

# Echocardiography in AVSD

Antigoni Deri

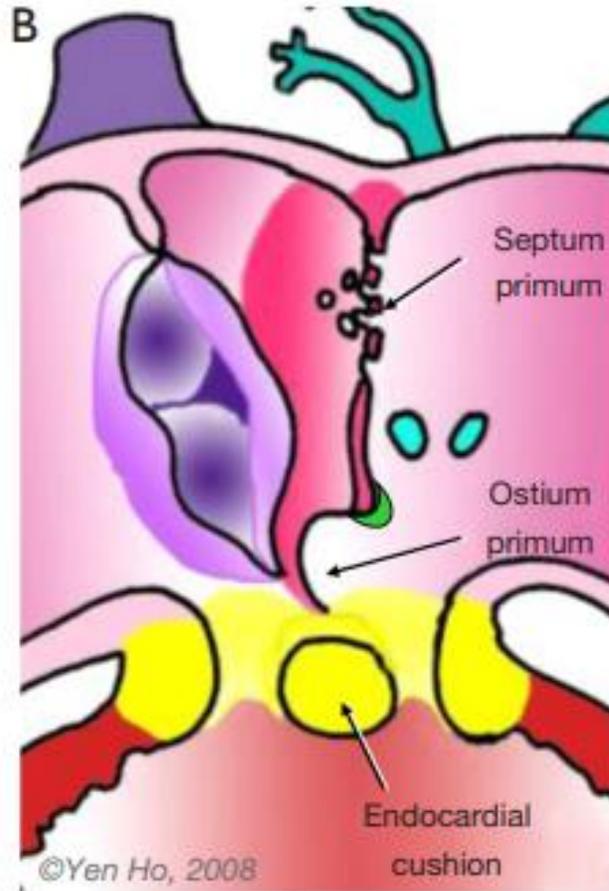
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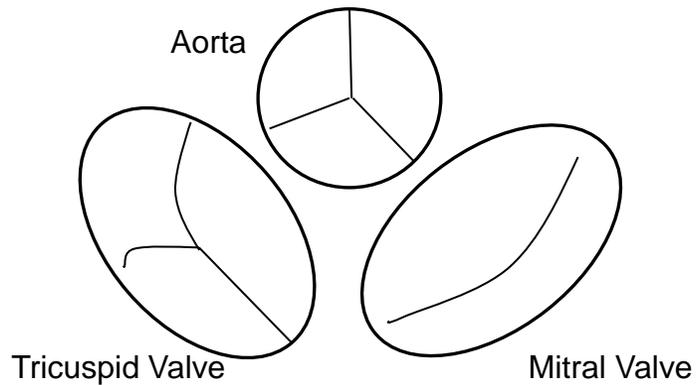
# Development



- Defect in mesenchyme formation
  - Abnormalities in development of superior and inferior endocardial cushions
  - Failure of formation of the AV septum
- Common junction/annulus
- Deficient AV septation

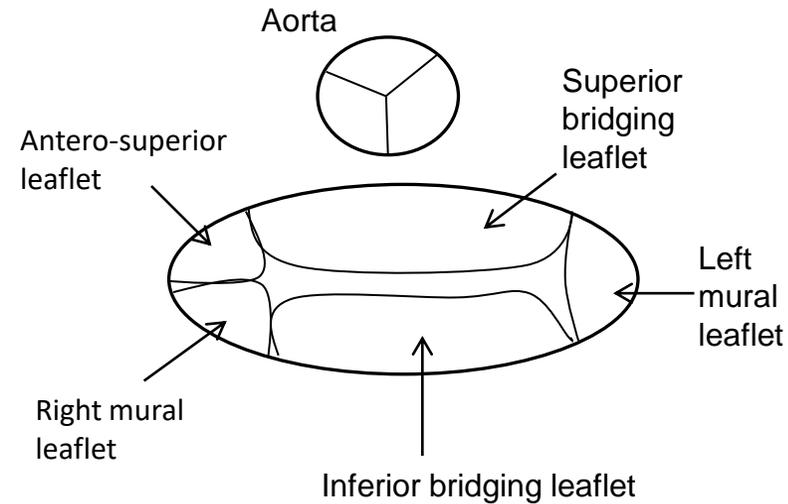
# Atrioventricular junction

## Normal Heart



- Two separate AV valves
- LVOT wedged
- Mitral valve has two leaflets

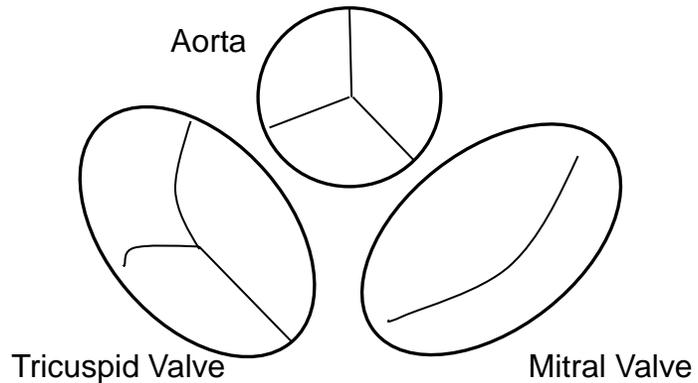
## AVSD



- Common AV annulus
- Unwedged LVOT
- The left AV valve is trileaflet

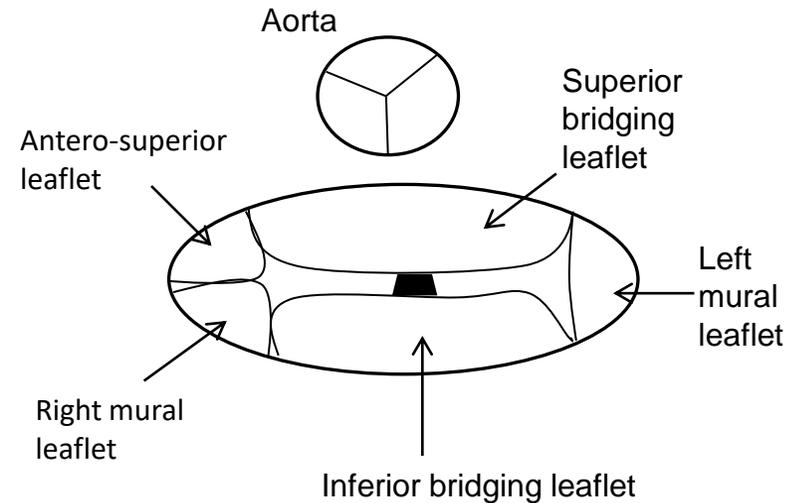
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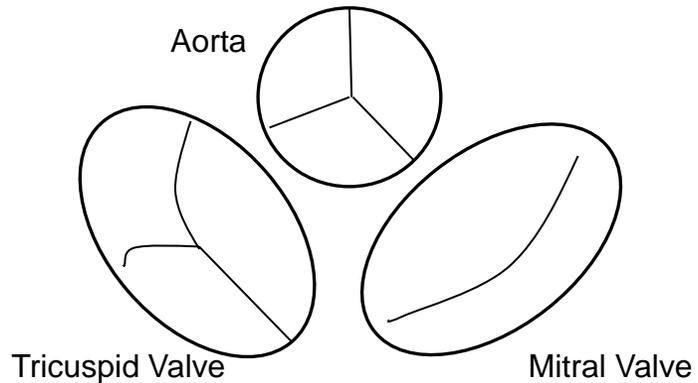
## AVSD



- Common AV annulus
- Unwedged LVOT
- The left AV valve is trileaflet
- Common or separate orifices

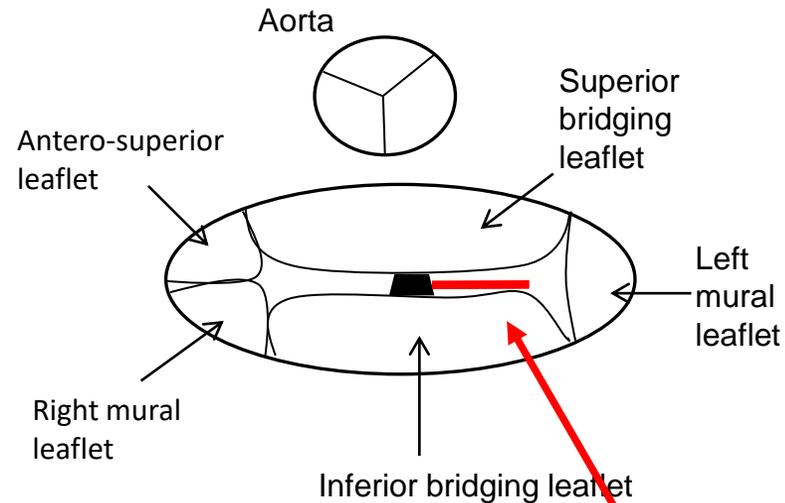
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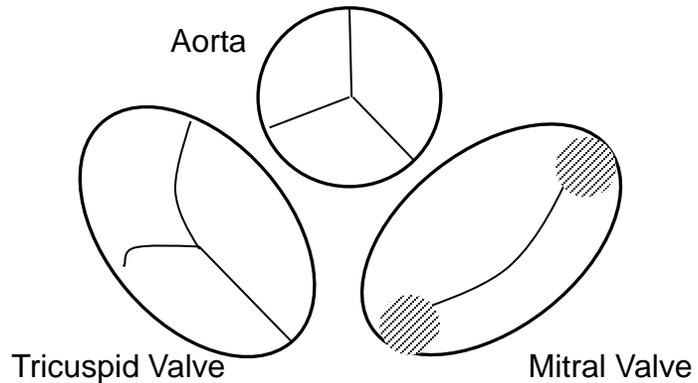
## AVSD



- Common AV annulus
- Unwedged LVOT
- The left AV valve is trileaflet
- Common or separate orifices
- Zone of apposition rather than 'cleft'

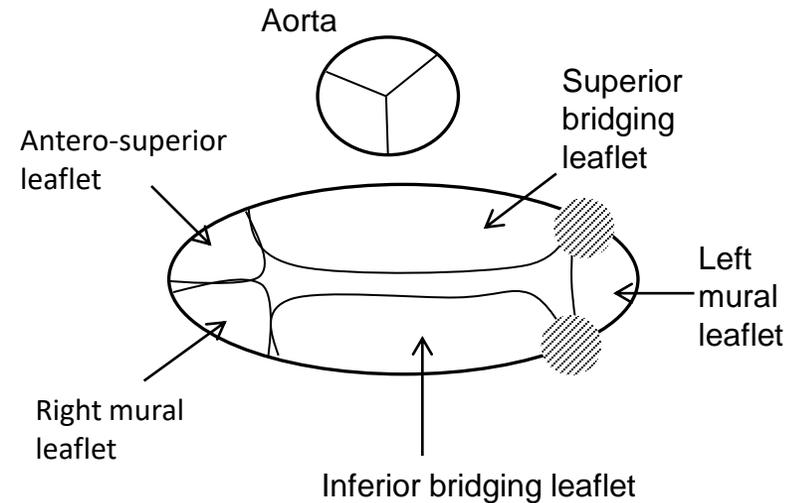
# Atrioventricular junction

## Normal Heart



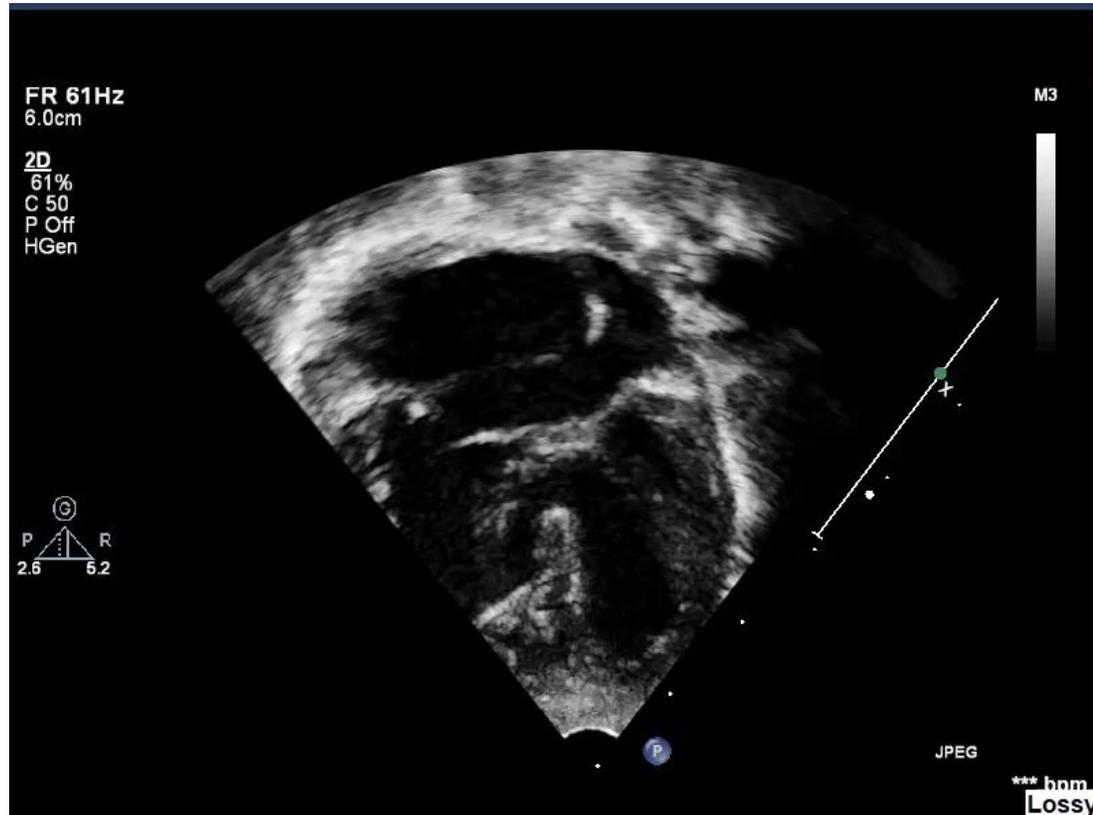
- Two separate AV valves
- LVOT wedged
- Mitral valve has two leaflets

## AVSD



- Common AV annulus
- Unwedged LVOT
- The left AV valve is trileaflet
- Common or separate orifices
- Zone of apposition rather than 'cleft'
- Supero inferior orientation of papillary muscles

