



“TRANSPOSITION OF GREAT ARTERIES” EMBRYOLOGICAL BASIS AND ITS CLINICAL IMPORTANCE

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ABSTRACT

Transposition of the great vessels is a type of congenital heart defect involving an abnormal arrangement of any of the great vessels such as the superior vena cava, pulmonary arteries and aorta. Congenital heart diseases concerning the primary arteries are further divided into sub-group called transposition of great arteries. There are different forms of anomalies like Dextro-transposition, Levo-transposition and Dextrocardia. It was first announced about two centuries ago, but the treatment became effective from the mid 21st century. Today the survival rate for infants is greater than 90%.

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Introduction

Transposition of the great arteries (TGA) is a congenital cyanotic heart defect. The large vessels that carry the blood from the heart to the lungs and to the body are "shifted". Most of the cases are related to the transposition of the arteries based on the manner they are shifted. This means the oxygen intake is decreased in the blood which is pumped from the heart to the body.

In normal hearts, the blood that returns from the body through the right side of the heart and pulmonary artery forward it to lungs to get oxygen. The blood then comes to the left side of the heart and exits the aorta to the body. In transposition, the blood goes to the lungs and then picks up oxygen, returns to the heart, and then flow right back to the lungs without ever going to the body. The blood from the body returns to the heart without picking up oxygen from the lungs. Symptoms appear at birth or very soon afterward. The severity of the symptoms depends on the type and size of the additional heart defects and the amount of blood can mix between the two abnormal circulations [1- 4].

The major anatomical classifications in the case of transposition depend on the relationship of the great arteries with each other. In approximately 60% of the patients, the aorta is anterior and to the right of the pulmonary artery, Dextro-transposition of great arteries (Dextro-TGA). In some cases the aorta may be on the anterior and to the left of pulmonary artery, Levo-transposition of great arteries (Levo-TGA) [3].

The very first case of TGA was recorded in 1797 by Matthew Baillie, a Scottish physician and pathologist. His most marvelous work is *The Morbid Anatomy of Some of the Most Important Parts of the Human Body* in 1793. Among TGA he is also credited for the discovery of Situs versus [4].

Incidence

The heart with atrio-ventricular concordance and ventriculo-arterial discordance represent 5-7% of all congenital heart disease, corresponding to an incidence of 20-30 per 100,000 live births. This is mostly seen in males than the females. Some of the reasons are rubella, or other viral infection. It is also occurred due to alcohol consumption and diabetes. Also it affects people who are over the age of 40 [5, 6].

Ontogenesis of the normal development of the Great vessels

During embryonic development, the truncus arteriosus is a structure present of the developing heart. It is an arterial trunk that originates from the ventricles of the heart and later divides into the aorta and the pulmonary artery. The Truncus Arteriosus is always linked along with bulbus cordis which give rise to the ventricles. The truncus arteriosus and bulbus cordis is aortic-pulmonary septum [6]. Truncus Arteriosus occurs when the blood vessel exiting from the heart fails to separate completely during the development, leaving a connection between the aorta and pulmonary artery. There is a hole at the bottom of the two chambers known as ventricular septal defect. In truncus arteriosus, deoxygenated blood and oxygenated blood are mixed together as it flows to the lungs and the rest of the body. As a result, a lot of blood goes to the lungs while the heart works harder to pump blood to the rest of the body [7].

Partition of the Truncus Arteriosus and bulbus cordis

While the partition of the primitive ventricle is taking place in the main part of the heart, the Truncus Arteriosus is being divided into two separate channels. In a manner similar to that of the endocardial cushions, mesenchymal cells derived from local endocardium and also from the region of the aortic arches invade the cardiac jelly.

This process starts in the ventral aortic root which is situated between the fourth and sixth arches. Continuing toward the ventricle, the division is effected by the Continuing toward the ventricle, the division is effected by the formation of longitudinal ridges of readily molded young connective tissue of the same type as that making up the endocardial cushions of the atrioventricular canal. These ridges, called trunco-conal ridges, bulge progressively farther into the lumen of the Truncus Arteriosus and finally meet to separate it into aortic and pulmonary channels. The trunco-conal ridge follows a spiral course, also called aortico-pulmonary septum the spiraling aortic-pulmonary septum position the aorta in the left ventricle and the pulmonary artery in the right ventricle [8].

