The Functionally 'Single' Ventricle

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Functionally Single Ventricle AND The Univentricular Atrioventricular Connection

Echocardiographic Diagnosis

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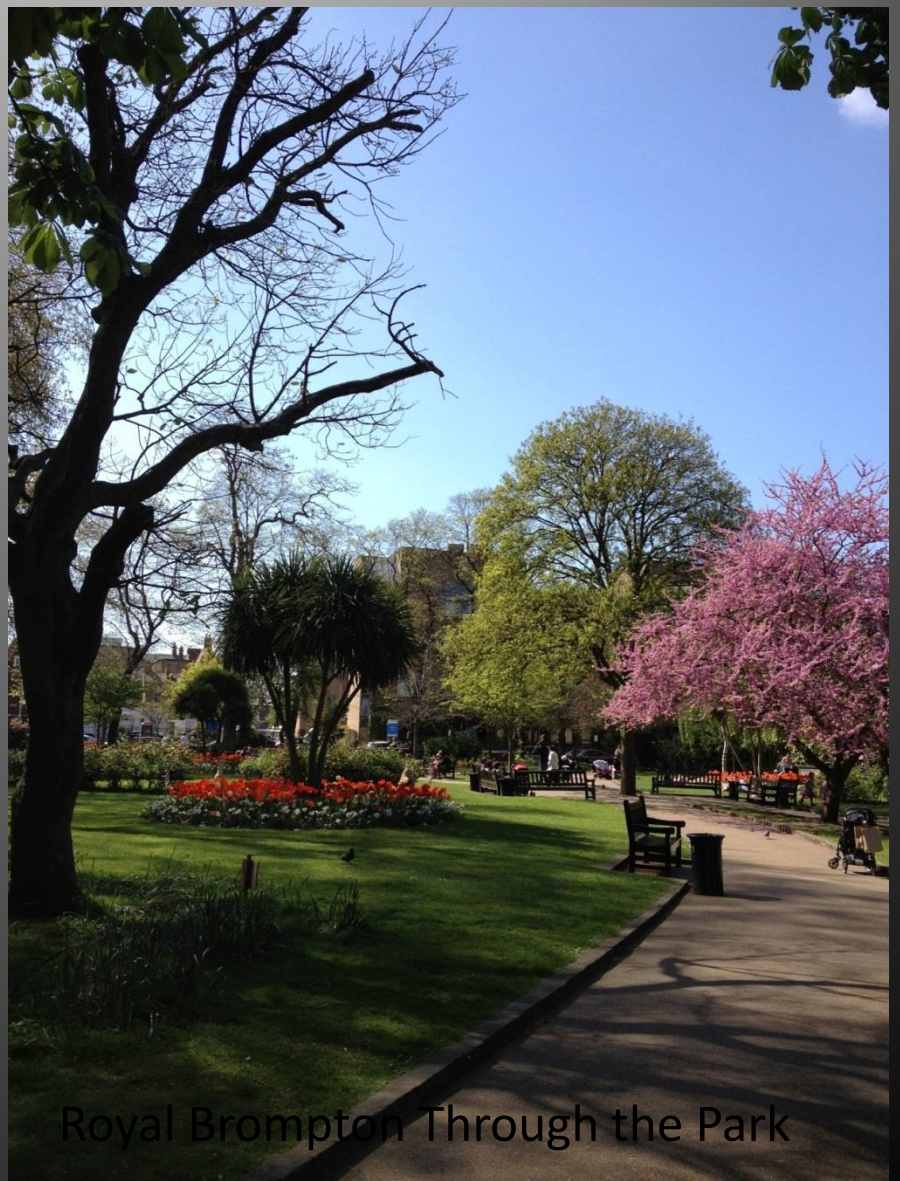


Royal Brompton Through the Park

The Functionally 'Single' Ventricle

Only one ventricle is capable of supporting the systemic circulation despite a biventricular Atrioventricular Connection

The Functionally 'Single' Ventricle

Or only one ventricle is capable of supporting the systemic circulation because the AV connection is Univentricular

The Functionally 'Single' Ventricle

- *Hypoplastic Left Heart Variants*
- *Pulmonary Atresia Intact Septum*
- *Unbalanced AVSD*

