Simplified TEE Assessment for Complex Adult Congenital Heart Disease

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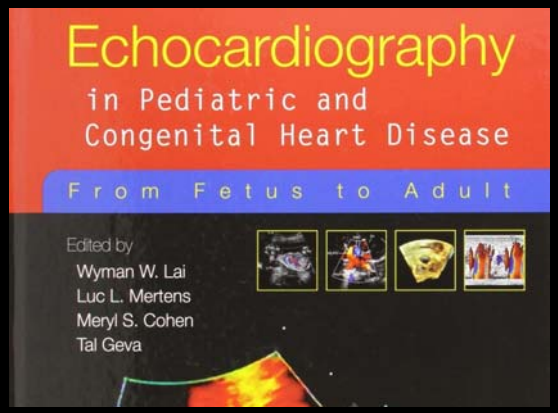
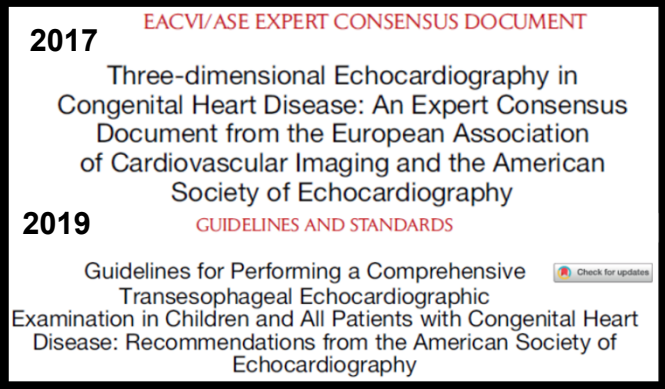
http://pie.med.utoronto.ca/TEE/

CHD Getting Started

Books/websites

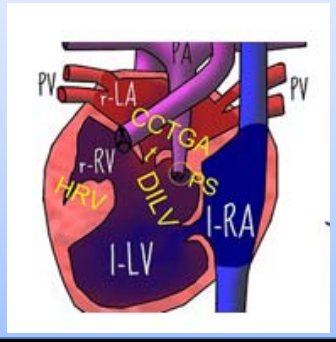


Guidelines



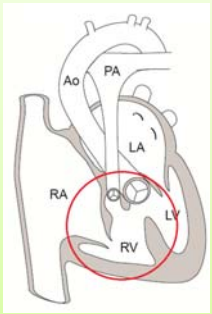
Experts

Anatomy



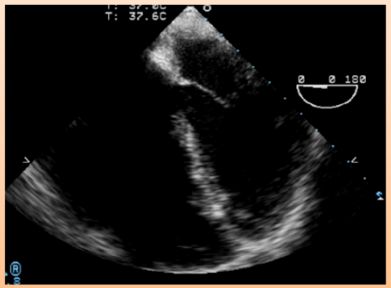
1

☑ TEE Checklist



2

TEE Images

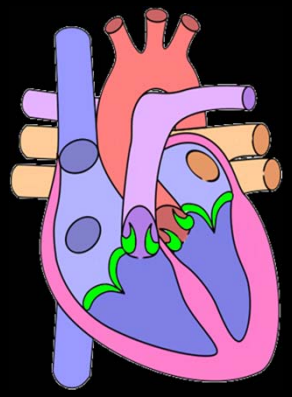


3

CHD Spectrum (AHA)

Simple
- Small ASD, VSD
- Mild PS
- Repair PDA, ASD, VSD

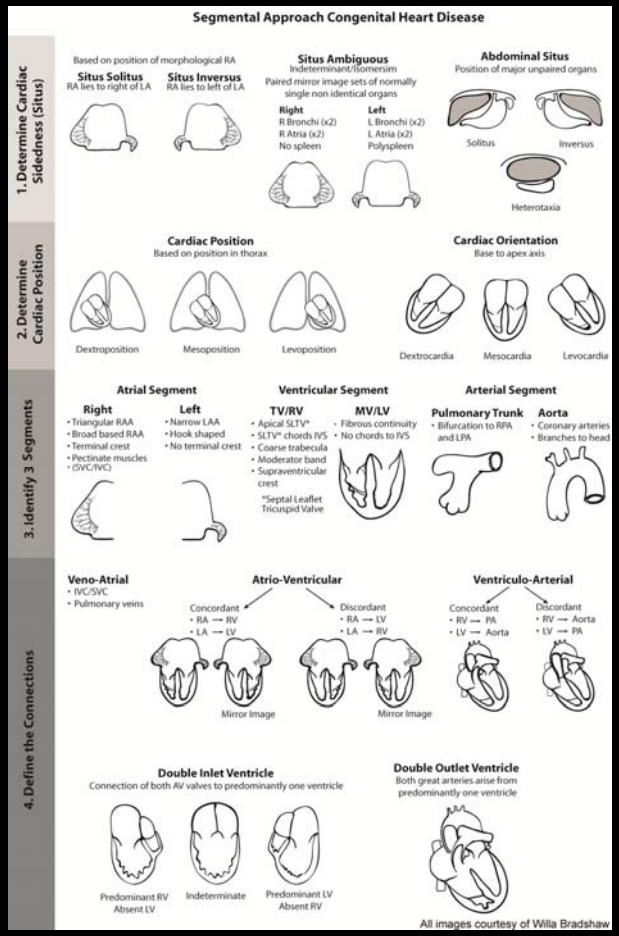
Moderate
- AV, MV
- AS (sub/supra)
- SOVA, Aorta LV fistula
- PAPVD
- AVSD
- Ebstein's
- TOF, RVOT infundibular
- PV regurge/stenosis
- PDA
- ASD primum, secundum, sinus venosus



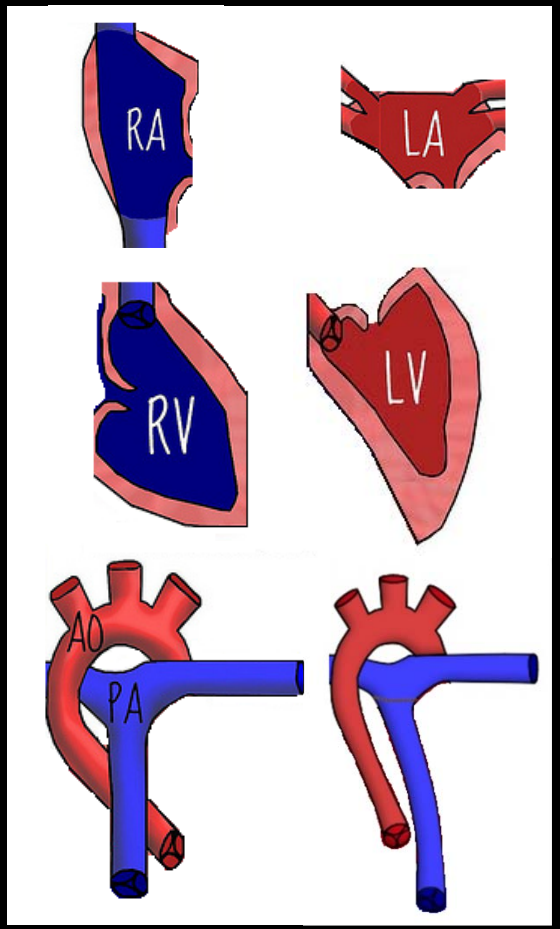
Complex
- All cyanotic hearts
- Double outlet ventricle
- Fontan
- Interrupted aortic arch
- Mitral atresia
- Single Ventricle
  - Tricuspid atresia
  - Double inlet ventricle
  - Pulmonary atresia
  - Hypoplastic L heart
- TGA (D-TGA, L-TGA)
- Truncus arteriosus
- Heterotaxy

Stout K, et al. J Am Coll Cardiol 2019;73:1494-1563.

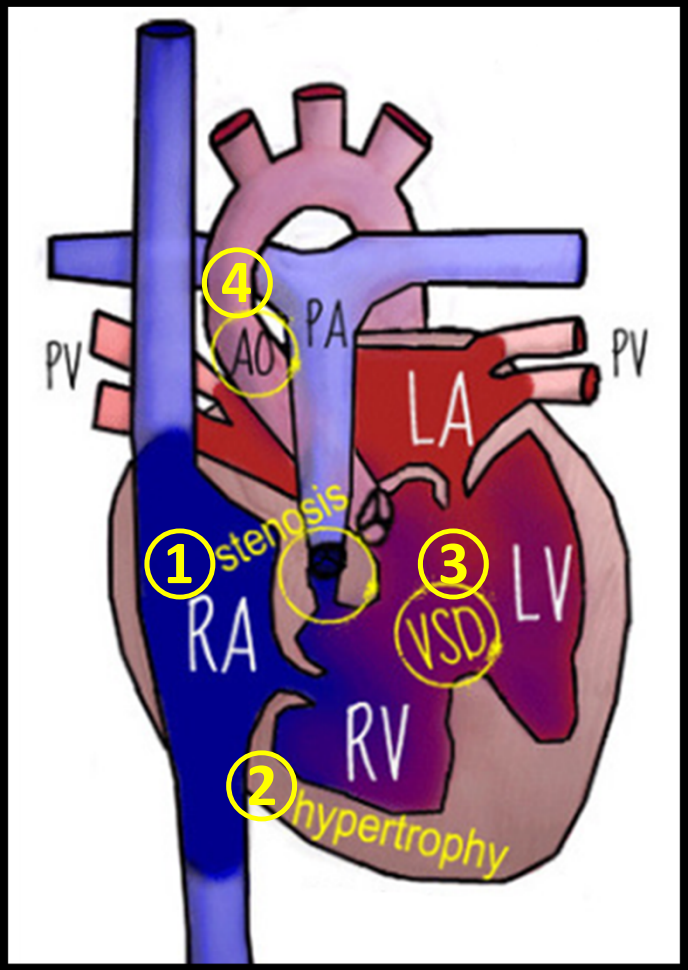
In The Beginning



- Terminology
  - Understand language
  - Use structure approach
  - Based on morphology
- Identify structures
  - Atria
  - Ventricles
  - Great vessels
- Secondary changes
- Associated anomalies
- Surgical modifications
- Where does the blood flow ?



Tetralogy of Fallot Anatomy



4 components

1. RVOTO (dynamic)
2. RVH
3. VSD
4. Overriding aorta (cyanosis)

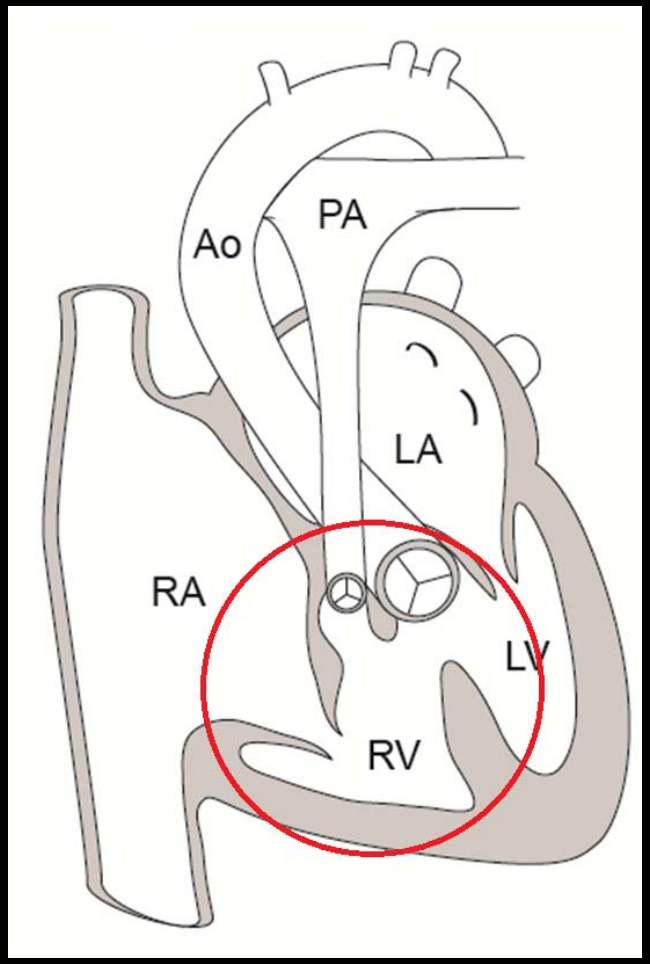
\+ ASD (pentalogy)

Unoperated
< 3% at 40 y.o.

Associated

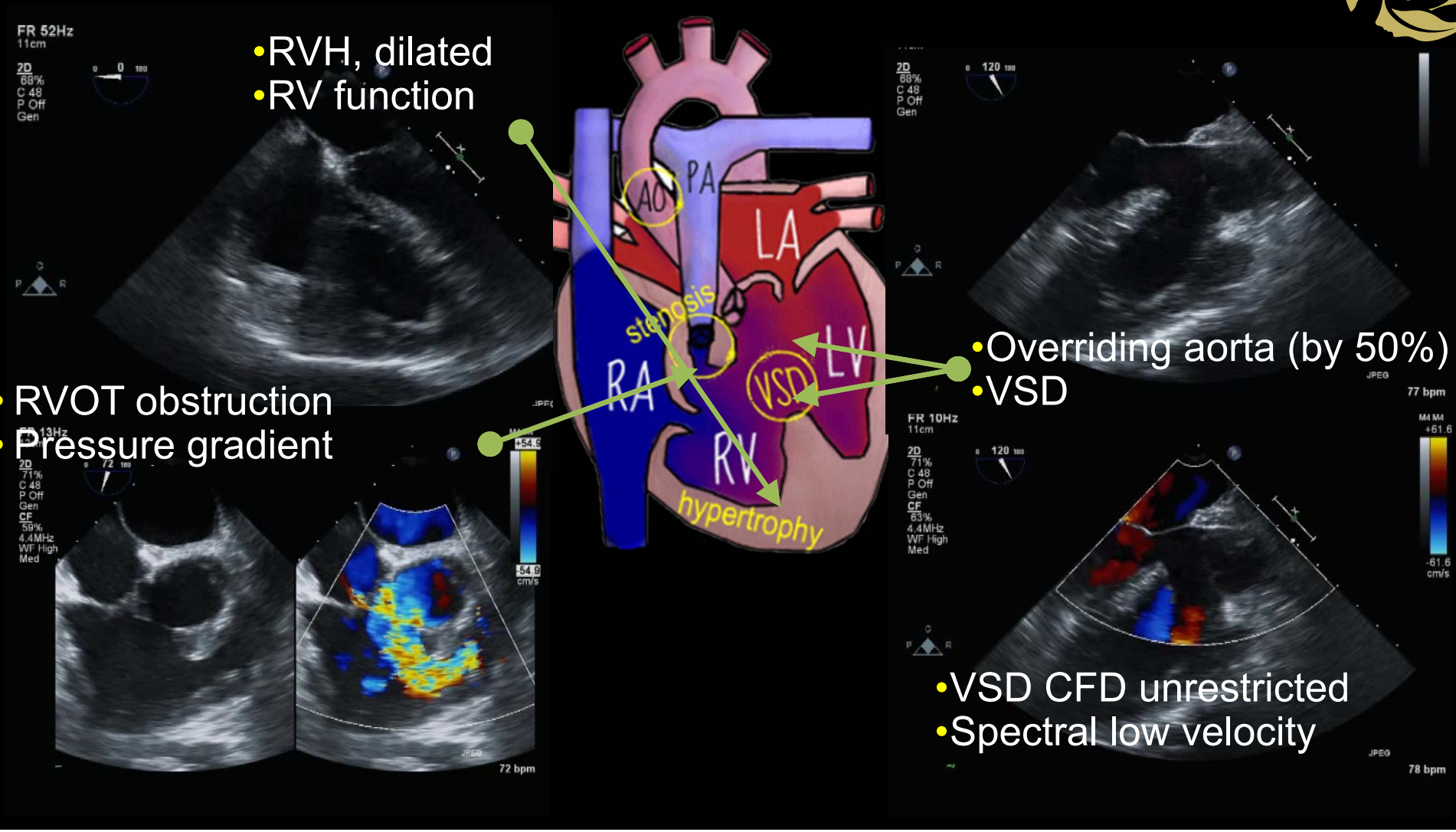
- R aortic arch
- PV atresia + VSD
- Coronary artery anomalies
- LSVC to coronary sinus
- AVSD = 'tet-canal' defect

TEE Checklist Uncorrected TOF



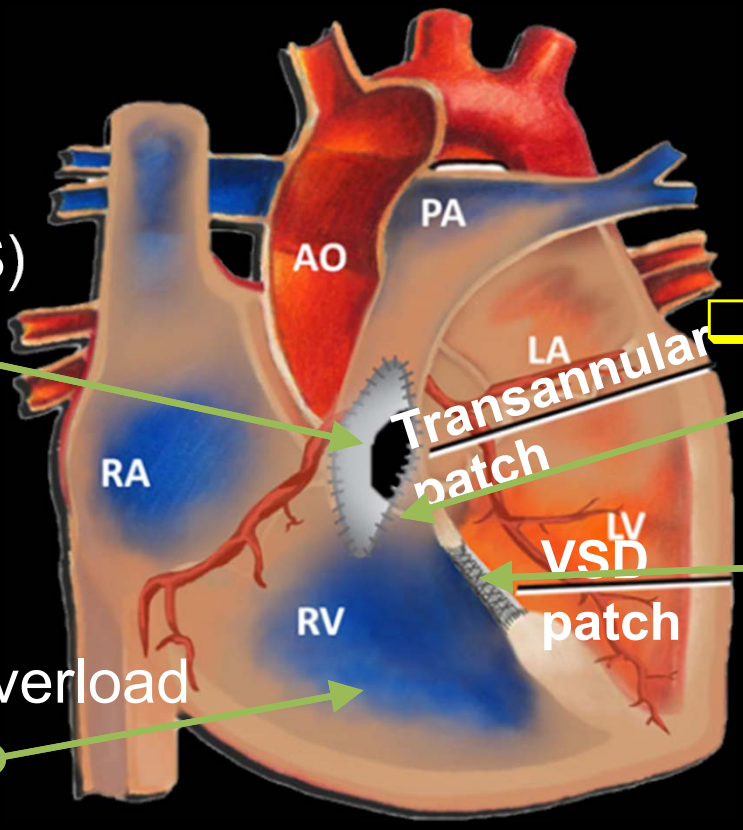
- VSD (size, location)
- Evaluate RVOT (sub/supra /valvar)
  - Morphology, obstruction, gradients
- Baseline ventricular function, RVH
- Aortic override /AV competence
- PA size
- Origin + course of coronary arteries
- Associated anomalies

