pulm.
(in normal heart in parasternal view aorta and pulm. should cross each other)
Aorta and Pulmonary relation.

In Short Axis View

appearance of “a double circle” is seen in TGA

In Short Axis View

appearance of “circle and sausage” is seen in normal heart
CCTGA

- Congenitally Corrected Transposition Great Artery (or ventricle inversion) occurs in < 1 % of all patients with congenital heart disease.
Pathology

- The physiology relation is normal

- In CCTGA the RA empties into anatomic LV through mitral valve and the LA empties into the RV through tricuspid valve. The RV is located to the Left morphology ventricle and the LV is located to the right morphology ventricle which is called VENTRICLE INVERSION.

- The great arteries are transposed, with the aorta arising from RV and the PA arising from LV.

- The aorta is located to the Left and anterior to the PA.
About 50% of CCTGA have dextrocardia.

In CCTGA theoretically no functional anomalies but most cases are complicated by associated intracardiac defect and arrhythmia.

The main issue is that the RV is now the systemic ventricle and the natural history is that it will start to fail in the 3rd-4th decade of life.
- In CCTGA morphological RV on the Left side.
- The best way to distinguish RV and LV is the septal attachment of Tricuspid leaflet to the septum (ventricle inversion)
- TV off-setting toward the apex seen on left side ventricle (RV)

- In normal heart the mitral valve can be seen clearly attached toward the lateral aspect of ventricle
- Short axis scan
  "double circle" is image

- 4 chamber view with ventricle inversion and trabeculation of RV at Left sided morphology ventricle
Different relation between TGA and CCTGA

TGA
Aorta is anterior and mainly to the Right Pulmonary

CCTGA
Aorta is anterior and mainly to the Left Pulmonary
Ebstein’s Anomaly

- Ebstein’s anomaly of the Tricuspid valve occur in < 1 % of all congenital heart defect
Pathology

- Downward displacement of septal and posterior leaflet of tricuspid valve into RV cavity (atrialized RV) and the functional RV cavity is small.

- Usually the septal leaflet is plastered toward septum and the anterior leaflet elongated (redundant, resembling like sail).

- May be associated with tricuspid regurgitation.

- ASD/PFO usually present and occasionally other defect such as PS, VSD might occur.

- Severe form of Ebstein usually will present at early age and requires early intervention.
Ebstein Anomaly

- Plastered septal leaflet to ventricle septum

- Redundant and elongated ant. Tricuspid leaflet

- Small functional of RV cavity
RVOT obstruction may present in severe form of Ebstein Anomaly
Coarctation Aorta

- Coarctation of Aorta occur in 8% to 10% of all cases of congenital heart defect. It is more common in males than female ratio 2:1.
Coarctation is a restrictive flow through the aortic arch.

Type of coarctation varies like long tubular hypoplastic arch, discrete aortic arch etc.

Coarctation is almost always in a juxtaductal position.

As many as 85% of patients with COA have a bicuspid aortic valve.
- Discrete type of coarctation with restrictive flow below left subclavian
Diastolic Run OFF from CW doppler

Chamber size usually dilated when patient grow
PULMONARY STENOSIS

- Pulmonary valve stenosis can occur as an isolated defect or as part of congenital heart disease.
Several types of Pulmonary Stenosis include:

1. Infundibular stenosis.
2. Valvular pulmonary stenosis.
3. Subvalvular pulmonary stenosis.
4. Supravalvular pulmonary stenosis.
- Infundibular stenosis (napkin-ring)

  - Stenosis occur mainly at Right ventricular outflow tract
  - The RV proximal to obstruction usually hypertrophied
  - Usually a thick rigid muscular seen at the level of the infundibulum ostium.
Valvular Pulmonary stenosis

- Occur at pulmonary valve

- Usually the pulmonary valve is thickened and domed in systole

- Post stenotic dilatation usually involve the main pulmonary artery and also Left pulmonary artery because of the jet flow through the stenotic orifice pulmonary valve to main pulmonary artery and also to Left pulmonary artery.
Subvalvular Pulmonary stenosis

- Subvalvular pulmonary stenosis occur between the RV and pulmonary conus

- Usually the stenosis at the level just below the pulmonary valve (proximal) and between the infundibulum. (discrete stenosis of the infundibulum ostium).
- Supravalvular Pulmonary Stenosis

- Supravalvular Pulmonary stenosis occur distal to pulmonary valve and may involve main pulmonary artery and the two pulmonary branches.

- Type of stenosis may be discrete membrane stenosis, tubular and may occur at numerous area of stenosis.
Suggestion grading of Pulmonary Stenosis according to the peak gradient.

- Peak gradient of Pulmonary stenosis less than 1/3 Systemic blood pressure is Mild PS.
- Peak gradient 2/3 of systemic blood pressure is moderate PS.
- Peak gradient of more than 2/3 systemic pressure is equal to severe PS.
Valvular PS

Infundibular PS

Supravalvular PS

Medial papillary muscle
Membranous septum
Outlet septum
TV
RA
PA
LA
Have a nice day
THANK YOU

HANIF OSMAN, RDCS