

The 2<sup>nd</sup> Contemporary Morphology Course with Specimens and 3D Print Models

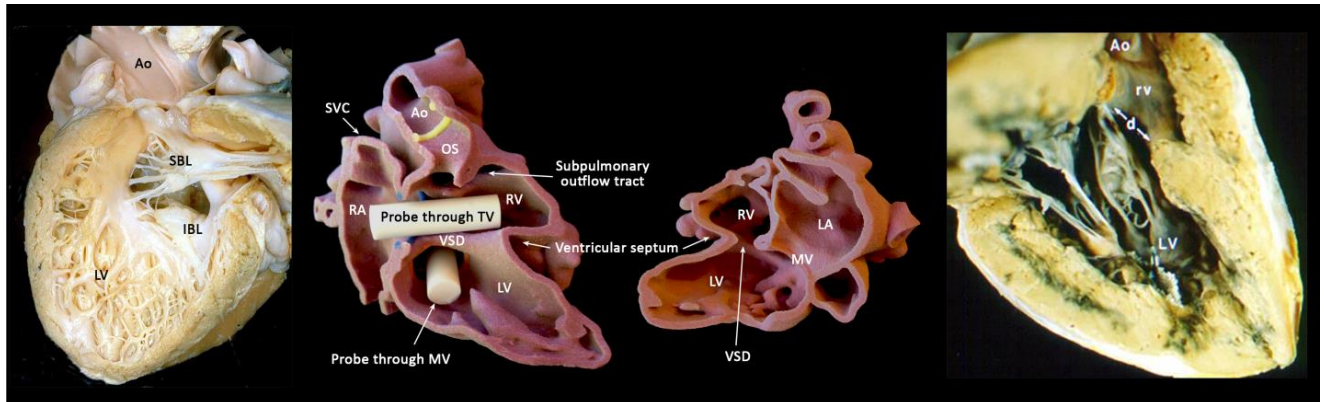
## CONGENITAL HEART DISEASES IN YOUR HANDS

### ♥ Abnormalities of the Atrioventricular Junction ♥

December 6-7, 2019

Peter Gilgan Centre for Research and Learning (PGCRL)

Hospital for Sick Children, Toronto, Canada



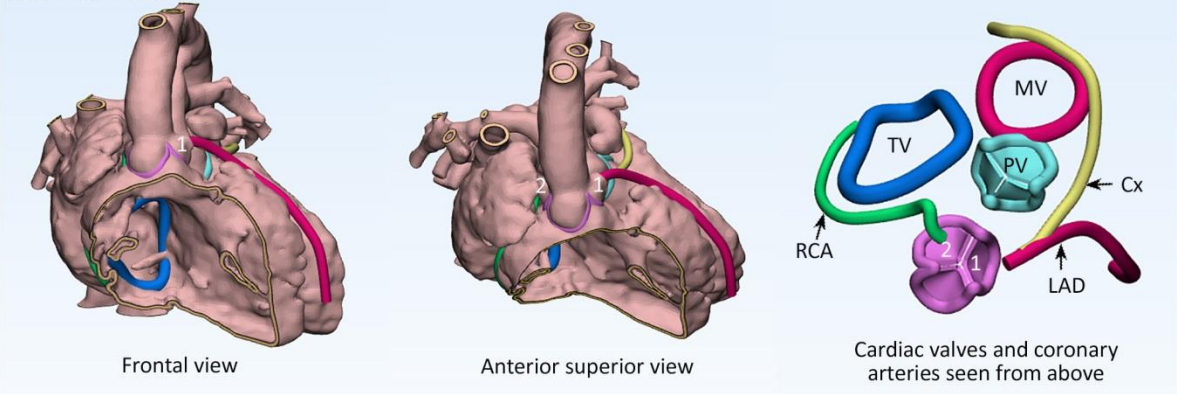
## TRANSPOSITIONS OF THE GREAT ARTERIES IN YOUR HANDS

- Hands-on Course with 3D Print Models -

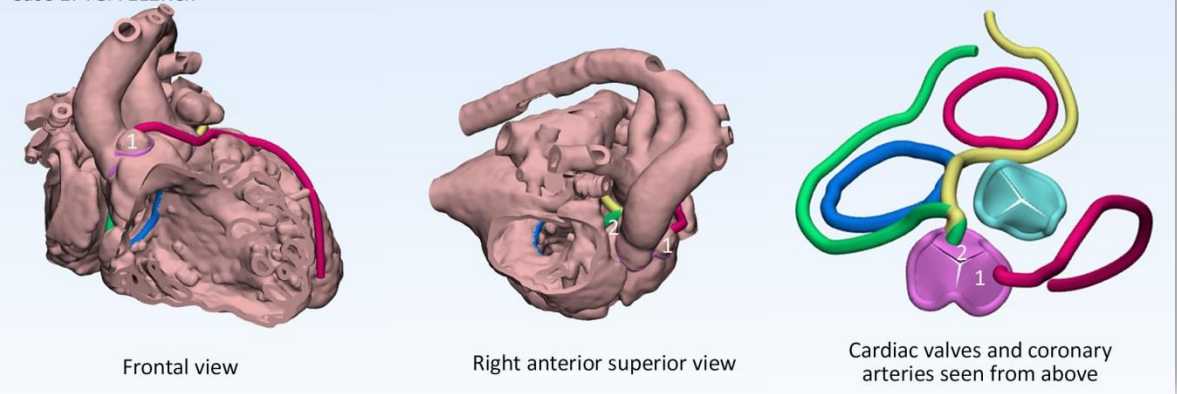
**CASES 1-3. Transposition of the great arteries with intact ventricular septum with different coronary artery branching patterns.**

❖ Source images: Contrast-enhanced CT angiograms obtained in end-systole (Asan Medical Center, Seoul, Korea).