TEE Uncorrected TOF



TEE Checklist Corrected TOF

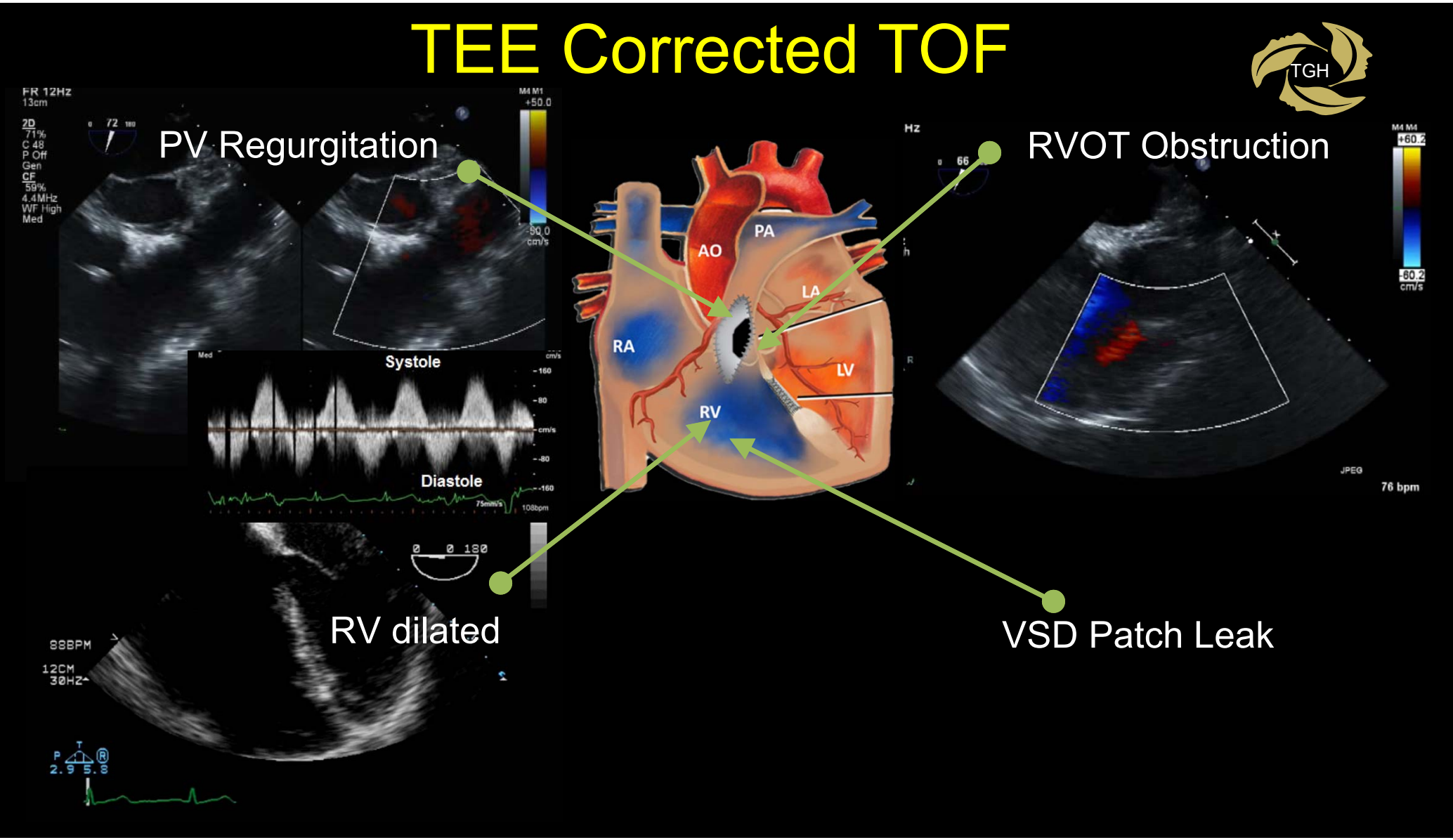
- Pulmonic valve (PI or PS)
  - Color Doppler
  - CW Doppler
- RV vol or pressure overload
  - Size + function
  - TR (RVSP)



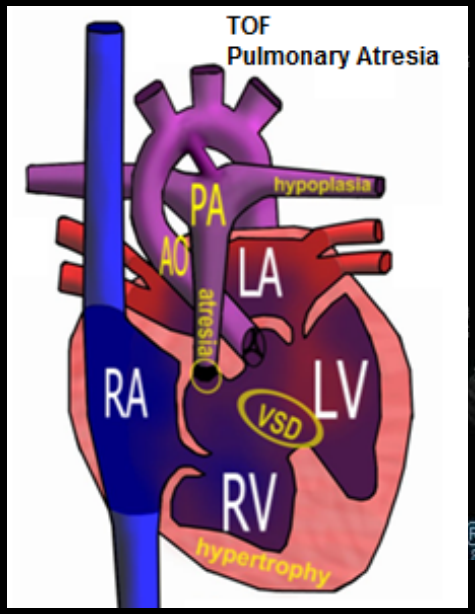
https://www.kidsheartshouston.com

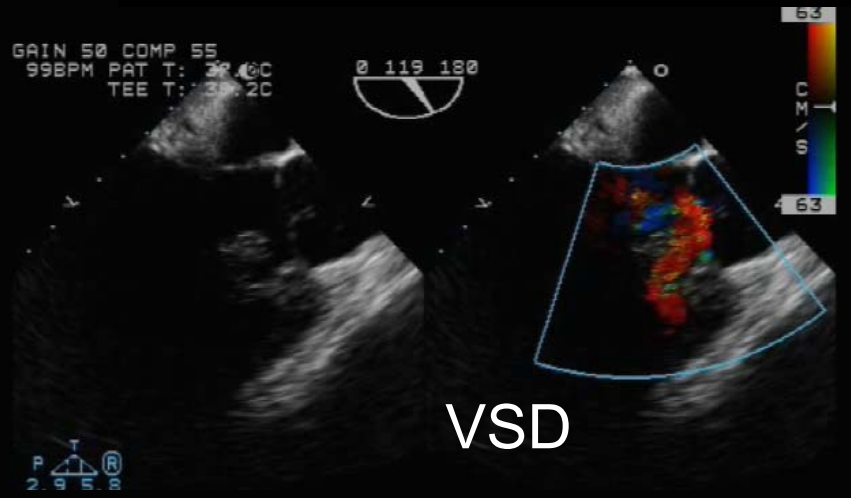
- RVOT obstruction
- VSD patch leak

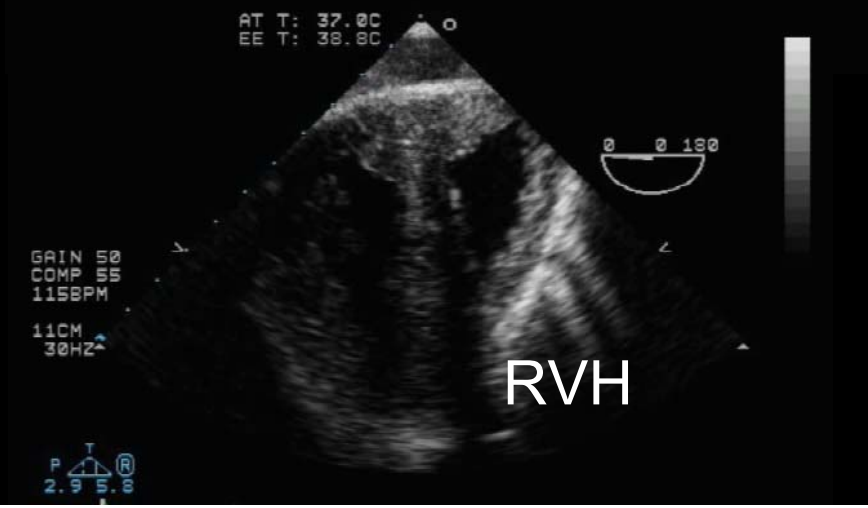
TEE Corrected TOF



Pulmonary Atresia + VSD

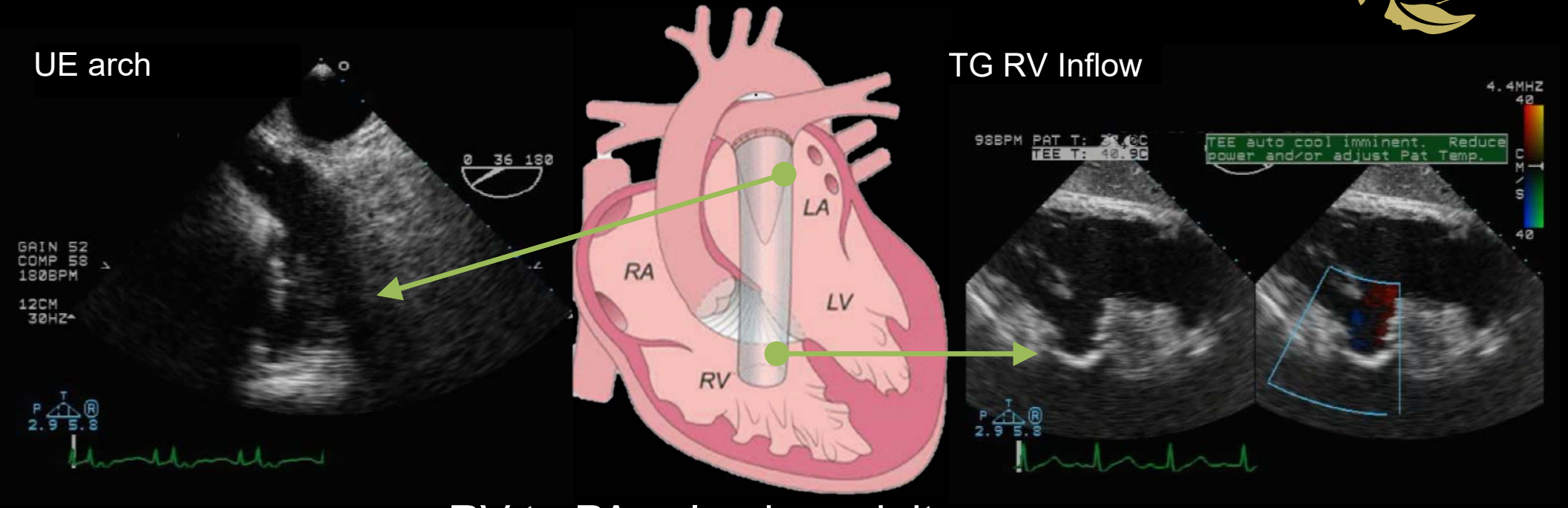






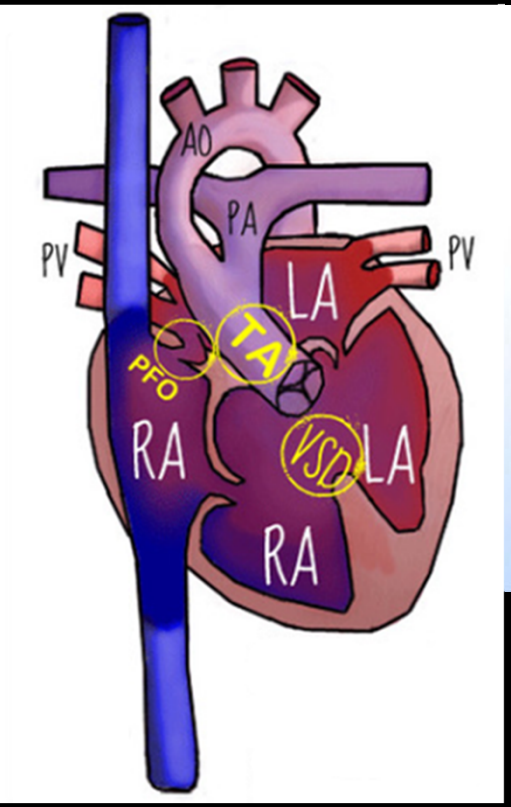
- Most severe form of TOF
- No RV to PA continuity
- Pulmonary blood flow variations
  - Confluent: RPA + LPA connected
  - Not confluent

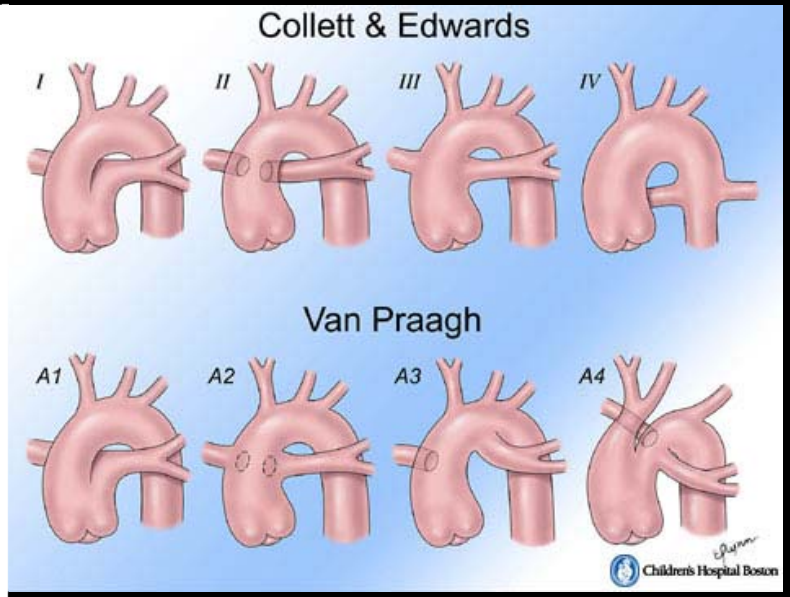
Rastelli Procedure



- RV to PA valved conduit
- If RV + TV well developed
- D-TGA, over-riding aorta (TOF), DORV
  - + VSD
  - + RVOTO (PS, PV atresia, muscle)