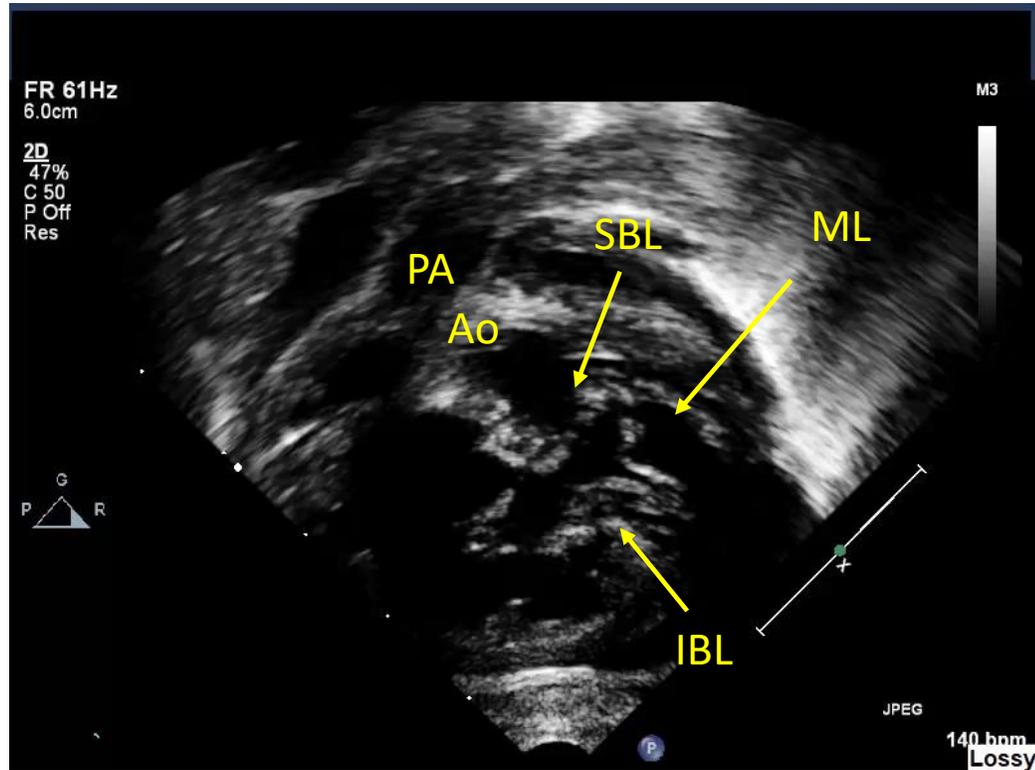
# Echocardiographic features of AVSD



4 chamber view of complete AVSD

- Lack of contiguity between the leading edge of the atrial septum and the crest of the ventricular septum
- Defect at the site of membranous AV septum
- Lack of off-setting of the right AV valve

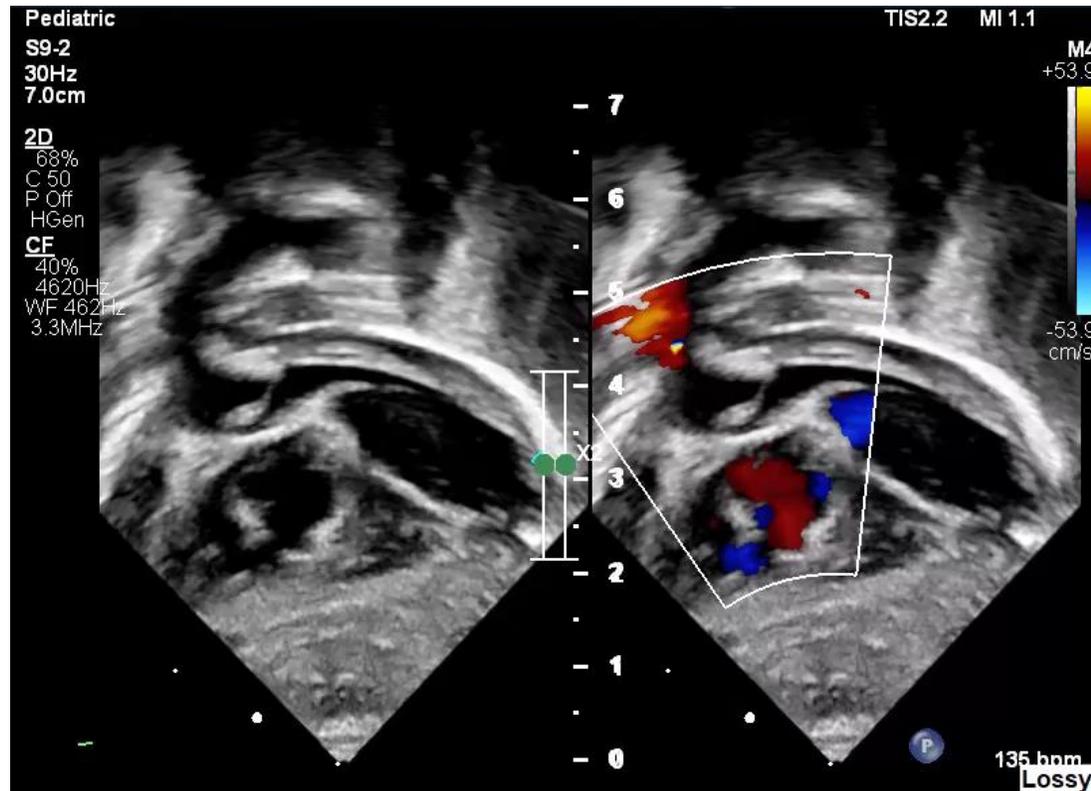
# Echocardiographic features of AVSD



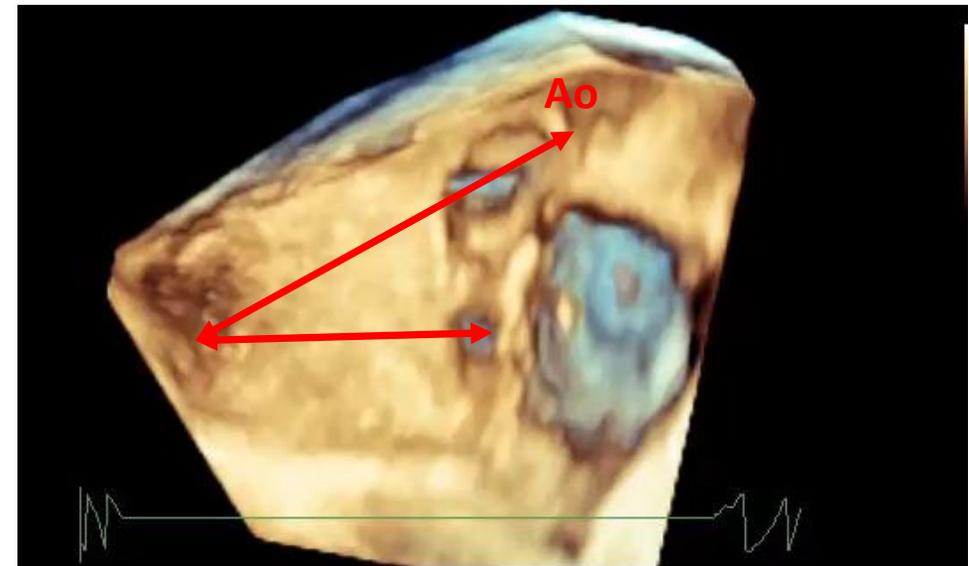
Subcostal LAO view

- Common AV junction regardless of the number of AV valvar orifices
- Aortic orifice is anterosuperior to the common junction
- No resemblance to normal MV and TV
- Five leaflets identified when valve is closed

# Echocardiographic features of AVSD



Subcostal LAO view  
Elongated LVOT (Goose-neck deformity)

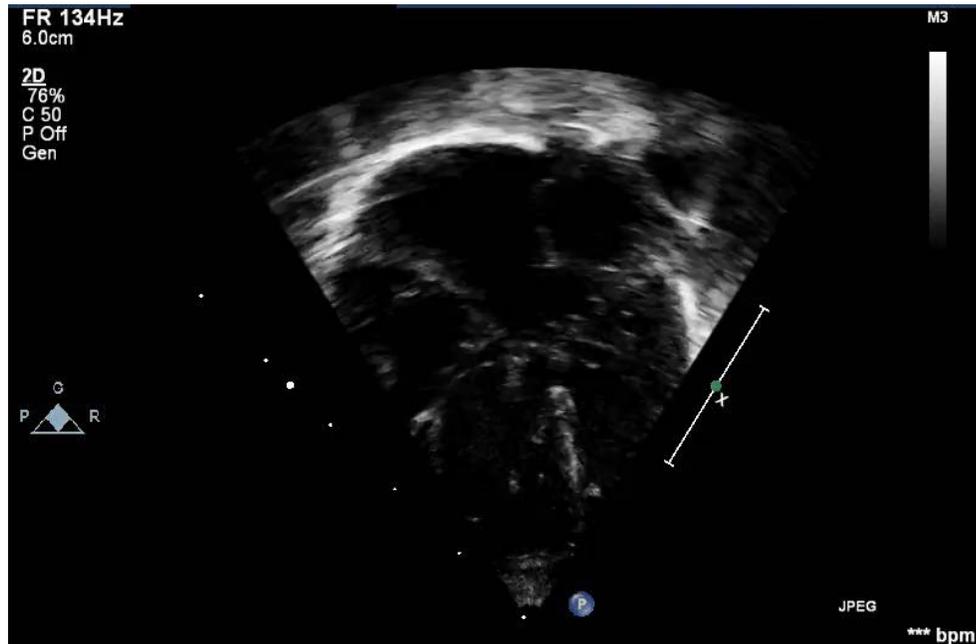


En face view of the V septum from the LV side

- Inflow < Outflow
- Substrate for LVOT obstruction, even post repair

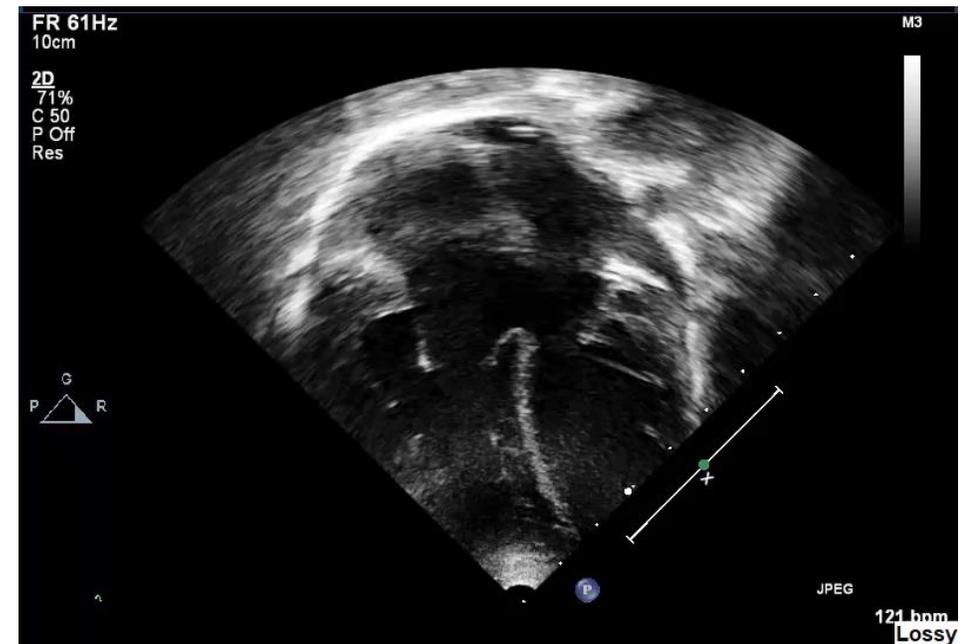
# Classification

## Complete



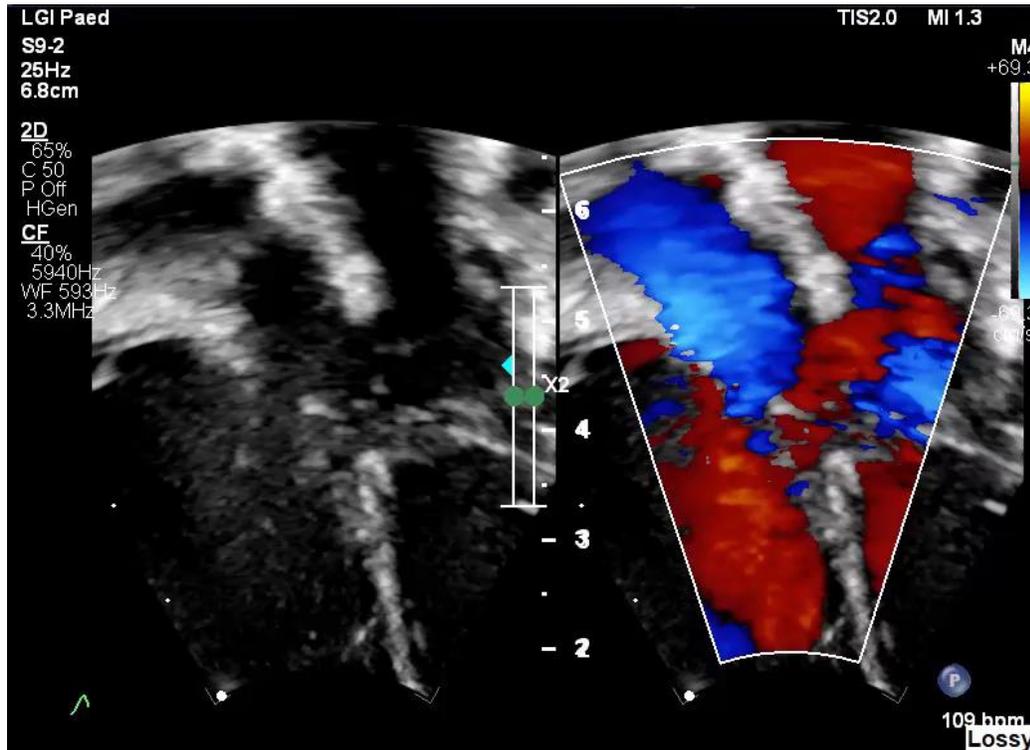
- Large ostium primum ASD and unrestrictive VSD
- Shunt at A and V level or
- Or exclusively at the ventricular level
- Single or two orifices
- Early repair