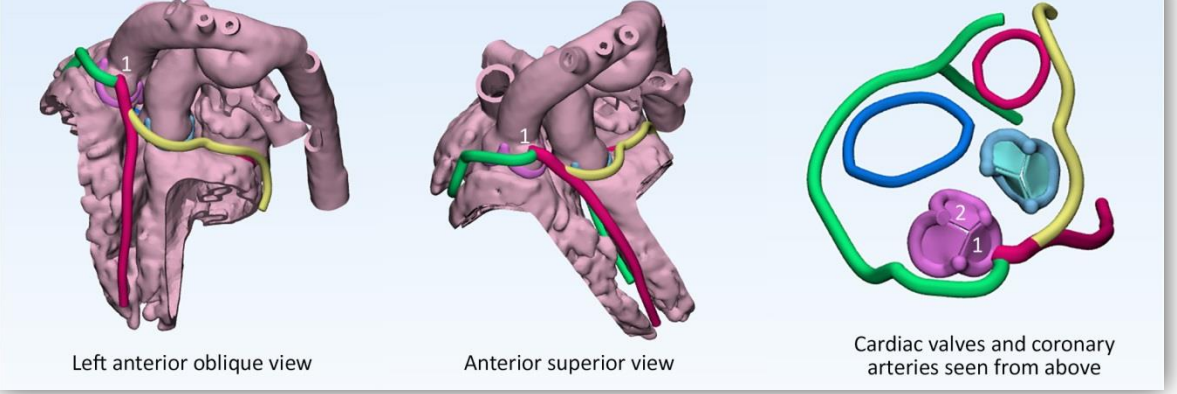
Case 1. TGA 1LCx2R



Case 2. TGA 1L2RCx

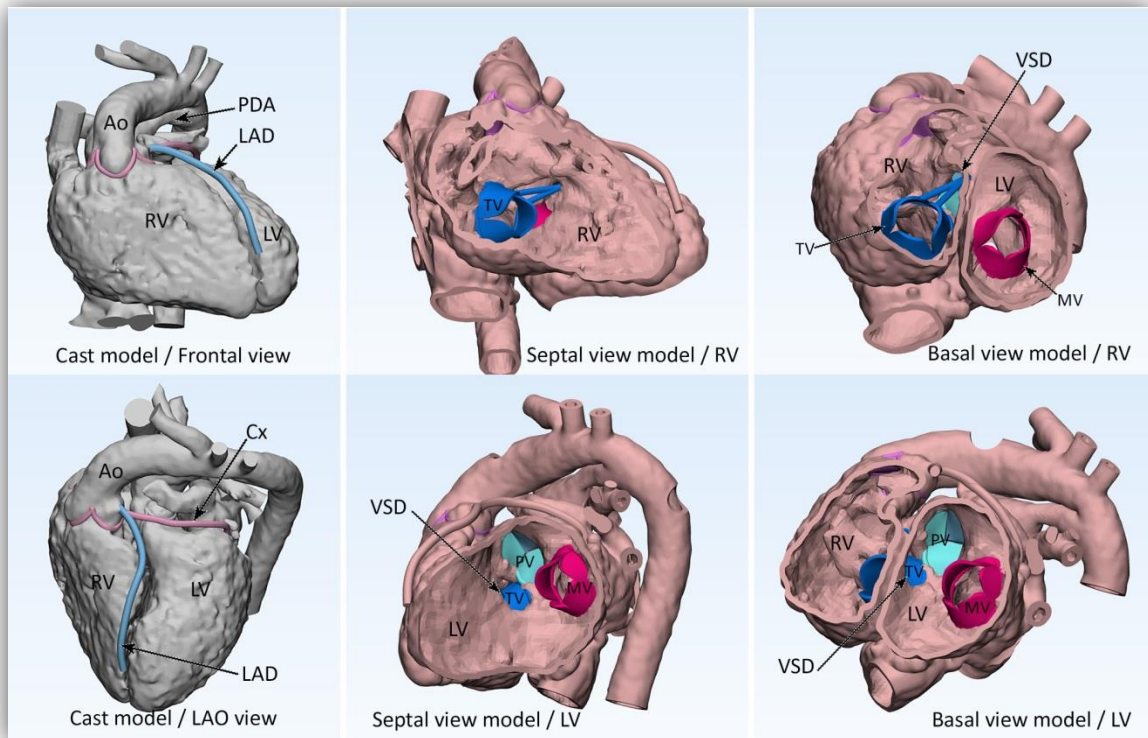


Case 3. TGA 1LCxR



#### CASE 4. Transposition of the great arteries with a perimembranous VSD and small PDA.

- ❖ Source images: Contrast-enhanced CT angiograms obtained in end-systole (Asan Medical Center, Seoul, Korea).

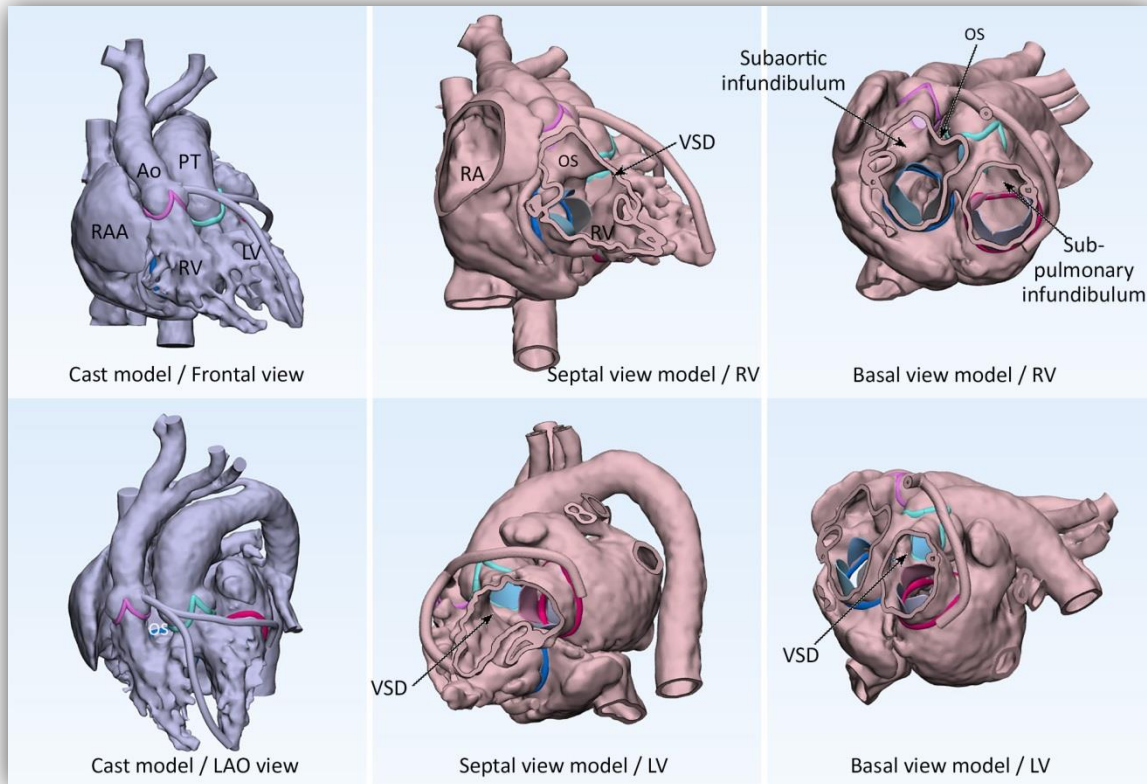


- ♥ Situs solitus, levocardia and left aortic arch
  - ♥ Atrioventricular concordant connection
  - ♥ Ventriculoarterial discordant connection : Aorta from RV through muscular infundibulum (aorto-tricuspid discontinuity) and pulmonary arterial trunk from LV without muscular infundibulum (pulmonary-mitral continuity).
  - ♥ An unrestrictive perimembranous VSD involving membranous septal area and adjacent muscular septum with the pulmonary and tricuspid valves in direct contact through the VSD. The atrioventricular conduction axis is expected to run along the posteroinferior margin of the VSD.
  - ♥ No significant (or minimal) overriding of pulmonary valve across the VSD.
  - ♥ The ventricular outflow tracts are parallel to each other.
  - ♥ The ascending aorta is directly anterior and slightly rightward in relation to the pulmonary arterial trunk
  - ♥ Usual coronary arterial origins and branching pattern (1LCx2R): The left coronary artery arises from the left-sided facing aortic sinus (Sinus 1) and bifurcates into the left anterior descending (LAD) and circumflex (Cx) coronary arteries. The right coronary artery arises from the right-sided facing sinus (Sinus 2) and courses along the right atrioventricular groove.
  - ♥ The aortic arch is unobstructed. There is a small PDA.
- ❖ Compare this case with Cases 5 and 6 for various degree of overriding of pulmonary valve across the VSD.