Truncus Arteriosus

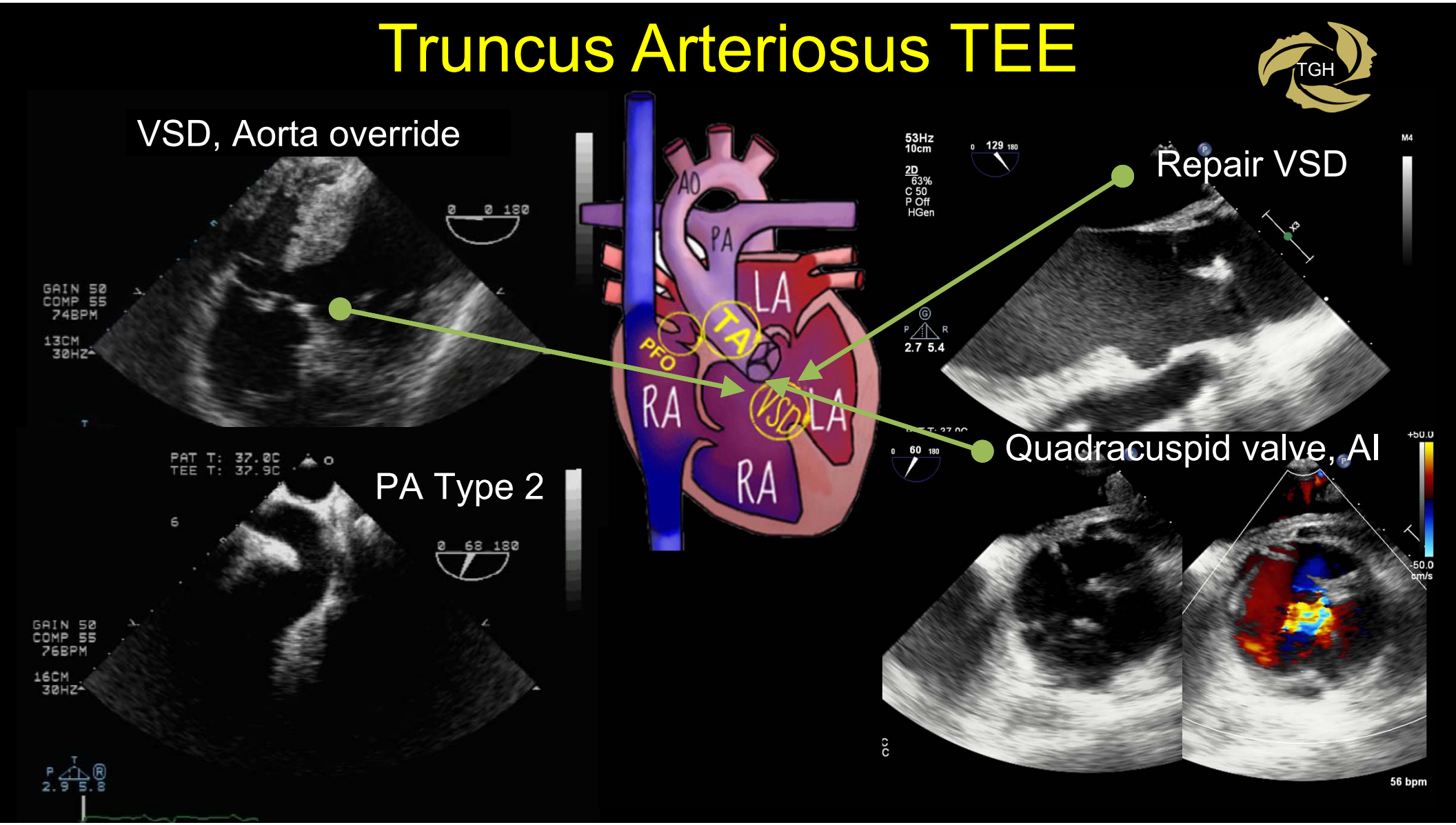




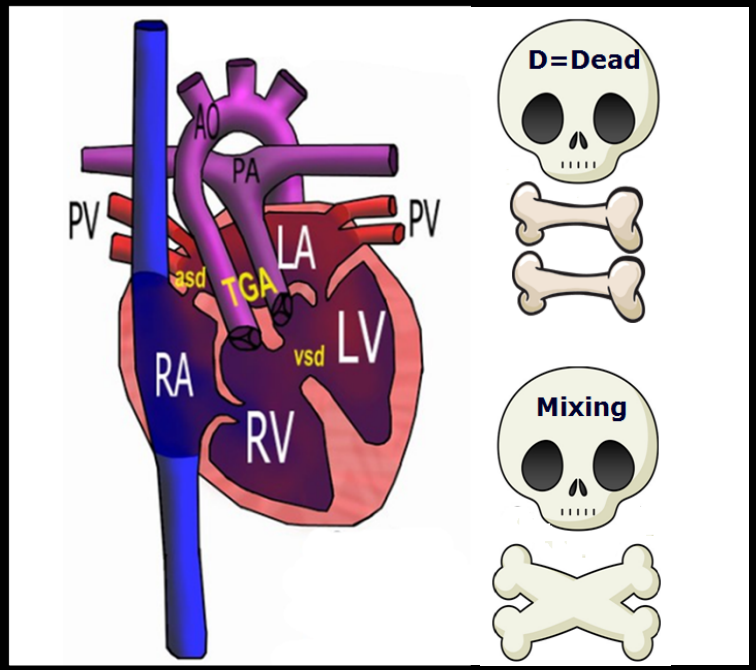
Type 1: main PA → RPA/LPA
Type 2: common trunk → RPA/LPA
Type 3: RPA ← common trunk
LPA ← PDA or aorta
Type 4: Arch hypoplasia, interrupt or coarctation + large PDA

- Single arterial trunk, gives rise to PA + aorta
- Single V-A valve
- Overriding VSD
- 4 types based on PA origin
- Associated: VA regurgitation, RAA, coronary ostia

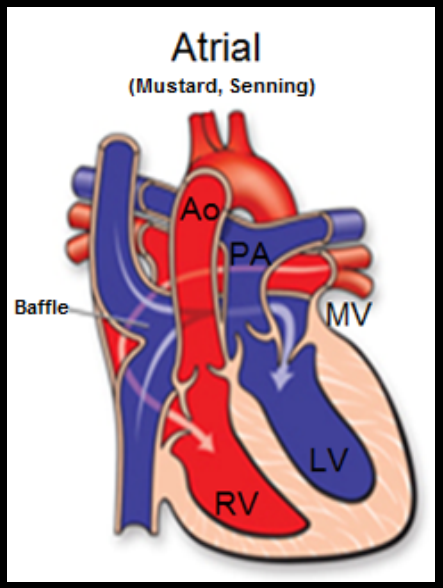
Truncus Arteriosus TEE



Transposition D-TGA Anatomy

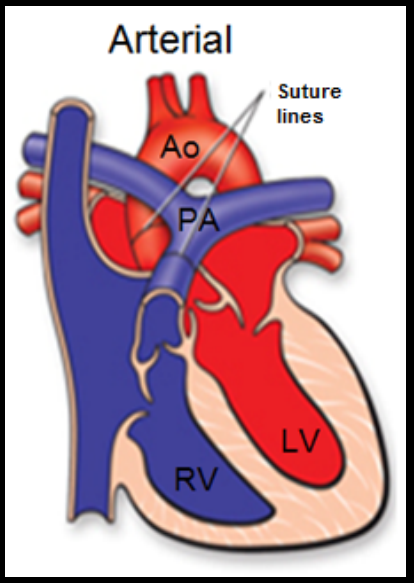


- Ventriculoarterial discordance
  - RV (systemic) → aorta
  - LV (pulmonic) → PA
- Mixing: PDA, ASD, VSD



Atrial Switch

- Systemic venous return to LV
- Pulmonary veins to RV

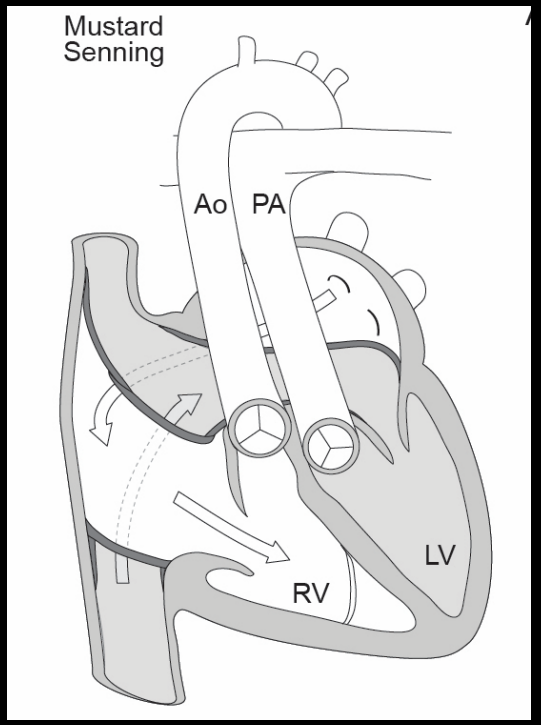


Arterial Switch

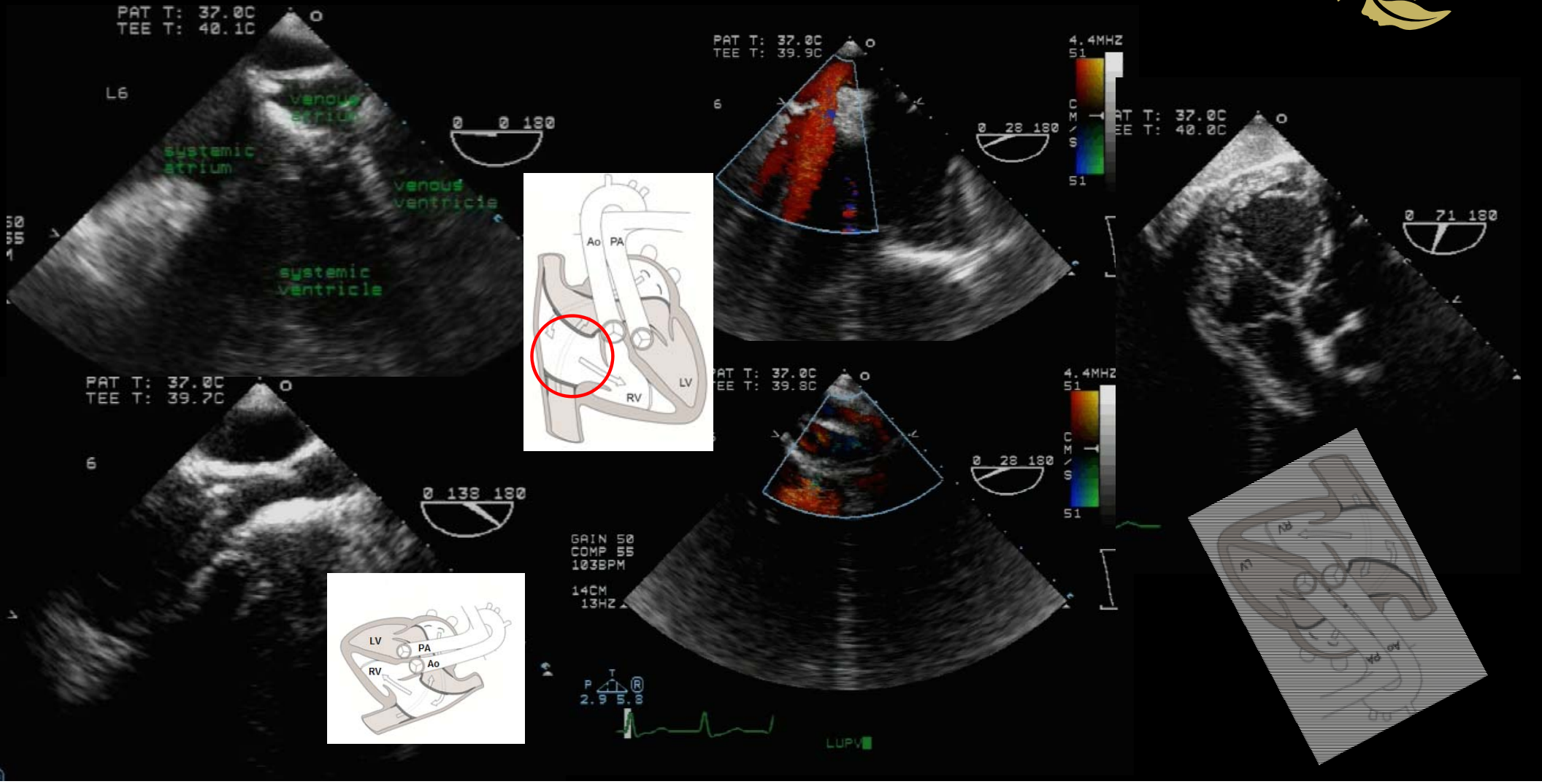
- PA + aorta switch
- Coronaries switch

TEE Checklist Mustard

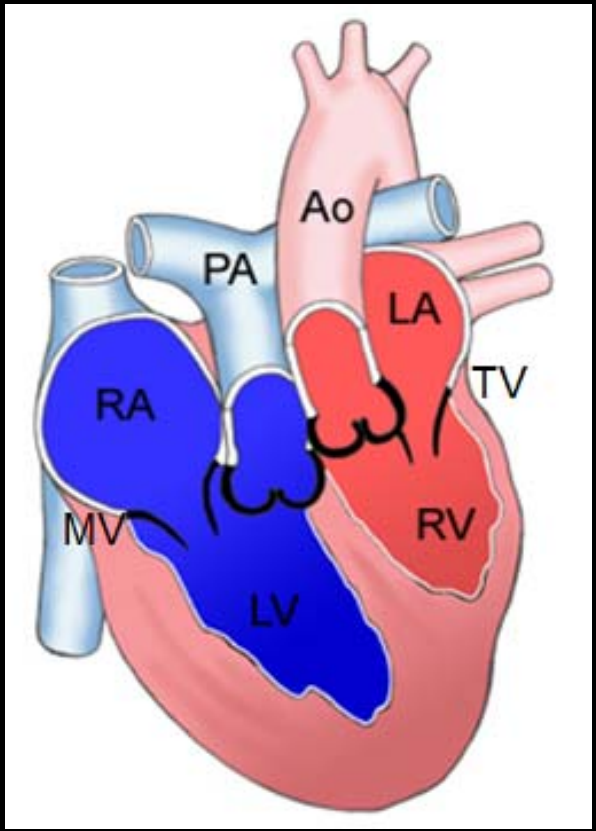
- Measure systemic (RV) size and function
- Assess systemic A-V valve (TR)
- Baffle obstruction by
  - Color Doppler
  - Spectral Doppler
  - Contrast
- PASP from MR
- Exclude LVOT obstruction



TEE Mustard


PAT T: 37.0C
TEE T: 40.1C
L6
venous atrium
systemic atrium
venous ventricle
systemic ventricle
Ao
PA
LV
RV
PAT T: 37.0C
TEE T: 39.9C
4.4MHZ
PAT T: 37.0C
TEE T: 40.0C
PAT T: 37.0C
TEE T: 39.7C
PAT T: 37.0C
TEE T: 39.8C
GAIN 50
COMP 55
103BPM
14CM
13HZ
LV
PA
Ao
RV
LUPV

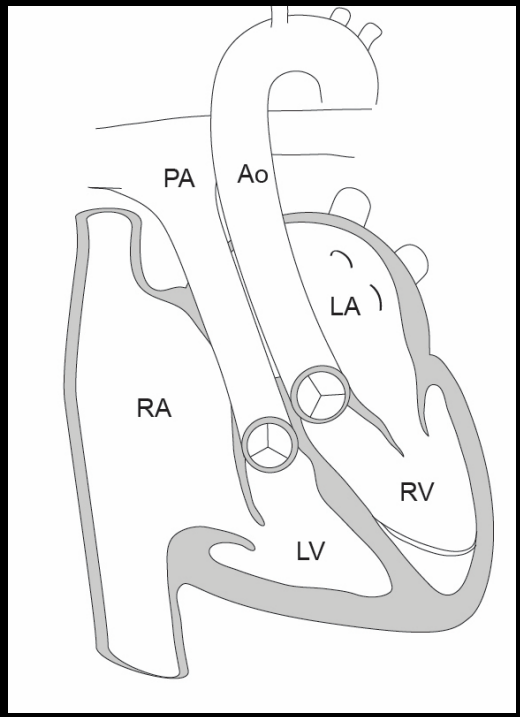
L or CC-TGA Anatomy



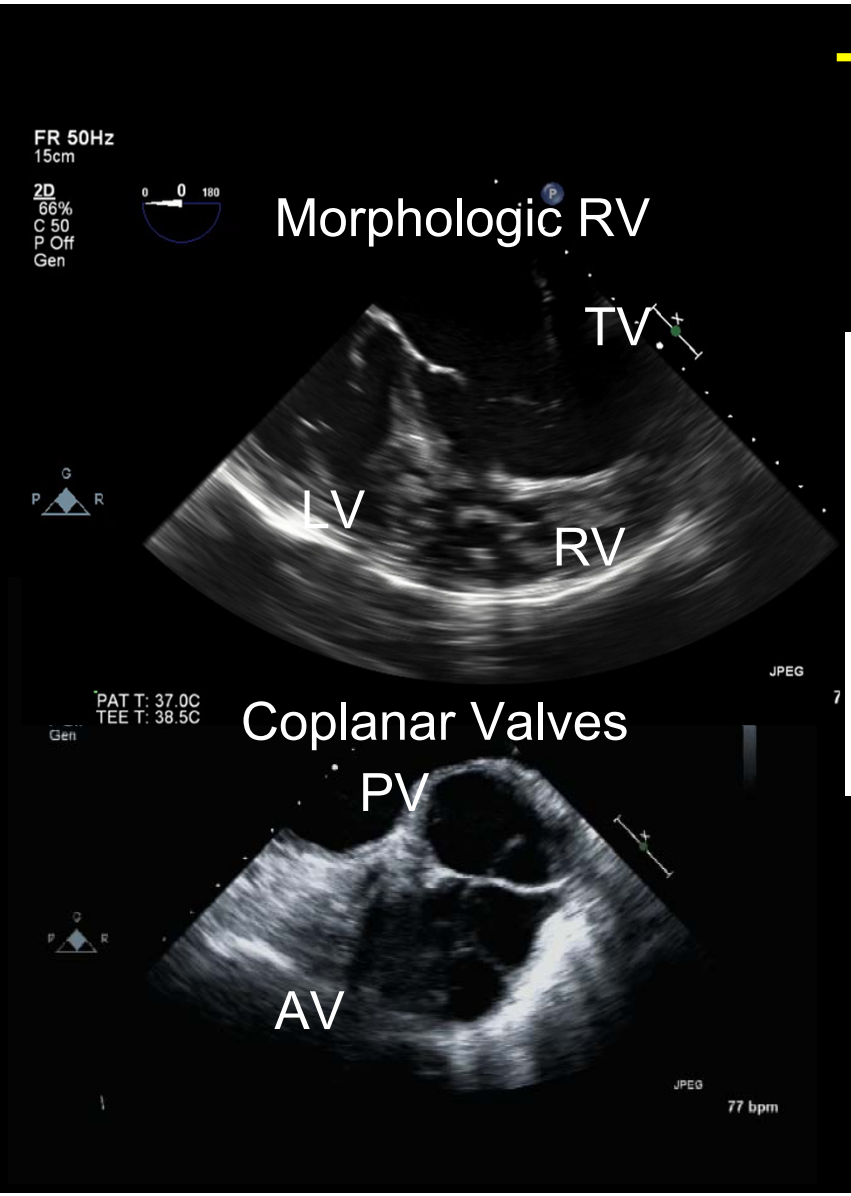
- Double discordance
  - AV: ventricles inverted
  - VA: great arteries transposed
- Systemic vent = morphologic RV
- Pulmonic vent = morphologic LV
- Parallel great vessels
- Associated anomalies
  - ASD
  - VSD
  - A-V valve abnormalities
  - Coronary artery anomalies
  - Arch anomalies