## Partial (Ostium primum ASD)



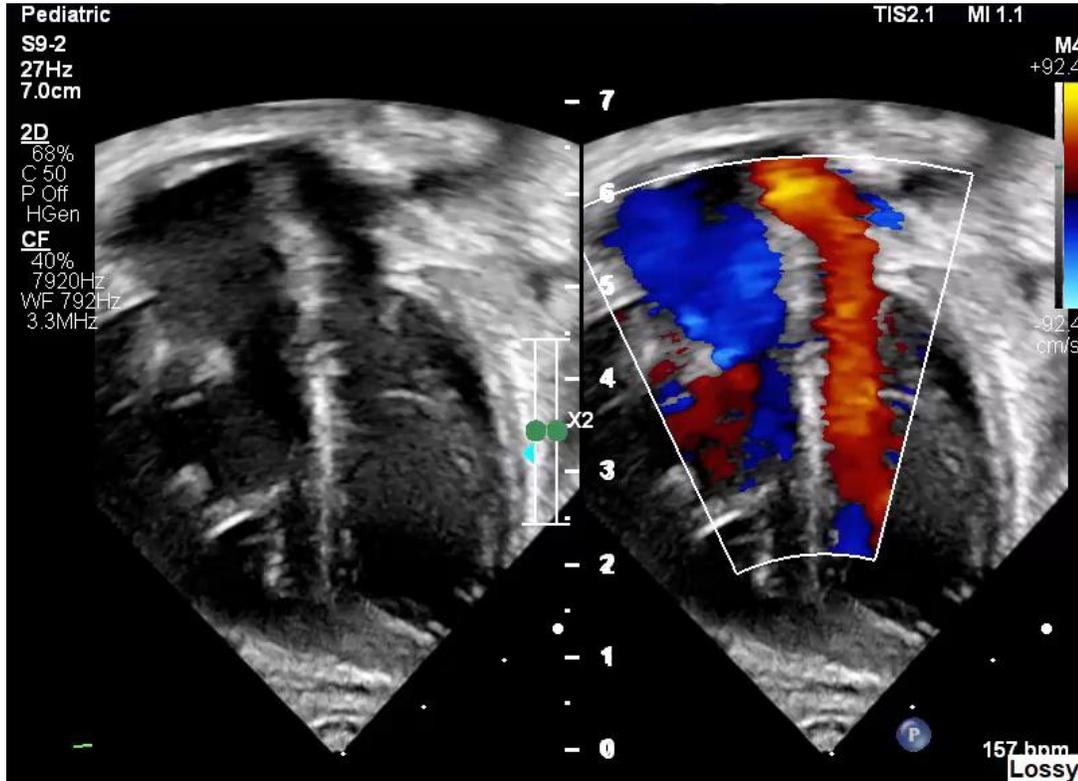
- AV valve adherent to the ventricular septum
- Annulus divided into two orifices
- ASD physiology of ASD

# Ostium primum ASD, restrictive VSD (‘transitional’)



- Chordal attachments to the crest of the septum
- Aneurysmal tissue
- Restrictive VSD
- Timing of repair depends on VSD size

# AVSD with no atrial component

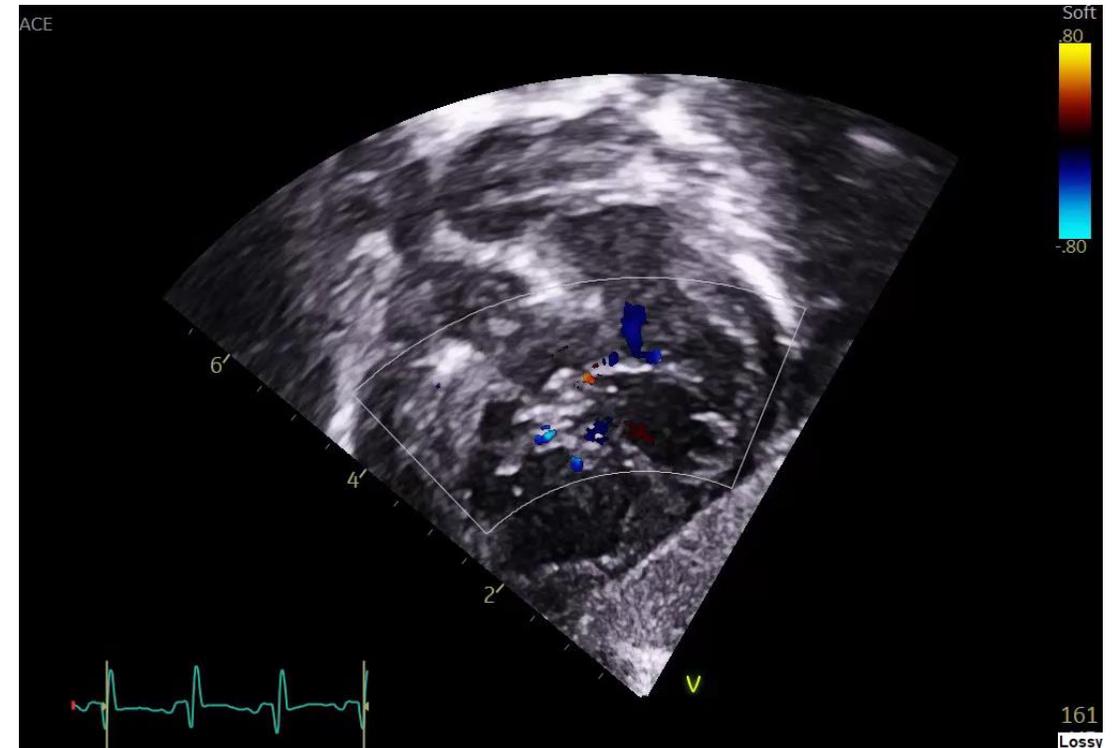
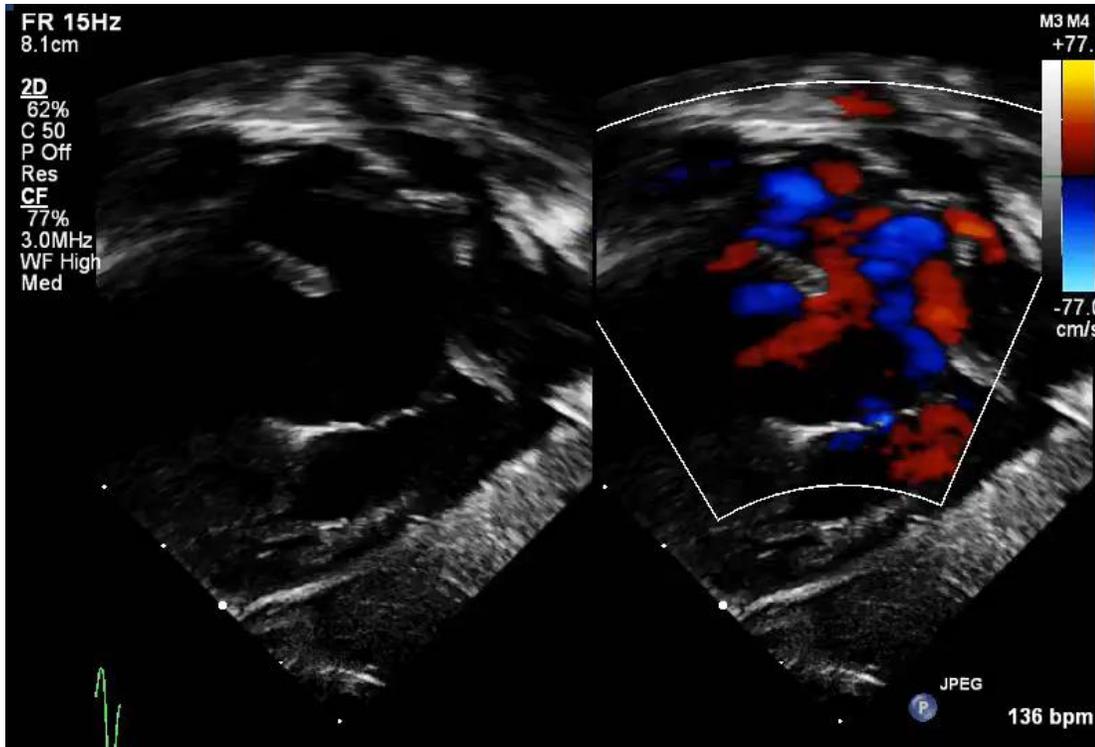


- The valve is adherent to the atrial septum
- Only shunt at the ventricular level
- Distinguish from inlet VSD: Common annulus and trifoliate left AV valve

# Echocardiographic assessment

- Sequential segmental analysis
  - Subcostal
  - Apical
  - Parasternal
  - Suprasternal views
- 
- Look for additional lesions
  - Complement with TOE and 3D

# Subcostal views



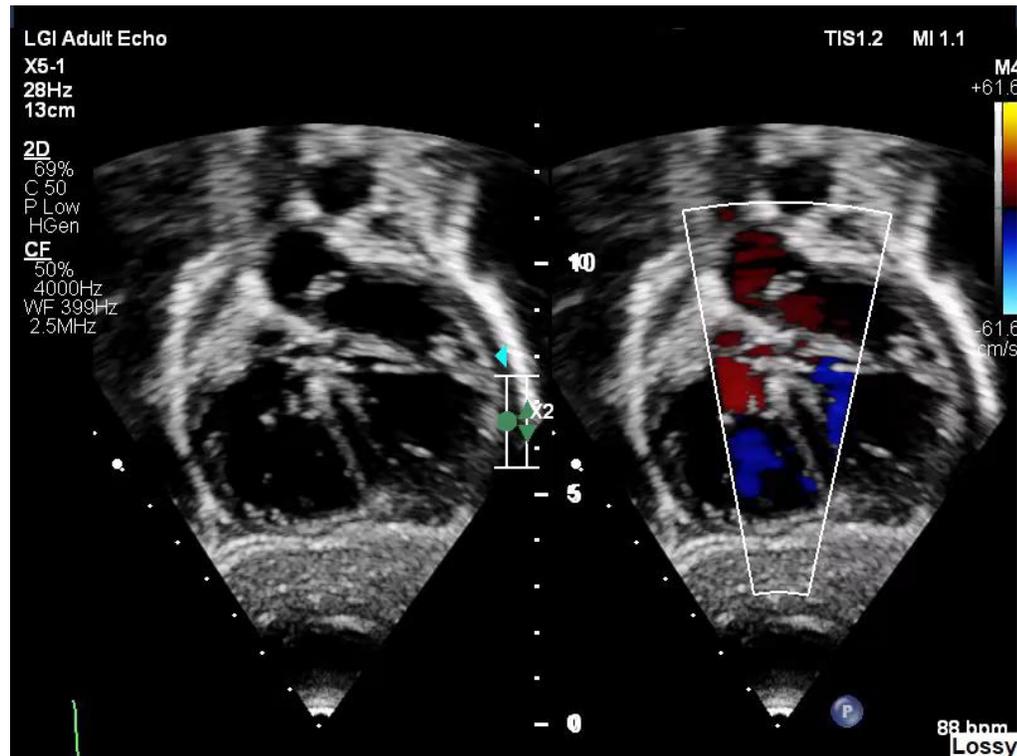
Assessment of atrial septum, additional ASDs

The size of VSD might be difficult to assess

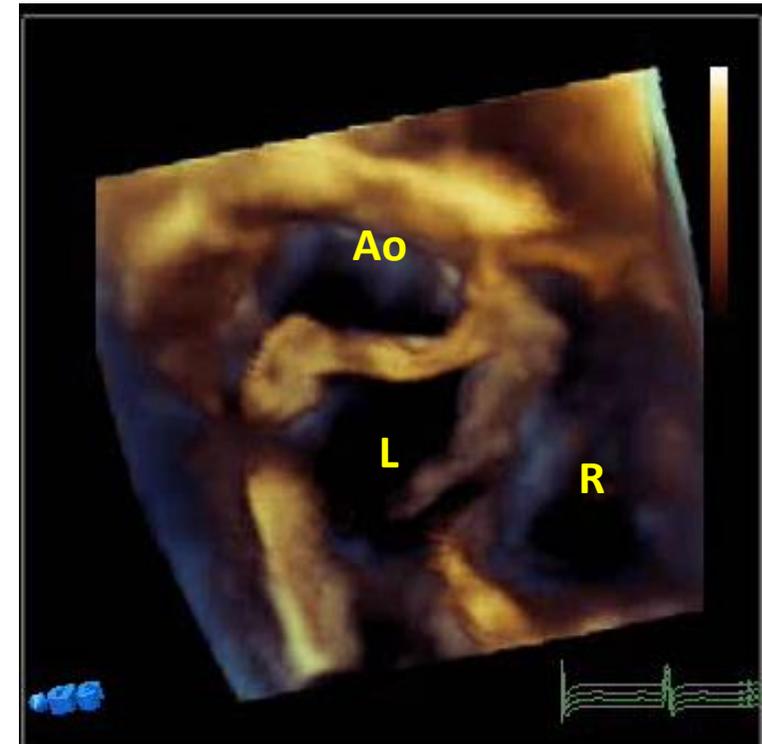
Common AV valve: number of orifices, mural leaflet, origin of regurgitation, balance LVOT

