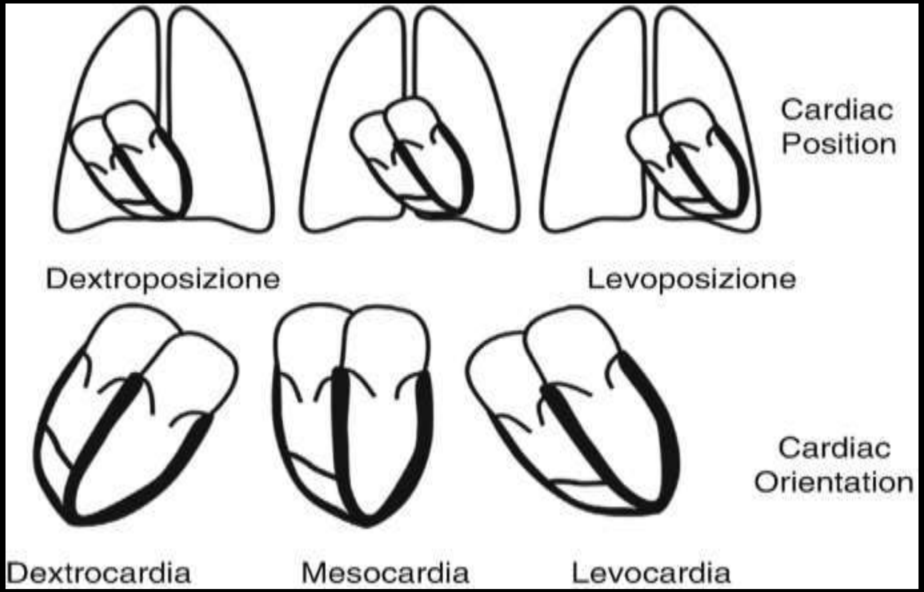
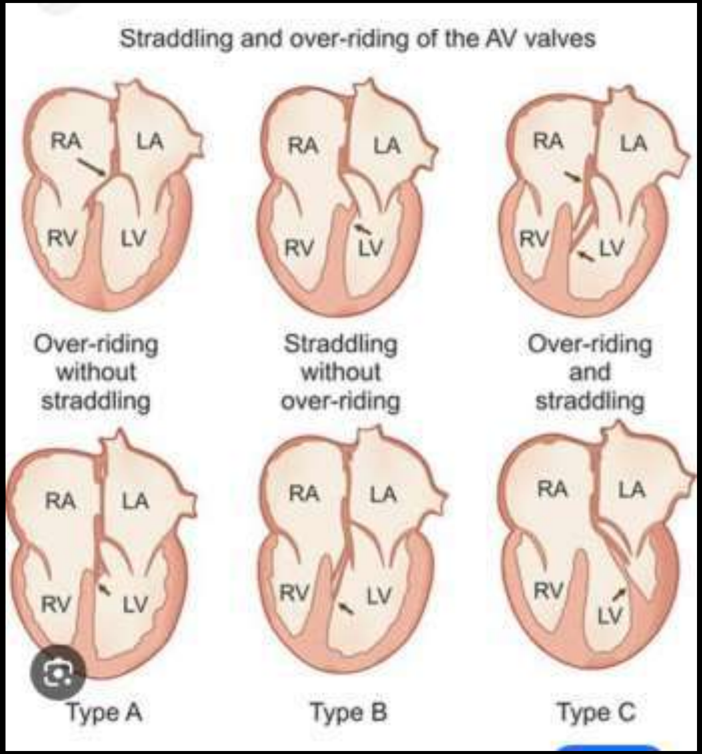
Multimodality imaging in ACHD.

Laman Eyvazli, MD
Central Custom Hospital

What we must know?

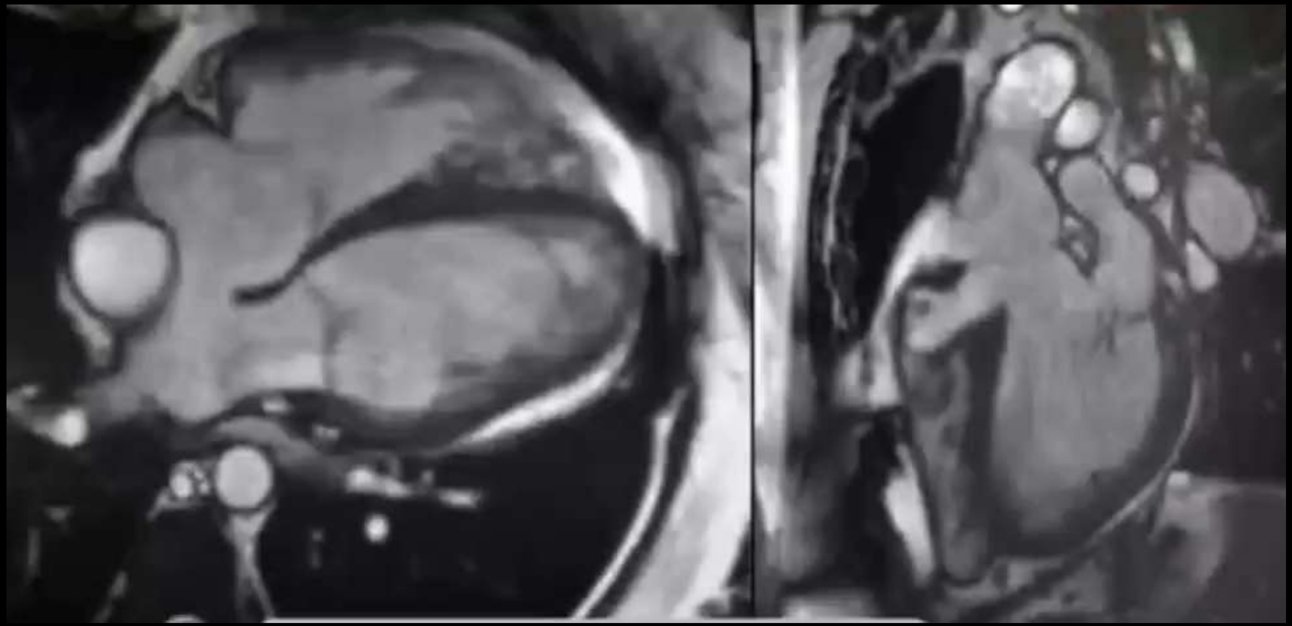
- Levocardia\ Mesocardia\ Dextrocardia- is related to the direction of the apex
- Isomerism – Both atriums in RA/LA
- Overriding –positioned directly over a IVS
- Stradling – AV valves cross over or straddle the IVS, connecting the both ventricles with chordae or papillary muscle
- Double outlet –major arteries originated from one ventricle, instead of one from each ventricle
- Double inlet – left and right atriums empty into the same lower chamber





Malalignment
– between IAS and iVS, between muscular and infindubular/conal
-Conal septum ant/post deviation
Ant deviation-> RVOTO-> pulmonar stenosis,peripheric pulmonar stenosis
Post deviation-> LVOTO->arcus hypoplasia, coarctation,interruption

Atrial septal defect



Types of ASD

- Secundum typ
- Primer typ- AV
- Sinus venosus SVC typ
- Sinus venosus IVC typ
- Unroofed coronary sinus ASD

-In most cases, the diagnosis is made using **transesophageal echocardiography (TEE)**.
-In complex cases or when there is suspicion of additional pathology, we may use **contrast-enhanced CT** as an additional diagnostic tool.
***Preoperative cardiac catheterization is mandatory**.
-For **secundum-type ASDs**, **percutaneous transcatheter closure (PTC)** can be performed. In such cases, the **rims** surrounding the defect must be adequate.
-Another important point is that the **device size should correspond to the total septal length** to ensure proper placement and closure."

Case 1. Secundum typ ASD

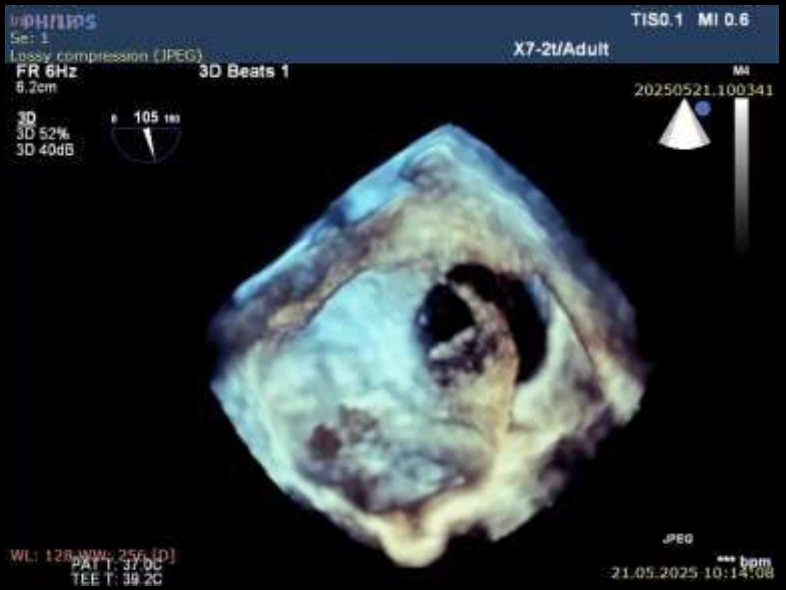
TEE from my archine

Case 2. **Sinus Venosus typ ASD+ Cardiac typ PAPVD**

***If the shunt in ASD is directed from right to left, Probability of PAPVD is high — pulmonary veins Must be checked.**

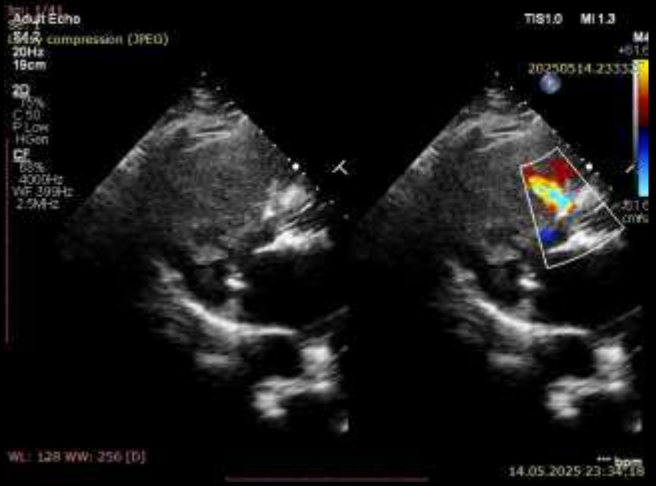
***In patients with sinus venosus type ASD, it is necessary to check the SVC flow after repair, as obstruction may develop.**