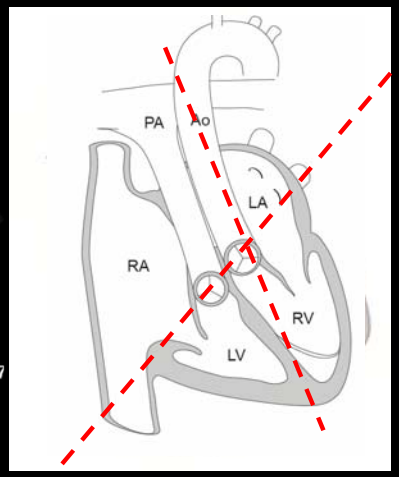
TEE Checklist L-TGA

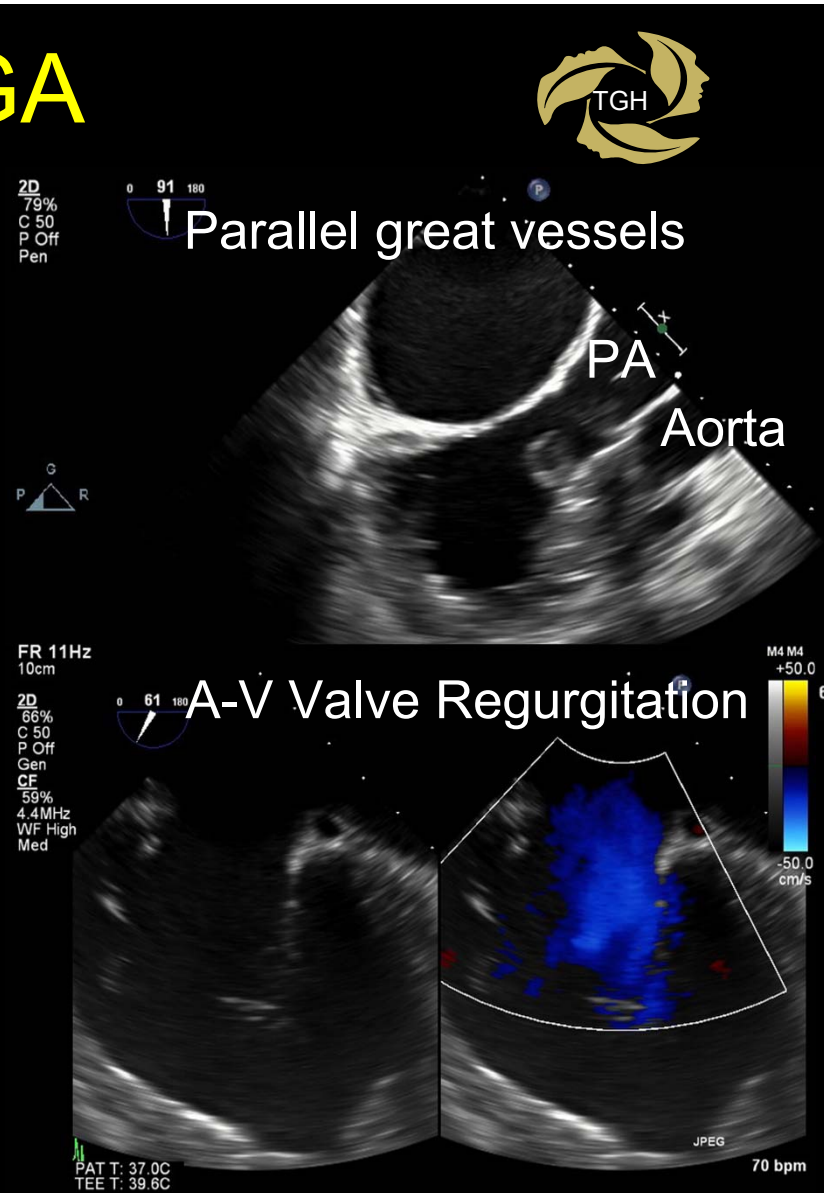
- Systemic ventricle (morphologic RV)
  - More apical positioned A-V valve
  - Chordae attached to septum
  - Moderator band
- Pulmonic ventricle (morphological LV)
  - Less apical position of A-V valve
  - No moderator band
- Coplanar great vessels
  - Anterior vessel is Aorta (and AV), coronaries
  - Posterior is PA (and PV), branching
- Associated anomalies
  - Pulmonic stenosis
  - VSD



TEE L-TGA


FR 50Hz
15cm
2D
66%
C 50
P Off
Gen
Morphologic RV
TV
LV
RV
PAT T: 37.0C
TEE T: 38.5C
Coplanar Valves
PV
AV
JPEG
77 bpm


PA
Ao
LA
RA
RV
LV

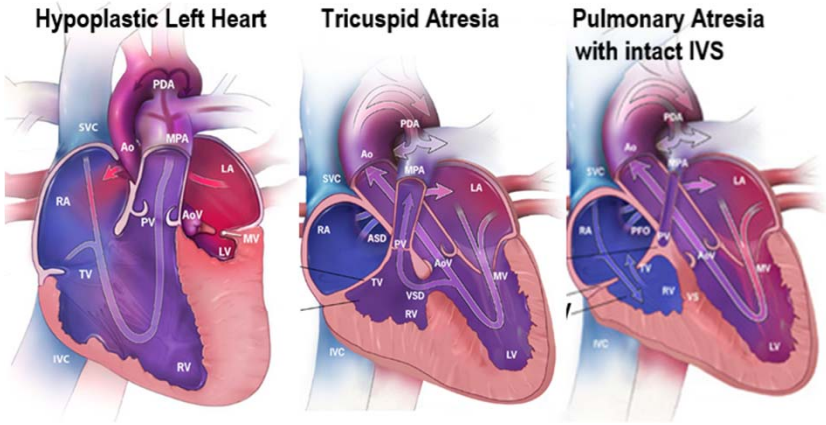

2D
79%
C 50
P Off
Pen
Parallel great vessels
PA
Aorta
FR 11Hz
10cm
2D
66%
C 50
P Off
Gen
CF
59%
4.4MHz
WF High
Med
A-V Valve Regurgitation
M4 M4
+50.0
-50.0
cm/s
JPEG
PAT T: 37.0C
TEE T: 39.6C
70 bpm

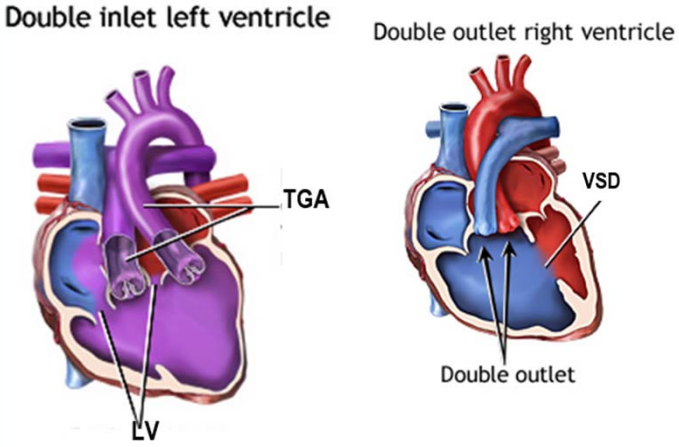
Definition Single Ventricle

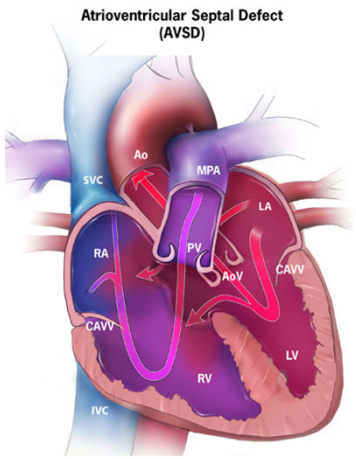
- Abnormal heart structure at birth
  - Anatomic: 1 functional ventricle
  - Physiologic: no sequential circulations (pulmonary/systemic)
- Several different CHD lesions
  - All have cyanosis
  - All require pulmonary blood flow
- Rare, 5/10,000 live births
  - Palliation procedures
  - Untreated survivors DILV/PS

Single Ventricle Anatomy

❶ Absent A-V Connection	❷ Double Inlet	❸ Common A-V Valve
Hypoplastic Left Heart Syndrome Tricuspid Atresia Pulmonary atresia intact IVS	DILV DIRV/DORV Indeterminant	Unbalanced A-V canal







❹ Heterotaxia

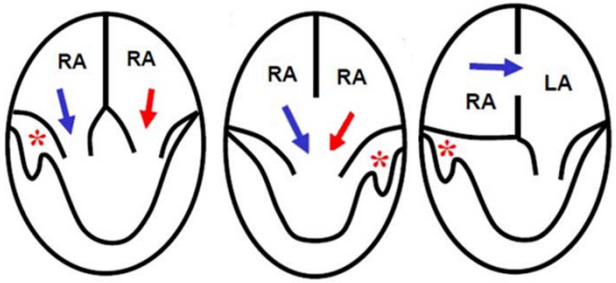
STS Congenital Heart Surgery Database

Common Associations

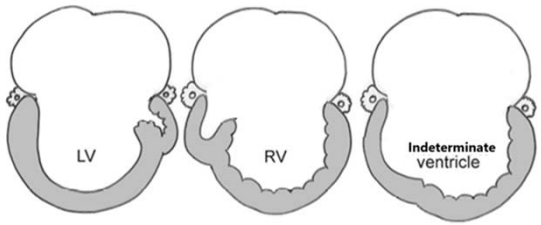
- VSD, PDA, ASD/PFO
- TGA
- Subpulmonary stenosis
- Persistent LSVC
- Interrupted IVC
- PAPVC
- CoA
- Right aortic arch

Anatomic Approach

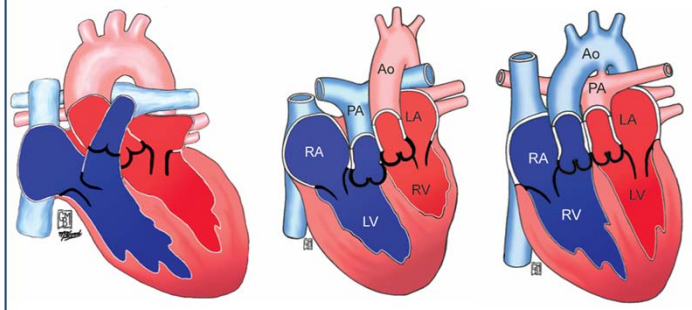
① Atrio-ventricular Connections	② Ventricular Morphology	③ Ventriculo-arterial Connections
• Double • Common • Absent/Single	• Left • Right • Indeterminant	• Concordant LV→ aorta • Discordant RV→ aorta



Double Inlet | Common Inlet | Single Inlet



Smooth Apex | Trabecula Apex | No IVS



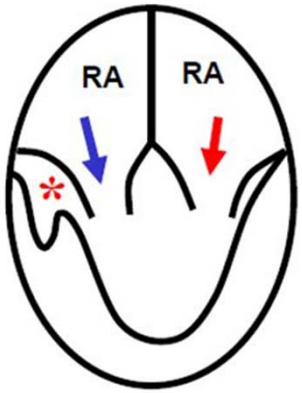
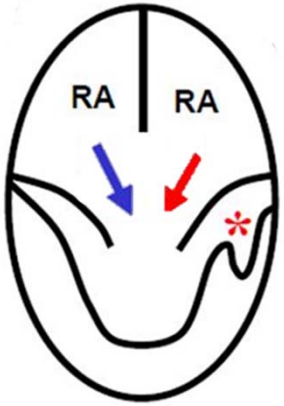
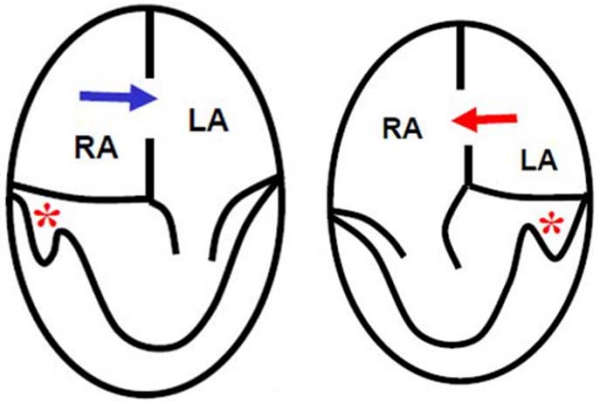
Concordant Normal | Discordant CC-TGA | Discordant D-TGA

Parallel Great Vessels

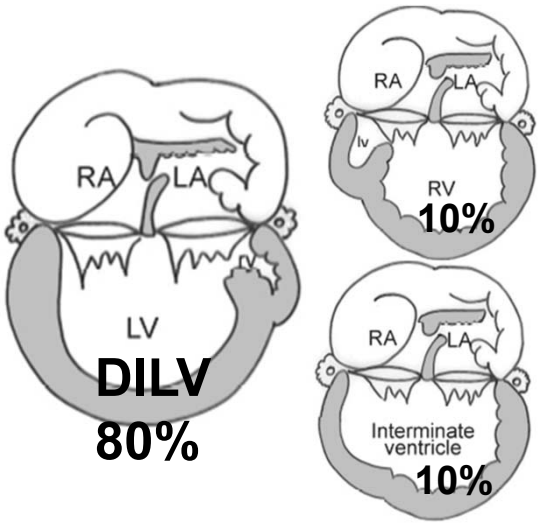
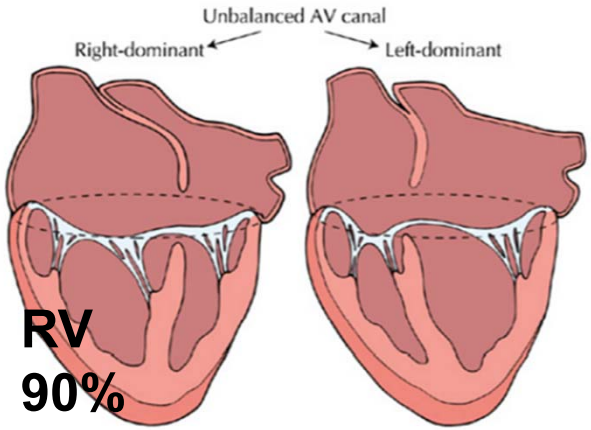
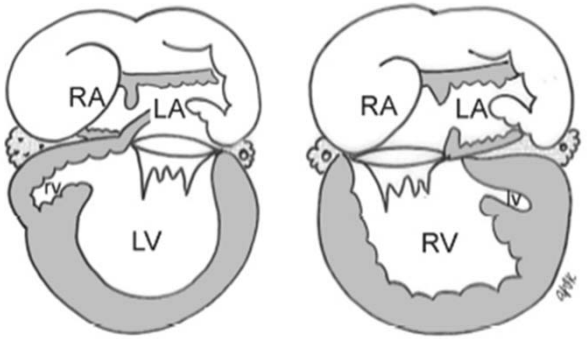
Atrioventricular Connections

Double	Common	Absent

• Both atria connect by two AV valves
• Mirror image MV
• Morphologic LV
	• Both atria connect by single A-V valve
	• Morphologic RV
		• Both atria connect by single A-V valve
		  • Mitral atresia
		  • Tricuspid atresia
		• Morphologic RV or LV

 **Double Inlet**	 **Common Inlet**	 **Single Inlet**

Atrioventricular Connections

Double	Common	Absent
• Morphologic LV (80%) • DILV • Mirror image MV	• Form of AVSD • Complete endocardial cushion • Unbalanced if common A-V valve predominantly over 1 Vent • Hypoplastic ventricle (90% RV)	• Atresia of R of L A-V valve • Either vent is hypoplastic • V-A connections concordant o discordant
 **DILV 80%**	 **RV 90%**	 **Tricuspid Atresia Hypoplastic RV** **Mitral Atresia Hypoplastic LV**

Single Ventricle Adult TEE

Ventricular Inflow

- ①Atria (2 or common)
- ②Atrioventricular valve (A-V)
  - Single, double, common
  - Function: regurgitation

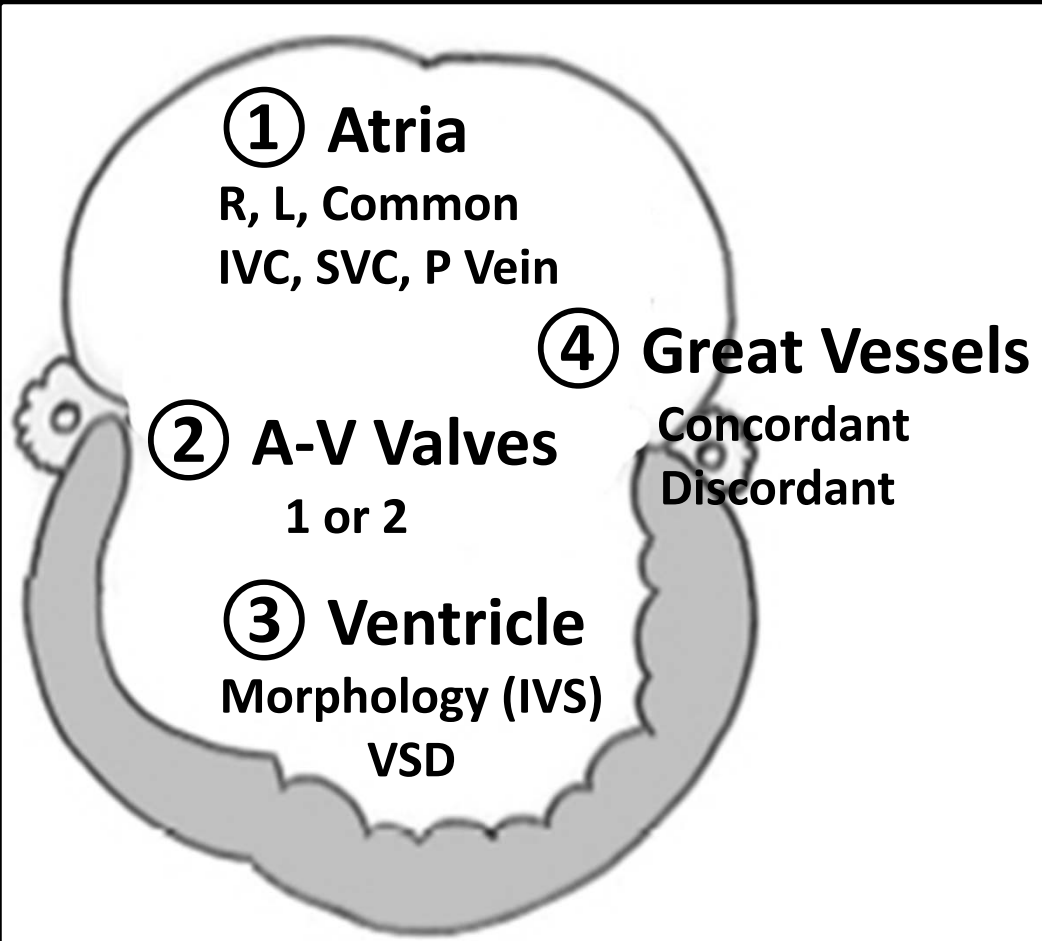
③Ventricle (ME, TG views)