# Partial AVSD

Subcostal LAO view



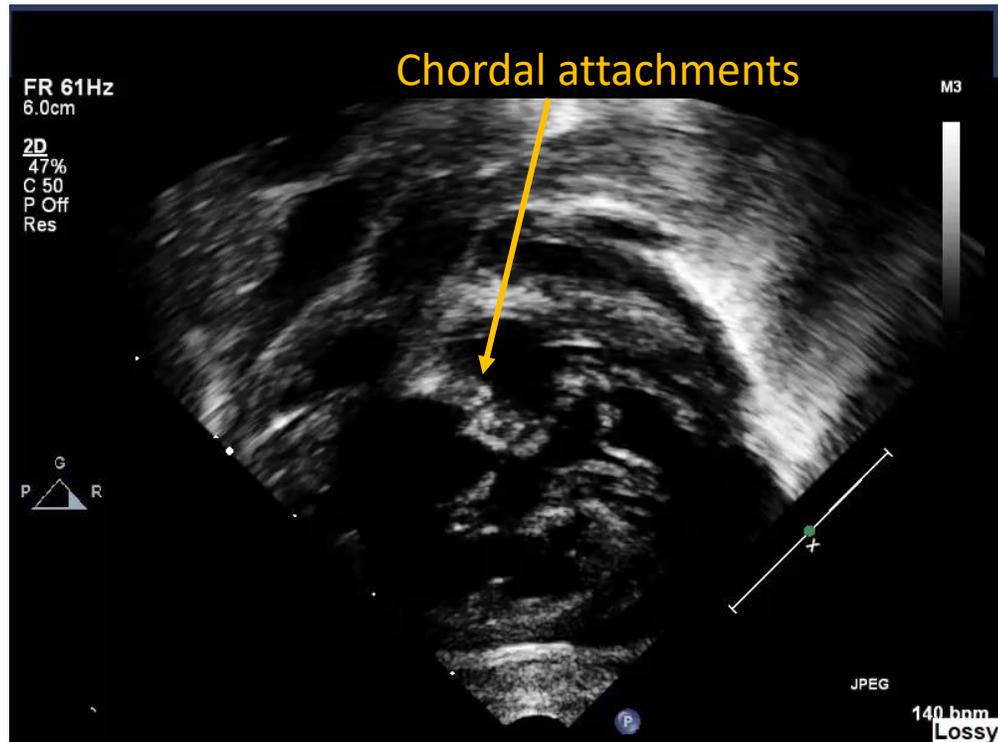
3D view from atrial side



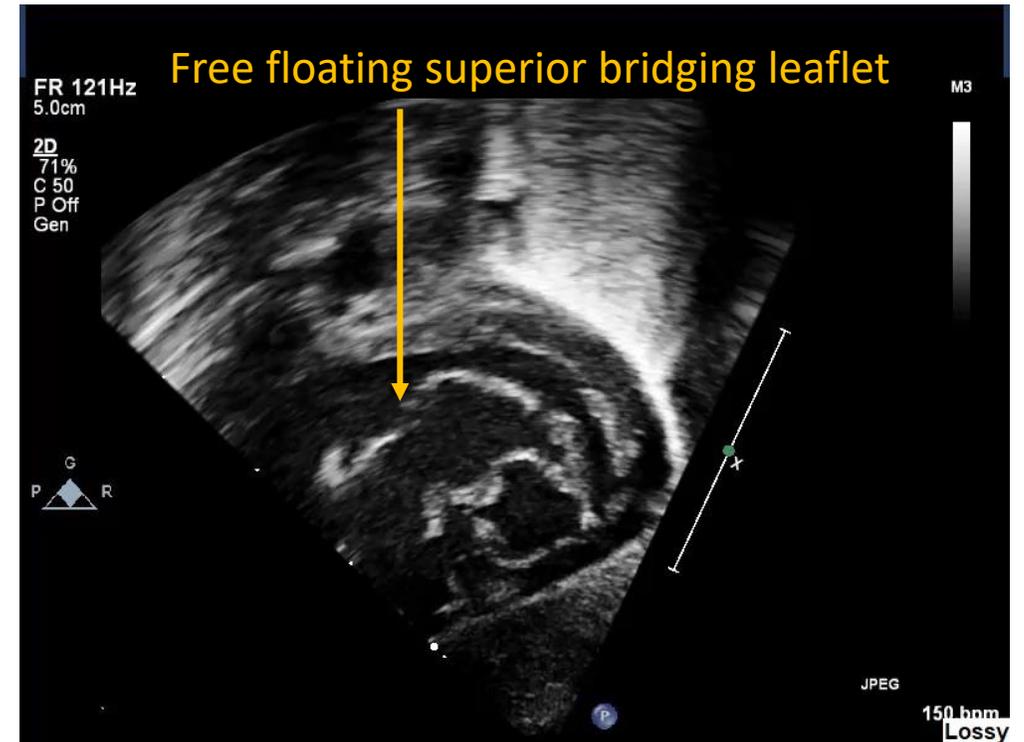
Trifoliate left AV valve – superior bridging leaflet perpendicular to the ventricular septum  
Separate orifices

# Rastelli classification

(commitment of the SBL to the RV)

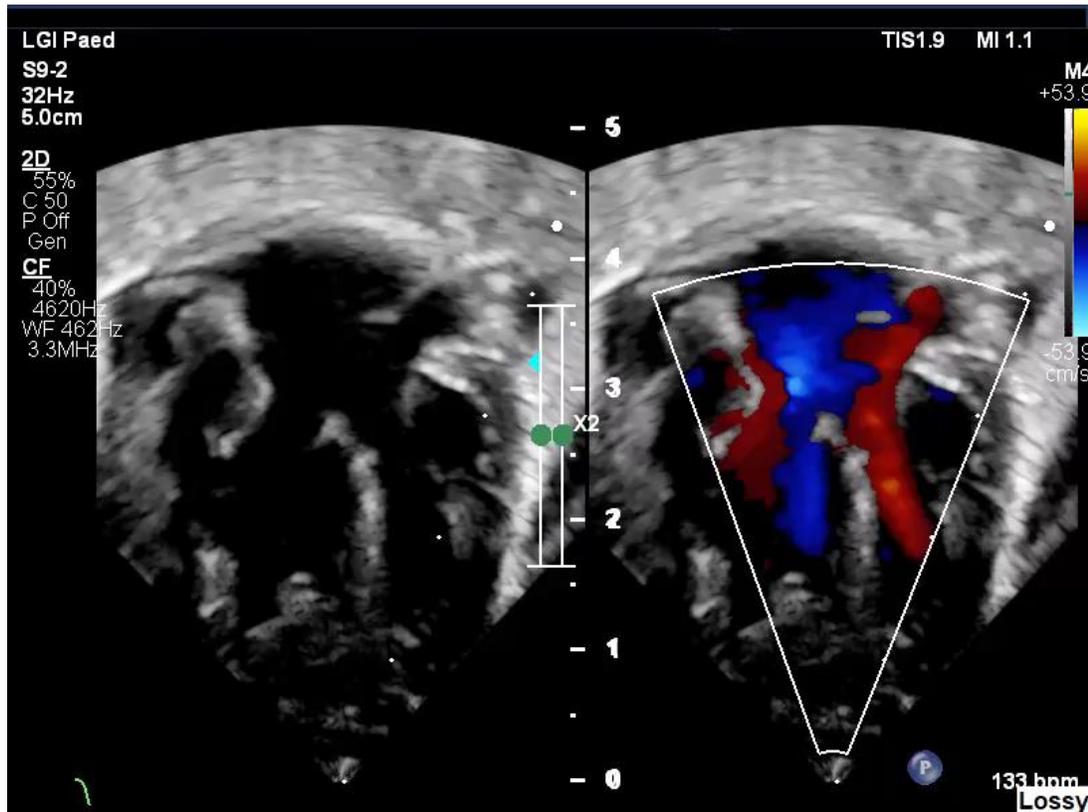


**Rastelli type A**  
Minimal bridging  
More vulnerable to LVOTO



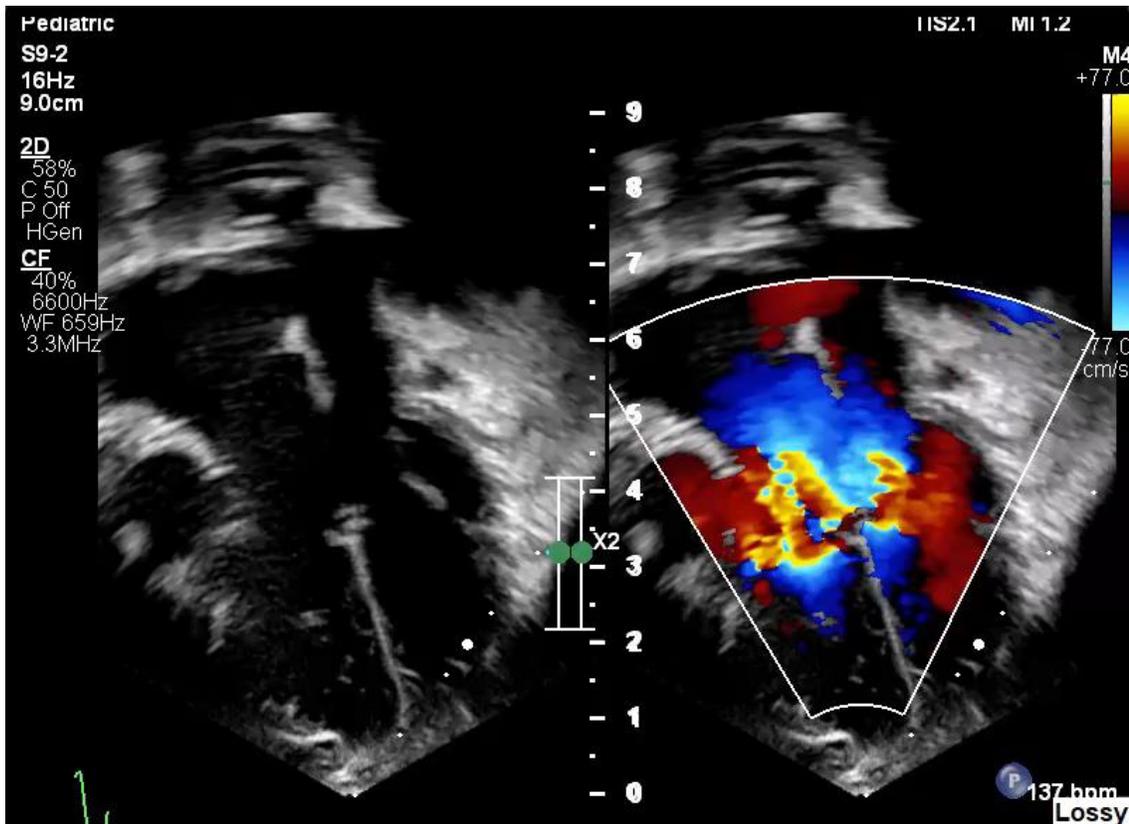
**Rastelli type C**  
Extreme bridging

# Apical



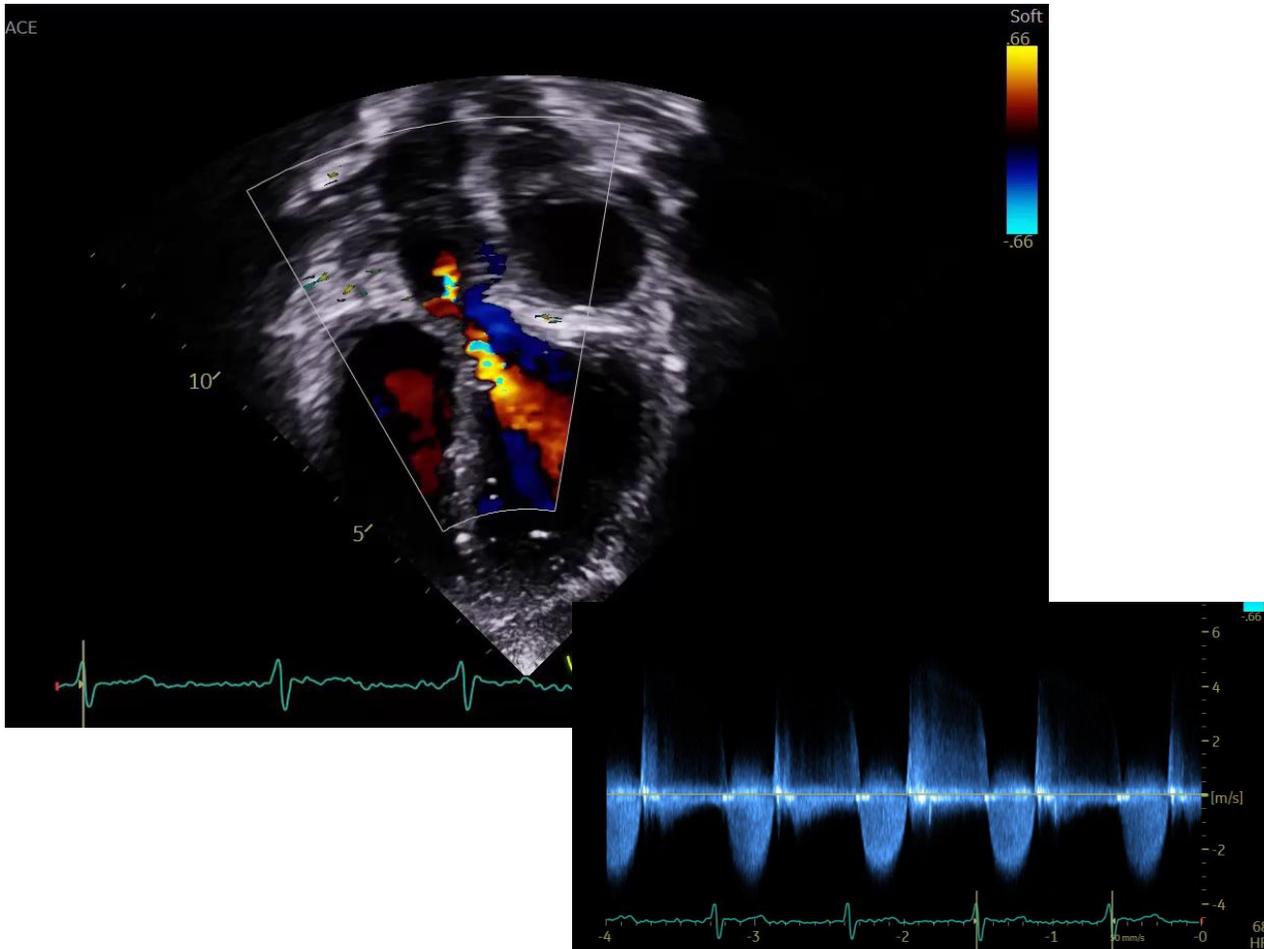
- Relationship of the AV valve in the AVSD
- Size of atrial and ventricular components
- Ventricular size and function