Case 3. Secundum ASD



TEE from my archine

Ventricular Septal defect – increased pulmonary flow



Case4. Outlet VSD

Types of VSD (Ventricular Septal Defect)

- Muscular VSD
- Perimenbranous VSD
- Inlet – AV position and posterior
- Outlet – Anterior-superior position
- Subarterial - double committed VSD- anterior position -check in Psax view 1-2 Oclock position

- **– What should we pay attention to?**

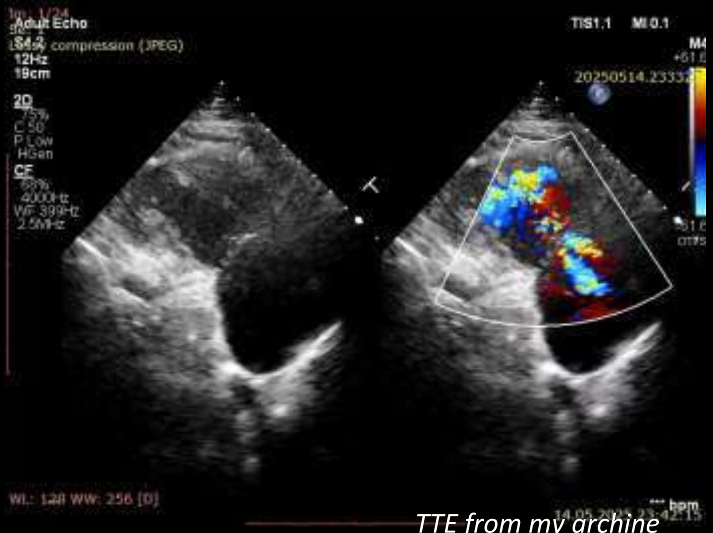
-Position and presence of additional defects: Assess using 2D imaging and color Doppler for location and any associated anomalies.
-Size (based on proportion to aortic annulus):

- Small VSD: < 1/3 of the aortic annulus
- Moderate VSD: 1/3–2/3 of the aortic annulus
- Large VSD: > 2/3 of the aortic annulus

Flow characteristics:

- Is the flow laminar or turbulent/restricted?
- Evaluate the pressure gradient across the defect.

- **Presence of outflow tract obstruction**:LVOTO? RVOTO?
- **Indirect signs**:
- Assess for volume overload or pressure changes.
- What are the heart chambers size ?
- RVSP / sPAP / Vp measurements:



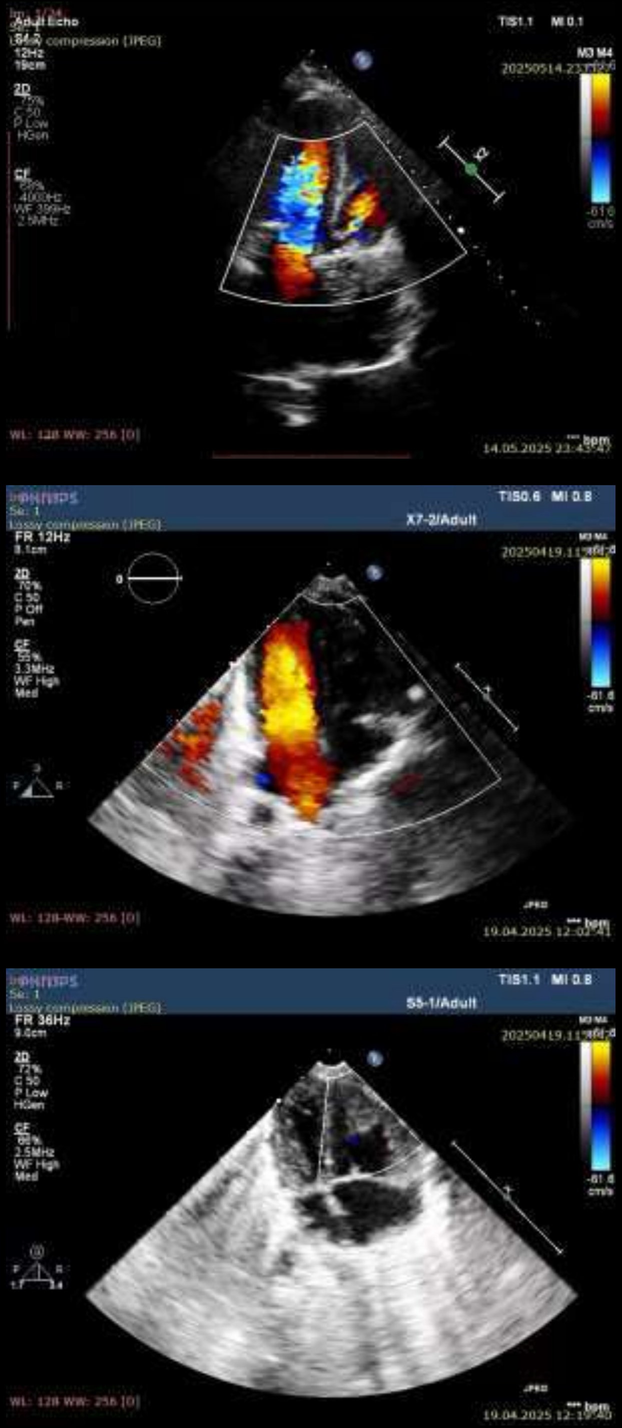
TTE from my archine

VSD flow directly impacts the lateral wall, leading to tissue proliferation

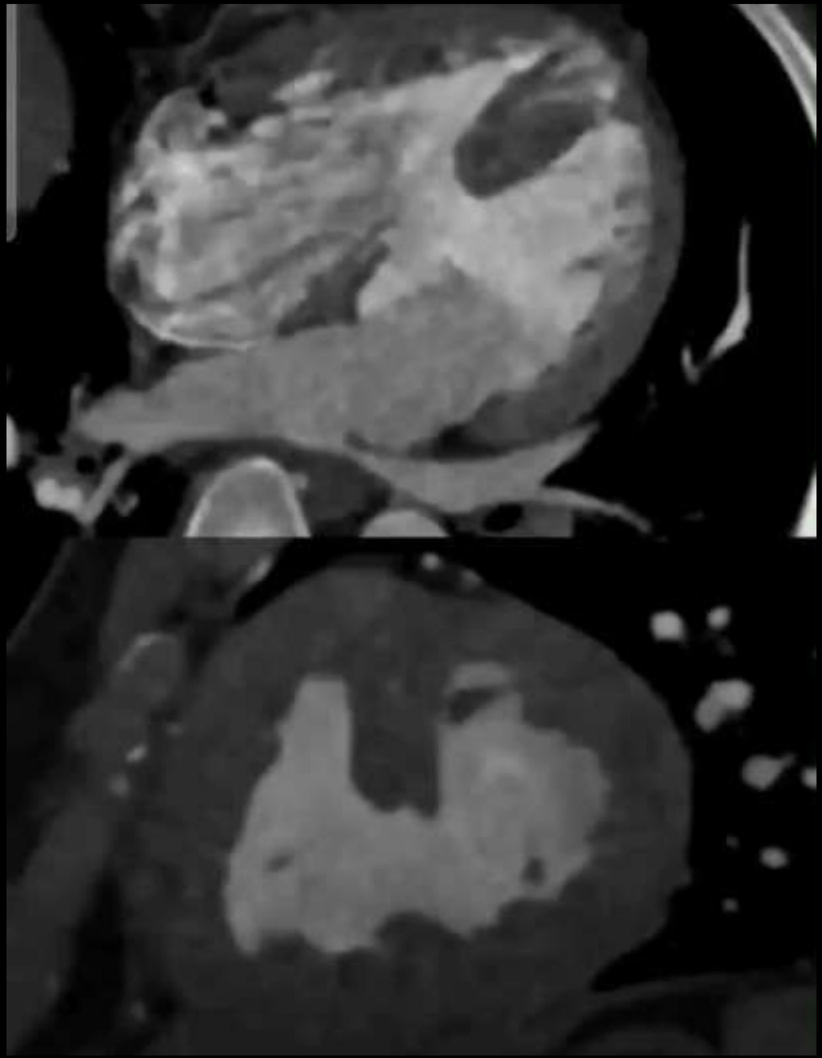
*A VSD is considered restrictive if the gradient is greater than 36 mmHg.”

VSD Long-Term Complications

- **Double Chambered Right Ventricle (DCRV)**:
  High velocity jet from VSD causes proliferation the RVOT endocardium.
- **Discrete Subaortic Stenosis**:
  This is related to the LVOT anatomy and the turbulent flow created by the VSD.
- **Aortic Valve Prolapse (AVP) / Aortic Insufficiency (AI)-** *is a compensatory mechanism where the leaflet prolapses in attempts to close the shunt. This leads to diastolic restriction, and the altered leaflet coaptation causes aortic insufficiency."*
- **Endocarditis**
- **Hemodynamic Consequences**:
  Since the left ventricle (LV) pumps blood toward the right side during systole, the right ventricle (RV) subsequently pumps blood directly into the pulmonary artery (PA), resulting in LV volume overload.
- **Patient Follow-Up:**
  We follow up these patients with ECG, echocardiography, Holter monitoring, and MRI.



Case 5. Muscular VSD



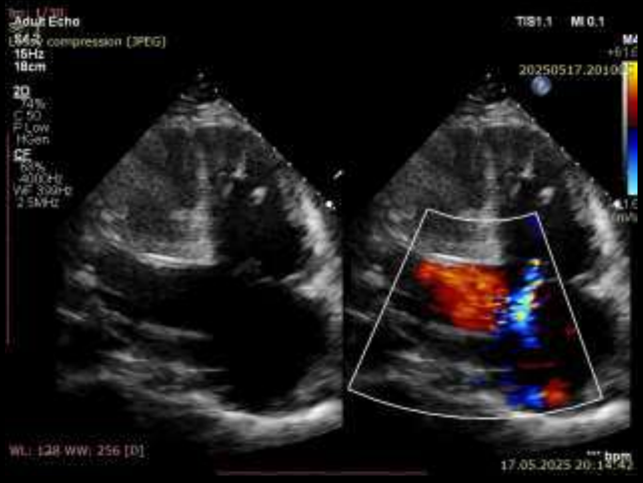
TTE from my archine

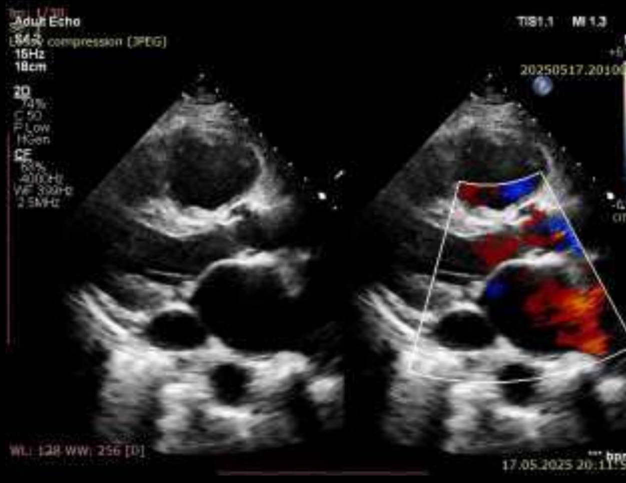
PAPVD\ TAPVD(R-L shunt)- increased pulmonary flow

PAPVD types

- Supracardiac – PV drained to SVC , innominate vein
- Cardiac – PV drained to CS\ RA
- Infracardiac – PV drained to IVC
- Mix typ

-PAPVD is generally diagnosed using contrast-enhanced computed tomography .
This is our sinus venosus ASD patient in which we have seen RUPV drained into RA.
-The diagnoses of unroofed coronary sinus and persistent left superior vena cava (PLSVC) have been ruled out.”
- *We consider that the extra pv is draining into the cs and we* recommended a contrast enhanced CT examination for the patient.”