**CASE 5. Transposition of the great arteries with a perimembranous outlet VSD with one third of pulmonary valve overriding the ventricular septum.**

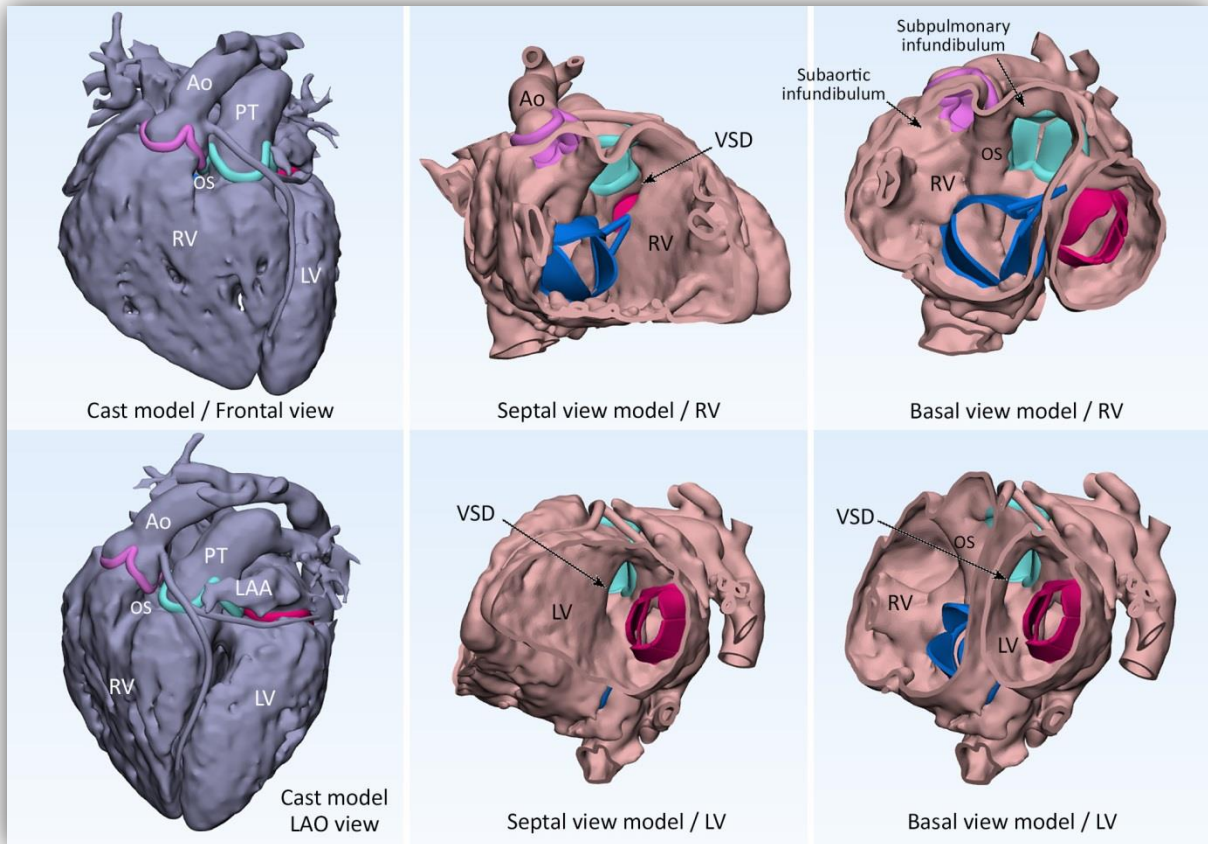
- ❖ Source images: Contrast-enhanced CT angiograms obtained in end-systole (Asan Medical Center, Seoul, Korea).



- ♥ Situs solitus / levocardia / left aortic arch
- ♥ Atrioventricular concordant connection
- ♥ A perimembranous VSD extending predominantly toward the outlet side.
- ♥ One third of pulmonary valve overriding the ventricular septum across the VSD. Note that the outlet septum (OS) is malaligned rightward and forward in relation to the rest of the ventricular septum. As a consequence, it is exclusively a right ventricular structure. The outlet septum is oriented parallel to the rest of the ventricular septum within the right ventricle.
- ♥ Bilateral infundibulum. Longer subaortic and shorter subpulmonary infundibulum.
- ♥ Aorta rightward and slightly anterior to pulmonary arterial trunk.
- ♥ Mildly narrow subaortic outflow tract. Severe tubular hypoplasia of the aortic arch with a large PDA.
- ♥ Usual coronary arterial origins and branching pattern (1LCx2R): The left coronary artery arises from the left-sided facing aortic sinus (Sinus 1) and bifurcates into the left anterior descending (LAD) and circumflex (Cx) coronary arteries. The right coronary artery arises from the right-sided facing sinus (Sinus 2) and courses along the right atrioventricular groove.
- ❖ Compare this case with Cases 4 and 6 for various degree of overriding of pulmonary valve across the VSD.

**CASE 6. Double outlet right ventricle with subpulmonary VSD (So-called Taussig-Bing malformation).**

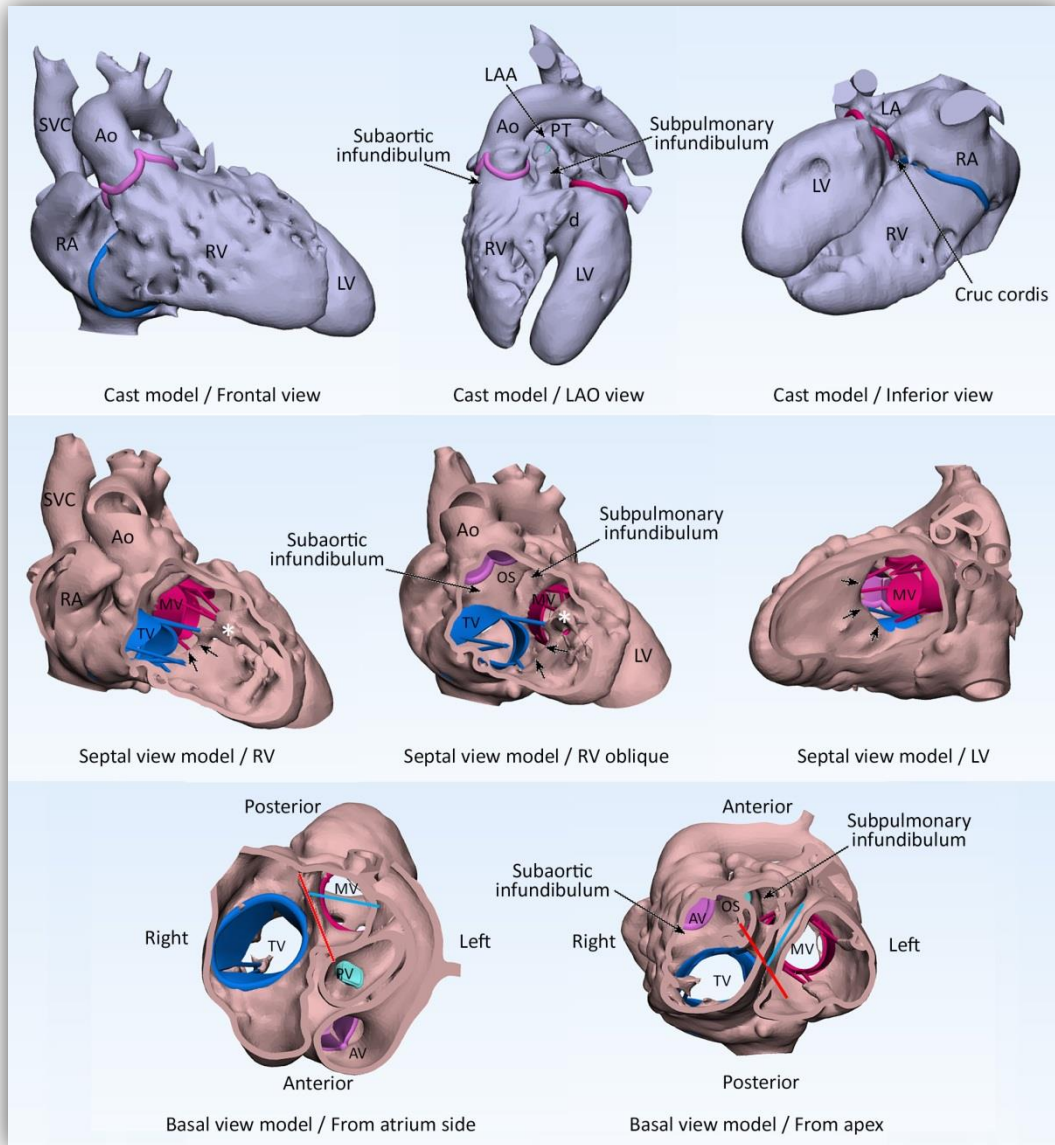
- ❖ Source images: Contrast-enhanced CT angiograms obtained in end-systole (Asan Medical Center, Seoul, Korea).