# Apical



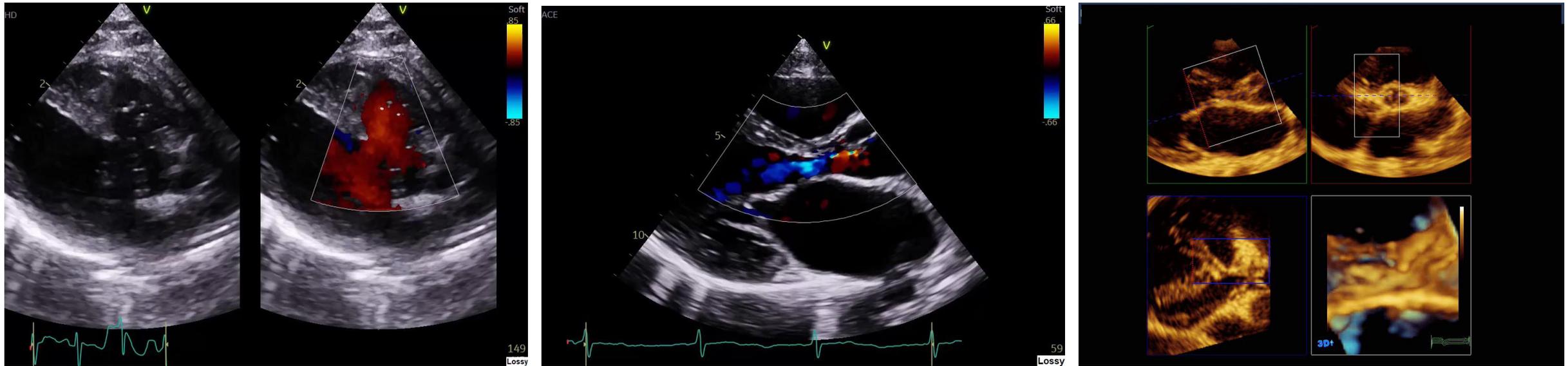
- Relationship of the AV valve in the AVSD
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- Degree and location of valve regurgitation

# Apical



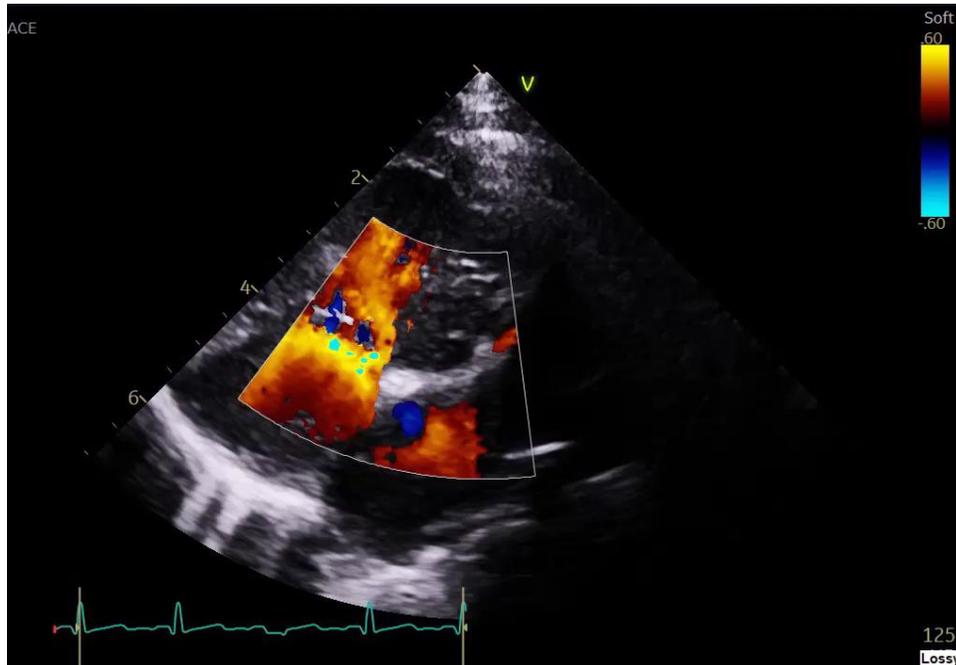
- Relationship of the AV valve in the AVSD
- Size of atrial and ventricular component
- Ventricular size and function
- Degree and location of valve regurgitation
- Assessment of LVOT

# Parasternal Long Axis View



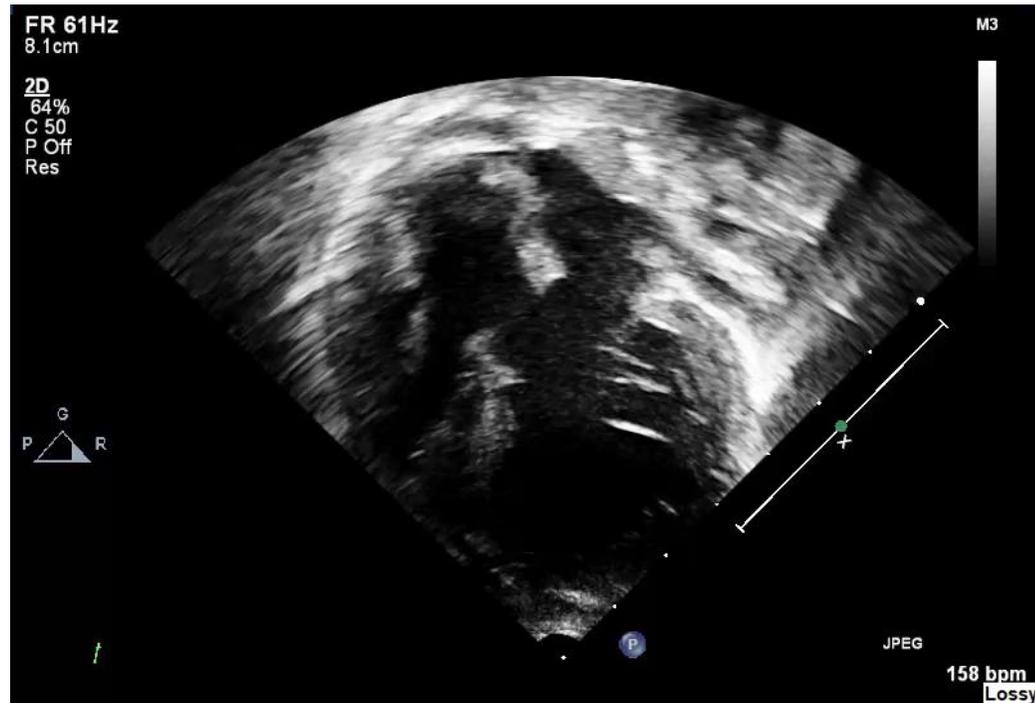
- VSD
- LVOT – elongation more prominent in separate R and L AV valves
- Mechanism of LVOTO

# Parasternal short axis view



- Assessment of ASD and VSD, additional VSDs
- Assessment of Left AV valve: trifoliate appearance, papillary muscles, origin of jets of regurgitation
- Septal flattening

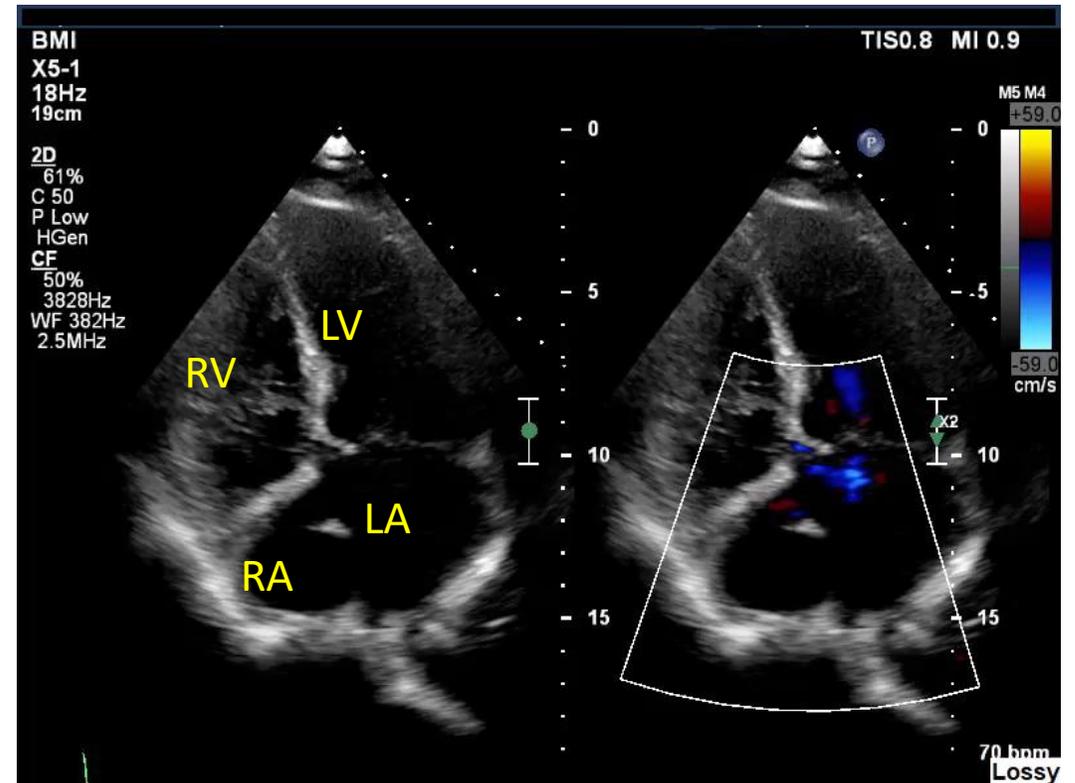
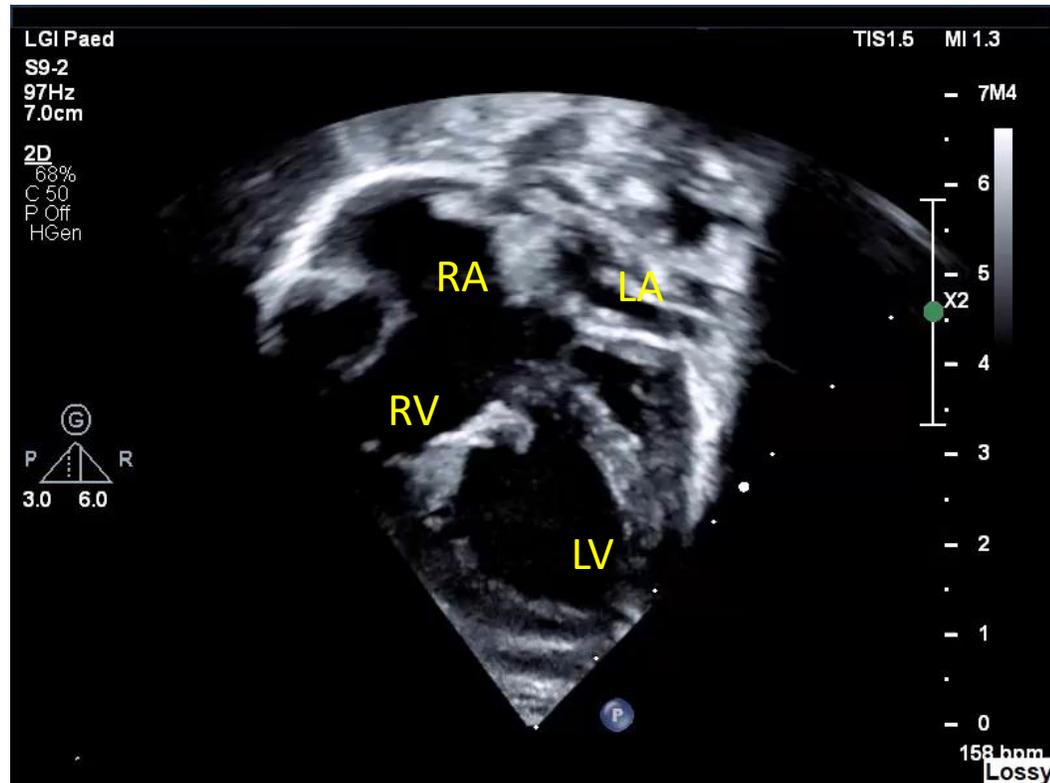
# Dominance of chambers