<<-Crab views

We can evaluate pulmonary veins in
* High psax view
* Suprasternal crab view

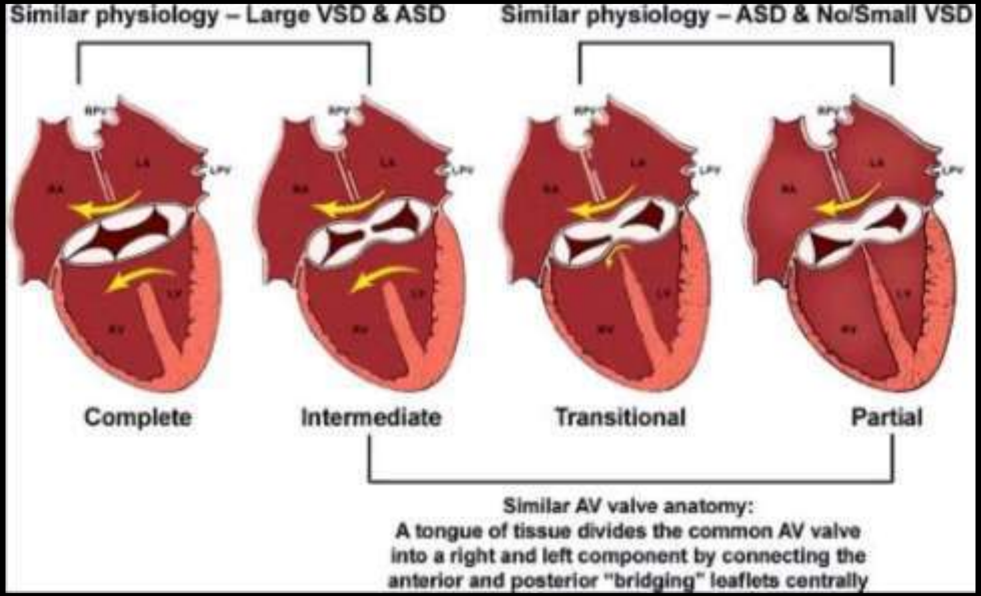
“Upper veins are shown in blue, and lower veins in red; this corresponds to the direction of blood flow.”

From my archive

Atrioventricular septal defect- increased pulmonary flow

Types

- Complete -primium ASD + common anulus+ Inlet VSD - Single annulus single orifice
- Intermediate – Primium ASD + large vsd +- single annulus double orifice
- Transient - primium ASD + seperated anulus + restricted/ absent VSD
- Incomplete - primium ASD + seperated anulus, no VSD



Specific signs

- AV valve leaflets are at the same level.
- No AV septum present.
- Aortic valve is located anteriorly.
- Elongation of the LVOT (Left Ventricular Outflow Tract).
- Counterclockwise rotation of the LV papillary muscles.
- Cleft in the AML.

Balance at the Ventricular Level:

•The common AV valve opens equally into both ventricles.
•Both ventricles are well developed.

Unbalance at the Atrial Level:

•The interatrial septum (IAS) is malaligned with the interventricular septum (IVS).
•Seen in conditions like DORV or DOLV .

Unbalance at the Ventricular Level:

•The common AV valve is more dominant toward one ventricle.
•Seen in conditions like DILV or DIRV .

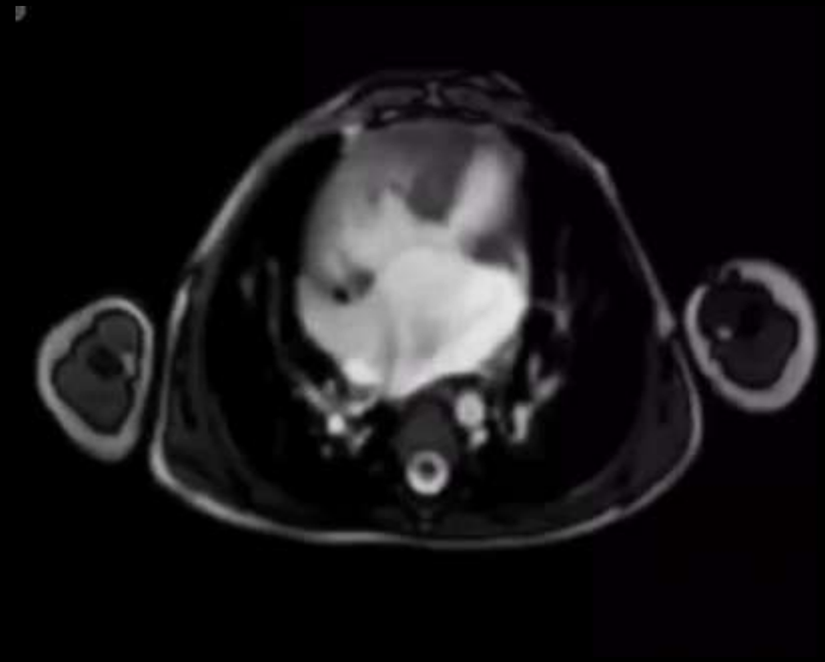
What Should We Pay Attention To?

•AV Valves: *Assess using subcostal en face view.*

•ASD/VSD: *Evaluate the size and direction of the shunt.*

•Ventricular Size and Function: *Assess development and contractility of both ventricles.*

•LVOT and RVOT: *Check for patency, obstruction, and anatomical alignment.*



Surgical Timing Based on Volumetric Parameters

•An RVEDV indexed to body surface area (RVEDVi) > 120–140 mL/m² is indicative of significant right ventricular volume overload.

Persistent RV volume overload is associated with an increased risk of:

- Right heart failure (RHF)
- Progression to pulmonary hypertension (PH)
- Arrhythmias and reduced exercise capacity

Surgical intervention is generally advised when RVEDVi exceeds 120–140 mL/m² to prevent irreversible RV dysfunction and adverse outcomes.

Interatrial Septal (IAS) Malalignment and Its Consequences (Unbalances AVSD patients)

•IAS Malalignment can result in abnormal streaming of pulmonary or systemic venous return, which may lead to:

- Double Outlet Right Atrium (DORA) or
- Double Outlet Left Atrium (DOLA)

These terms refer to both atrial inflows predominantly draining into a single ventricle .

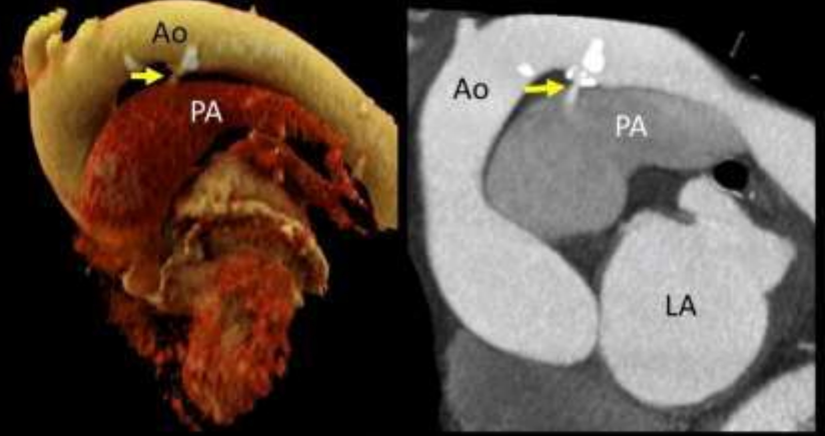
•This abnormal inflow pattern leads to unbalanced ventricular loading, where one ventricle receives the majority of the inflow:

- *Over time, this results in hypoplasia (in opposite ventricle) of the underloaded ventricle due to decreased volume stimulus during development.*

Postoperative Findings in Patients with Corrected Atrioventricular Septal Defects (or Similar Conditions):

1. Residual Left Ventricle to Right Atrium (LV-RA) Shunt:
    1. *May persist due to incomplete closure or patch dehiscence.*
    2. *Can lead to volume overload of the right atrium and right ventricle.*
    3. *Requires monitoring; significant shunts may necessitate reintervention.*
2. Atrioventricular (AV) Valve Insufficiency or Stenosis:
    1. ***Insufficiency:*** *Often due to valve dysplasia, malcoaptation, or leaflet tethering.*
    2. ***Stenosis:*** *Can result from postoperative scarring or valvular thickening.*
3. Subaortic Stenosis:
    1. *Can be a residual or progressive finding post-repair.*
    2. *Often due to fibromuscular ridges or tunnel-like narrowing below the aortic valve.*
    3. *May contribute to left ventricular outflow tract (LVOT) obstruction, requiring surgical revision if significant.*

Patent ductus arteriosus- increased pulmonary flow



Patent ductus arteriosus „Botalli" (**PDA**) with persistent connection between pulmonary artery (PA) and Aorta (Ao) resulting in a left-right shunt (→).

1.Classic typ origin DescA?,Angle <90^0 , course –straight (IVS-PA)
2.Intermediate typ? Origin DescA, Angle - 90^0-135^0 , course- curved
3. Vertical typ origin below the aortic arch,course vertically (PA VSD/TOF)course- curved
4.Duct from Aortic Arch to single lung – origin aortic arch
5.PDA from subclavian/innominate artery to single lung
6. PDA from subclavian/innominate artery to both lung

The **volume of blood flow through the PDA** is **critically important**.