  (with hypoplastic RV or LV)
- *With Straddling Mitral or Tricuspid Valve*
- *Miscellaneous with hypoplastic LV or RV*
- *Hearts with Univentricular AV Connection*

The Univentricular AV Connection

The atria connect predominantly to only one ventricle

Univentricular AV Connection Types

Absent Right AV Connection

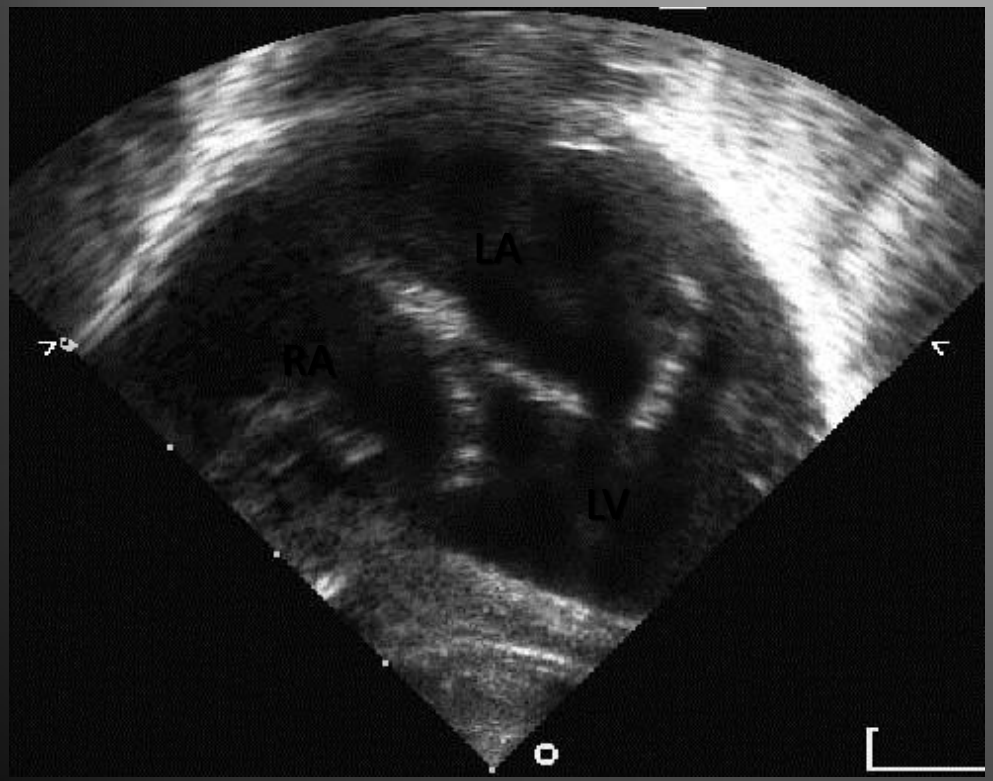