Balanced at ventricular level

AV valve equally distributed over well developed ventricles

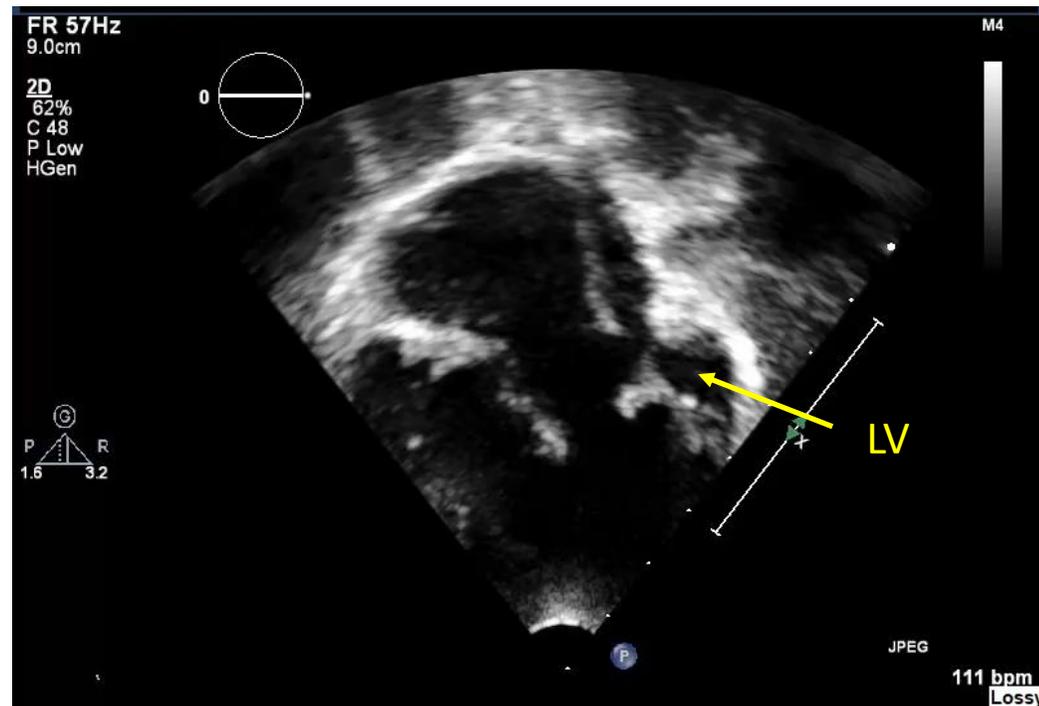
# Unbalanced at atrial level



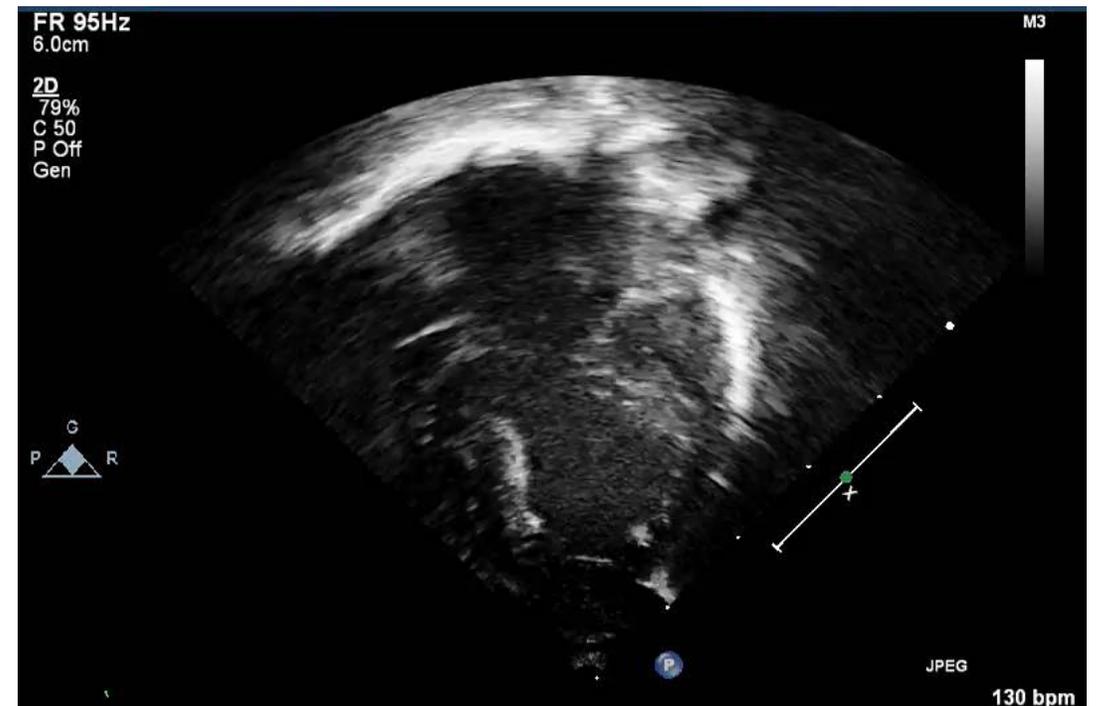
- Malalignment of atrial septum in relation to the ventricular septum
- Double outlet atrium
- Opposite ventricle might be hypoplastic

# Unbalance at ventricular level

**RV dominance**

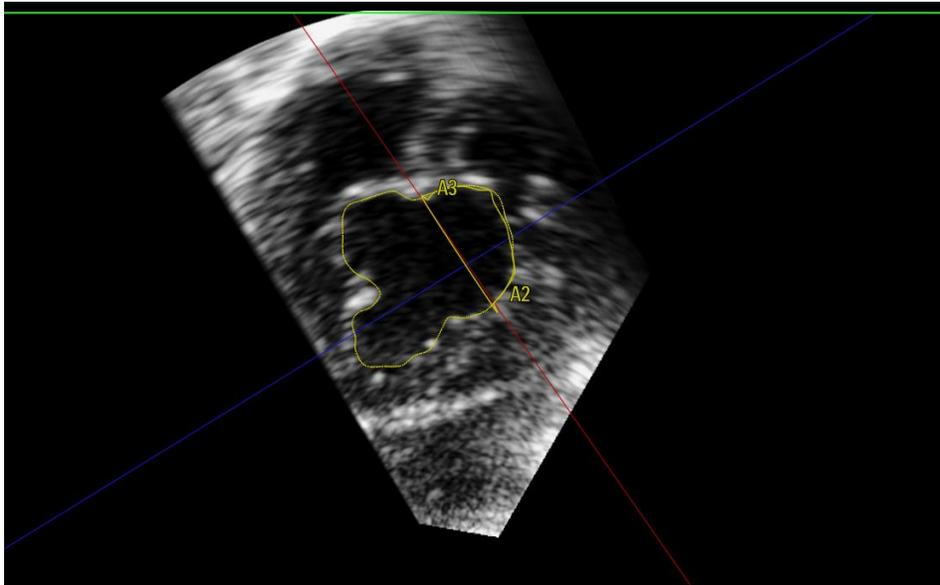


**LV dominance**



AV valve opens predominantly to one ventricle  
Hypoplasia of contralateral ventricle and structures  
Can be challenging to septate

# Modified AV valve index



area of LAVV/area of total AVV

0.4-0.6: balanced AVSD

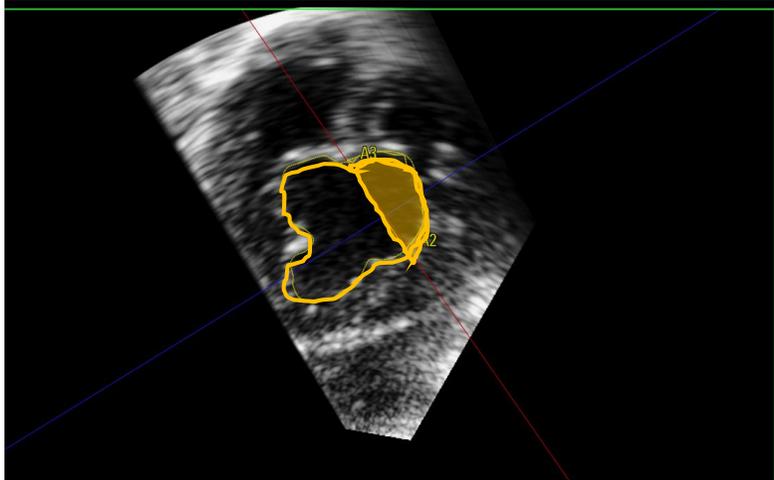
$\leq 0.4$ : Right dominance

heterogeneity of surgical strategy

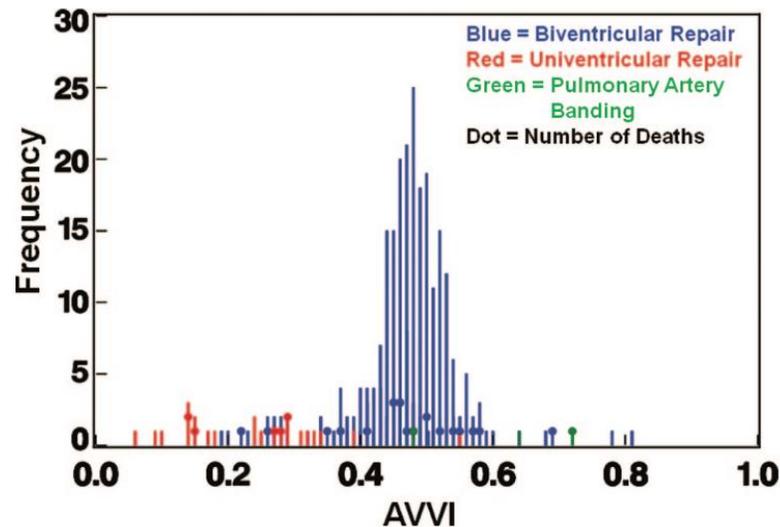
increased surgical risk

$\geq 0.6$ : Left dominance

# Modified AV valve index



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0.4-0.6: balanced AVSD

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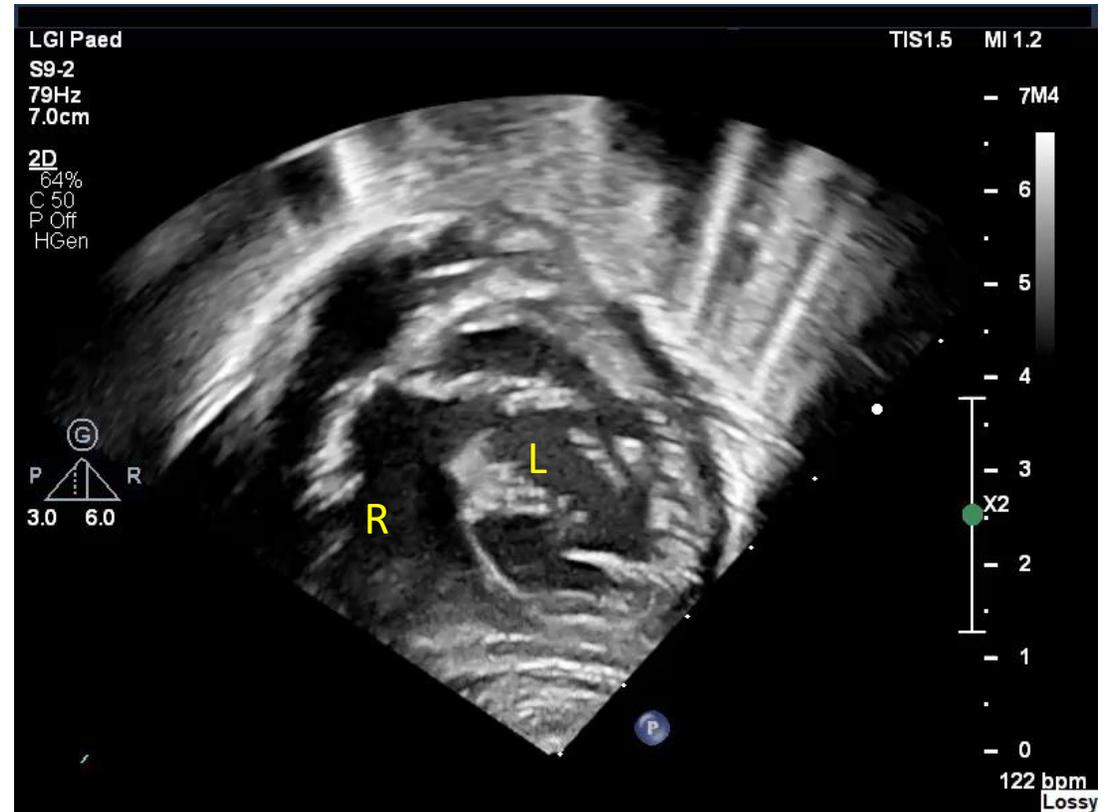
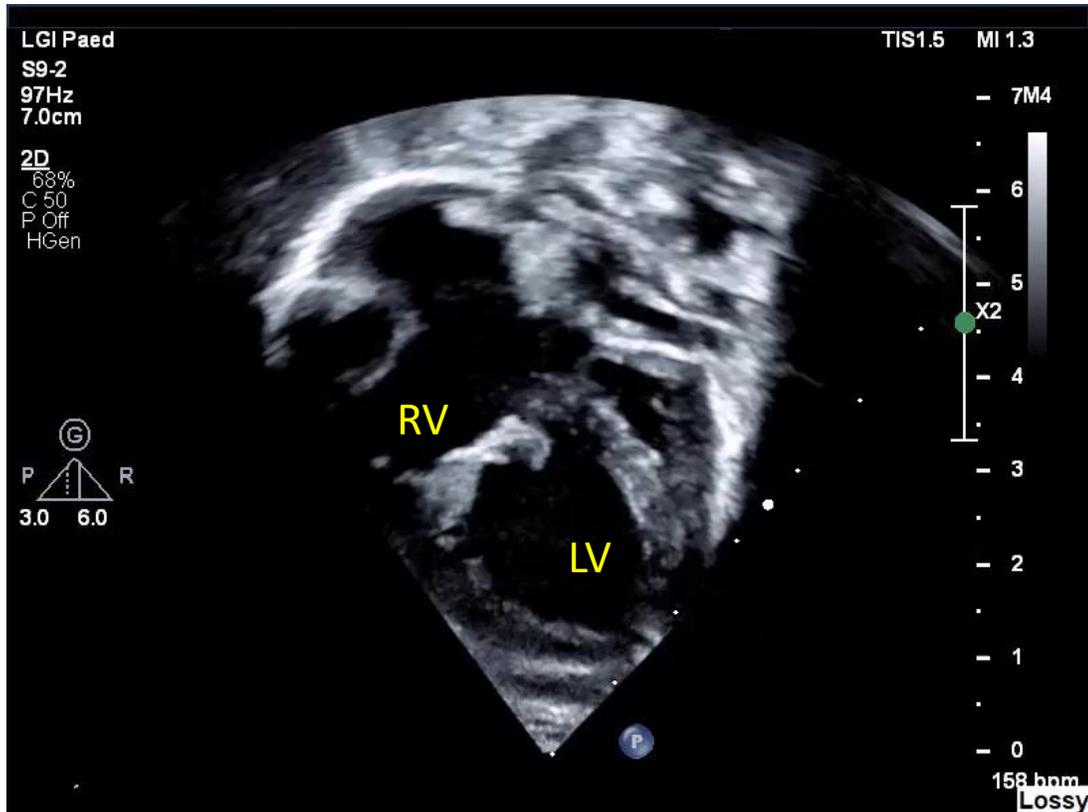
heterogeneity of surgical strategy

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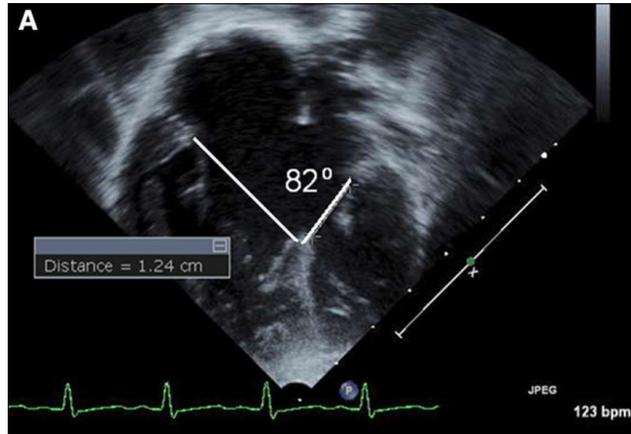
*Jegatheeswaran A et al. Circulation. 2010*