- **Excess flow** → risk of **pulmonary hypertension** and **CHF**
- **Insufficient flow** (in ductus-dependent cases,PA, **severe PS**→ **cyanosis**

Silent PDA < 1.5mm hemodinamically insignificant
Small 1.5-3mm Mild L-R shunt
Modereta 3-5mm Moderate shunt (volume overload)
Large PDA >5mm Significant shunt (HF,PH)

Possible Complications of Late PDA Closure:

Reduction of Ejection Fraction (Geometric Remodeling and Myocardial Stunning)

1. Chronic volume overload causes geometric remodeling of the left ventricle.
2. Sudden elimination of the volume load after PDA closure can lead to transient myocardial stunning and a decrease in EF.

Aortic Insufficiency (Due to Volumetric Changes)

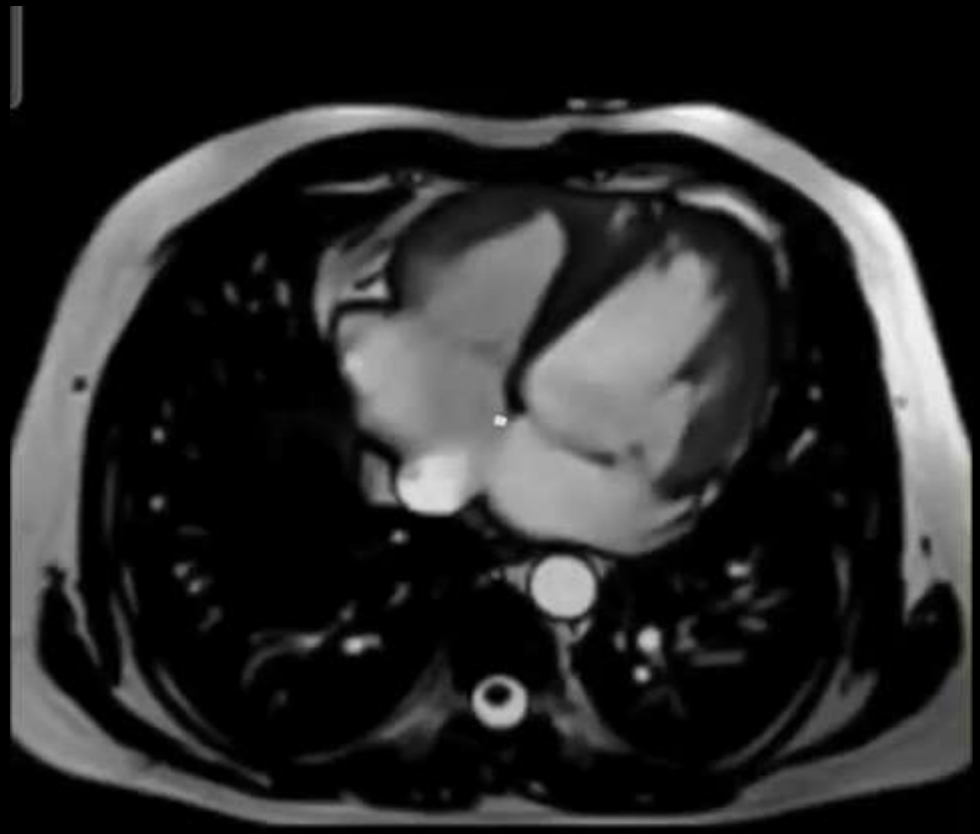
1.Abrupt changes in left ventricular loading conditions may affect aortic valve leaflet coaptation.
2.The aortic valve, adapted to chronic volume overload, may become insufficiency after PDA closure due to altered geometry.

TTE from my archine

AP window

It is the same phisiology with PDA , but proximal Ascending aortic level

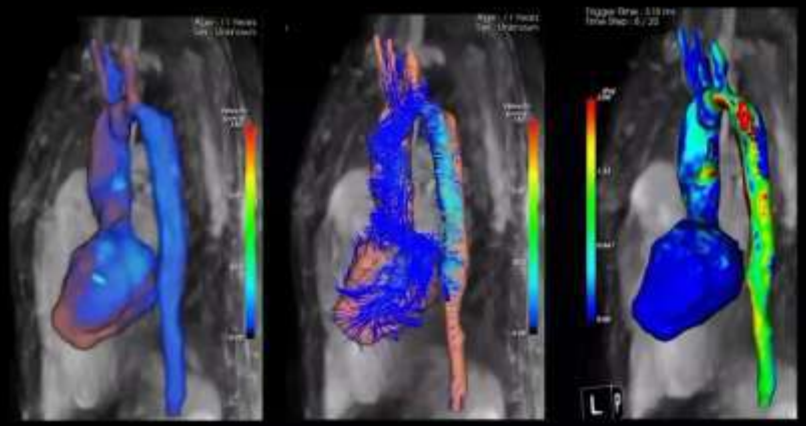
Type I / proximal: the communication is located between the ascending aorta and the pulmonary trunk.
Type II / distal: the defect, in a spiral curve, involves the pulmonary bifurcation at the level of the right pulmonary artery (RPA). In this type of defect, there can be hypoplasia or even aortic arch interruption due to the significant deviated flow from the aorta towards the RPA during fetal life.
Type III is characterized by total absence of the aortopulmonary septum resulting from the combination of proximal and distal defects.



CMR – RVEDVi >120-140ml/m2 - Volume overload requiring surgery

Clinical presentation is dependent on the size of the defect and on the associated lesions.

Arcus pathologies



- Coarctation

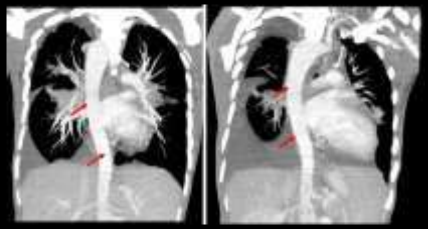
 -Preductal - proximal to the ductus arteriosus

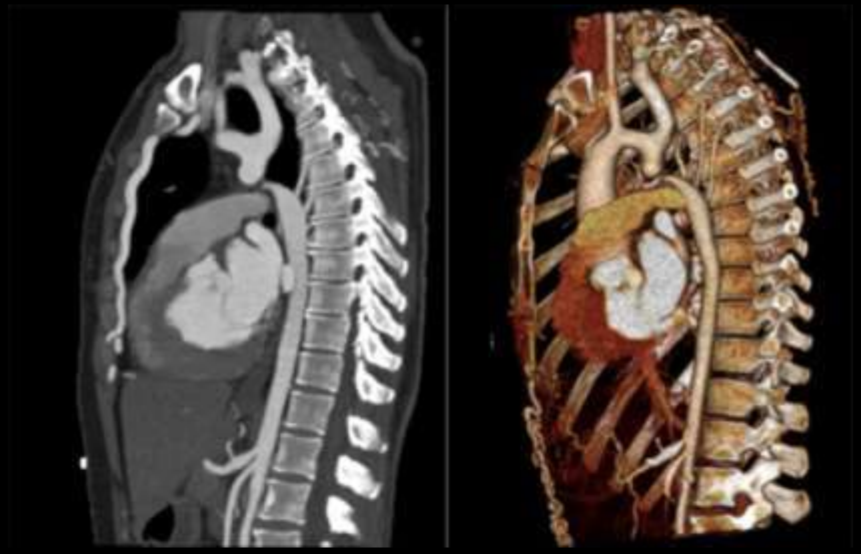
 -Ductal - Narrowing occurs **at the site of the ductus arteriosus**.

 -Postductal - **distal to the ductus arteriosus**

- Interruption - complete discontinuity between the ascending and descending aorta.
- Double aortic arch - Both right and left aortic arches persist, forming a vascular ring around the trachea and esophagus.

- Right aortic arch - The aortic arch courses to the **right side** of the trachea instead of the left.



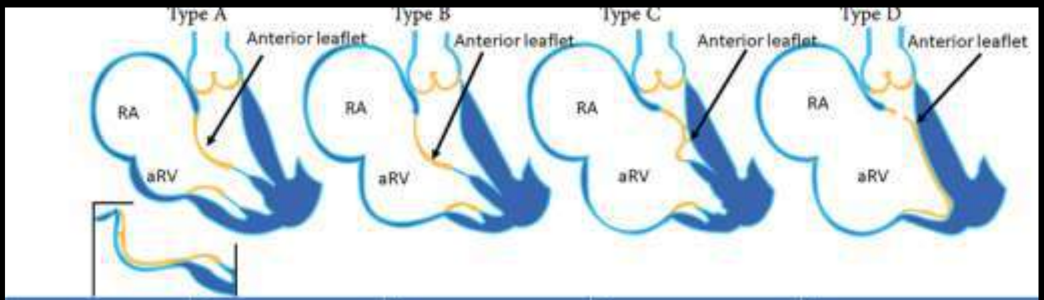


*The left aortic arch is positioned at approximately the 1–2 o’clock direction, while the right aortic arch lies around the 11–12 o’clock position.

Ebstein (R-L shunt) (RV Miopathy) and Ebsteinoid anomaly (ccTGA,ivs/pa-da)

Failure of delamination

*Inoperated adult in A/ B level



	A	B	C	D
Septal/posterior leaflet displacement	Mild	Moderate	Severe	Extremely severe (tricuspid valve sac)
Anterior leaflet	Normal morphology and mobility	Abnormal attachments with normal mobility	Partial adhesions to the ventricular free wall and restricted motion	Severe adhesions to the ventricular wall
"Atrialized" ventricle	Small/preserved function	Moderate/reduced function	Large/reduced function	Very large /minimal function
Functional ventricle	Normal/preserved function	Reduced/preserved function	Small/reduced function	Infundibulum/depressed function

-*Discplacement index 8mm/*m2
-True anulus
-Tricuspid insufficiency and stenosis
-RV?- atrialized RV?- RVOTO?
-The higher the RVSP , the more **functional the RV** tends to be, indicating preserved contractility and forward flow capacity.
Conversely, a low RVSP may favor **functional atresia** or severely impaired RV function.
-Pulmonary valve ?Peripheric pulmonary branch? PDA?BT shunt?
-IAS ? IVS ? Additional defects?
-IVC ? HV reverse flow ? **Hepatic reverse flow may not be seen until late stages** due to initially high RA compliance, so it does NOT exclude RV failure.
RV failure may be **"silent" early on** due to compensatory mechanisms.