- ♥ Situs solitus / levocardia / left aortic arch
- ♥ Atrioventricular concordant connection
- ♥ Non-perimembranous VSD involving the outlet component of the ventricular septum.
- ♥ Both arterial trunks arising from the right ventricle. Note that the outlet septum (OS) is malaligned rightward and located in the central part of the right ventricular outflow tract. It is exclusively a right ventricular structure. The outlet septum is oriented parallel to the rest of the ventricular septum within the right ventricle.
- ♥ Bilateral infundibulum.
- ♥ Side-by-side arterial trunks with aorta rightward and slightly anterior to pulmonary arterial trunk.
- ♥ Mild tubular hypoplasia of the aortic arch.
- ♥ Usual coronary arterial origins and branching pattern (1LCx2R): The left coronary artery arises from the left-sided facing aortic sinus (Sinus 1) and bifurcates into the left anterior descending (LAD) and circumflex (Cx) coronary arteries. The right coronary artery arises from the right-sided facing sinus (Sinus 2) and courses along the right atrioventricular groove.
- ❖ Compare this case with Cases 4 and 5 for various degree of overriding of pulmonary valve across the VSD.

## CASE 7. Double outlet right ventricle with straddling and overriding mitral valve.

❖ Source images: Contrast-enhanced MR angiograms obtained in end-diastole (SickKids, Toronto)

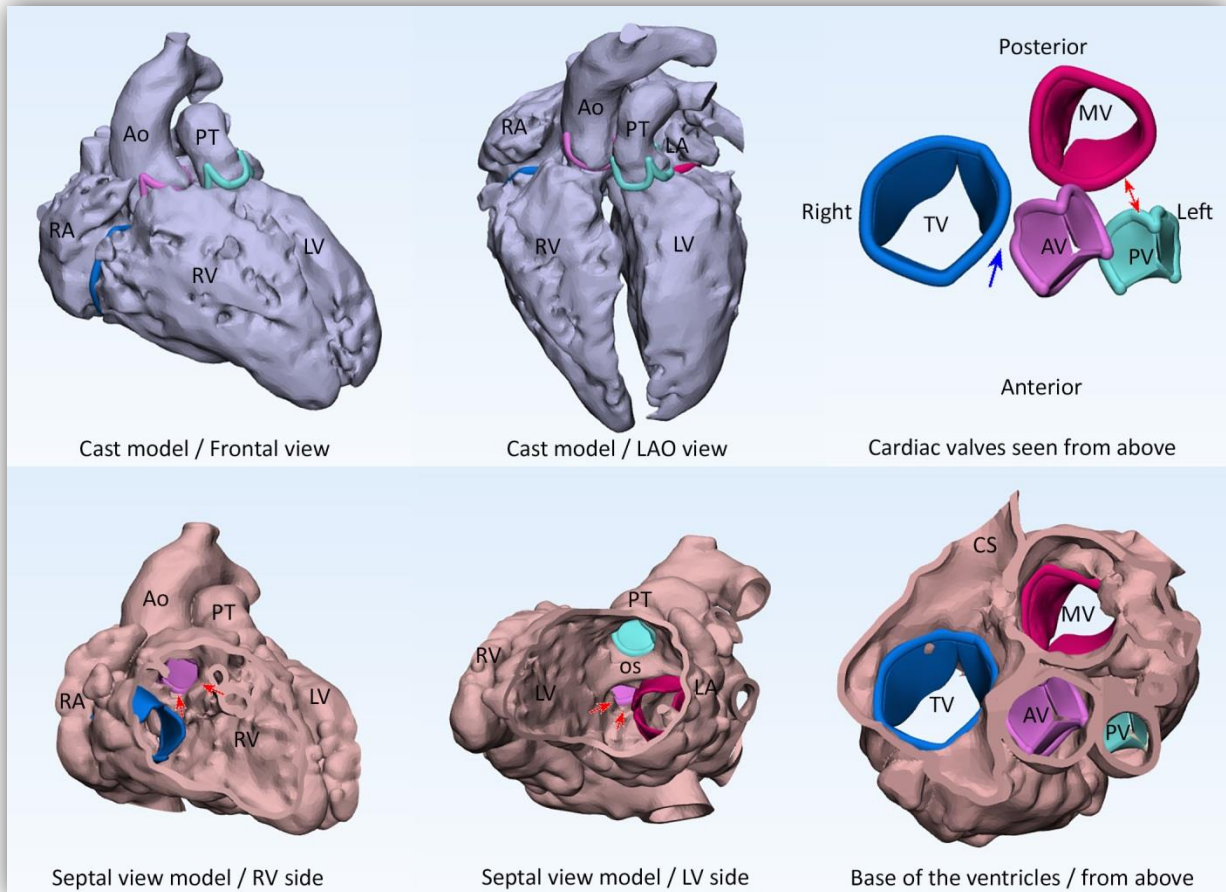


- ♥ Situs solitus / levocardia / left aortic arch
- ♥ Leftward malalignment of the anterior aspect of the ventricular septum (green line in Basal view model in lower column) in relation to the atrial septal plane (red line in Basal view model) with the posterior aspect hinged to the crux cordis. Note that the interventricular septum is kept aligned with the crux cordis (Cast model / Inferior view in right panel of upper column). The mitral valve annulus overrides the malaligned ventricular septum and the chords from the mitral valve insert to the papillary muscle (white asterisk in Septal view model / RV and RV oblique in middle column) that arises from the right ventricular septal surface.
- ♥ Double outlet right ventricle with bilateral infundibulum. Muscular tunnel narrowing of subpulmonary infundibulum.