Absent Left AV Connection

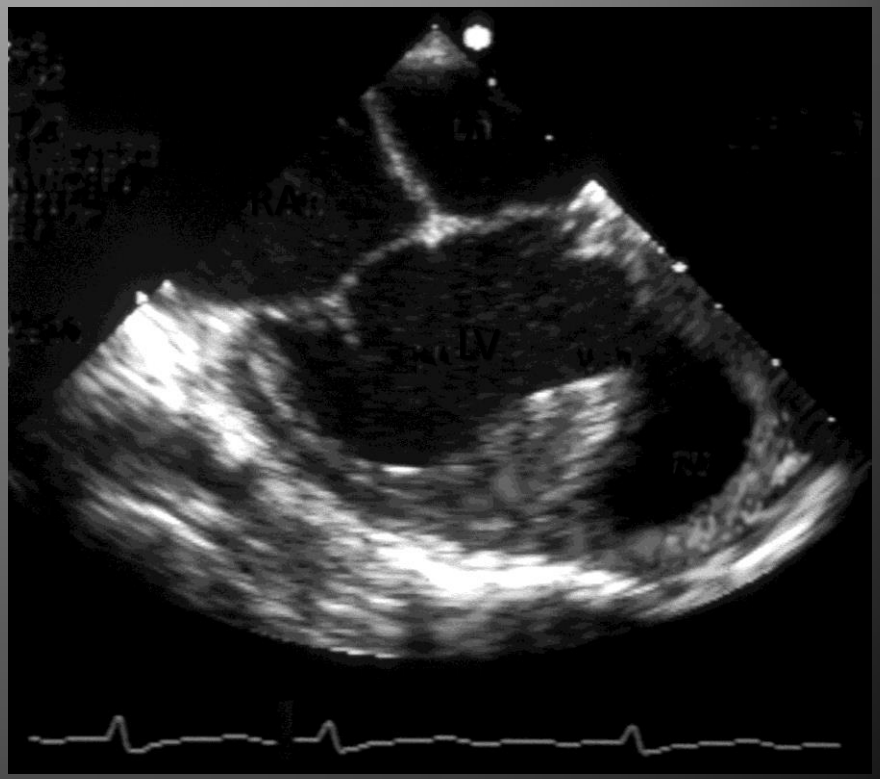
Double Inlet

Double Inlet with 2 AV Valves

DOUBLE INLET VENTRICLE

2 AV VALVES



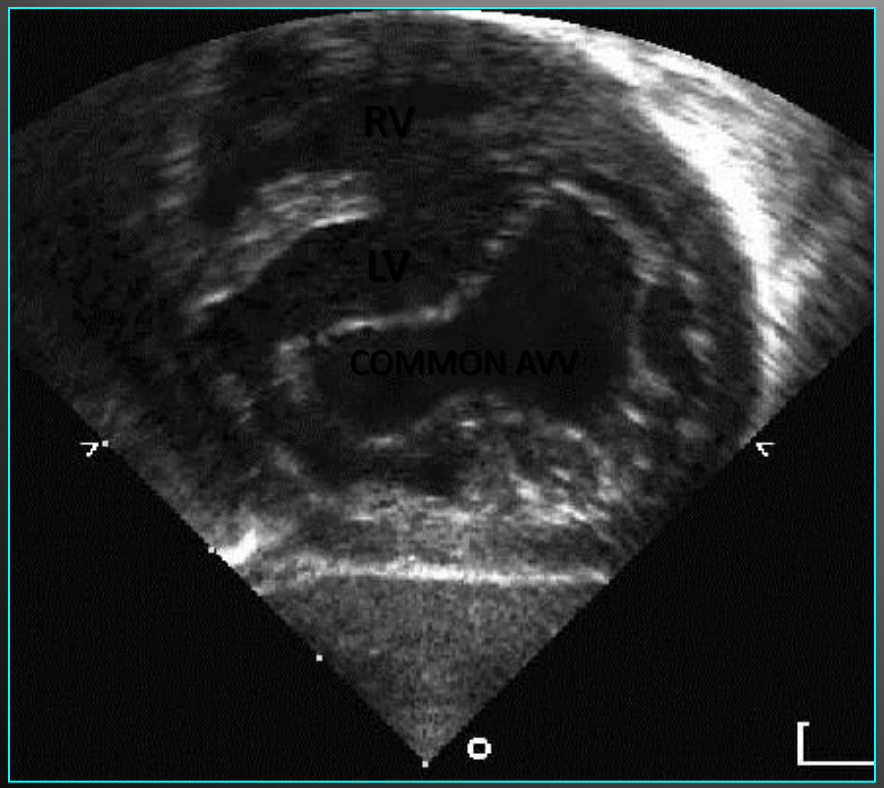


Double Inlet with Common (Single) AV Valve

DOUBLE INLET VENTRICLE

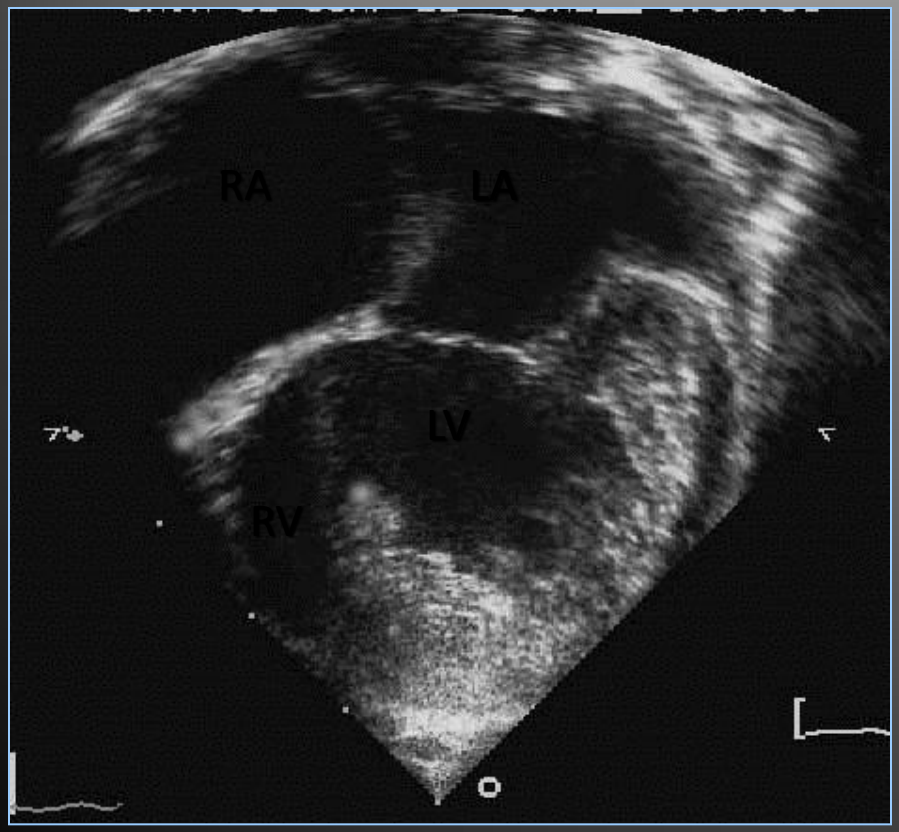
COMMON AV VALVE/ COMMON JUNCTION