Two Important point:

1. Can the Right Ventricle (RV) Provide Antegrade Pulmonary Flow?

This depends on:

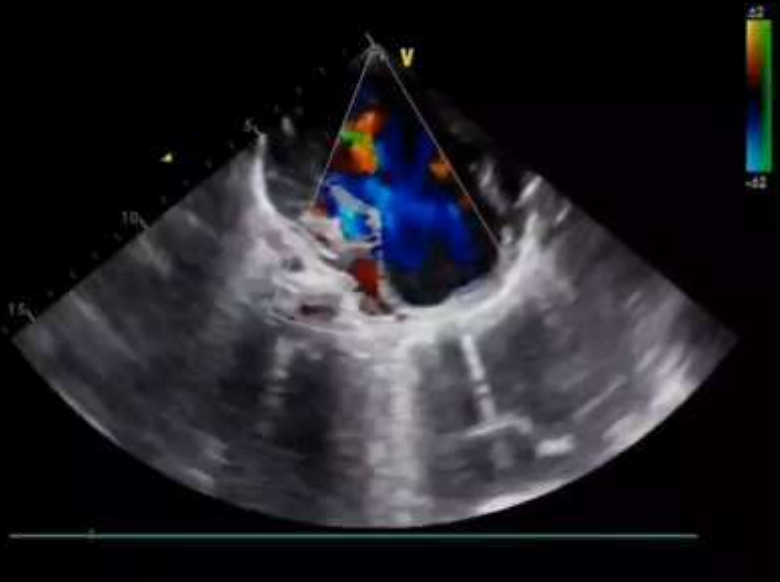
•RV size and function
•Presence and severity of **pulmonary valve stenosis or atresia**
•Integrity of the right ventricular outflow tract (RVOT)

If RV antegrade flow is possible, it can maintain pulmonary circulation without complete reliance on shunts or collaterals.

How to Maintain Pulmonary Blood Flow (Pulmonary Flow Maintenance)?

Options include:

•**Antegrade flow from RV** (if feasible)
•**Patent Ductus Arteriosus (PDA)** or surgical shunts in cases of PA or PS
•**Pulmonary artery reconstruction or augmentation** to optimize flow
•In complex cases, staged surgeries (e.g., RV-PA conduit, Glenn, Fontan) are planned to maintain balanced pulmonary circulation.



Difference between Ebsteinoid valve

-Dilated annular size
-RV miopathy (thinning+dilatation)
-ATV sail like appearance (compensation mechanism)
-atrialized RV

TV dysplasia difference from ebstein

- Normal annular insertion
- Thick, immobile leaflets with short chordae
- No atrialized RV

CMR- RV measurement and function evaluation

CMR –preferably RVEDVi >50ml/m2, <50ml/m2 limit surgical option
RVEF> 40-45%,
EF<40% increasing surgucal risk
TRF>30-40%

Starnes procedure-TV closure- Prevents severe TR
RV fenestration- helping reduce systemic venous congestion
ASD enlargement - R_L shunt maintain systemic outpul
RA plication- reduce atrial size, decrease arrhythmia risk

Tricuspid atresia (R-L shunt)

Types of Tricuspid Atresia Based on RV Development

- Monopartite –only inlet portion
- **Bipartite - inlet+ trabeculated-apical portion**
- **Tripartite - inlet+ trabeculated-apical+ outlet**

- **Specific Signs of Tricuspid Atresia (TA)**
- **1. Small Right Ventricle (RV) Cavity**
- *Due to absence of direct inflow from the right atrium.*
- Degree of hypoplasia depends on:
  - Presence of a VSD
  - Presence or absence of pulmonary outflow
- **2. Right Ventricular Hypertrophy (RVH)**
- *If there is subpulmonary obstruction or pulmonary atresia, RVH may develop due to pressure overload*.
- **3. Poor Development of Right Ventricular Outflow Tract (RVOT)**
- *RVOT is often* ***underdeveloped or atretic*** *due to lack of normal flow.*
- In some types, the **pulmonary valve and main pulmonary artery are extremely hypoplastic or atretic**.
- **4. Severe Pulmonary Atresia**
- In many cases, **pulmonary atresia is complete** and **pulmonary blood flow depends on a PDA** or surgical shunt.
- Severity of pulmonary atresia influences cyanosis and urgency of intervention.

Central shunt** is preferred over the traditional* ***(BT) shunt (Avoids Coronary steal phenomenon)

- Ductus dependent PA circulation -To keep PDA open- if necessary PDA stenting
- Pulmonary circulation with VSD
- **Small VSD → inadequate pulmonary flow**, leading to **cyanosis**
- **Large VSD** → May permit sufficient flow or risk of pulmonary overcirculation
- IF ASD size is not adequate- Balon atrio septostomy

TA patient managemant strategies

Monopartite –**Early Phase:**
•**Balloon atrial septostomy** (*to ensure adequate atrial level mixing*)
•**PDA stenting** (*to maintain pulmonary blood flow*)
Late Phase:
•**Starnes Procedure:** Tricuspid valve closure →Followed by **Fontan pathway**
Bipartite--> early phase
•Pulmonary balloon valvuloplasty - *to enhance RV-pulmonary connection*
•ASD closure-(*to direct blood through the RV*)
•If oxygen saturation (SpO_2) remains <90% - bidirectional Glenn + IVC-RV + PA-
Tripartat- PDA stent , Pulmonary balloon valvuloplasty , ASD closure

Why Atrial Septostomy Is Not Performed in IVS + PA (Bipartite) Patient?
• Encourage **antegrade flow through the TV and RV to PA**.
• Preserve or enhance the potential for **biventricular or 1.5 ventricle circulation**.

<u>Biventricular repair only possible if RV Volume/function adequate in CMR</u>
RVEDVi<30ml/m2 severe hypoplastic-single palliation preferred
30-50ml/m2- borderline
>50ml/m2 candidate for biventricular repair

IVS+ PA-intact IVS + pulmonary atresia (R-L shunt)

Additional LV Pathologies in IVS-PA Patients due to the altered hemodynamics - LVNC,EMF, LVOTO, LVdysfunction

Anterior angulation

- RVOT is closed- stenotic/atretic.
- RV hypoplastic - The smaller is the TV, the smaller is the RV
- Pulmonary circulation PDA dependent (typ1)
- Mostly confluent PA
- Normal arborization in most patients
- Rarely non confluent PAs- bilateral PDA- rarely MAPCA
- **Atrial Septal Defect (ASD)** is present in most cases
    - *Ensures right-to-left shunting and systemic output*
- TV insufficiency dilated anulus- dicplacement -> Ebstein like/displastic appearance
- TV stenosis –hypoplastic, obstructed anulus, thickened valve leaflets, dysplastic chordae ,

Abnormal papillary muscles – leads to hypoplastic RV

- Coronary anomalies – Presence of **RV–coronary sinusoids** or **fistulas** seen on ventriculogram

TTE from my archine

VSD + PA– decreased pulmonary flow

Prevalent Types of Pulmonary Blood Flow Patterns

1. Pulmonary Atresia – Ductus-Dependent Pulmonary Circulation-syanotic group.

Management options :

- **PDA stenting** or
- **Systemic-to-pulmonary shunts** such as:**Blalock-Taussig (BT) shunt/ Central shunt**

2. Free Pulmonary Flow – Heart Failure Group

- *As PVR **drops over the first few weeks**, **excessive pulmonary flow** may lead to **symptomatic heart failure**.*
- **Pulmonary artery (PA) banding** is often performed to: **Restrict pulmonary blood flow- Reduce pulmonary overcirculation and pulmonary hypertension**

3. Pulmonary Stenosis (PS) with Balanced Pulmonary Flow- **saturations around 80–85%**.

-D TGA + TA
- Subaortic VSD + PS
-Large ASD
- Hypoplastic RV,RPA and LPA

Key Parameters Evaluated in Patients with Pulmonary Atresia and MAPCAs

- Presence and size of native PA-*diameter, confluence, and distribution*
- Number and anatomy of MAPCAs
- Origin, course and distribution to lung segments
- Stenosis, turtuosity or narrowing
- Suitability for Unifocalization – *caliber and length*
- Whether complete repair (*VSD closure + RV-PA conduit*) is feasible or needs staging

Why Atrial Septostomy Is Not Performed in IVS + PA (Bipartite) Patient?

- Encourage **antegrade flow through the TV and RV to PA**.
- Preserve or enhance the potential for **biventricular or 1.5 ventricle circulation**.

Fallo-

VSD –usually large and anterocephal malalignment->RVOTO
RVH - chronic pressure overload
Overriding Aorta- is displaced over the VSD, receiving blood from both ventricles
Pulmonary stenosis - subvalvular/valvular/supravalvular
MAPCA

Important Point in (TOF)
- **Pulmonary artery (PA) size and arborization** directly influence:

•Surgical timing
•Type of repair (complete vs. staged)
•Suitability for shunt or conduit

* **Palliative Preoperative Percutaneous Procedure: RVOT Stenting**

* **Preoperative Must-Know: Coronary Artery Course-**
defining the coronary artery anatomy is critical *before any interven*tion

-Postop complications
1. Severe Pulmonary insufficiency
2. Residual VSD / Additional VSD **->>** *clinical findings is mild PH, O2 desaturation*

- Persistent left-to-right shunting
- RV volume overload
- Heart failure symptoms
- Pulmonary overcirculation

-**How Are These Patients Followed Up Post-Repair**
1. **RV Volumetric Assessment**
2. **Flow Regime -Pulmonary Regurgitation**

Accurate volumetric analysis is essential for determining the timing of PVR

PRF> 25-40% moderate to significant regugitation
PRF> 40-50% indicates severe pulmonary regurgitation

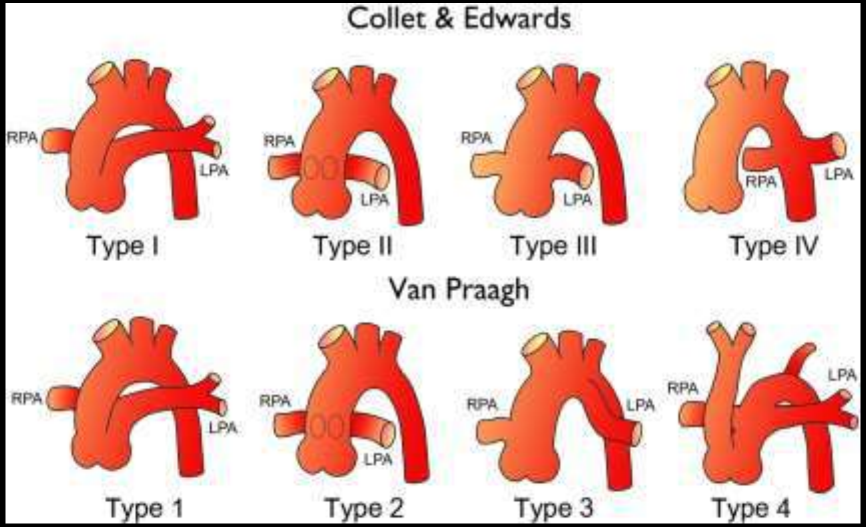
RVEDVi> 160ml/m2 RV dilatation
RVEDVi> 170-180ml/m2 threshold for considering PV
RVESVi>80-90ml/m2 may suggest impaired RV systolic function
RVEF < 45-50%