**CASE 8. Transposition of the great arteries with the aorta posteriorly located and overriding the ventricular septum (So-called posterior transposition).**

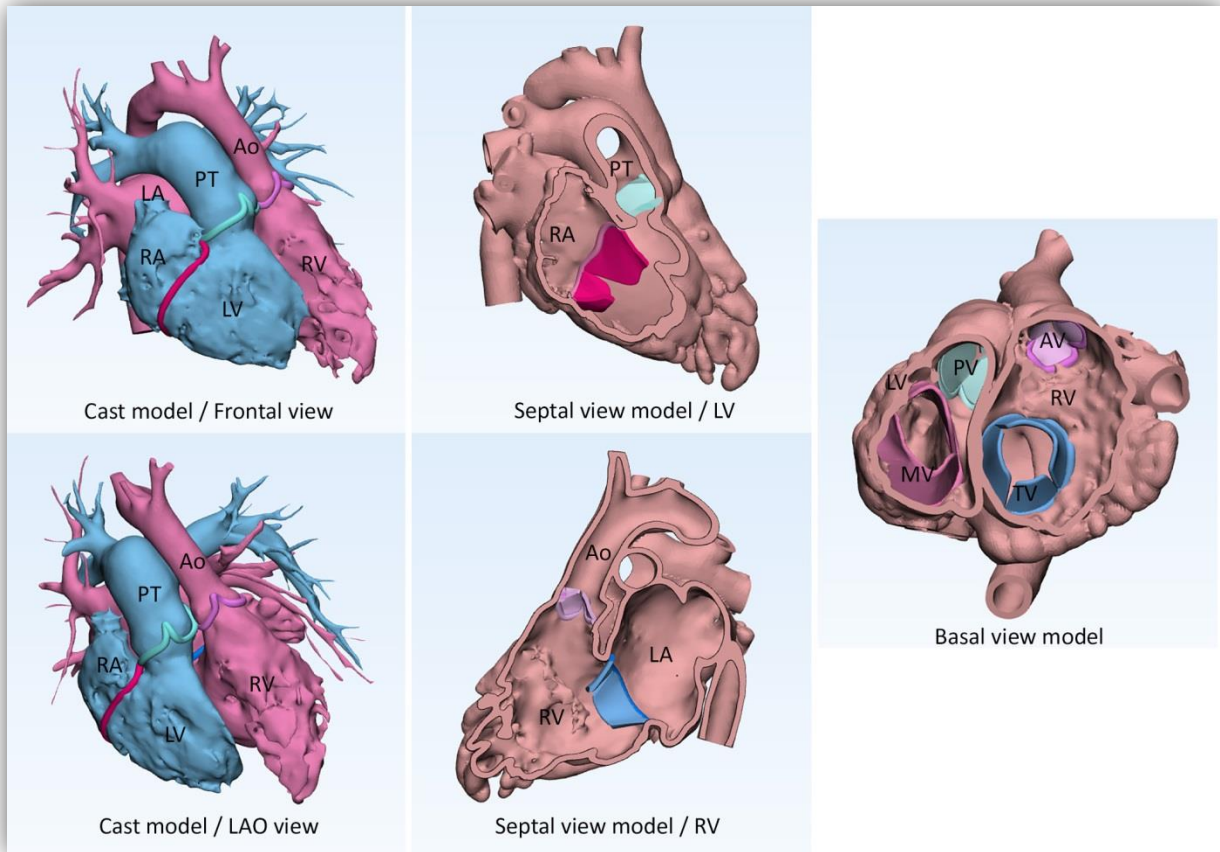
❖ Source images: Contrast-enhanced MR angiograms obtained in end-diastole (SickKids, Toronto)



- ♥ Situs solitus / levocardia / left aortic arch
- ♥ Side-by-side relationship of the arterial trunks with the aorta on the right and slightly posterior to pulmonary arterial trunk (right panel, upper column). Note that the aortic valve arises from both ventricles without muscular infundibulum without gap between the tricuspid and aortic valve annuli (blue arrow in right-hand figure of upper panel). The pulmonary valve is supported by a muscular infundibulum with a gap between the pulmonary valve and mitral valve annuli (double-headed red arrow in right panel, upper column).
- ♥ A VSD (red arrows) involves the membranous and outlet parts of the septum immediately below the aortic valve.
- ♥ The aortic valve (colored in pink) overrides the ventricular septum.
- ♥ The left ventricular outflow tract is divided into the right posterior subaortic and left anterior subpulmonary outflow tract by the outlet septum (OS). This case is a variant of double outlet left ventricle.

## CASE 9. Congenitally corrected transposition of the great arteries in levocardia .

- ❖ Source images: Contrast-enhanced MR angiograms obtained in end-systole (Pontifical Catholic University, Santiago, Chile)



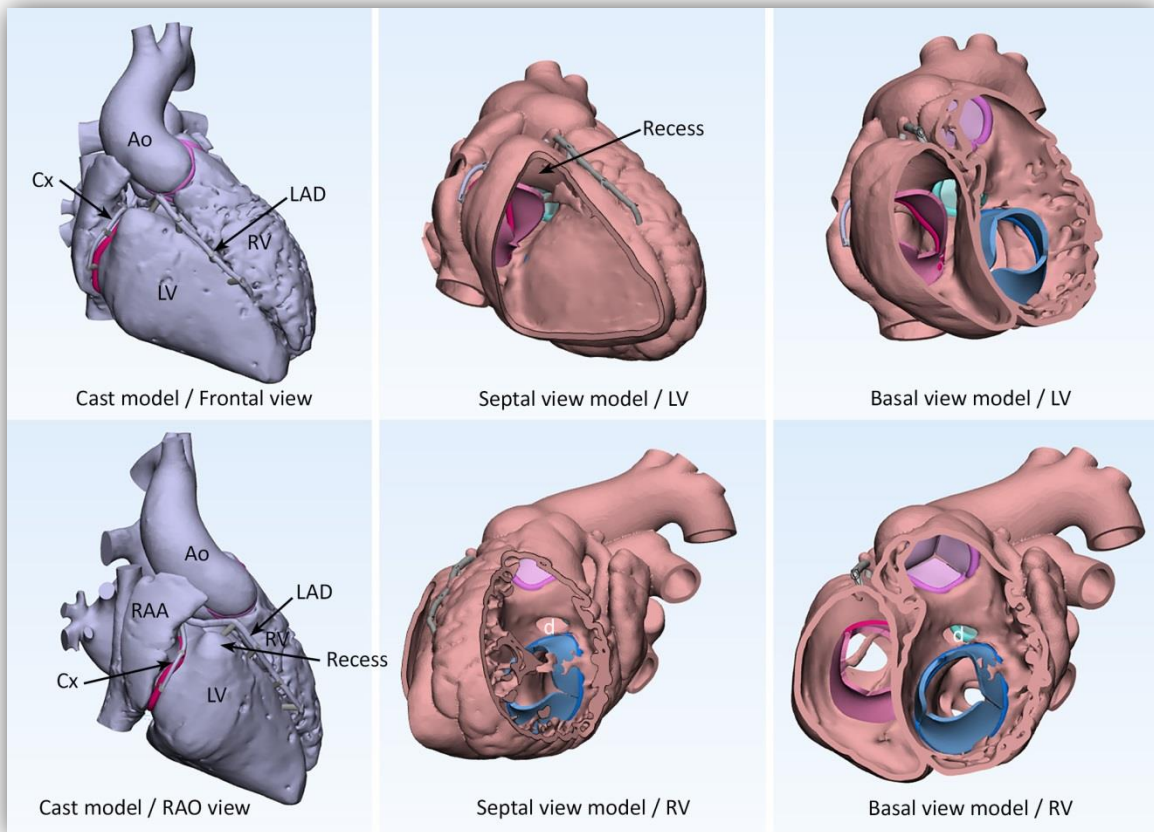
- ♥ Situs solitus / levocardia / right aortic arch with mirror-image branching pattern
- ♥ Atrioventricular discordant connection.
- ♥ Pulmonary arterial trunk arising from right-sided left ventricle with the mitral valve in direct contact with the pulmonary valve.
- ♥ Aorta arising from left-sided right ventricle through muscular infundibulum.
- ♥ Parallel ventricular outflow tracts
- ♥ Classic great arterial relationship for congenitally corrected transposition with the aorta on the left and anterior to the pulmonary arterial trunk.