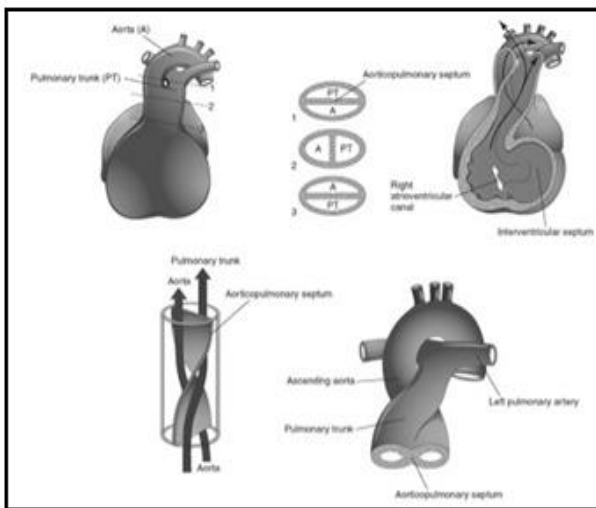


Fig 1: Normal embryology of the heart .

Onto-genesis of the abnormal development of the Great vessels

The congenital anomalies are the most common causes for the death in infants less than one year of age [9-18]. During the stage of partitioning of the bulbus cordis and truncus arteriosus, the aortic-pulmonary septum fails to follow a spiral pattern. The non-spiral septum results in failure of the conus arteriosus to develop during transformation of the bulbus into the ventricles [19]. Historically, knowledge of the embryogenesis of any heart defects was difficult to attain because of the lack of models based on the subjected anomalies. It was really difficult to obtain definite model of the malformations since it is not consistent in animals as well as in humans.

During the eight weeks of development of the heart, the abnormal starts at the mid of the developing period. It allows the aorta and pulmonary artery to get attached to the incorrect chamber. There is not just one type of abnormal switching, but many. These anomalies are named based on their way of switching of the pulmonary trunk and the aorta.

The two clinical conditions based on the morphological defect of the great arteries are:

Dextro-TGA, (D-TGA) results in twisting of the heart tube towards the right and also due to the abnormal growth and development of the bilateral sub arterial conus. Dextro-TGA is due to the abnormal development of the bilateral sub-arterial conus. The sub-pulmonary conus is reabsorbed, which allows for posterior shifting of the pulmonary valve and

development of fibrous continuity between the pulmonary and mitral valve.

The unabsorbed sub-aortic conus make the aortic valve to move anteriorly where it abnormally engages with right ventricle [20, 21].

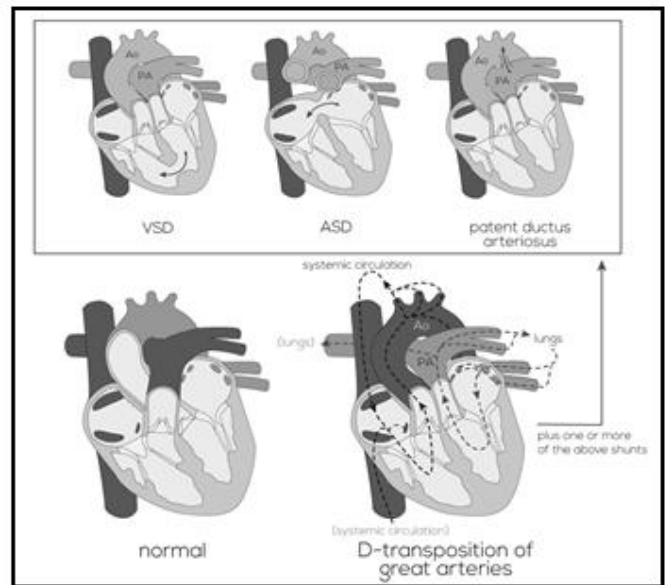


Fig 2: Diagrammatic representation of D-TGA heart.