Truncus arteriosus

Diagnosis Gold standart- CT
Follow up- Echo, CMR

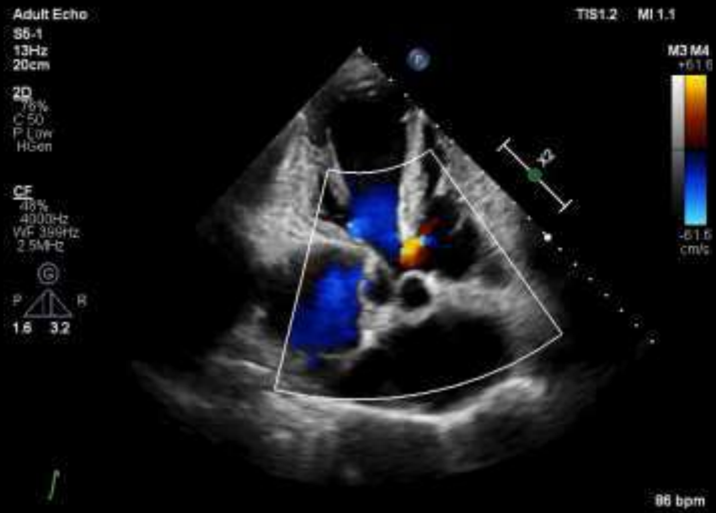
- **Type I:** Both pulmonary arteries originate from a short common pulmonary trunk.
- **Type II:** The pulmonary arteries arise separately from the posterior aspect of the truncus arteriosus.
- **Type III:** The pulmonary arteries have separate origins from the lateral aspects of the truncus arteriosus.
- **Type IV:** Pulmonary atresia with ventricular septal defect (VSD); the pulmonary arteries are absent or supplied by collateral vessels.

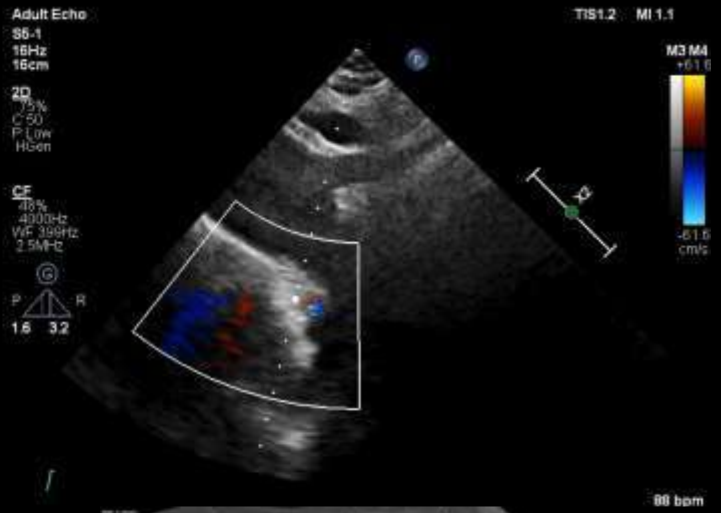
Van Pragh;
Type l - Collet type l, Type ll- Collet type ll and lll
Type lll - One pulmonary artery arises from the truncus; the other is supplied by collateral vessels.
Type lV - Associated with **interrupted aortic arch** and other arch anomalies.

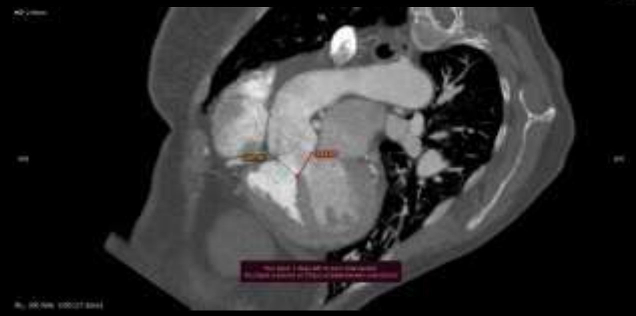


CMR and CT values
- Ventricular volumes - *guides surgical timing, checks for dilation, failure*
- Ejection fraction - *reduced EF impact prognosis*
- Truncal valve assessment - *moderate truncal valve regurgitation requires surgical intervention*
- Pulmonary artery origins - *helps define type and surgical approach*
- Branch PA size – *Determines feasibility of surgical repair or conduit placement*
- Aortic arch anatomy - *important in surgical repair plan*
- Great vessel relationship
- LGE - *Poor prognostic indicator post repair*

RVEDVi> 100ml/m2 –dilation
RVEDVi>150ml/m2-significant dilation
LVEF/RVEF< 50% dysfunction
Truncal valve regurgitant fraction >30-40% moderate to severe







Rastelli procedure: *<u>Closure of VSD +RV -PA conduit</u>*

Post Op Complications
- Conduit dysfunction -CMR
- Residual VSD
- Truncal valve insufficiency/stenosis-CMR
- Pulmonary hypertension
- Arrythmias
- Endocarditis risk
- Ventricular remodeling -LV systolic dysfunction-CMR
- Branch PA stenosis –CMR

TTE from my archine

TGA

D-TGA ventriculo-arterial discordance
RA-RV-Ao, LA-LV-PA
L-TGA atrio-ventrucular and ventriculo-arterial discordance
RA-LV-PA, LA-RV-Ao

- Parallel circulation
- Need blue blood get to the lungs and pink blood to get to the body

<u>What is the initial management of TGA?</u>

-Start prostaglandin E1 infusion to maintain ductus arteriosus patency

- atrial-level mixing is typically more effective -> balloon atrial septostomy

Early phase- arterial switch Jaten operation - *the great arteries are anatomically corrected*

Late phase- atrial switch operation - *redirects blood flow at the atrial level*.

<u>What Complications Might an Adult Cardiologist Encounter in TGA patients ?</u>

- RV dysfunction
- TV insufficiency
- Baffle obstruction –*commonly SVC*
- Baffle leaks- *can cause hypoxia and increase stroke risk*
- Pumonary hypertension

-ccTGA IVC-RA-LV-PA/ PV-LA-RV-Ao
-Ebstein deformity of LV sided TV valve

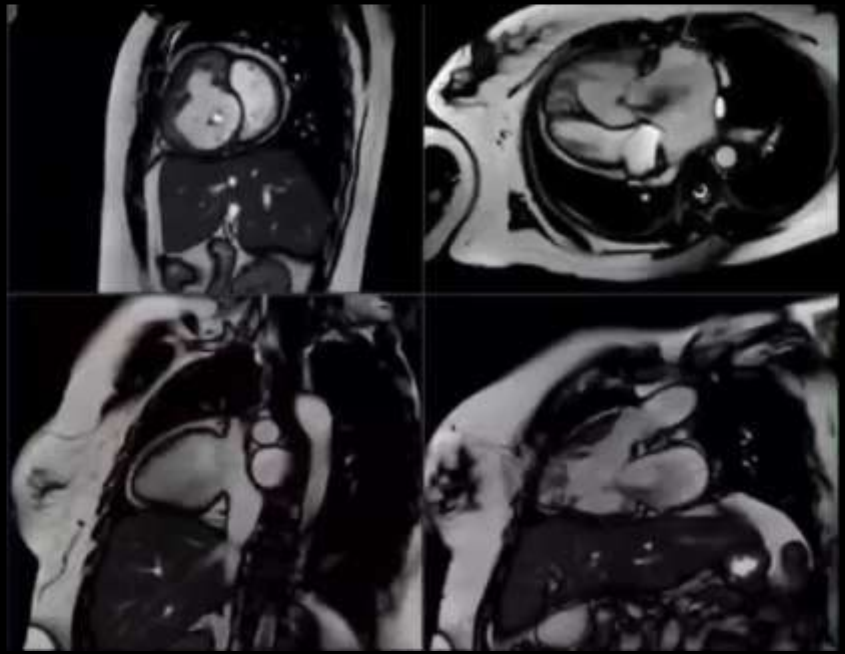
*** The preferred site for mixing is the atrial level — an atrial septal defect (ASD).**
* Even if the **patent ductus arteriosus (PDA)** is small and there is **no ventricular septal defect (VSD)**, this can still provide **adequate mixing** to maintain systemic oxygenation

TGA

HOW do we differentiate?

- LV Shape
- Mitral-aortic continuity
- Mitral-pulmonary continuity in TGA
- Position of the Chordae Tendineae
- LAA morphology
- Coumadin ridge

- RV Shape, trabeculation
- The septal leaflet of the TV is directly attached to the interventricular septum
-TV is located more apically than the mitral valve
-According to RAA morphology
- Crista terminalis, Chiari network, eustachian and tebesian valve in RA



Tips &Tricks

- **If the aorta is located anterior and to the *left*** → it suggests **L-TGA**
- **If the aorta is located anterior and to the *right*** → it indicates **D-TGA**
- **Anterior malalignment** of the conal septum may lead to **right ventricular outflow tract obstruction**
- **Posterior malalignment** may result in **left ventricular outflow tract obstruction**
- The **arterial switch operation** should ideally be performed **within the first 21 days of life**, while the **left ventricle is still conditioned** to support systemic circulation.
- The **pulmonary artery becomes the neoaorta**, so:
- It must have a **normal structure**, with no **stenosis**, **insufficiency** or **abnormal valve morphology** (e.g., **no dysplastic or bicuspid pulmonary valve**)