- ❖ Compare this case with Cases 10 and 11.



**CASE 10. Congenitally corrected transposition of the great arteries with a small perimembranous VSD .**

- ❖ Source images: Contrast-enhanced CT angiograms obtained in end-systole (Asan Medical Center, Seoul, Korea).

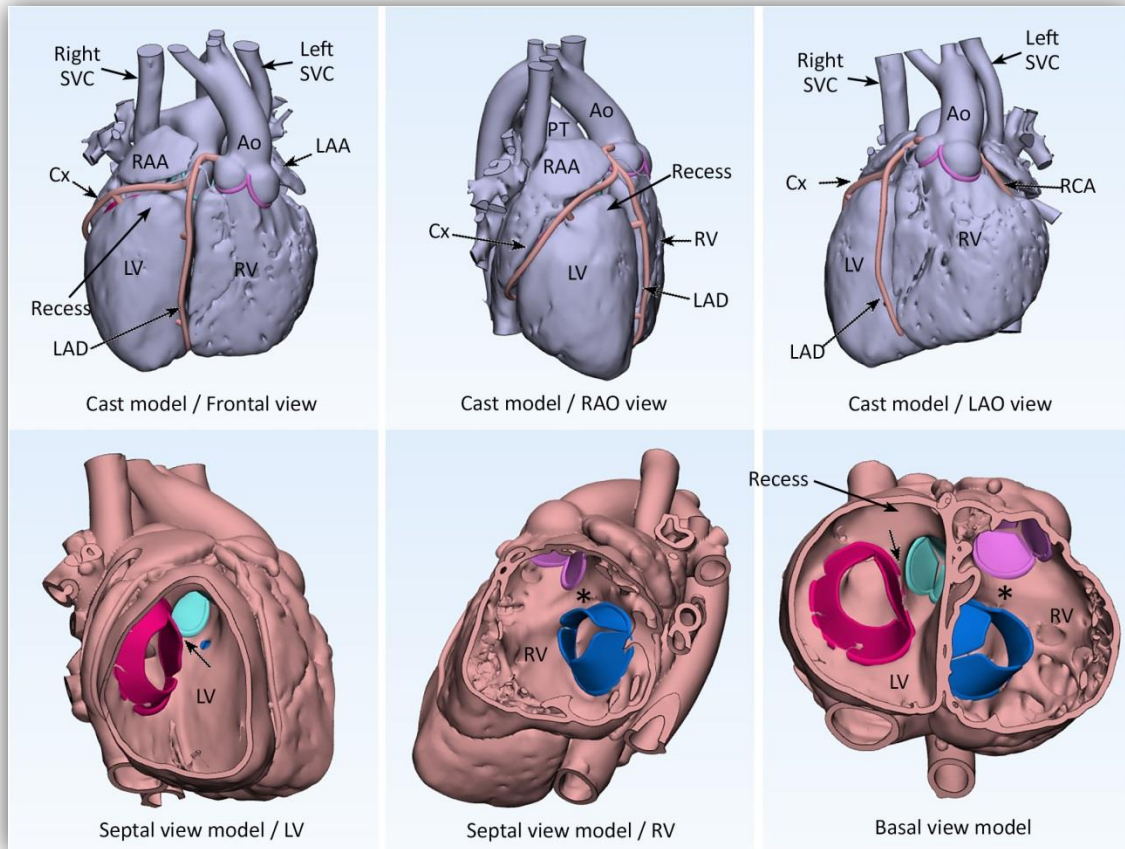


- ♥ Situs solitus / levocardia / left aortic arch
- ♥ Atrioventricular discordant connection.
- ♥ Small perimembranous VSD.
- ♥ A recess of left ventricle protruding antero-superiorly in the space formed by the ascending aortic root, pulmonary arterial trunk and right atrial appendage.
- ♥ Pulmonary arterial trunk arising from right-sided left ventricle through a short infundibulum.
- ♥ Aorta arising from left-sided right ventricle through a long muscular infundibulum.
- ♥ Parallel ventricular outflow tracts. Subpulmonary outflow tract is mildly encroached on in the wedged position without discrete stenosis.
- ♥ Aorta directly anterior to pulmonary arterial trunk.
- ♥ Usual coronary arterial origins and branching pattern for congenitally corrected TGA (1R2LCx). Left facing aortic sinus (Sinus 1) giving rise to the right coronary artery coursing along the left-side tricuspid valve annulus. Right facing sinus (Sinus 2) giving rise to the left coronary artery that bifurcates into the left anterior descending (LAD) and circumflex (Cx) coronary arteries.

- ❖ Compare this case with Cases 9 and 11.