- Size, function
- Right, left or indeterminate

④Outflow vessels

- Relation ventricle (normal, TGA)
- Function semilunar valves
- Size vessels

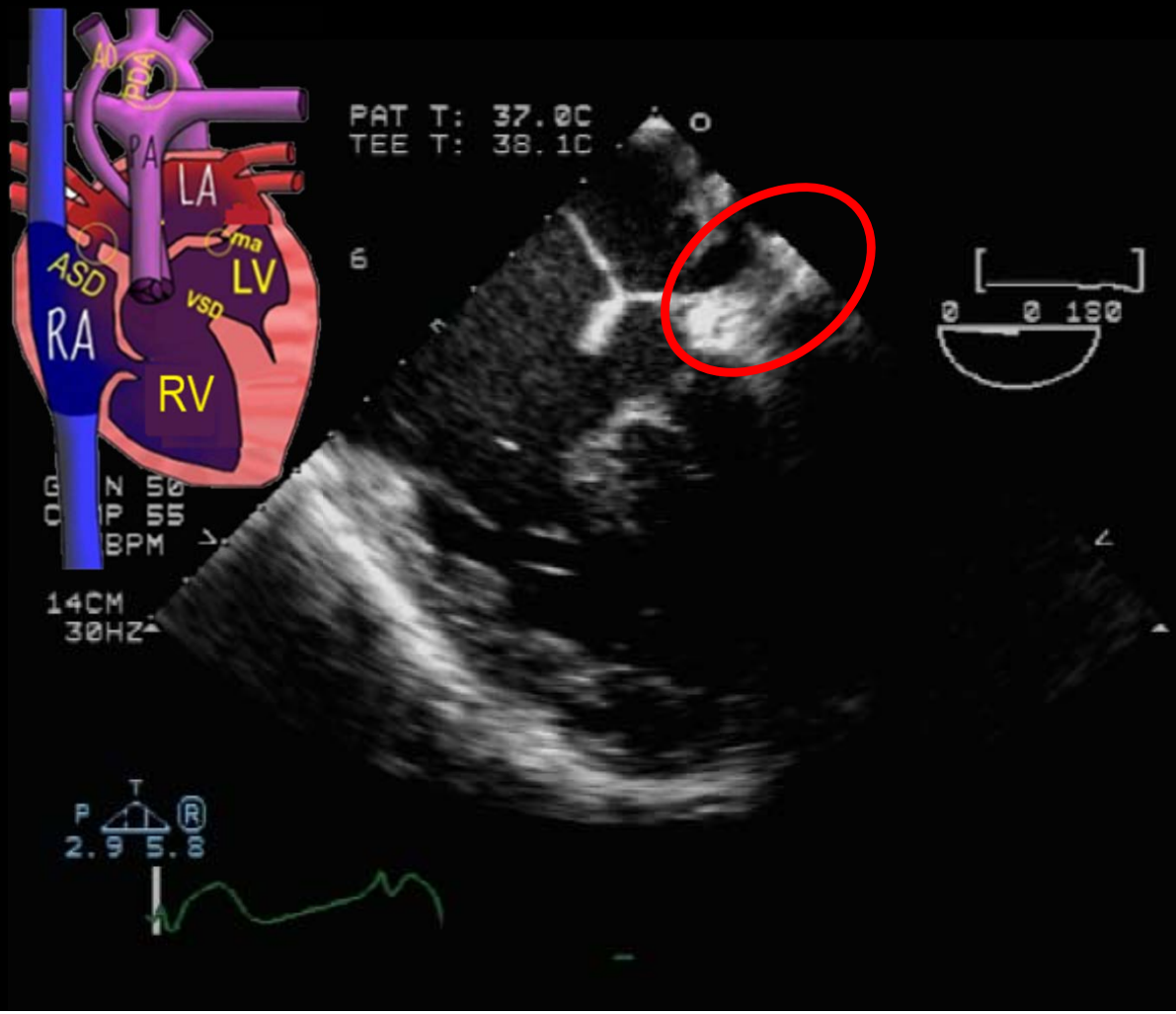
- Palliative procedures
  - Shunt: ASD, Glenn, Blalock-Taussig
  - Fontan



Hypoplastic Left Heart Syndrome

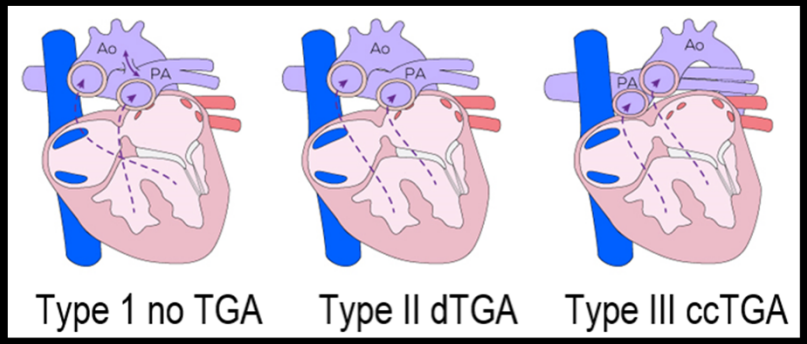
- Underdeveloped L heart
  - LV small non-functional
  - Atresia aortic / mitral orifice
  - Hypoplasia ascending aorta

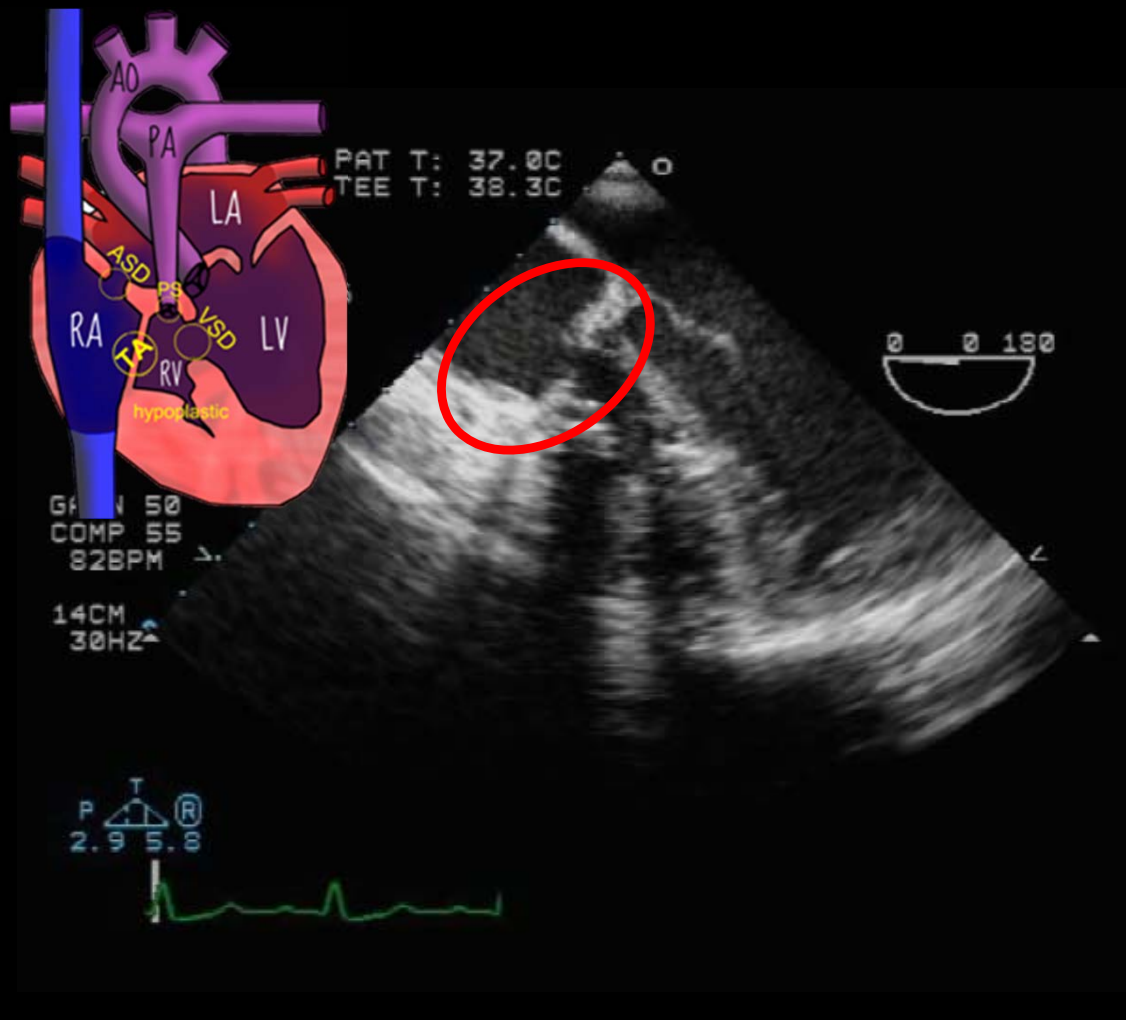
- Pulmonary venous return: LA → ASD → RA
- Systemic + pulmonary venous mix RA, RV → large MPA
- Systemic flow by PDA



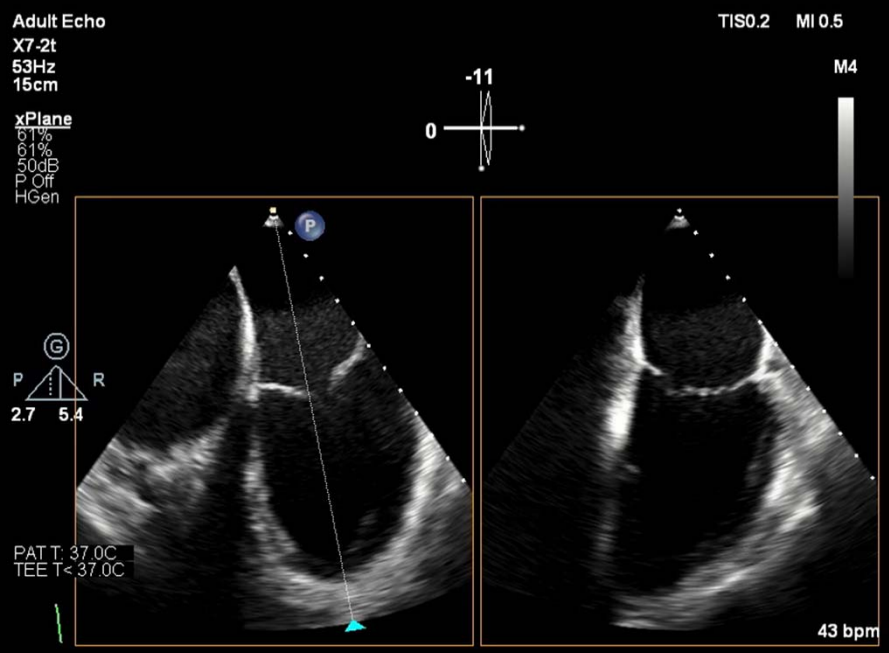
Tricuspid Atresia Anatomy

- Atresia tricuspid orifice
- Underdevelop R heart
  - RV small non-functional
  - Hypoplasia PA, or PS
- All ASD → venous blood mixes
- 3 types based on outflow
- Subtype (8): obstruction to PBF
  - A. Pulmonary atresia
  - B. Pulmonary stenosis/hypoplasia
  - C. No obstruction to PBF
  - – Commonest: 1B no TGA+ PS + VSD

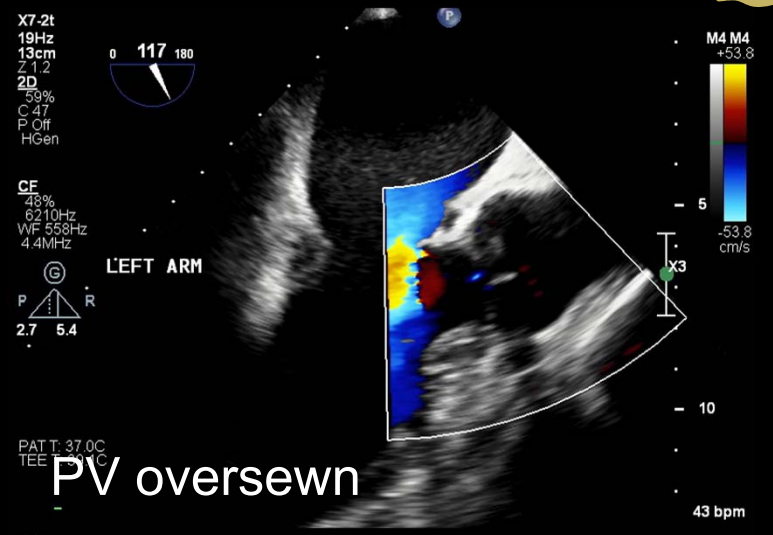




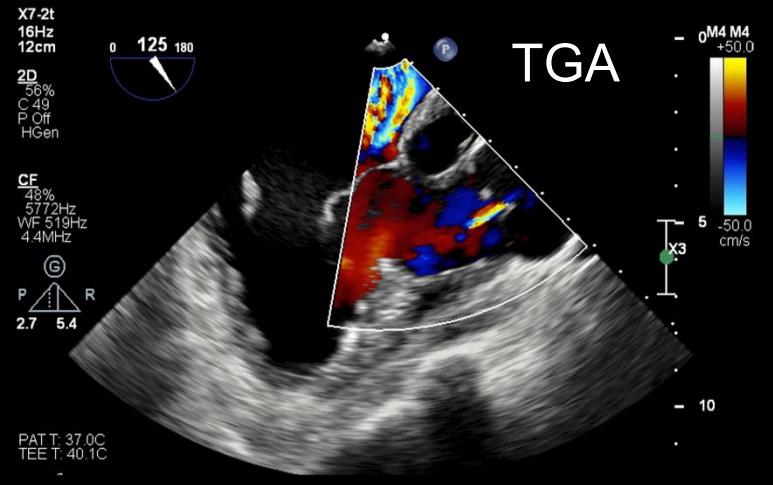
Tricuspid Atresia + TGA TEE



Single Ventricle
LV morphology
Double outflow
Parallel (TGA)