***Coronary artery anatomy must be clearly defined before surgery.**

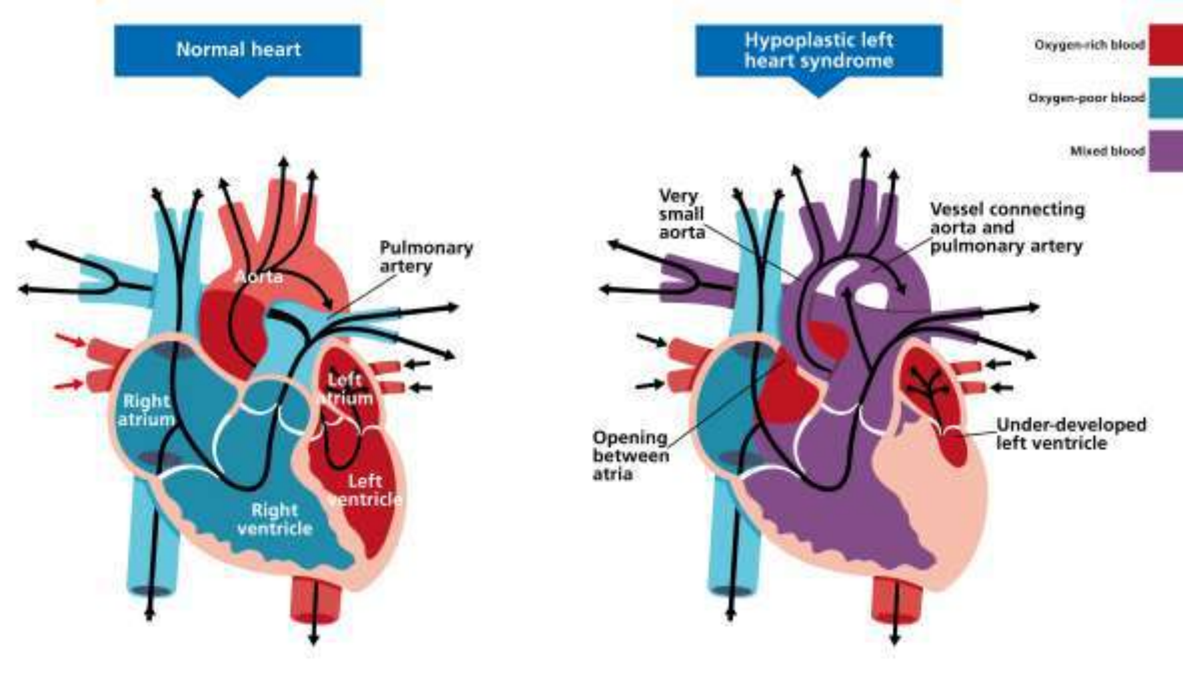
Hypoplastic left heart disease- Fontan candidate

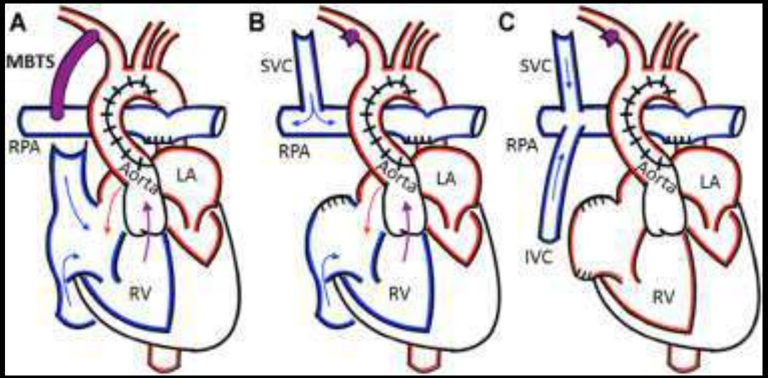
Left heart valves atresia/hypoplasia caused HLHS

-Worst Prognosis in Left-Sided Obstructive Lesions

Mitral Stenosis + Aortic Atresia

- **There is inflow into the LV (via the mitral valve), but no outflow (due to aortic atresia).**
- *This causes **severe pressure buildup**, leading to **LV distention and dysfunction***
- *Without an **outflow tract**, the **left ventricle must be decompressed**.*
- *The stagnant flow and high pressure can lead to **endocardial fibroelastosis***





1. Systemic Circulation depends on ASD and PDA

•*In **hypoplastic left heart syndrome (HLHS)** and similar pathologies, the **systemic output** is entirely dependent on:*

- **ASD** → for left atrial decompression
- **PDA** → for supplying the **aorta** and **coronary arteries**

If the ASD Is Restrictive presented as Cyanosis

•*A **restrictive ASD** impairs **pulmonary venous drainage**, raising **LAP**, causing **pulmonary congestion and severe cyanosis**.*

PDA Is Vital for Both:

•**Systemic circulation**

•**Coronary perfusion**

*Without it, **severe ischemia and rapid deterioration** occur.*

Preoperative Assessment

LV,MV, mitral anulus,LVOT- hypoplastic

Arcus – hypoplastic/interrupted

Aortic valve – atresic/stenotic,Mitral valve - atresic/stenotic

Borderline LV- Mitral valve and aortic valve annuli are approximately 50% hypoplastic compared to normal.

RV must support both pulmonary and systemic circulation, preoperative RV function is critical in determining outcomes

Tricuspid Valve Function:

- **Tricuspid regurgitation**, which can severely affect forward flow
- **Annular dilation** or **valve dysplasia**, common in longstanding RV volume overload

Surgical Management

- Stage l - palliative operation- Norwood procedure- neoaorta + arcus reconstruction+sano shunt - preferred over the BT shunt, as the BT shunt may cause coronary steal phenomenon) +atrial septotomi

- Interstage highest mortality stage – balancing systemic and pulmonary circulations is challenging.

- Coarctation and ASD restriction may develop

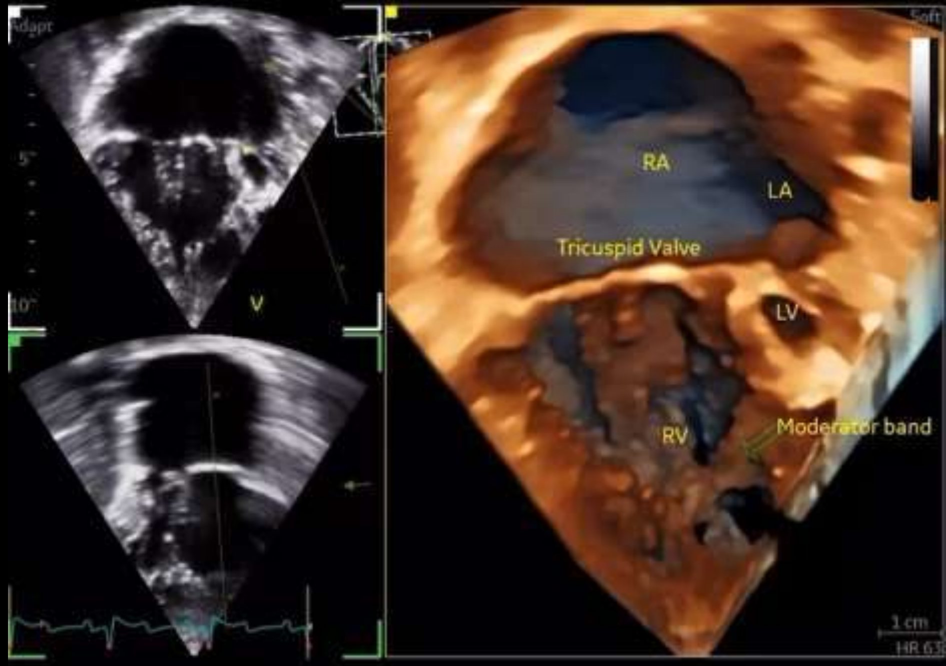
- Stage ll - During the Glenn stage, the AV valve should be evaluated. If there is significant insufficiency of the right AV valve, surgical repair should be performed.

- Glenn – reduces volume load on single ventricle and improves long term outcomes

- Stage lll - Fontan operation

- Fenestrated – to reduce pressure and improve cardiac output in the short term

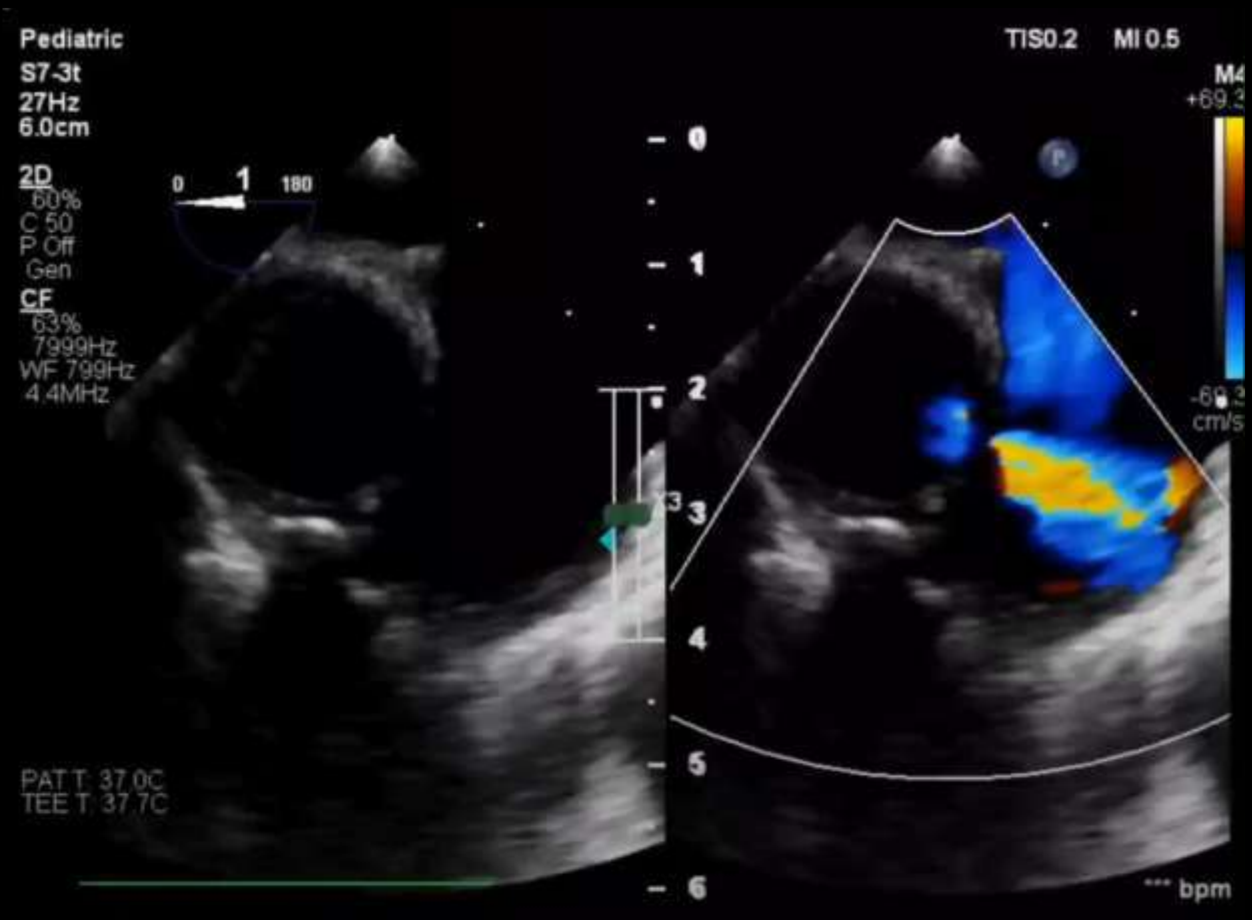
- Entire venous return bypasses the heart and flows passively into the lungs