DOUBLE INLET LEFT VENTRICLE

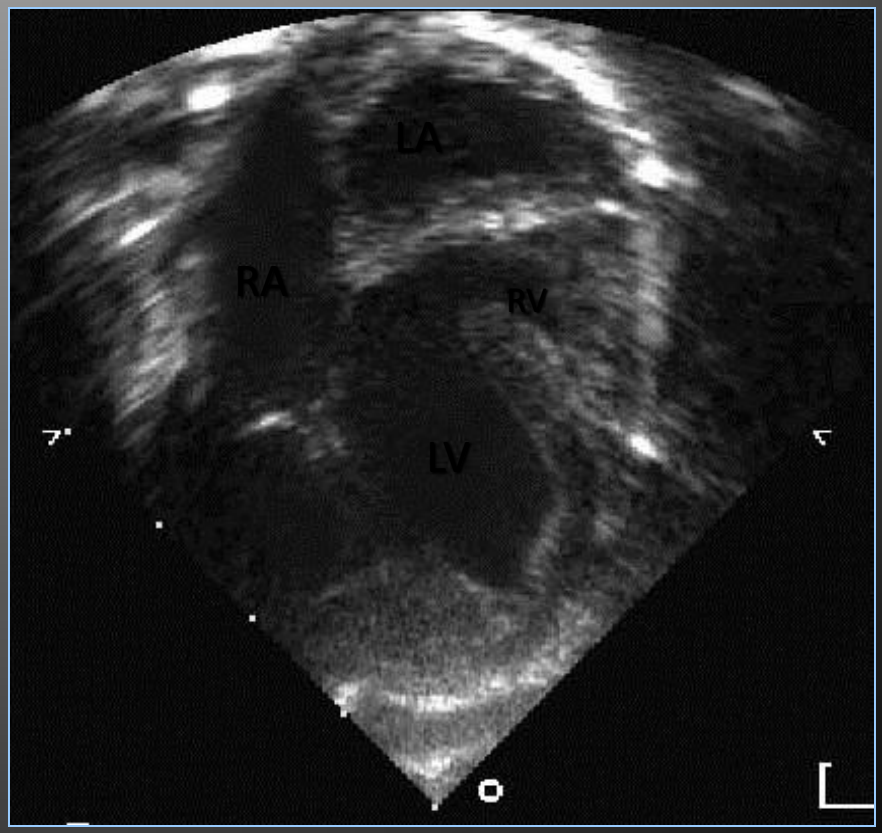


ABSENT AV CONNECTION

ABSENT RIGHT



ABSENT LEFT



UNIVENTRICULAR AV CONNECTION

- *ANY ATRIAL ARRANGEMENT*
- *ANY VENTRICULOARTERIAL CONNECTION*
- *MANY ASSOCIATED ANOMALIES*

Morphology of Dominant Ventricle:

Right

Left

Solitary Indeterminate

How can we determine ventricular morphology?