# AVVi might not be enough



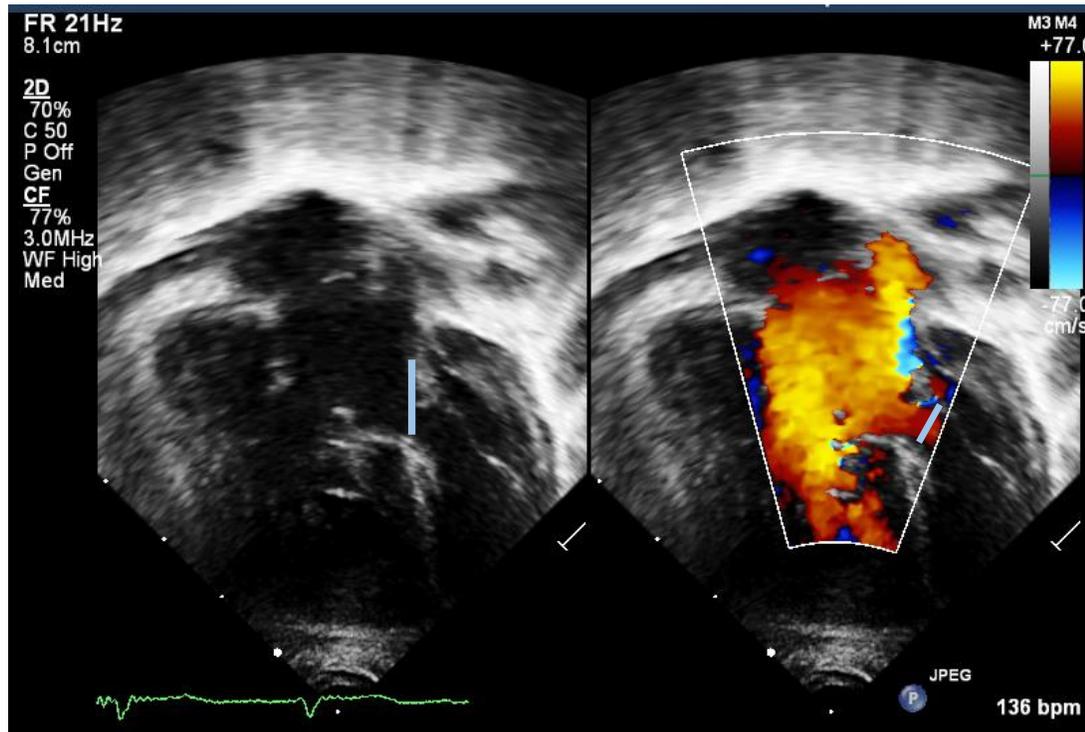
A favourable AVVi might not correlate with the ventricular volumes

# RV/LV inflow angle



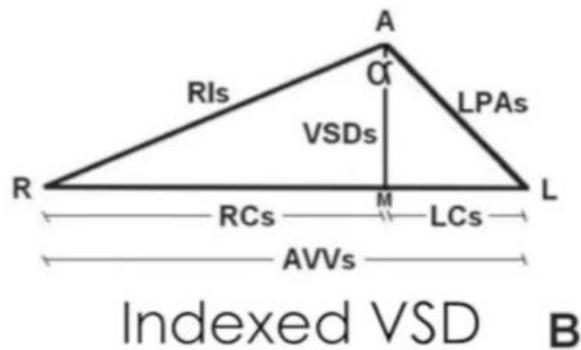
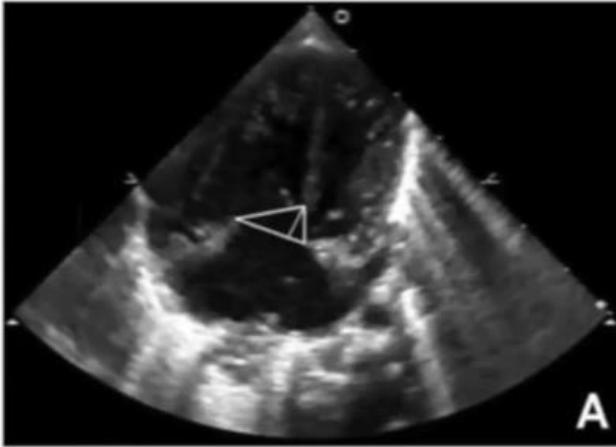
- The angle between the base of the RV and LV free wall using the crest of the ventricular septum as the apex of the angle
- Degree of LA override over the right AV valve also a significant parameter

# LV inflow index (LVII)



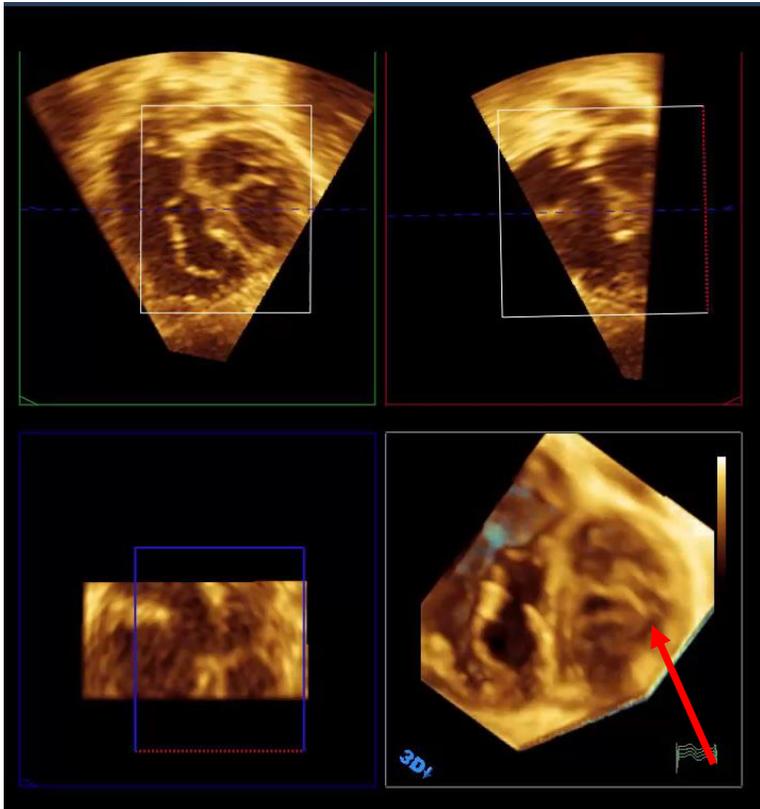
- Secondary LV inflow/2D LAVV annulus
- In cases of mild/mod LV hypoplasia, a greater LVII predicted survival after biV repair in patients with R dominant unbalanced AVSD.
- No patient with  $LVII < 0.5$  survived

# Indexed VSD

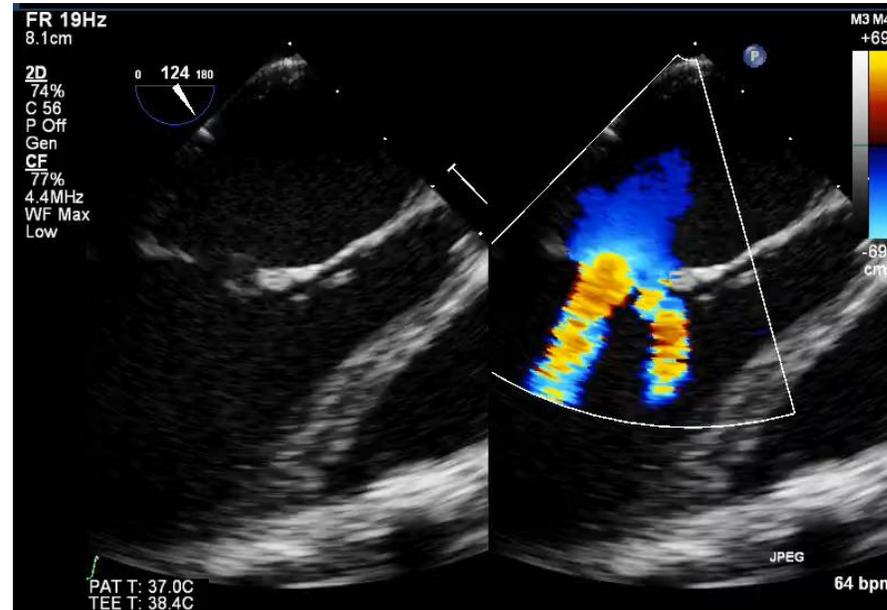


- Combined use of iVSD and AVVi helps clarify surgical decision-making
- For iVSD  $< 0.2$ , biV repair may be recommended
- For  $> 0.5$ , uniV palliation might be a reasonable strategy
- Smaller defects are more likely to survive biV repair

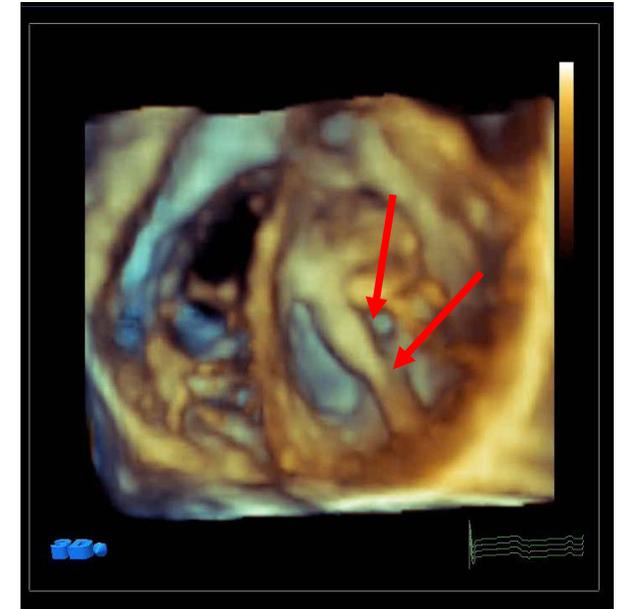
# Abnormalities of the left AV valve



Partial AVSD. MPR and 3D demonstrate hypoplastic mural leaflet

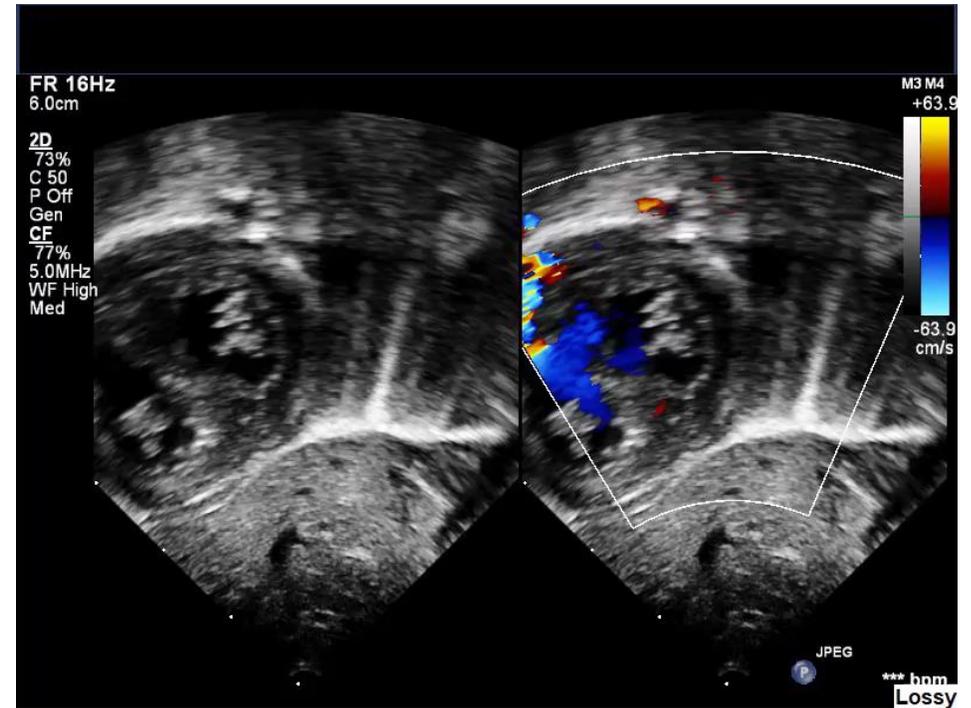
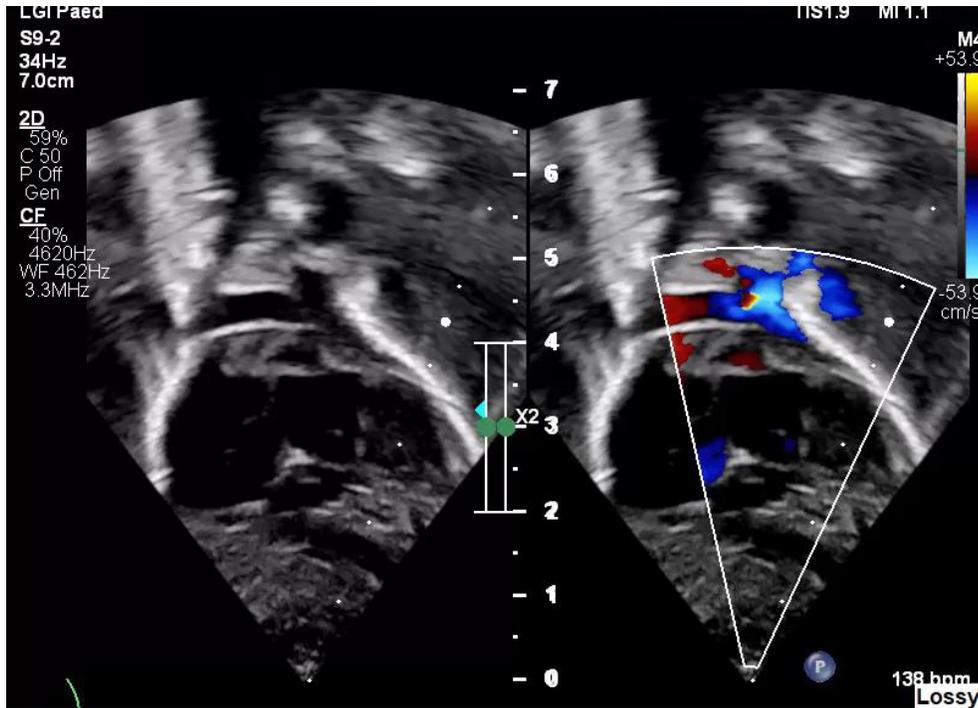


