Dextro-Transposition of the great arteries (D-TGA)

Dextro-Transposition of the great vessels has the aorta arising from the right ventricle (RV) and the pulmonary artery (PA) arising from the left ventricle (LV). This is referred to as ventriculoarterial discordance. D-TGA is the most common form of transposition of the great arteries and in this disorder the ventricles are oriented so that the right ventricle is positioned as normal to the right of the left ventricle but the origin of the aorta is anterior and rightward to the origin of the pulmonary artery.

Dextro-Transposition of the great arteries (D-TGA) is a lesion characterized by atrioventricular concordance and ventriculoarterial discordance, in which the aorta arises from the right ventricle and the pulmonary artery from the left ventricle. Two parallel circuits of blood flow exist such that the path of blood follow is as follows:

1. Systemic veins to IVC and SVC to RA then through the Tricuspid valve into the RV then into the Aorta and back to the systemic veins. (Systemic veins ➔ IVC and SVC ➔ RA then ➔ Tricuspid valve ➔ RV ➔ Aorta ➔ back to the systemic veins.)

2. From the lungs venous blood drains via the Pulmonary veins to left atrium then through the mitral valve into the left ventricle to the pulmonary artery to the lungs and then through the pulmonary veins back into the left atrium. (Lungs ➔ Pulmonary veins ➔ LA ➔ Mitral valve ➔ Left Ventricle ➔ Pulmonary artery ➔ Lungs ➔ Pulmonary veins ➔ LA )

Note the **Atrioventricular valves are ALWAYS attached/associated with their respective ventricles (MV with LV, TV with RV).** This is true with both D-TGA and L-TGA!
Note the two parallel circulations above exist such that the first circulation sends deoxygenated from the systemic venous system back to the systemic arterial circulation and the second circulation sends oxygenated blood from the pulmonary venous system back to the pulmonary arterial system.

With a prevalence of around 2.3-4.7 per 10,000 live births, D-TGA accounts for about 3% of all congenital heart disease and about 20% of all cyanotic heart disease. The specific embryology for D-TGA is not fully delineated, but it is thought to be due to abnormal growth and development of the bilateral subarterial conus. Initially there is bilateral subarterial conus present at around one month of development. Normally, the subaortic conus and subpulmonary conus are present in the first month of gestation as the great arteries are positioned superior to the right ventricle. Next, the subaortic conus is resorbed at around 30 to 34 days into gestation, and this allows for migration of the aortic valve inferiorly and posteriorly into its normal position above the left ventricle. The Subaortic conal resorption results in the characteristic fibrous continuity between the mitral and aortic valve within the left ventricle. The pulmonary valve retains its association with the right ventricle due to the persistence of the subpulmonary conus or infundibulum = muscular right ventricular outflow tract (RVOT).

Unlike normal development, in D-TGA the subpulmonary conus (NOT the subaortic conus) is abnormally resorbed, which allows for posterior movement of the pulmonary valve and the development of fibrous continuity between the pulmonary and mitral valve resulting in the PA arising from the
LV. Due to the unabsorbed subaortic conus the aortic valve moves anterior and rightward where it abnormally associates with the morphologic right ventricle.

With D-TGA there is NO subpulmonary conus (fibrous continuity b/w the MV and pulmonic valve exists) and there is an abnormal subaortic conus (infundibulum) between the RV and the aorta due to failure of the subaortic conus to resorb. This results in the aorta being anterior and rightward of the pulmonary artery and the aorta arises from the RV whereas the PA arises from the LV and the pulmonic valve has fibrous continuity with the mitral valve. This is referred to as dextro or D-transposition because the looping of the ventricles during cardiac morphogenesis that positions the RV on the right and the LV on the left is Normal = rightward = Dextro (to distinguish this from L-TGA)

With normal cardiac development, the aorta is positioned posterior and to the right of the main pulmonary artery, where it arises from the left ventricle, but with D-TGA the aorta is positioned anteriorly and rightward where it arises from the right ventricle and has a subarterial conus (infundibulum) similar to the normal right ventricular outflow tract. D-TGA is often associated with other cardiac defects:

VSD – 50% of patients with D-TGA

LVOT obstruction = SubPulmonic obstruction -25% of patients with D-TGA

Variable coronary anatomy- important for arterial switch operation.
Echocardiography for D-TGA:

D-TGA is generally diagnosed by two-dimensional echocardiography. A transthoracic subcostal view shows the main pulmonary artery dividing into the left and right pulmonary arteries after it arises from the posterior left ventricle, and a transthoracic short-axis or parasagittal plane shows the aorta rising anteriorly from the right ventricle. Notably both the aortic and pulmonic valves appear in the same plane (you can see the short axis of both valves simultaneously).