Levo-TGA or congenitally corrected transposition of the great arteries (CC-TGA) or (L-TGA) is due to looping of the primordial heart tube to the left instead of the right. In case of Levo-TGA, looping takes place in the left side (Levo or L-loop) leading to abnormal positioning of the ventricles and abnormal connections among the atrium, ventricle, pulmonary artery and aorta [22].

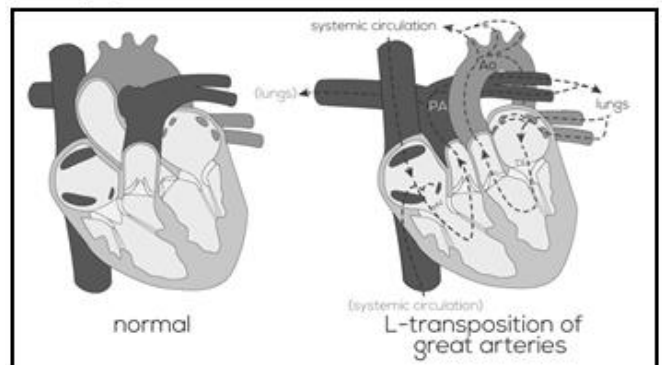


Fig 3: Diagrammatic representation of L-TGA heart.

In Situs-Inversus, mal-position of the heart can be explained with its development. There are two rotational movements involved during the development of the heart. Based on the researches done, in the first phase of embryonic development, there is a 180° rotation around a transverse body axis, which brings the heart to the future thoracic region. During the second rotation around the sagittal and longitudinal axis, it results in the heart being properly positioned in the mediastinum.

Discussion

In order to prove the existence of transposition, numerous experiments were done in order to get more acquainted with this disease. So the attempts were done all over the globe. During the time 1957-1964 in California there were 742 recorded cases on transposition of great arteries. Among them 669 were dead and the remaining 75 were alive thus concluded the study. Copies of all death certificates of the 8-

year period 1957-1964 in which the cause of death was coded as congenital disease (International Statistical Classification 754.0 to 754.7) were secured from the Bureau of Vital Statistics of the California State Department of Public Health.' In addition, the California Adaptation of International Statistical Classification Code 759.5 for Multiple Congenital Defects was utilized to screen for additional congenital heart cases. Fetal deaths were due to the malformations of the cardiovascular system from 1959 to 1963. Of 79 fetal cases, eight were of transposition of the great arteries. The life span in transposition anomalies is very low compared to other anomalies. It lasts from first month of birth to one year from birth.

There are other types of anomalies in the transposition. One such study taken was, Thirty-three cases of corrected transposition of the great vessels were analyzed. Among them, 31 of corrected transposition in situs solitus, and the remaining two of corrected transposition in situs inversus (Dextrocardia). In 18 of these 33 cases, of which 64% were males, diagnosis was confirmed by operation. In all this studies and experiments it has been proven that the transposition of great arteries is rare, congenital as well as fatal. In the case of Dextrocardia the whole position of the circulatory system is reversed but it doesn't affect the normal circulation, so it is a harmless anomaly, but in case if the patient has to undergo a transplantation of the heart it is very difficult to the surgery since they have to reverse the whole schematic, so people with Dextrocardia should be more careful than people with Sinus Solitus.

Conclusion

Heart is one of the fragile organs in the body that needs care all around the clock. Even a slight ignorance in taking care of it can lead to fatal issues. Transposition of great arteries is one among the congenital anomalies of the heart. As the name indicates it is seen in infants between the first month and the first year after birth. Sometimes it is not fatal and the person lives a long life. Whereas in some cases; it is fatal because of their living styles, habits and the condition of their abnormality. Since this anomaly is rare, there isn't much awareness in some parts of the world. In some countries, they do have a counter measure to this disease. In this article, I have done a review of this anomaly in my own words. This article mainly summarizes the anomalies of the great arteries based on its embryological significances. This article states the different types of transposition of the great arteries. Though this anomaly sounds deadly, due to latest improvements in the field of modern technology, this disease can be cured if necessary.

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