## CASE 11. Congenitally corrected transposition of the great arteries in mesocardia.

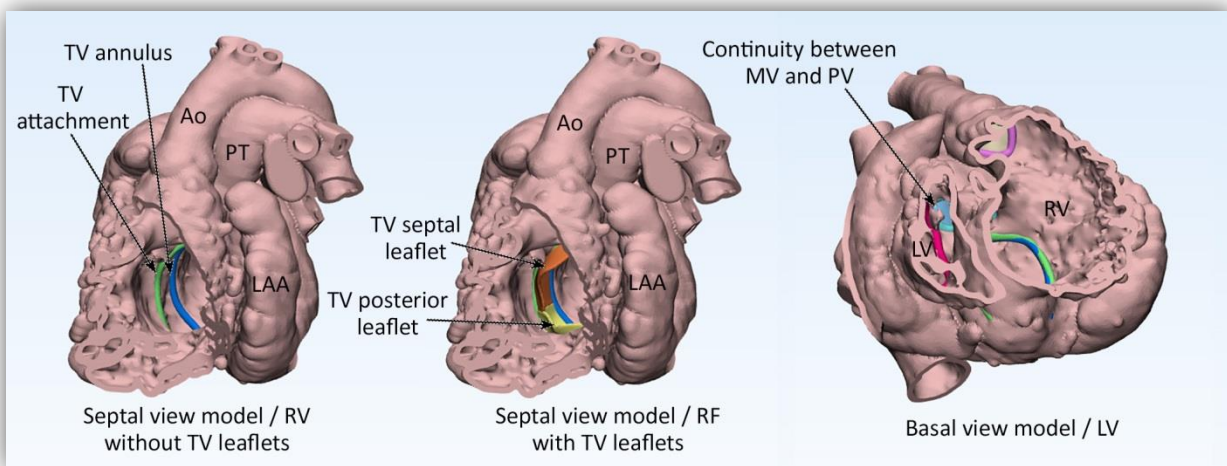
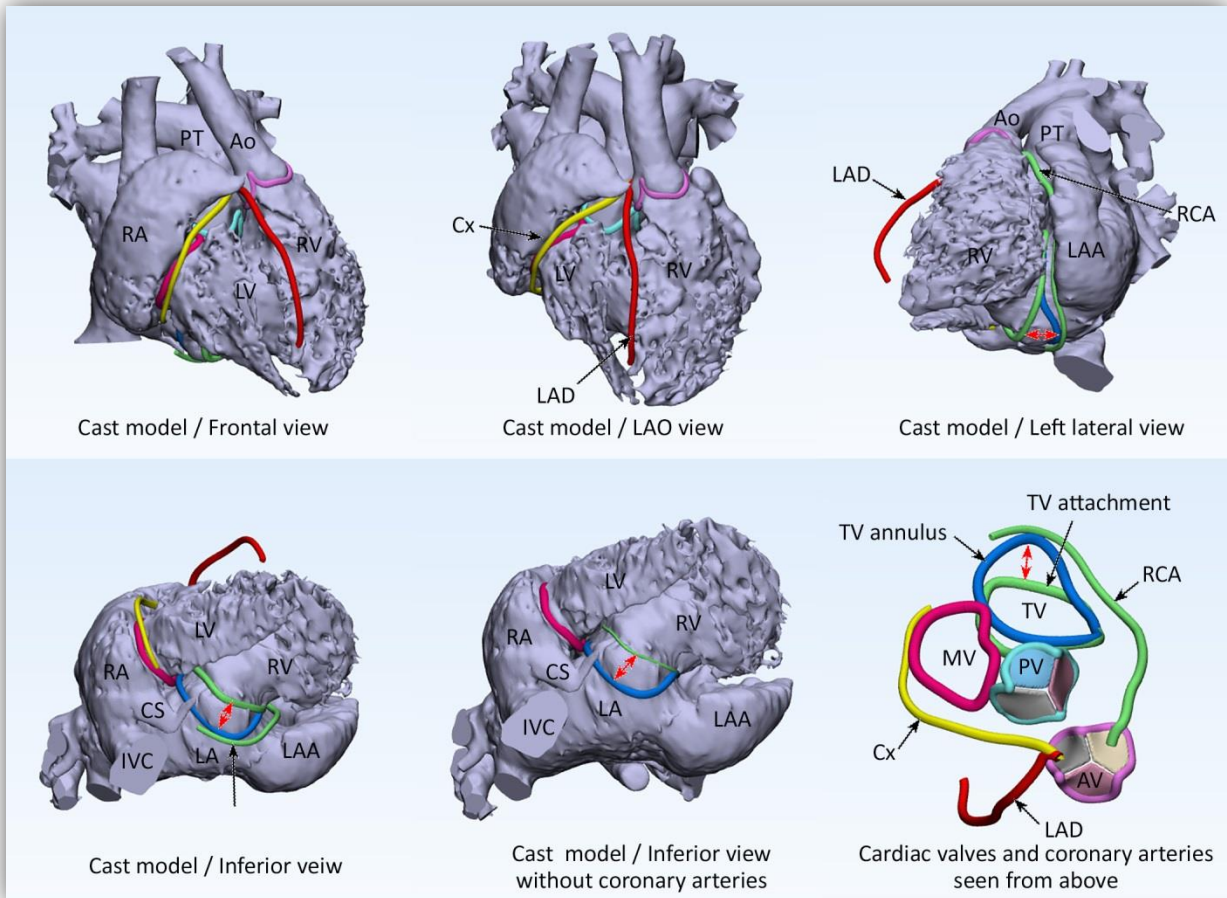
❖ Source images: : Contrast-enhanced MR angiograms obtained in end-diastole (SickKids, Toronto)



- ♥ Situs solitus / mesocardia / left aortic arch
  - ♥ Atrioventricular discordant connection.
  - ♥ A recess of left ventricle protruding antero-superiorly in the space formed by the ascending aortic root, pulmonary arterial trunk and right atrial appendage.
  - ♥ Pulmonary arterial trunk arising from right-sided left ventricle with pulmonary valve in direct contact with mitral valve (arrow in left and right hand panels in lower column).
  - ♥ Aorta arising from left-sided right ventricle through a long muscular infundibulum (asterisk in middle and right hand figures in lower column).
  - ♥ Parallel ventricular outflow tracts.
  - ♥ Classic great arterial relationship with aorta left anterior to pulmonary arterial trunk.
  - ♥ Usual coronary arterial origins and branching pattern for congenitally corrected TGA (1R2LCx). Left facing aortic sinus (Sinus 1) giving rise to the right coronary artery coursing along the left-side tricuspid valve annulus. Right facing sinus (Sinus 2) giving rise to the left coronary artery that bifurcates into the left anterior descending (LAD) and circumflex (Cx) coronary arteries.
  - ♥ Bilateral SVCs with left SVC connecting to dilated coronary sinus.
- ❖ Compare this case with Cases 9 and 10.

**CASE 12. Congenitally corrected transposition of the great arteries with perimembranous VSD and Ebstein's malformation of left-sided tricuspid valve.**

❖ Source images : Contrast-enhanced MR angiograms obtained in end-systole (SickKids, Toronto)