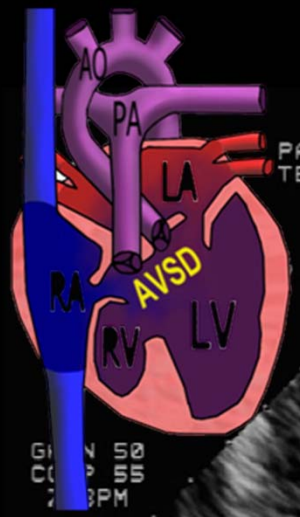


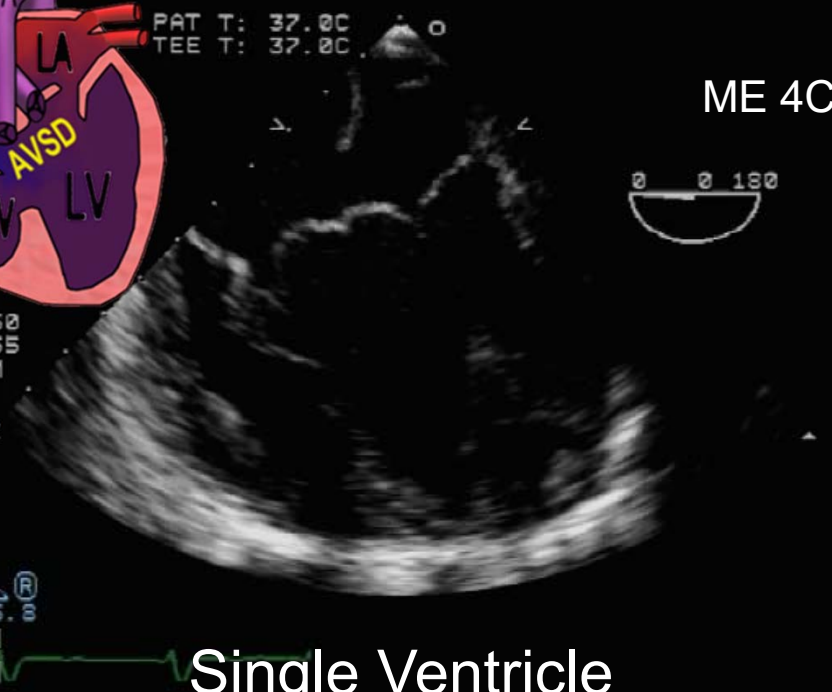
PV oversewn

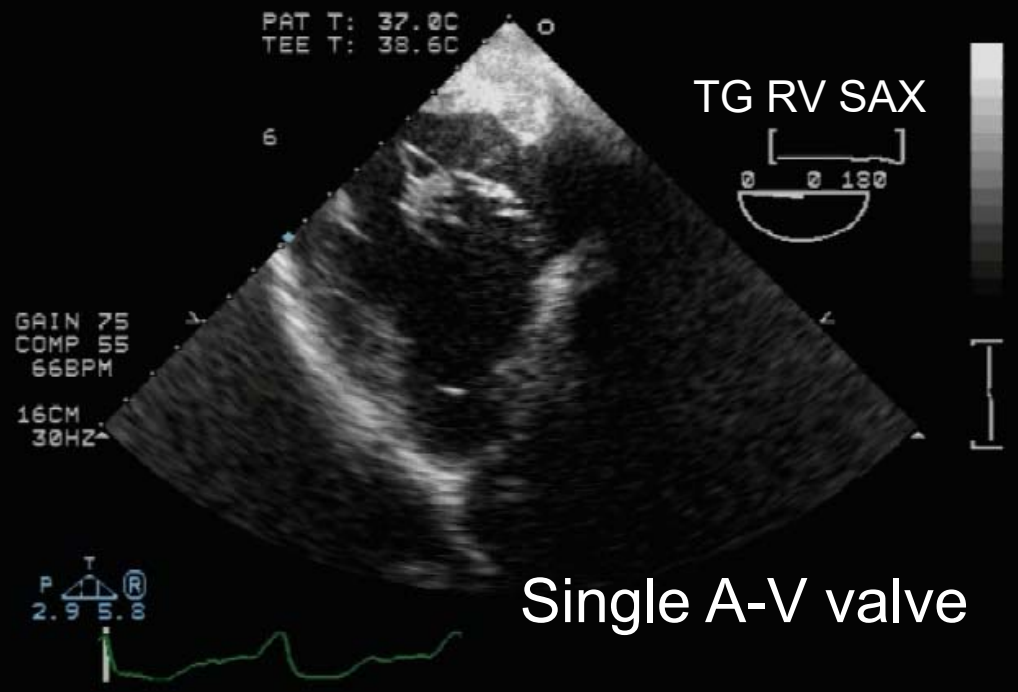


TGA

Unbalanced AVSD





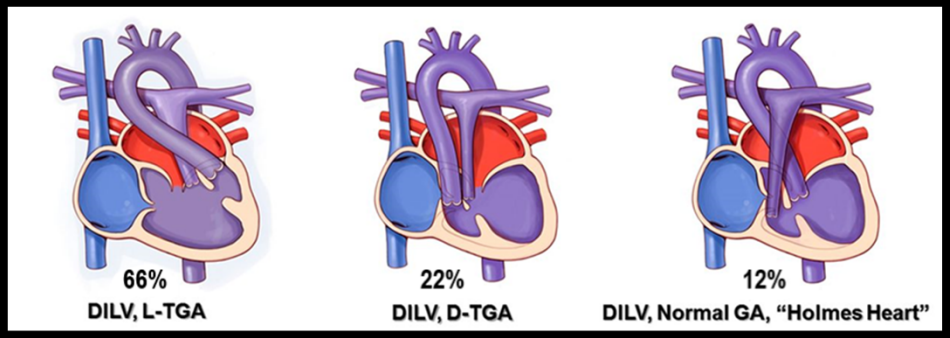


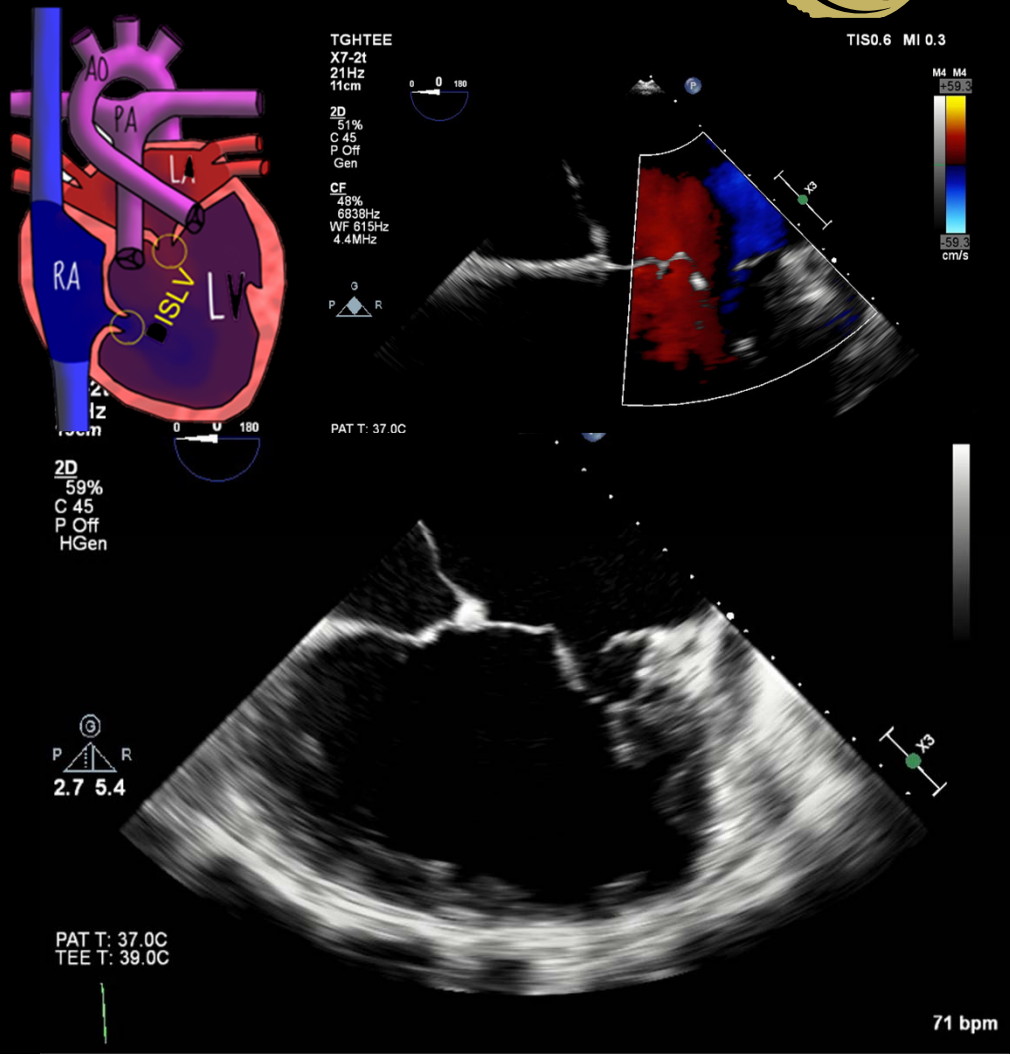
Single Ventricle
RV morphology
Single A-V inflow
ASD, VSD

Single A-V valve

Double Inlet Ventricle

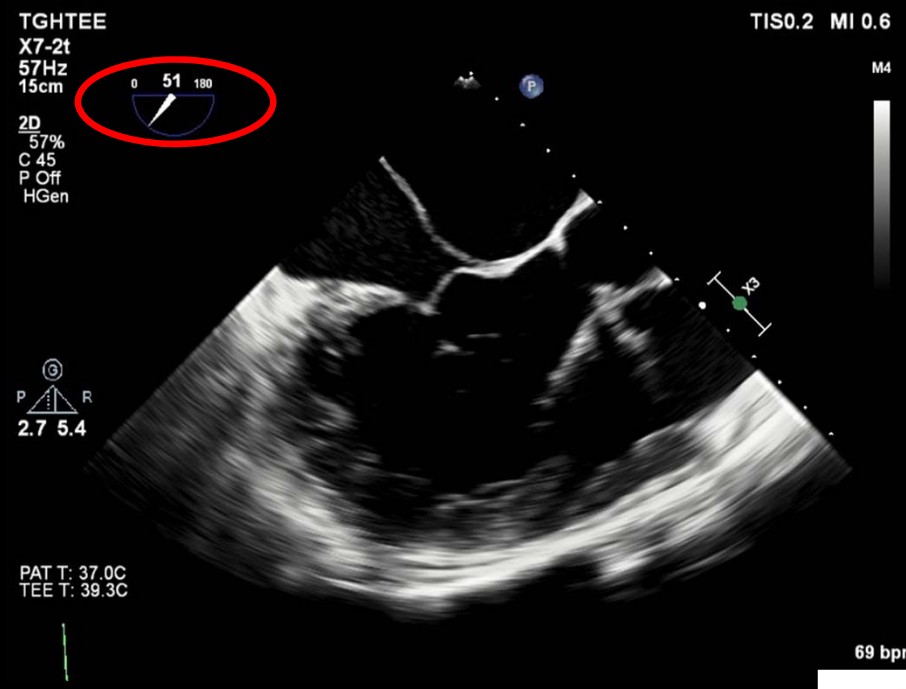
- Commonest single ventricle
  - DILV 80%
  - DIRV + indeterminate 5-10%
- 2 A-V valves (mirror image)
- Outflow based on great arteries
  - Normal
  - CC-TGA
  - D-TGA

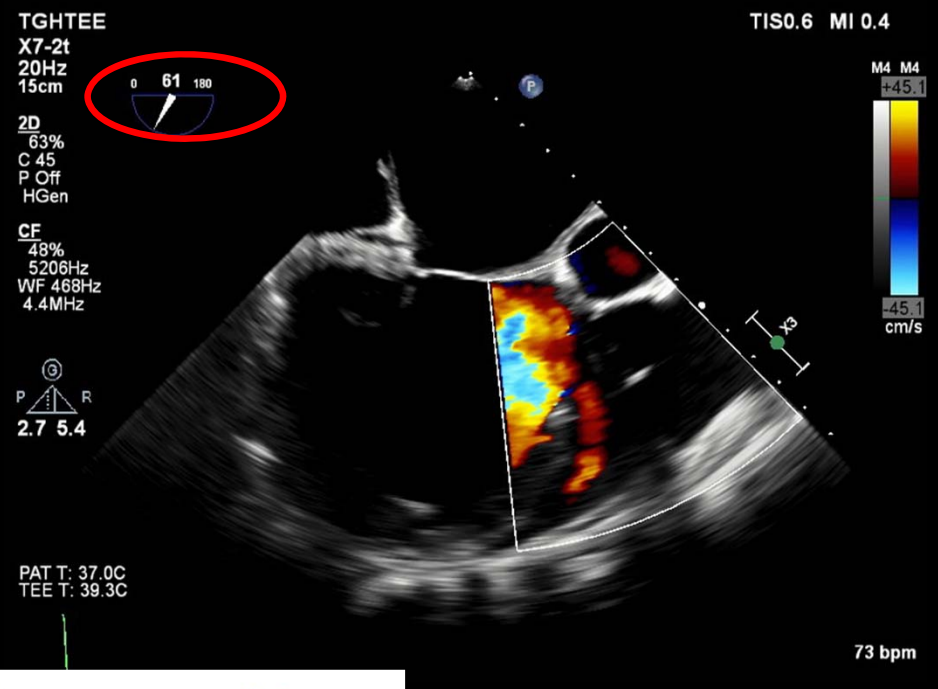




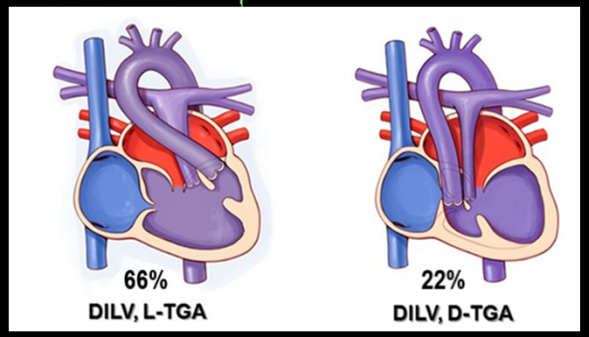
Potcrucha, JT et al. Heart 2016

DILV + TGA



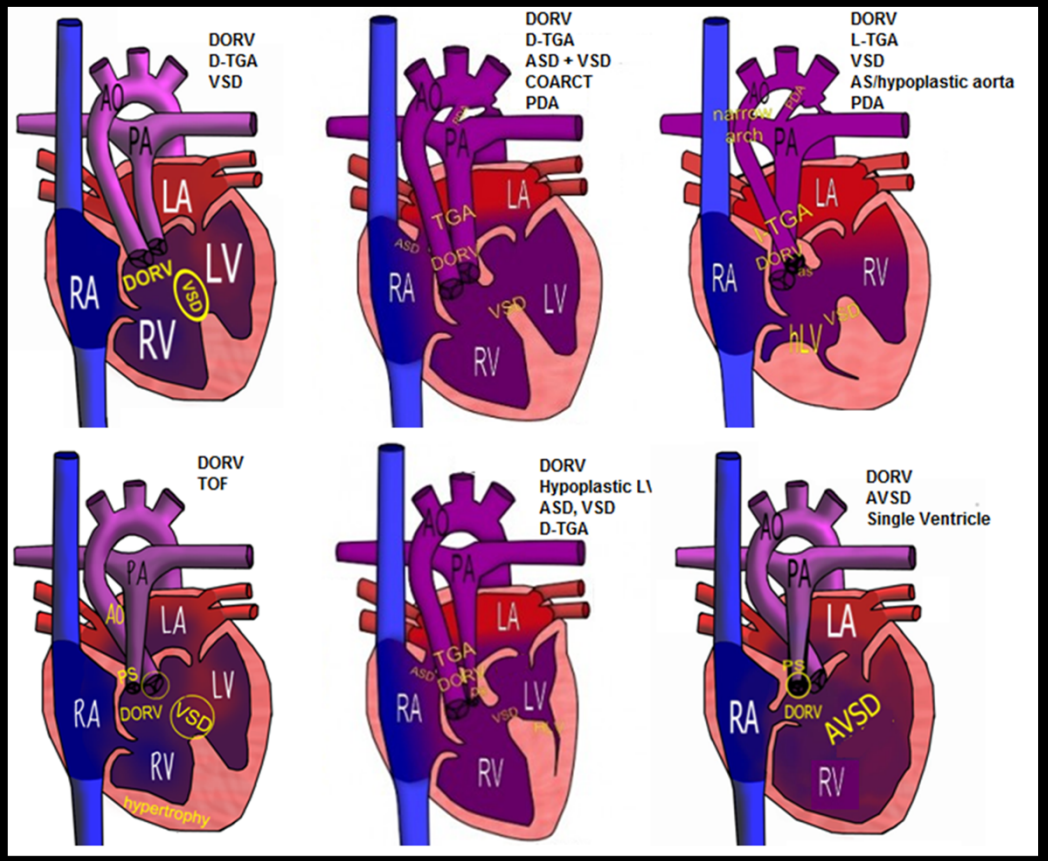


DILV
Hypoplastic RV
TGA
VSD

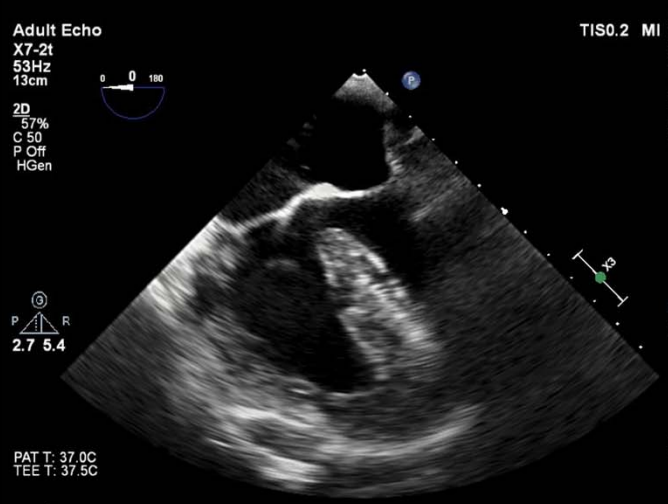


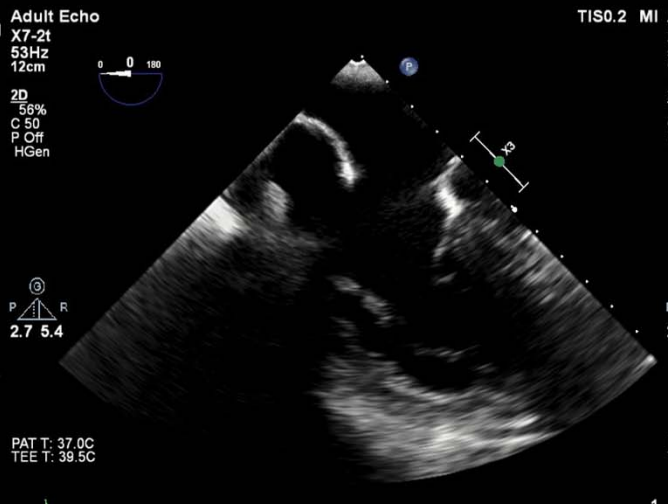
Double Outlet RV Anatomy

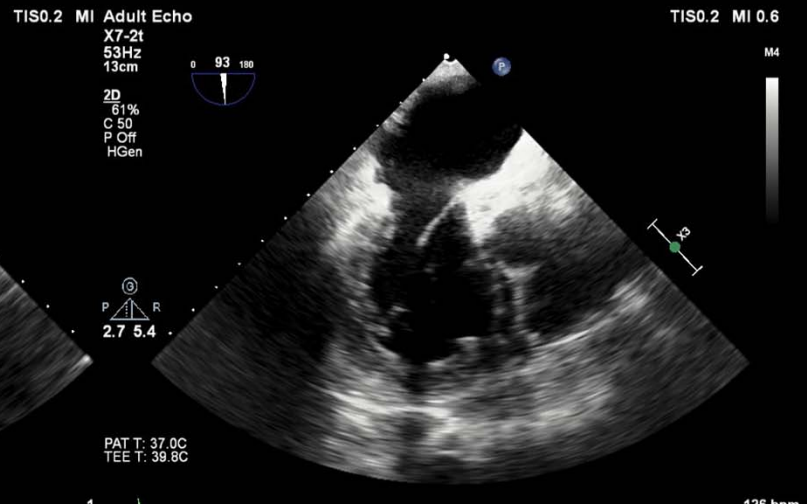
- VA connection both great arteries align with RV
- Broad spectrum: similar features to TOF, TGA
- VSD present:
  - Subaortic
  - Subpulmonary (Taussig-Bing)
  - Doubly committed
  - Non-committed (remote)
- Great Vessels
  - Side by side
- Physiology
  - VSD, TOF, D-TGA, single ventricle