Fontan evaluation

-**Fontan Evaluation Checklist**

1. **Is there a fenestration?**
    1. *Check for the presence and size of fenestration between the Fontan circuit and atrium.*
2. **Tunnel assessment**
    1. *Evaluate the extracardiac or lateral tunnel for patency, narrowing, or thrombus.*
3. **Anastomosis assessment**
    1. *Inspect all surgical connections:*
        1. *SVC-PA (Glenn)*
        2. *IVC or conduit-PA (Fontan)*
4. **Flow and turbulence**
    1. *Use **Color Doppler** to detect abnormal flow patterns and areas of turbulence.*
5. **CW and PW Doppler**
    1. *Measure flow velocities and pressure gradients within the circuit.*
6. **Hepatic vein flow**
    1. ***Increased flow with inspiration** is normal.*
    2. ***Reversed flow** may indicate elevated Fontan pressures or poor compliance.*



Fontan Contraindications & Risk Factors

•**Systolic dysfunction:**

*Patients with poor ventricular contractility **cannot maintain adequate systemic circulation**, making them **unsuitable for Fontan physiology**.*

•**Severe Aortic Insufficiency (AI):**

*Leads to **volume overload** and **compromised systemic systolic function**, further worsening cardiac output.*

•**Elevated Pulmonary Vascular Resistance (PVR):**

*Since Fontan circulation relies on **passive pulmonary blood flow**, elevated PVR prevents efficient flow into the lungs — a **major contraindication** for the Fontan.*

Single ventricle

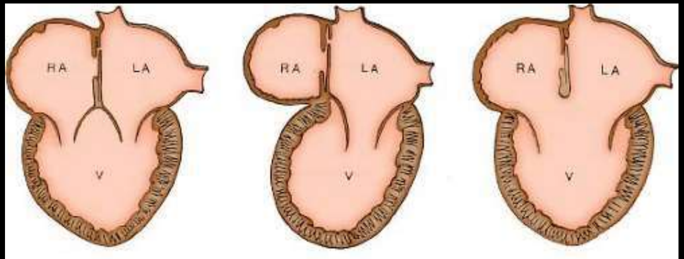
- Tricuspid atresia
- PA\IVS –monopartite
- Unbalanced AVSD
- DI LV \RV
- DORV- When a **remote VSD** can not be established a direct connection between the **aorta** and the **left ventricle.**
- Mitral atresia
- HLHS
- Large VSD

Univenctricular atrio-ventricular connection types

- **Double Inlet:** Both atria connect predominantly or exclusively to a single ventricle.
- **Single Inlet:** Only one atrium connects to the ventricle, or there is a single atrioventricular connection.

Single AV valve, single ventricle physiology

- **Common Inlet:** A single common atrioventricular valve provides inflow to one or both ventricles (AVSD).



Double inlet Single inlet Common inlet

Key Points to Evaluate in Follow-Up Echocardiography

- Is the **systemic ventricle** a **left** or **right ventricle**?

Pulmonary Outflow Tract

- **Is pulmonary flow adequate**?

Aortic Outflow Tract

- Is aortic flow adequate?

Valve Insufficiencies

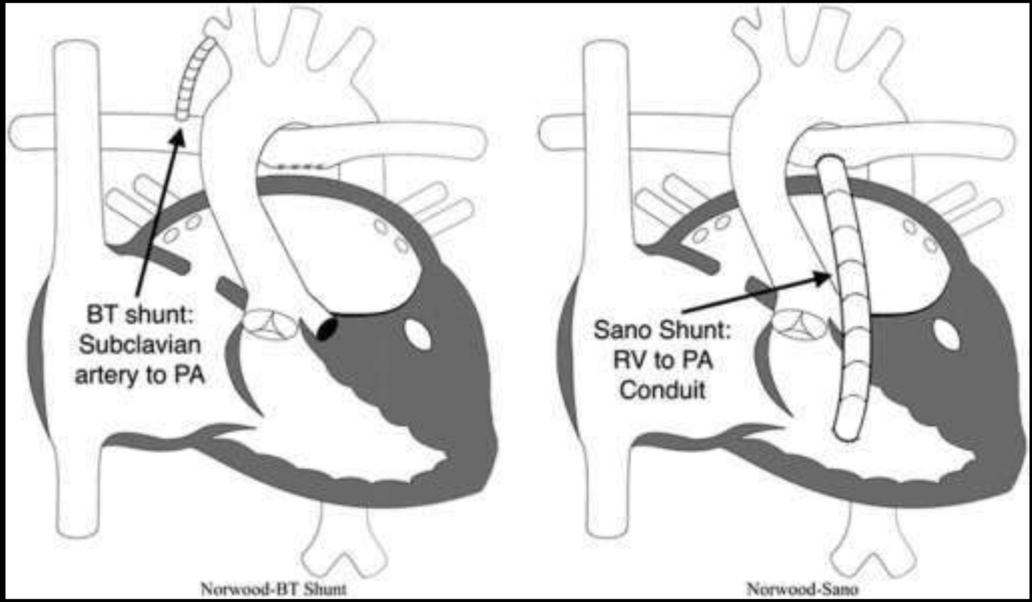
- Are there any significant **valve regurgitations**?

Systemic Ventricular Function

- Is the systemic ventricular function preserved?

Shunts evaluation

- BT shunt – *connect subclavian to PA*
- Sano Shunt – *between RV - PA*
- Glenn – *SVC is directly anastomosed to the PA*
- Fontan - *Glenn + IVC + hepatic veins to PA (no CS)*
- Kawashima (left isomerism patient) – *Anastomosis of IVC to PA (no HV and CS)*



Assessment of Shunt Flows
•**2D Echocardiography** – *for anatomical visualization*
•**Color-Flow Doppler** – *to detect flow direction and turbulence*
•**Pulsed-Wave (PW) Doppler** – *for measuring flow velocity at specific points*
•**Continuous-Wave (CW) Doppler** – *to assess high-velocity jets and pressure gradients*
•**Pressure gradient estimation across shunts or pulmonary artery bands (PAB)**

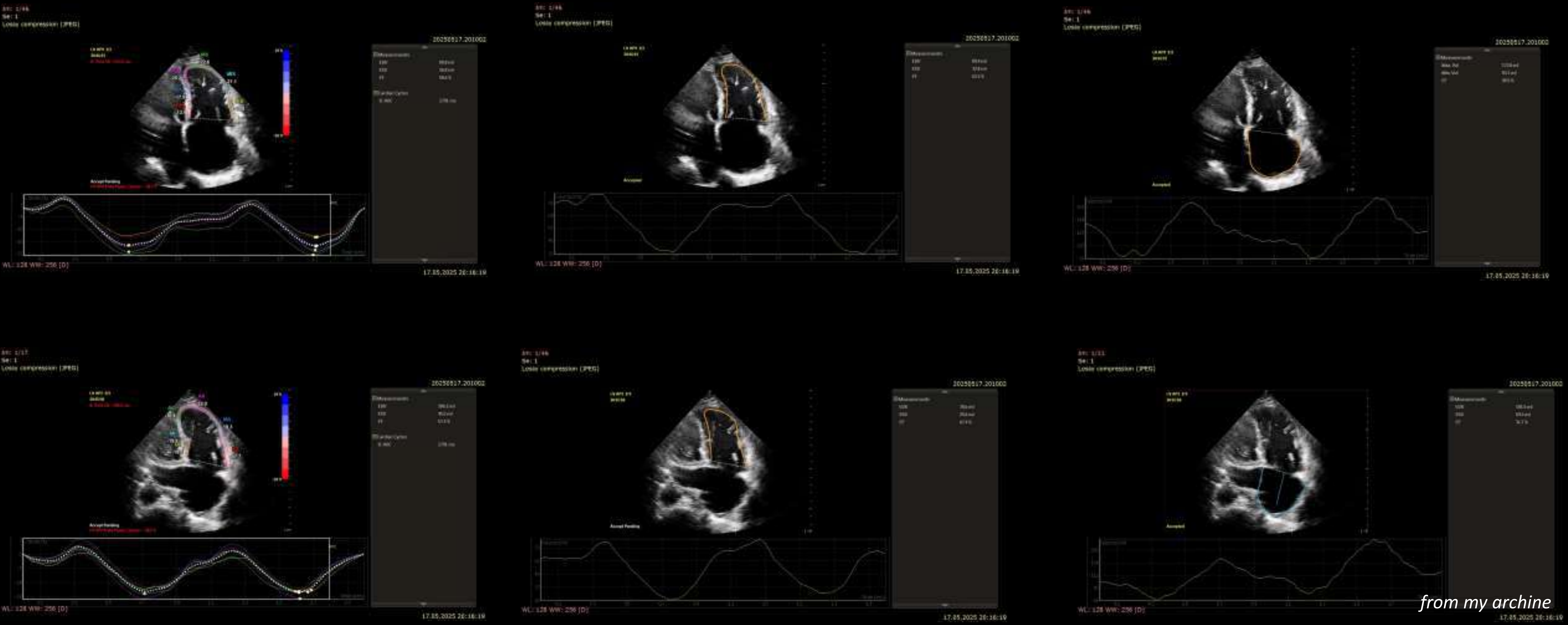
If Echocardiography Is Inadequate: Contrast enhanced CT

SVC flow

Shunt Function Assessment - The presence of diastolic reverse flow is evaluated using descending aorta (Desc Ao) Doppler.
•If diastolic reverse flow is absent, it indicates that the shunt flow is insufficient or absent.

Why Diastolic Reverse Flow?
•During **diastole**, blood normally flows forward through the aorta into the systemic circulation.
•However, if a **shunt is functioning well**, some blood will flow **backward (reverse flow)** into the pulmonary circulation via the shunt during diastole.

*Follow up -Echo controls every 6 month , CMR –every 2 years



from my archine

RV evaluation- **TAPSE** and **TDI** have **important limitations** in the assessment of overall **RV function**:

TAPSE and **TDI** primarily assess the **longitudinal motion of the lateral RV wall**.

Do not provide information about:

•The **septal** wall

•The **RV outflow tract (RVOT)**

•The **apex** or **global contractility**

*__Strain imaging__ and **volumetric assessments** (such as 3D echocardiography or cardiac MRI-gold standart) provide a **more comprehensive and reliable evaluation** of RV performance.

Thank you for your attention!