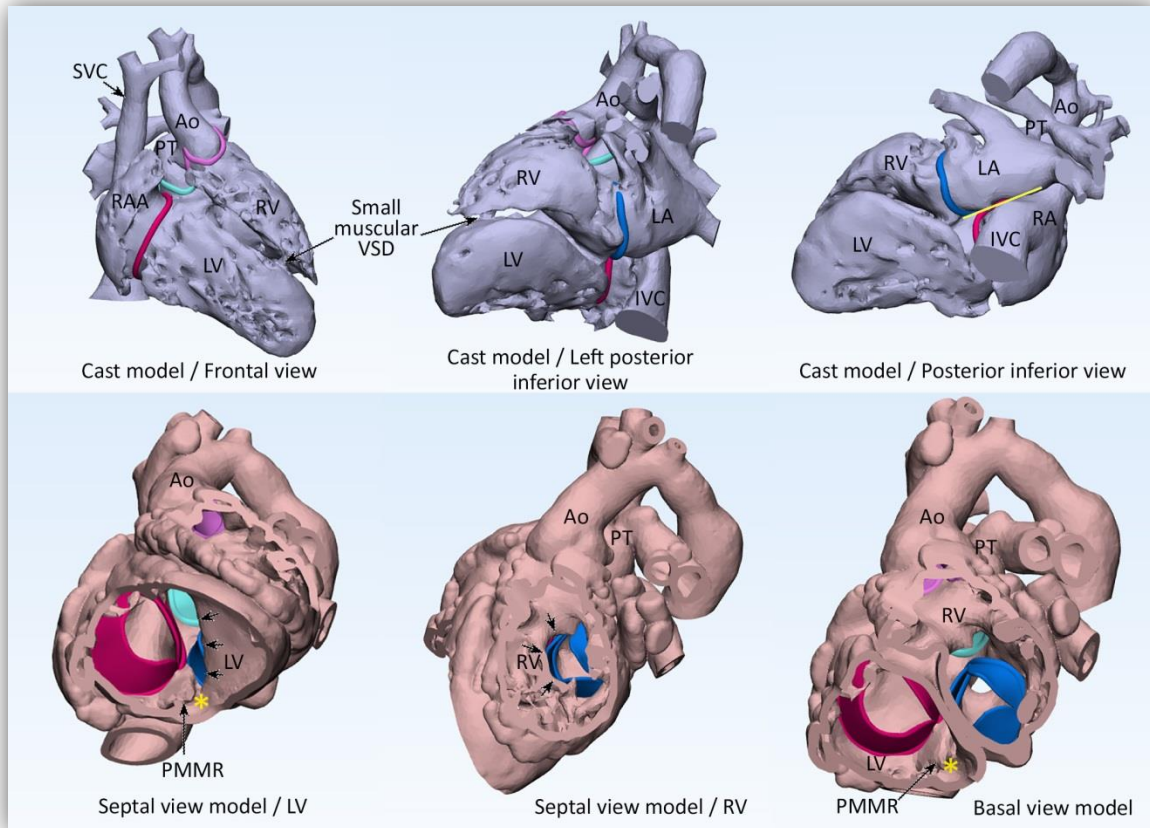




- ♥ Situs solitus / levocardia / left aortic arch
- ♥ Atrioventricular discordant connection.
- ♥ Small right-sided left ventricle and large left-sided right ventricle. Perimembranous VSD.
- ♥ Apically displaced attachment (marked with green line) of septal and posterior leaflets of tricuspid valve with atrialization of a small part of the right ventricular inlet (double-headed red arrow). The anterior leaflet is not sail-like as seen in Ebstein's malformation occurring with concordant atrioventricular connection.
- ♥ Pulmonary arterial trunk arising from right-sided left ventricle without muscular infundibulum.
- ♥ Aorta arising from left-sided right ventricle through a long muscular infundibulum.
- ♥ Parallel ventricular outflow tracts. Subpulmonary outflow tract mildly encroached on in the wedged position without discrete stenosis.
- ♥ Aorta anterior and slightly left to pulmonary arterial trunk.
- ♥ Usual coronary arterial origins and branching pattern for congenitally corrected TGA (1R2LCx).

### CASE 13. Congenitally corrected TGA with overriding and straddling left-sided tricuspid valve.

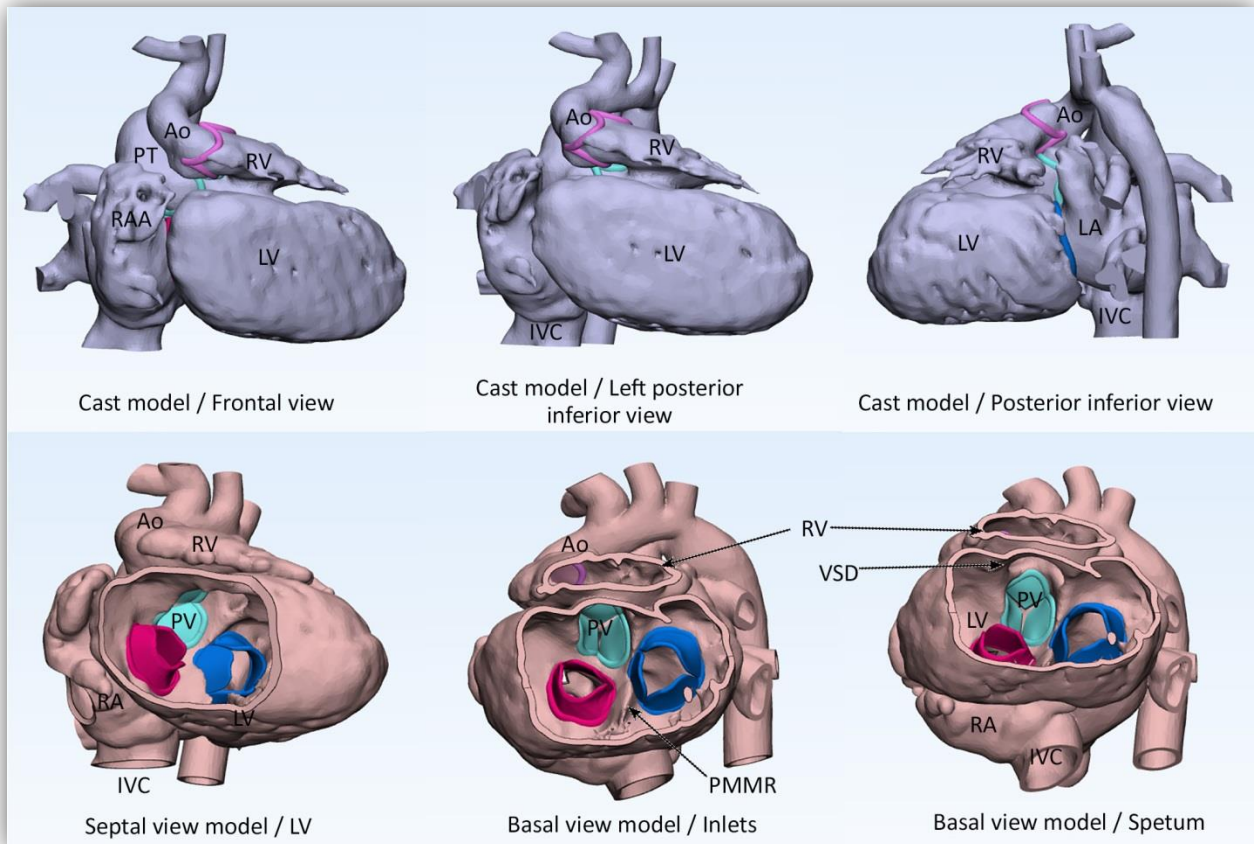
❖ Source images: : Contrast-enhanced MR angiograms obtained in end-diastole (SickKids, Toronto)



- ♥ Situs solitus / levocardia / left aortic arch
  - ♥ Atrioventricular discordant connection.
  - ♥ Large VSD (black arrows in lower column figures) extending from inlet to outlet. The ventricular septum is malaligned leftward from the crux cordis where a septal remnant is seen as a muscular ridge called posteromedian muscle ridge (PMMR) in the posterior wall of the left ventricle. There is a small gap (yellow asterisk) between this ridge and malaligned ventricular septum. A small part of the tricuspid valve overrides the ventricular septum. Straddling chords, not shown. Note that the posterior part of the ventricular septum is not aligned with the atrial septal plane (marked by yellow line in right panel of upper column).
  - ♥ Mildly hypoplastic right ventricle located superior and leftward in relation to left ventricle.
  - ♥ Pulmonary arterial trunk arising from right-sided left ventricle with pulmonary valve in direct contact with mitral valve.
  - ♥ Aorta arising from left-sided right ventricle through a muscular infundibulum.
  - ♥ Focal constriction in main pulmonary arterial trunk due to pulmonary artery banding.
  - ♥ Classic great arterial relationship with aorta left anterior to pulmonary arterial trunk.
- ❖ Compare this case with Cases 7 and 14.

## CASE 14. Double inlet left ventricle with transposition of the great arteries.

- ❖ Source images: Contrast-enhanced CT angiograms obtained in end-systole (Asan Medical Center, Seoul, Korea).



- ♥ Situs solitus / levocardia / left aortic arch
  - ♥ Both right and left atrioventricular valves connecting to a large main chamber of left ventricular morphology.
  - ♥ Severe leftward and superior malalignment of the ventricular septum with the small right ventricle located on the top of the left ventricle. Remnant of inlet ventricular septum called posteromedian muscle ridge (PMMR) seen between the right and left atrioventricular valves.
  - ♥ A muscular VSD.
  - ♥ Pulmonary arterial trunk arising from right-sided left ventricle with pulmonary valve in direct contact with atrioventricular valves.
  - ♥ Aorta arising from hypoplastic right ventricle through a muscular infundibulum.
  - ♥ Tubular hypoplasia of the aortic arch.
  - ♥ Classic great arterial relationship with aorta left anterior to pulmonary arterial trunk.
- ❖ Compare this case with Case 13.