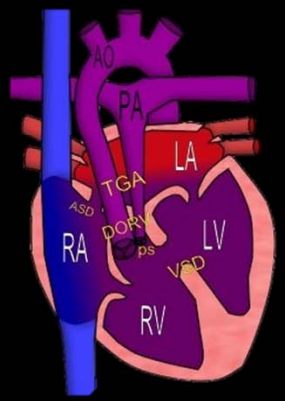


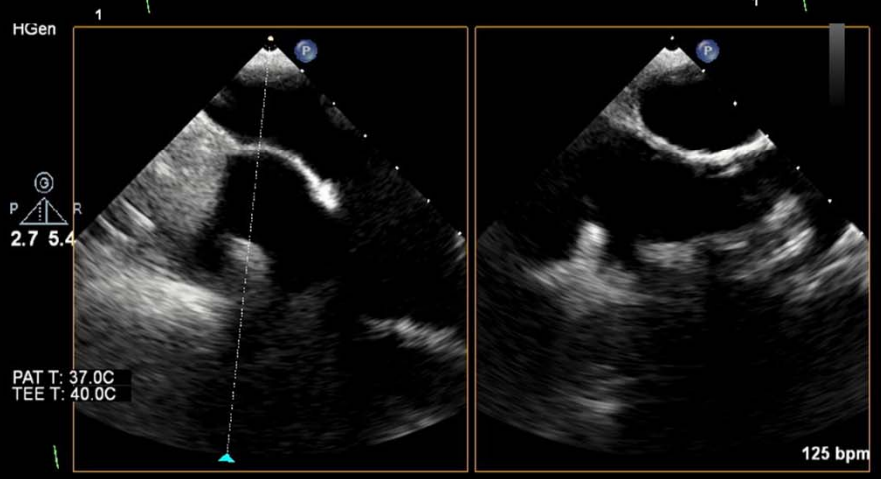
Double Outlet Ventricle











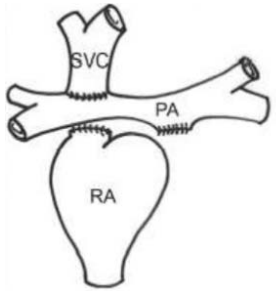
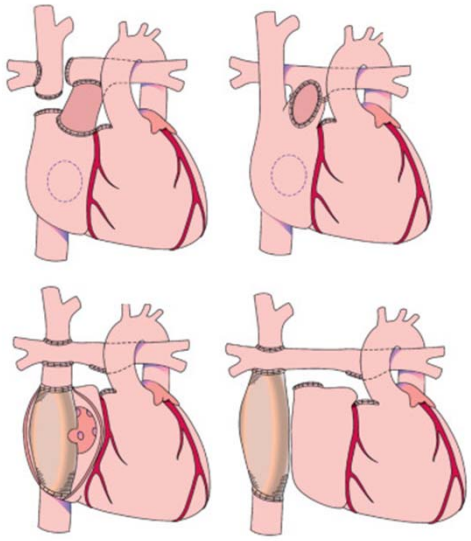
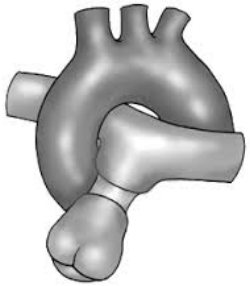
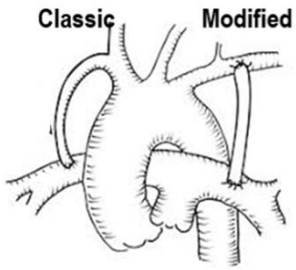
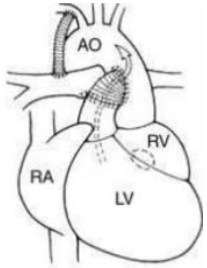
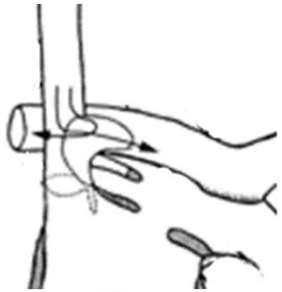
DORV
TGA
PS (75%)
ASD, VSD
Lateral Fontan

Goals of Palliative Surgery

Avoid Failing Fontan

- Unobstructed systemic blood flow (Qs)
  - maximize O_2 delivery + minimize ventricular hypertrophy
- Limit pulmonary blood flow (Qp)
  - minimize volume overload + pulmonary hypertension
- Unobstructed pulmonary venous return
  - minimize secondary pulmonary hypertension
- Minimize pulmonary artery distortion
- Avoid dysrhythmias

Surgical Options Single Ventricle

<table>
<tr><th colspan="2">Stage 1 (weeks)</th><th>Stage 2 (2-6 months)</th><th>Stage 3 (1-2 years)</th></tr>
<tr><td colspan="2">Adequate Qp, Qs</td><td>Limit PBF (PVR)</td><td>Separate Qp. Qs</td></tr>
<tr><td>Unobstruct VBF</td><td>Obstruct VBF</td><td rowspan="2">Bidirectional Glenn (BDG)
SVC → RPA, end to side
Remove BT shunt
  </td><td rowspan="2">Fontan
  </td></tr>
<tr><td>Ø PS → PA Band
  </td><td>Ø PS → DKS
  </td></tr>
<tr><td>+ PS → BT shunt
  </td><td>+ PS → Norwood
  </td><td>Hemi-fontan
Maintain SVC + RA continuity
SVC → RPA
Patch RA
  </td><td></td></tr>
</table>

Norwood Procedure

Relieve systemic obstruction

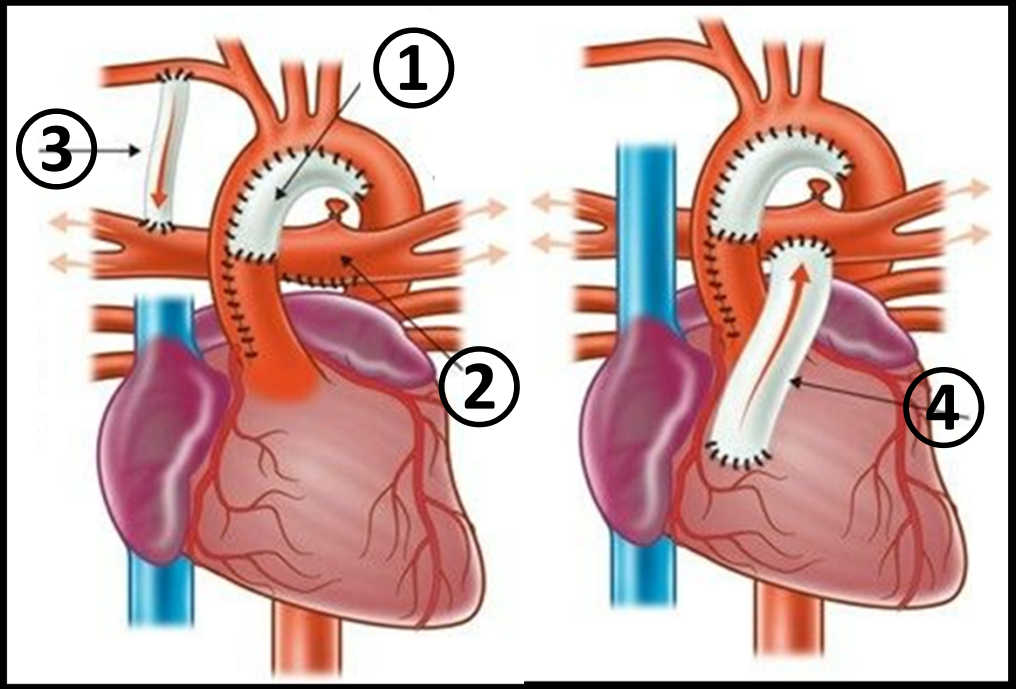
① Reconstruct aorta from PA trunk + patch

② Close PA + PDA

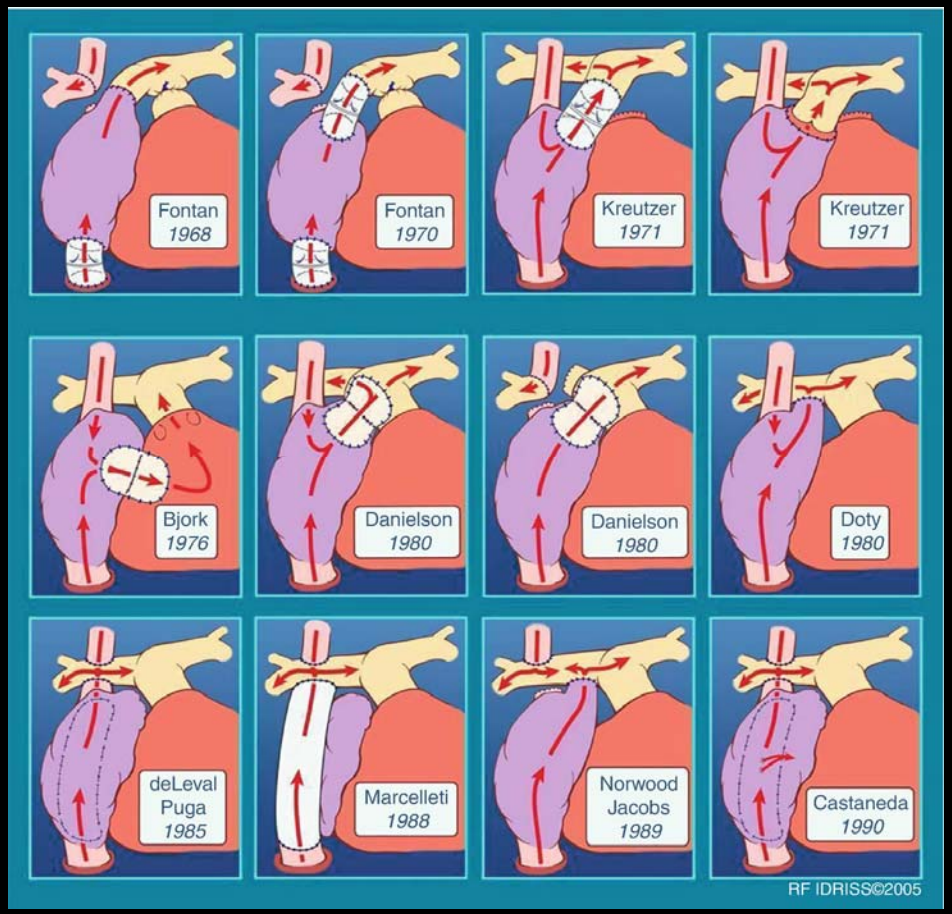
③ BT shunt (SCA → RPA)

④ Sano shunt (RV → MPA)