How can we determine ventricular morphology?

'Easily'

Morphology of Dominant Ventricle

- *Left*
  (with anterior rudimentary right ventricle)

- *Right*
  (With posterior rudimentary left ventricle)

- *Indeterminate*
(without rudimentary ventricle)

Double inlet ventricle

Double inlet Left Ventricle

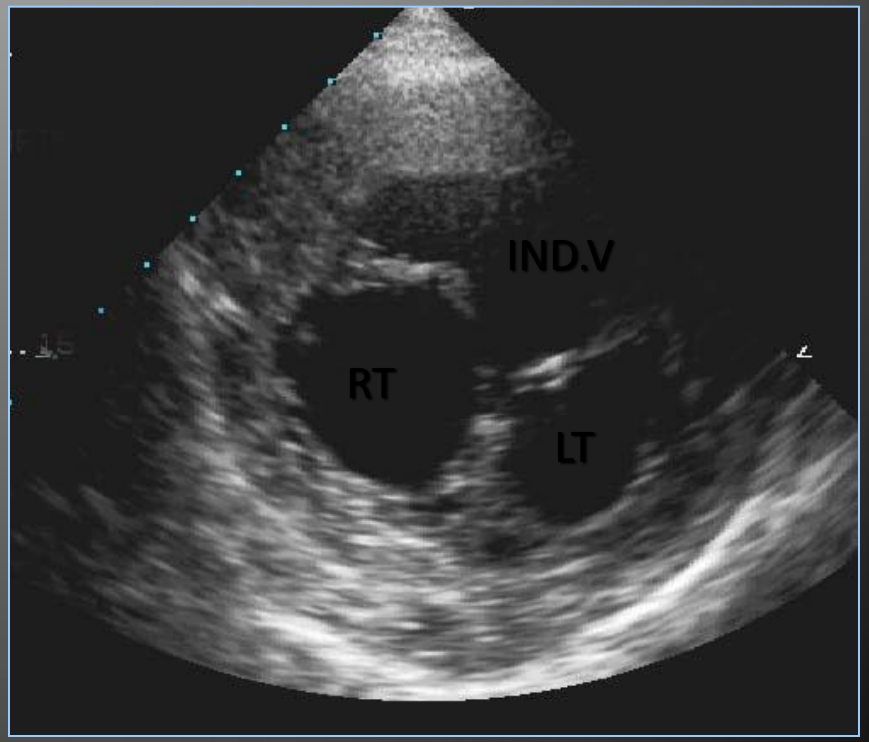
Rudimentary RV

Right or Left Sided

Double Inlet Right Ventricle

Double Inlet Solitary indeterminate Ventricle

SOLITARY INDETERMINATE VENTRICLE



Mode of AV Connection

Imperforate

Stenotic

Straddling AV Valve

ABSENT AV CONNECTION

- *ABSENT RIGHT (MOSTLY TRICUSPID ATRESIA)*
  - *SOLITUS (> 95%)*
  - *LA to LV (95%)*
  - *CONCORDANT VA CONN.(60%)*

- *ABSENT LEFT*
  - *SOLITUS (>95%)*
  - *RA to RV (50%)*
  - *RA to LV (50%)*

Absent Right AV Connection

Absent Left AV Connection

In Classical Tricuspid Atresia Which of following is true?

1. The dominant ventricle is RV
2. Most frequent associated anomaly is PS
3. An imperforate valve membrane lies between LA & RV
4. Situs inversus is found in 25%
5. Right aortic arch is characteristic

In hearts with double inlet ventricle which of these statements is true?

1. A common AV valve is found in the majority
2. Double inlet RV occurs in 15-20%
3. 5 sets of papillary muscles are usually found in DILV
4. The right sided AV valve is tricuspid
5. Coarctation of the aorta occurs typically when the ventriculo-arterial connection is discordant

In 'Univentricular Hearts' Typical late complications are?

1. AV valve regurgitation
2. Aortic regurgitation
3. Ventricular Dysfunction
4. Premature coronary artery disease
5. Atrial and/or ventricular arrythmias