In both types of TGA, the aortic and pulmonic valves are in the same plane and the great arteries are parallel and do not cross as is expected in normal cardiac anatomy.

The echocardiographic testing should systematically delineate atrioventricular or ventriculoarterial connections, the presence or absence of other commonly associated cardiac anomalies, such as a ventricular septal defect (VSD), as well as the coronary artery anatomy.

Other echo findings that need to be elucidated include the following:

- ASD-needed for intercirculatory mixing. Restriction to flow assessed.
- VSD- presence of a VSD increases the risk that there may be associated aortic arch anomalies, such as a coarctation.
- Atrioventricular valves
- Ductus arteriosus
- Coronary anatomy- important prior to arterial switch operation

Unlike many other forms of congenital heart disease D-TGA is NOT associated with any one common genetic abnormality.
D-TGA (Reverse differential cyanosis)
Levo-Transposition of the great arteries (L-TGA)

Congenitally corrected TGA = Levo- or L-looped transposition of the great arteries (L-TGA). This is a rare form of congenital heart disease characterized abnormal position of the ventricles where the LV is on the right and the RV is on the left, thereby resulting in both atrioventricular and ventriculoarterial discordance. It is also sometimes referred to as ventricular inversion or double discordance.

L-TGA by itself (no other defects) is “physiologically corrected” because there is normal in series flow of deoxygenated blood to the lungs and oxygentated blood to the systemic circulation. Despite this correct series circulation, these patients have a systemic RV and are thus at increased risk of heart failure.
L-TGA results from abnormal leftward looping of the heart causing the morphologic LV to be positioned on the right and the morphologic RV to be positioned on the left. In this scenario blood flows as follows:

Systemic veins to IVC and SVC to RA through the mitral valve into the LV then through the pulmonic valve into the PA and to the lungs then into the pulmonary veins into the LA then through the tricuspid valve into the RV and from the RV into the aorta. (Systemic veins→IVC & SVC→RA→mitral valve→LV→pulmonic valve→PA→lungs→pulmonary veins→LA→tricuspid valve→RV→aorta→systemic circulation→systemic veins)

L-TGA is associated with other cardiac lesions in >90% of cases. The most common associated lesions include the following:

- VSD; 70-80% of patients with L-TGA
- Pulmonary outflow obstruction (LVOT obstruction); 30-60%
- Tricuspid valve abnormalities: up to 90% of patients with L-TGA
- Mitral valve abnormalities: 55% of L-TGA patients
- Complete heart block = most common arrhythmia in patients with L-TGA (conduction system is abnormal and unstable)

In both types of TGA, the aortic and pulmonic valves are in the same plane and the great arteries are parallel and do not cross as is expected in normal cardiac anatomy.

L-TGA is diagnosed by recognizing the location of the TV and the RV. The
RV is ALWAYS associated with the tricuspid valve and the RV is characterized by coarse trabeculations, which differentiate it from the morphologic LV with its fine trabeculations and two well defined papillary muscles.

Note the **Atrioventricular valves are ALWAYS attached/associated with their respective ventricles (MV with LV, TV with RV).** This is true with both D-TGA and L-TGA!
Atrioventricular valves are ALWAYS attached/associated with their respective ventricles (MV with LV, TV with RV).
How do you recognize the different cardiac chambers?

**RIGHT ATRIUM (RA):**
- Usually contains entry of the vena cavae and the coronary sinus
- Crista terminalis separates the RA into anterior and posterior parts, and from it arise the numerous pectinate muscles, located throughout the right atrial wall and in the RA appendage (only found in the LAA of the LA).
- Often a Eustachian valve is present near the entry of the IVC
- RA appendage is more pyramidal shaped and often larger than the larger than the finger like LAA.
- Also seen from the RA side of the interatrial septum is the limbus of the fossa ovalis (formed by the septum secundum), a muscular ring on the superior and lateral sides of the fossa ovalis.