Partial AVSD. Double orifice left AV valve



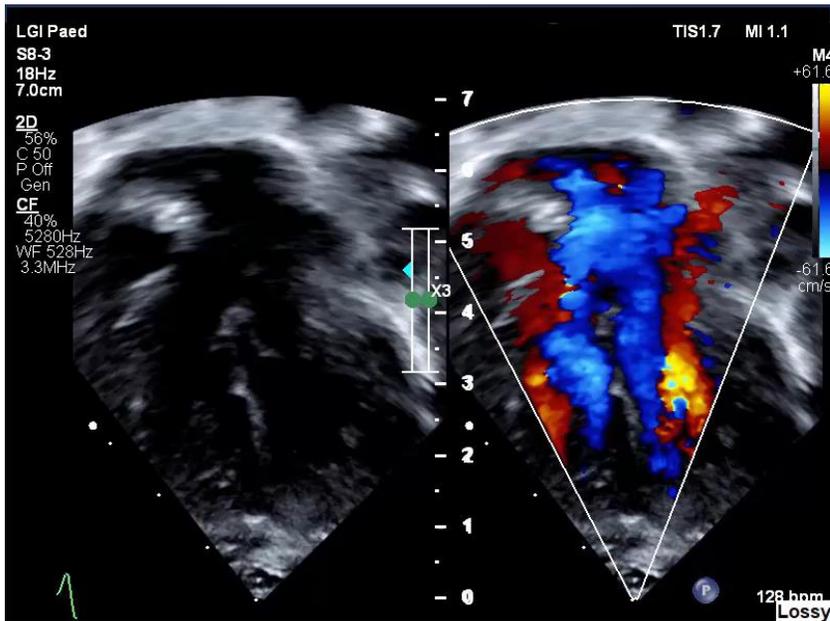
- Hypoplastic/absent mural leaflet
- Double orifice LAVV
- Abnormal pap muscles (fused/hypoplastic/single)

# AVSD + Tetralogy of Fallot



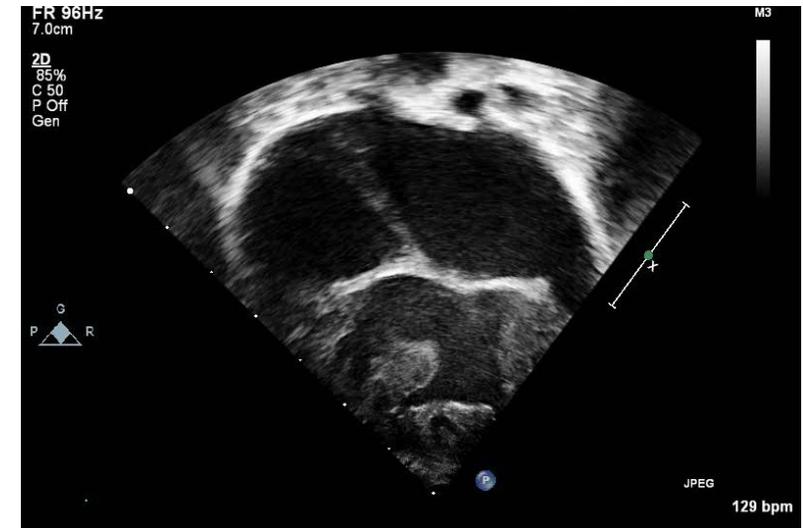
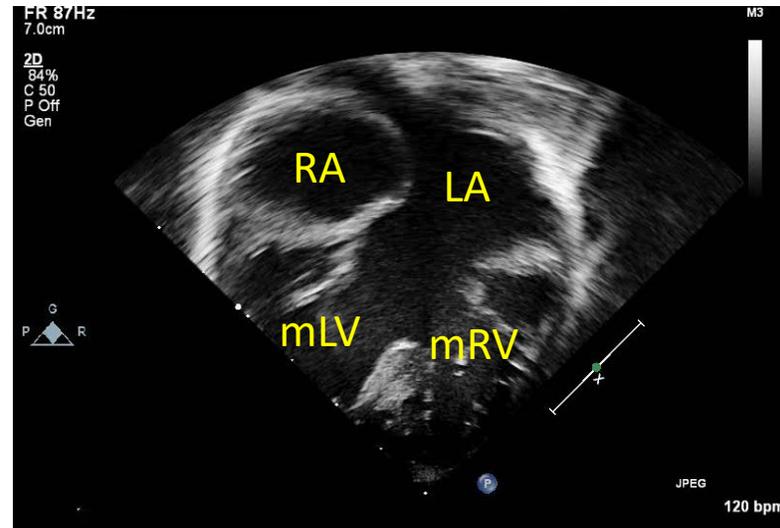
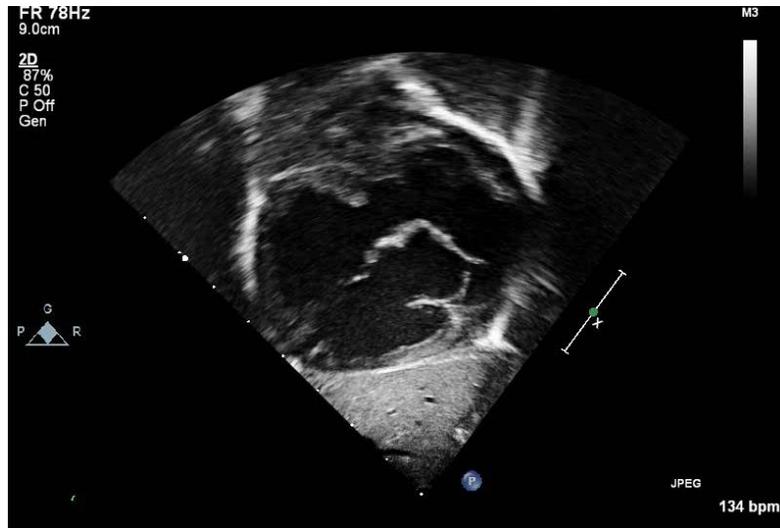
Subcostal views: AVSD and anterior deviation of the outlet septum with RVOTO

# AVSD and Isomerism



Patient with left atrial isomerism, small left AV valve and coarctation of the aorta

# AVSD in complex anatomy

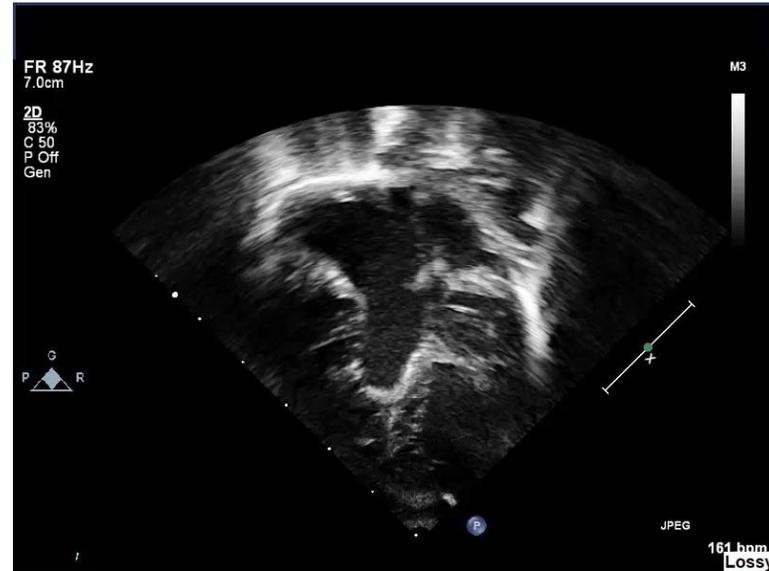


Dextrocardia, AVSD, Double outlet LA, DORV, malposed great arteries

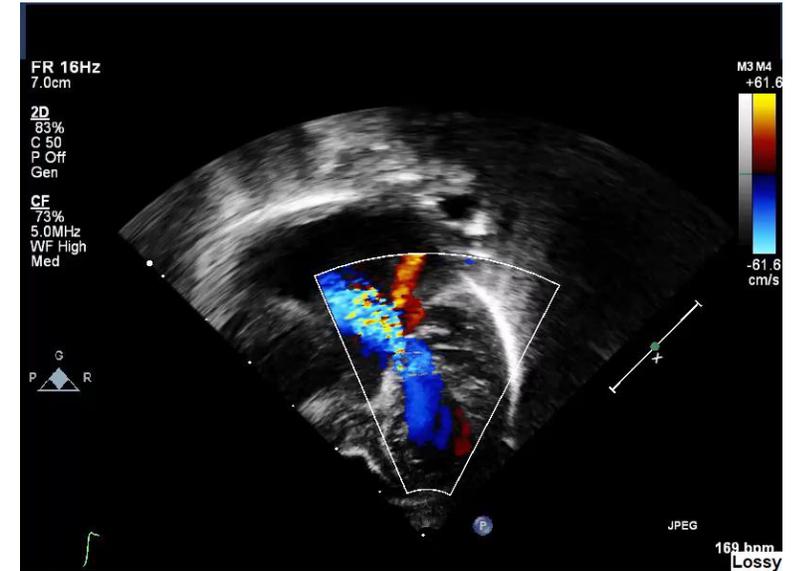
# Ebstein malformation in AVSD



Subcostal LAO view

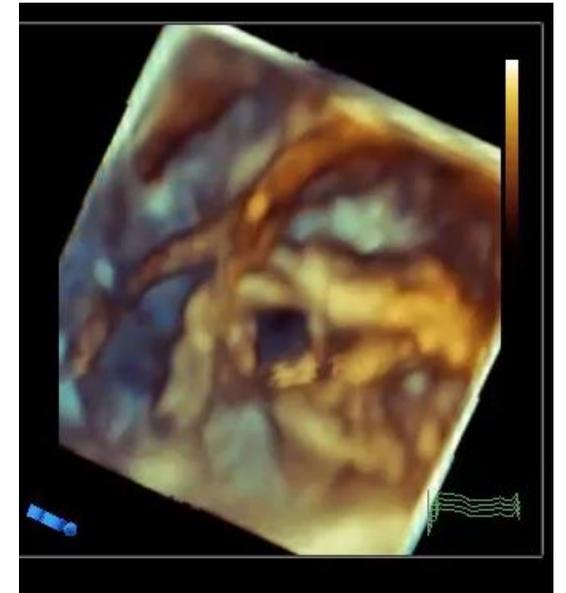
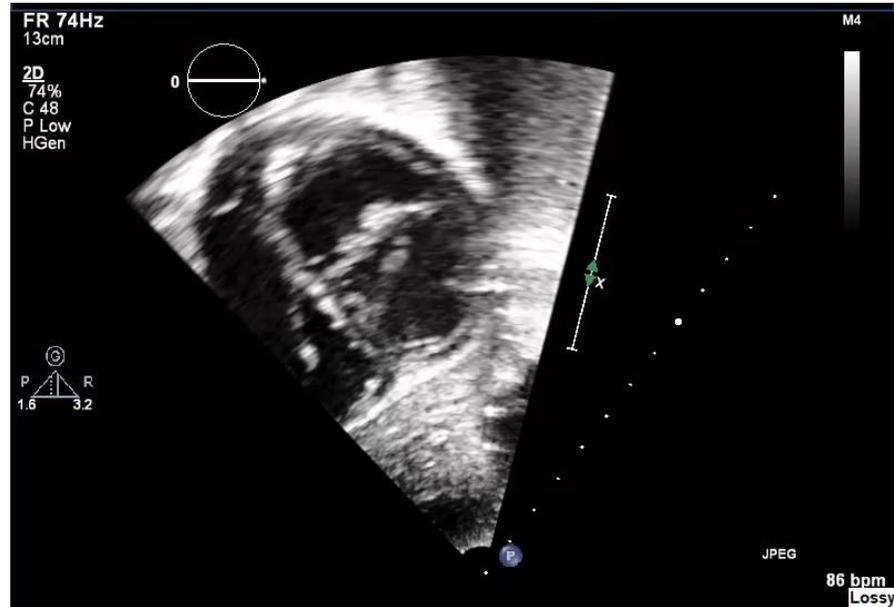
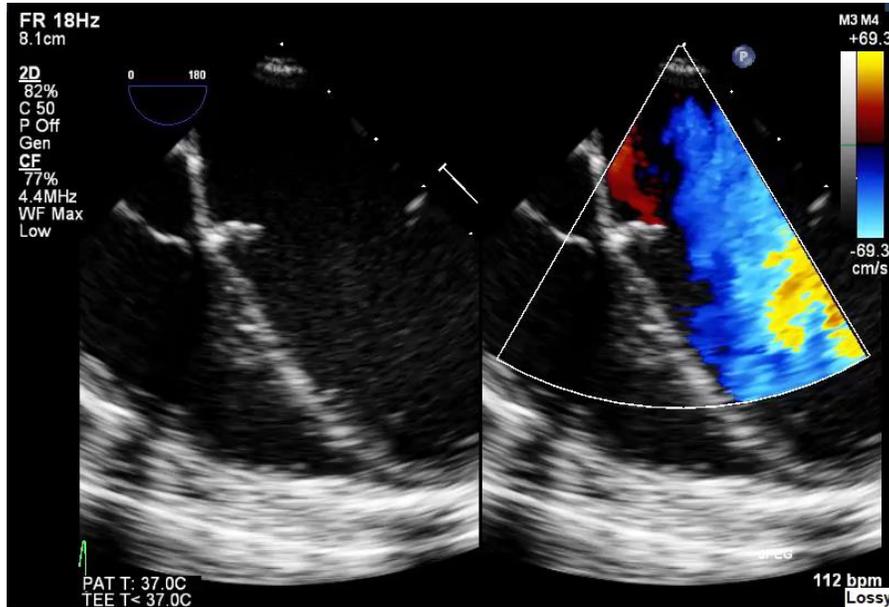


Apical 4 chamber



Failure of delamination of superior and inferior bridging leaflets from the RV aspect of the septum  
Only seen in the setting of separate orifices

# Intra op imaging & follow up checklist



- Residual shunts
- AVVR or stenosis
- LVOTO
- RV pressure
- Function

# Conclusions – Echocardiography in AVSD

- **Common anatomical features:**

Common AV junction/annulus

Unwedging of the aorta – narrow LVOT, vulnerable to obstruction

LAVV with 3 leaflets

- **Variability:**

Number of orifices

Level of shunting (size of atrial and ventricular components)

Balance/unbalance at atrial and ventricular levels

- Residual lesions are common (LAVV stenosis/regurgitation, residual VSDs, LVOTO)
- Echocardiographic assessment with transthoracic, transoesophageal echo and 3D for diagnosis, intraop and follow up
- Sequential segmental approach and assessment for any associated abnormalities

# Special thanks to the Leeds congenital echo team

Dr Grazia Delle Donne, Paediatric Cardiologist

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Sam Bainbridge

Sophie Bancroft

Sara Moore

Hannah Shaw

Rosie Brakefield

Laura Duffy

Lea Andriasyan



**LEEDS CONGENITAL**  
**HEARTS**