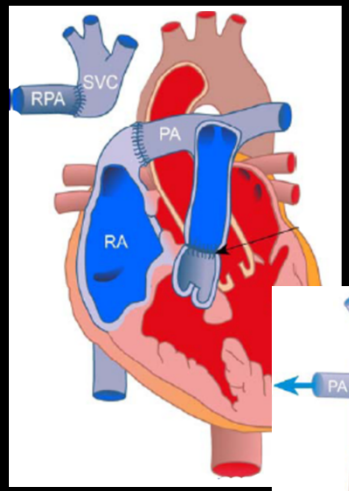
-improve coronary flow



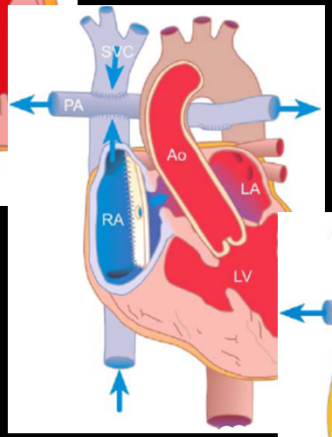
Fontan Variants



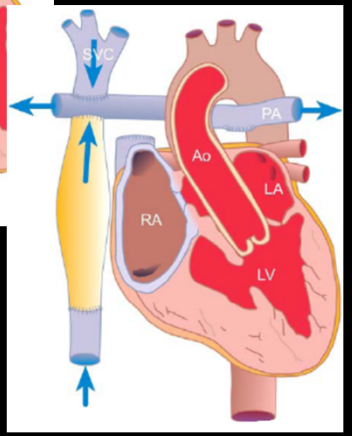
Atrial-Pulmonary < 1990



Lateral Tunnel 1990-2000

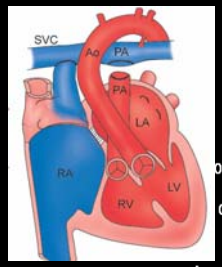


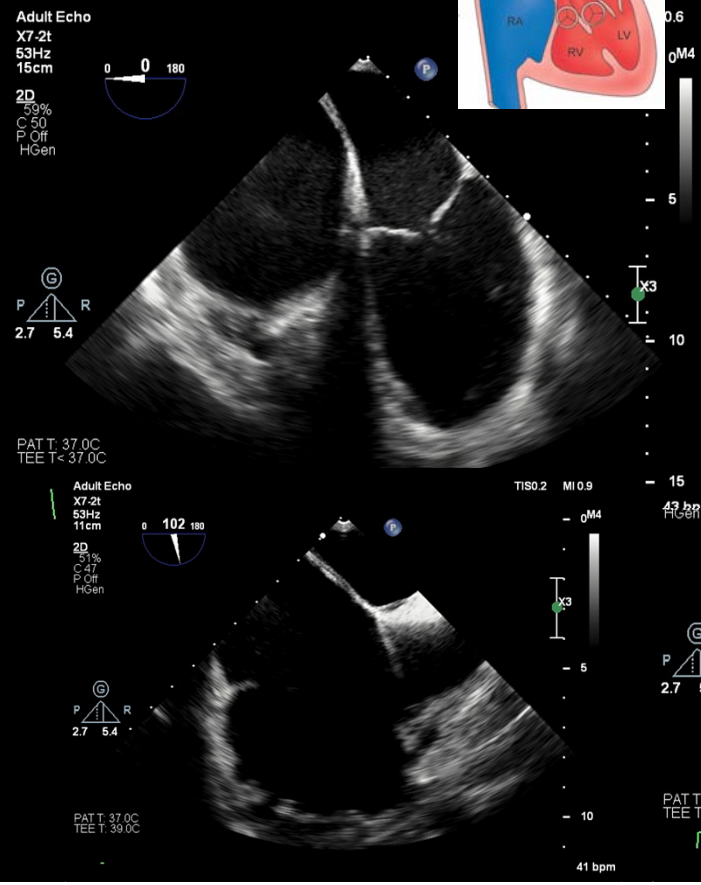
Extracardiac > 2000



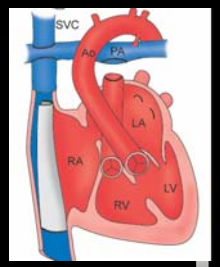
Fontan Variants TEE

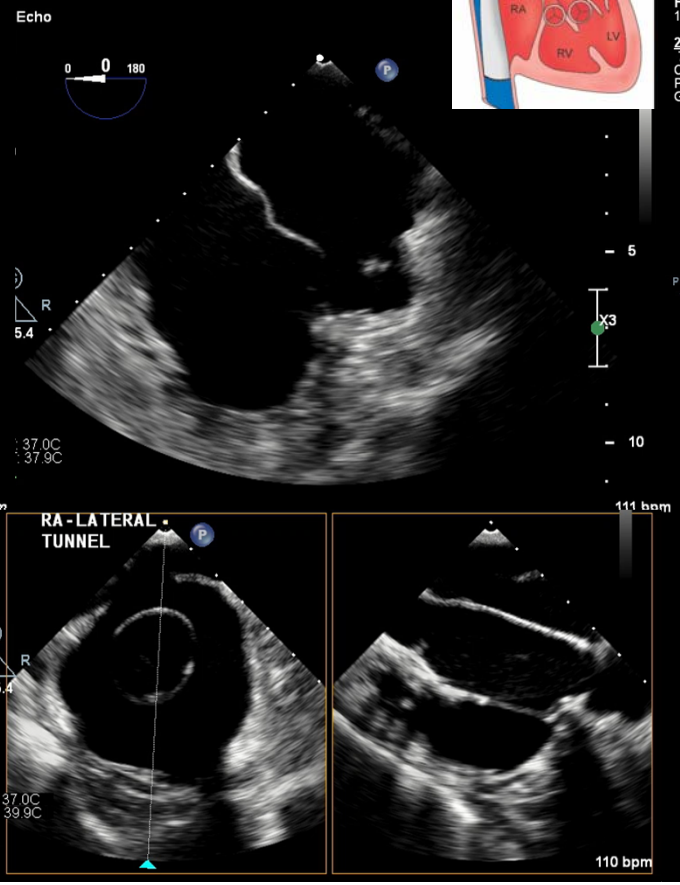
AP Fontan



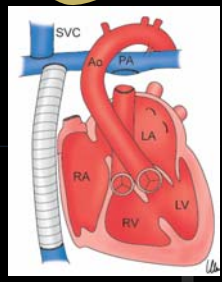


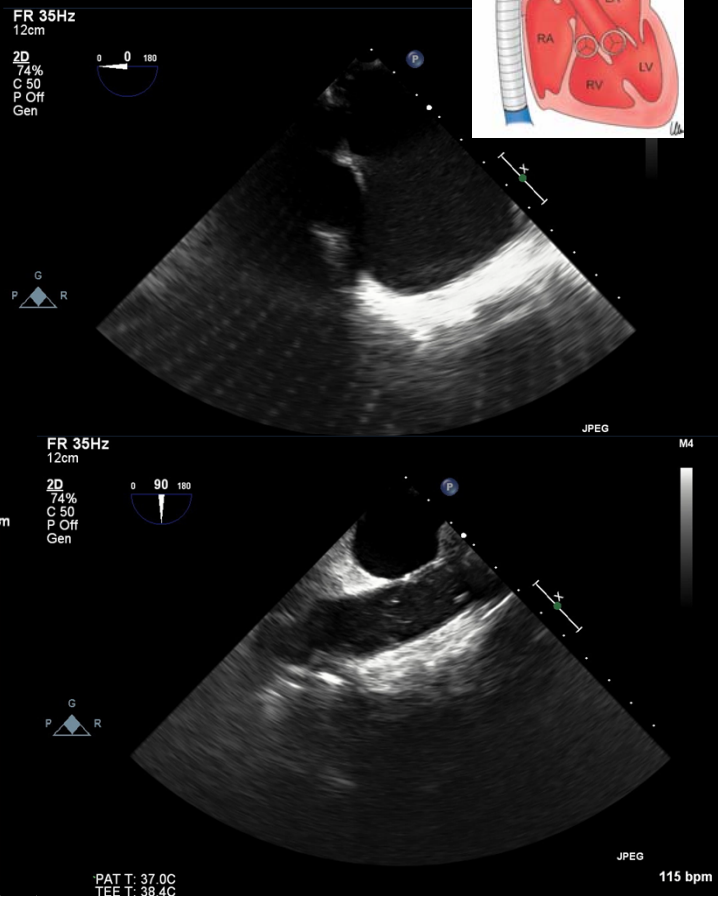
Lateral Tunnel





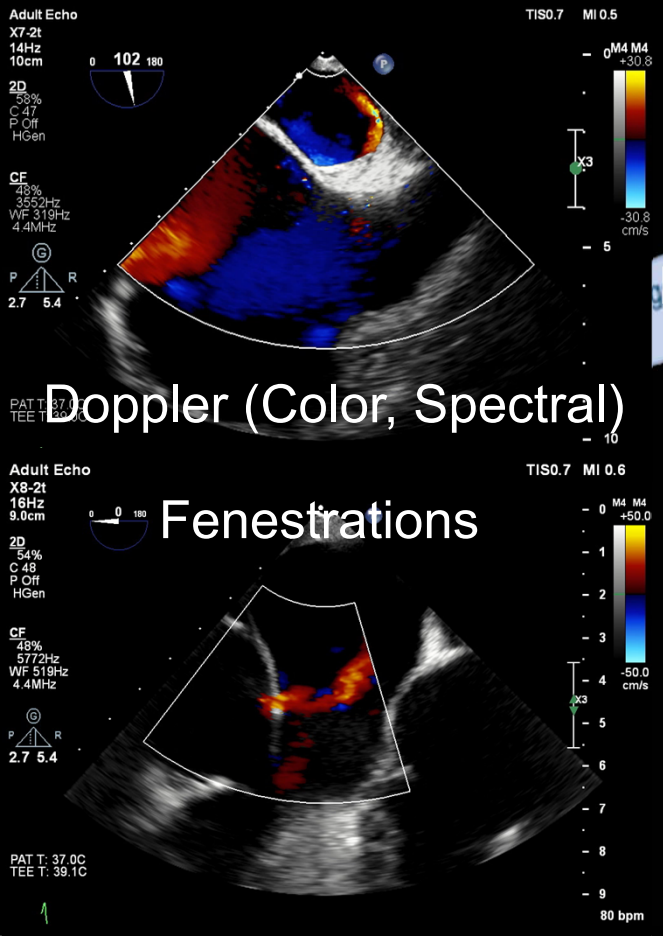
Extracardiac



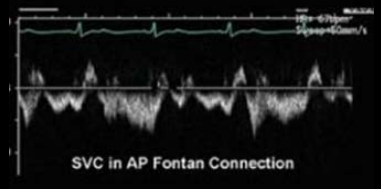


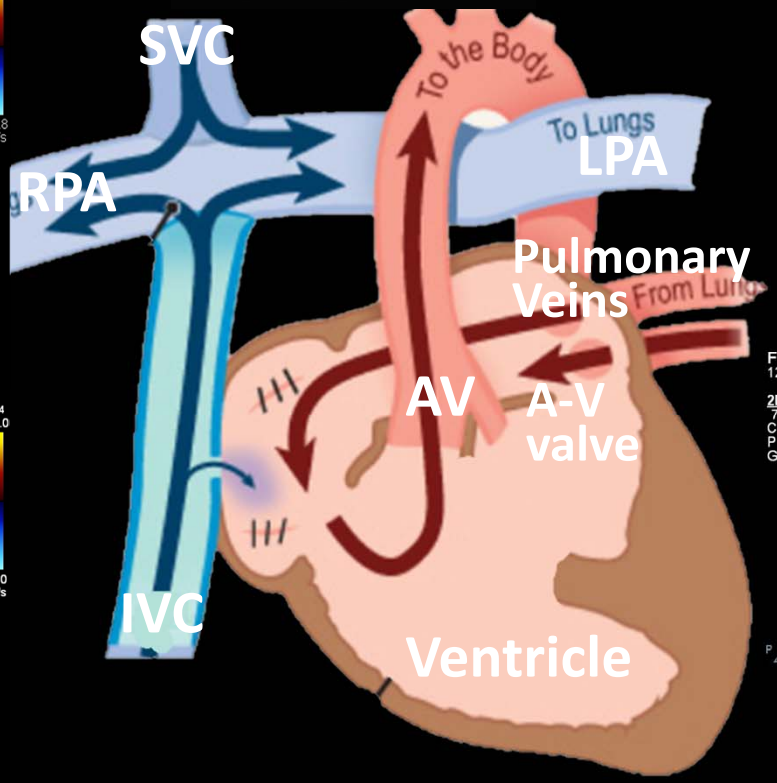
Fontan Assessment

Circuit Patency

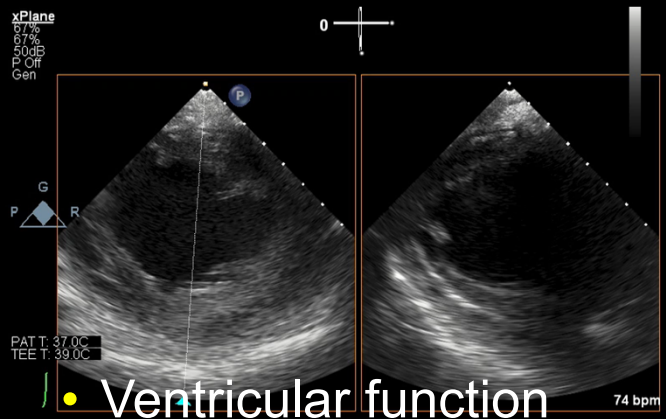


- Doppler (Color, Spectral)
- Fenestrations





Systemic Circuit



- Ventricular function
- A-V valve assessment
- Aorta outflow

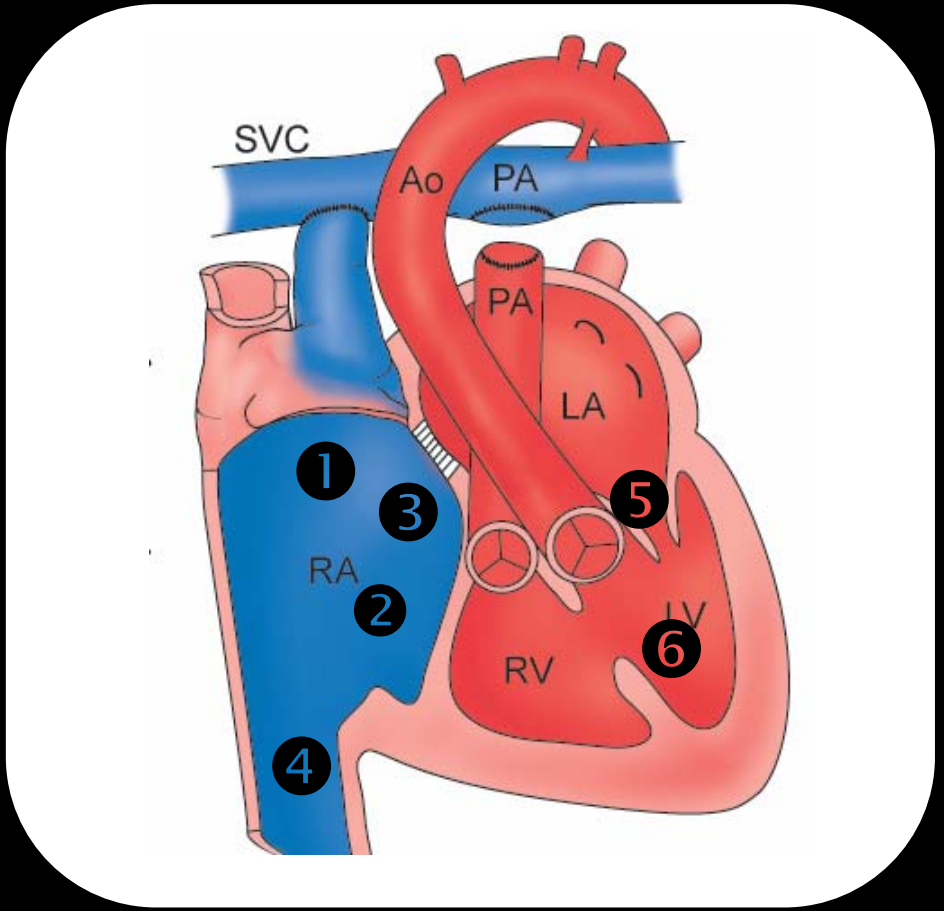
Fontan Dysfunction

Cardiac

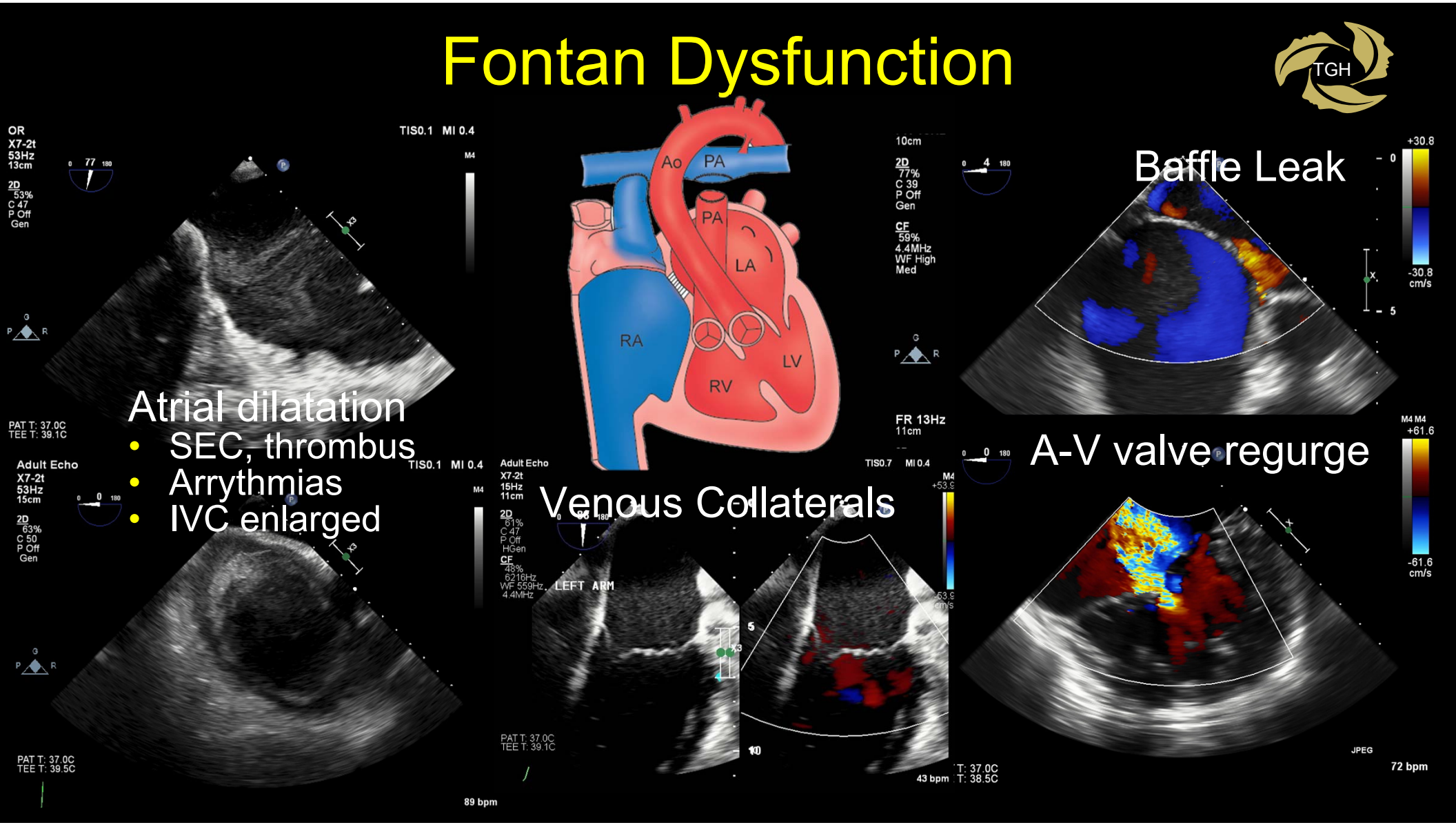
- ① Atrial dilatation
  - ② SEC, thrombus
  - ③ Arrythmias
  - ④ IVC enlarged
- ⑤ A-V valve dysfunction
- ⑥ Ventricle
  - Systolic/Diastolic dysfunction
- Collaterals
- Baffle Leak
- Obstruction

Non Cardiac

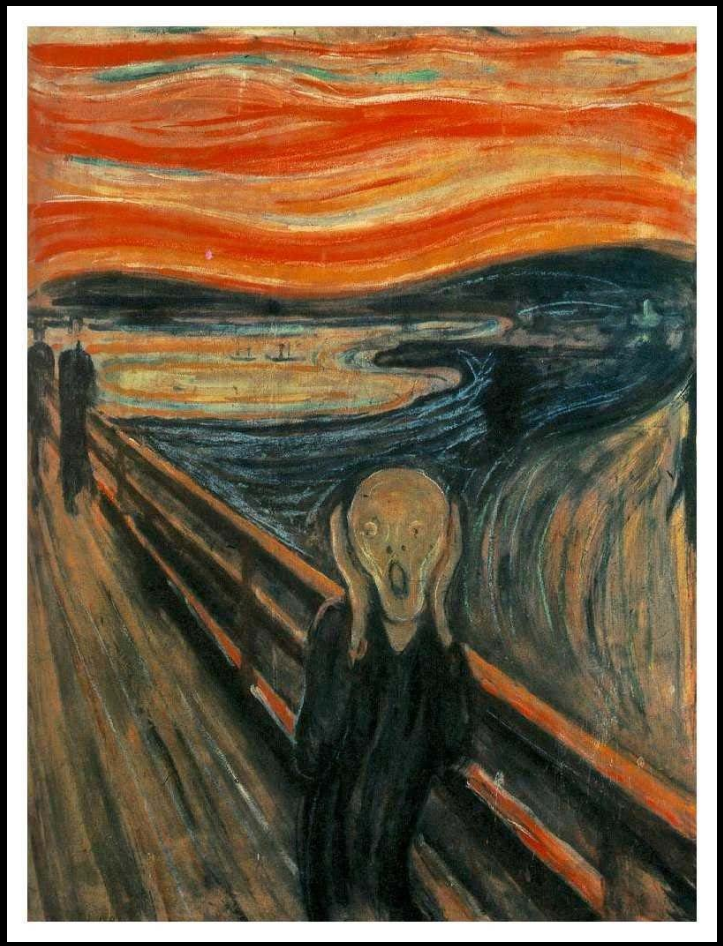
- Ascites or pleural effusion
- Liver abnormalities



Fontan Dysfunction



Summary



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Sorry