**LEFT ATRIUM (LA):**
- LA Appendage is a narrow thin and finger-like structure, which is distinct from the larger pyramidal-shaped RA appendage
- Usually receives the pulmonary veins (not always; ex. TAPVR, PAPVR)
- NO pectinate muscles outside of the LA appendage (pectinate muscles are present in the RA outside of the RA appendage).
ATRIOVENTRICULAR VALVES MITRAL (MV) AND TRICUSPID (TV) VALVES:

Note the **Atrioventricular valves are ALWAYS attached/associated with their respective ventricles (MV with LV, TV with RV).**

This is true with both D-TGA and L-TGA!

- Usually (exception = AV canal defect, MV and TV at same level) the septal component of the TV is more apically placed than the septal component (anterior leaflet) of the mitral valve.
- TV has septal chordal attachments and at least 3 papillary muscles
- MV has two distinct papillary muscles (usually) and NO septal attachments.
- MV orifice is more oval or fish-mouthed
- TV orifice is more triangular
- TV has 3 leaflets (duh!) and the mitral valve has two

**LEFT VENTRICLE (LV):**

- LV is ALWAYS attached to the MV. If you see a MV you have an LV!
- Smoother wall (less trabeculations than the RV)
- Two well defined papillary muscles (usually)
- Less apically displaced septal (anterior) attachment of the MV
- NO muscular outflow tract = conus = infundibulum thus allowing for a fibrous continuity between the mitral-aortic valve (usual) or the mitral-pulmonic valve (D-TGA).
- LV is usually circular in cross section

**RIGHT VENTRICLE (RV):**
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-RV is ALWAYS associated with the TV! Important! If you see the TV you have an RV!

-TV (see above) has 3 papillary muscles, 3 leaflets, more apical septal leaflet, etc.

-RV is more trabeculated than the LV

-RV has multiple papillary muscles (usually 3, unlike the two of the LV)

-RV has lots of muscle bundles, septal, moderator and parietal bands, trabeculations.

-often crescentric in short axis, but not always
Terminology explaining common congenital cardiac lesions is included below:

Remember:

___Position = heart position in the chest

___Cardia = location of the apex

Location/position of the heart within the chest can be:

Levoposition = normal (on the left)

Dextroposition = on the right

Mesoposition = in the middle

Location/position of the cardiac apex can be:

Levocardia = position of the cardiac apex on the left

Dextrocardia = position of the cardiac apex on the right

Mesocardia = position of the cardiac apex in the middle
Visceral Situs is determined by the position of the thoracic and abdominal visceral and can be:
- **solitus** = normal (liver on right, spleen on left etc.)
- **inversus** = mirror image of normal or
- **ambiguous** = bilateral right sided morphology or bilateral left sided morphology.

Visceral Situs **inversus** = a right-to-left reversal of the thoracic and abdominal viscera

Cardiac situs is generally determined by the position of the morphologic right atrium (RA), thus we refer to this as atrial situs

**Atrial Situs** can be:

Atrial Situs **solitus** (normal; RA on right, LA on left),

Atrial Situs **inversus** (mirror image; RA on left, LA on right),

Atrial Situs **ambiguous** (Heterotaxy):

- right-sided isomerism (asplenia syndrome) where left-sided structures are absent and right-sided structures are duplicated (2 morphologic RAs), or

- left-sided isomerism (polysplenia syndrome) where right-sided structures are absent and left-sided structures are duplicated (two morphologic LAs).

Cardiac structures are defined by specific morphologic features and not by location within the chest.

The atrioventricular valves (AV valves) always empty into the appropriate/correct ventricle

(tricuspid valve to the morphologic right ventricle and mitral to the left) no matter how the cardiac anatomy is arranged.
Remember:

The atrioventricular valves (Tricuspid & Mitral valves) are ALWAYS associated with the correct ventricle.

The tricuspid valve is ALWAYS associated with the Right Ventricle (RV) and the mitral valve (MV) is always associated with the LV.

If you see a MV it will ALWAYS be connected to the LV and if you see a TV it will ALWAYS be connected to the RV.

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