Clinical History:

We describe the case of a 30-year-old female G1P0A0 who presented to our department for a routine second trimester ultrasound at 22 weeks of gestation.

No history of consanguinity. No medical or surgical history.

Uneventful pregnancy so far, with normal previous blood tests and foetal ultrasounds.

Imaging Findings:

Ultrasound was performed following the guidelines presented by the International Society of Ultrasound in Obstetrics and Gynecology (ISUOG) for screening of the foetal heart.

Analysis of the four-chamber view was of primordial importance.

Foetal situs and cardiac axis were normal.

There were four identified cavities, however, the moderator band and a heavily trabeculated wall, the hallmarks of a right ventricle, were not seen anteriorly close to the chest. Instead, the ventricle closer to the chest wall had a smoother contour and proved to be the left ventricle (Fig.1).

Further analysis of the heart chambers demonstrated that the left atrium, receiving the pulmonary veins, was connected to the morphological right ventricle through the tricuspid valve (Fig. 2).

Outflow tract analysis demonstrated corrected transposition of great arteries with the aorta rising from the morphological right ventricle and the pulmonary trunk rising from the morphological left ventricle. Both vessels were lying parallel to each other.

Discussion:

Congenitally corrected transposition of the great vessels, also known, as “double discordance” is a rare congenital anomaly with incidence reported as 1 in 33000 live births, representing about 0.05% of cardiac congenital anomalies [1, 2]. It is characterised by atrioventricular and ventriculoarterial discordance.

During embryological development of the heart, the primary heart tube undergoes a left turn instead of the normal right turn, resulting in discordant atrioventricular and ventriculo-arterial connections. In this setting, a morphological right atrium (mRA) will be seen connecting through the mitral valve to a morphological left ventricle (mLV) leading into the pulmonary trunk. A morphological left atrium (mLA) will be seen connecting through the tricuspid valve with a morphological right ventricle (mRV) into the aorta (Fig. 3).

It is unknown why such an abnormal twisting of the primary heart tube occurs, however studies have shown some
increased incidence due to environmental factors (hair dye and air pollutants) [3] as well as among families with previous cases of congenitally corrected transposition of the great arteries, with recurrence risks of 2% to 5%.

This “double discordance” of the heart cavities, when isolated, will be completely asymptomatic because of a restored normal physiology and a correct oxygen rich blood flow, hence the term “corrected”. However many associated anomalies (most commonly ventricular septal defects and obstruction of the pulmonary artery outflow tracts) have been described and their seemingly “normal” physiology resulted in many cases in heart failure and cardiac rhythm anomalies. Other associations include Ebstein’s anomaly and coarctation of the aorta [4].

Multiple long-term complications have been described. First, there is an abnormal position of the atrioventricular node and abnormal course of the conduction axis, resulting in an increased risk of developing heart block over time (2%/year) reaching 30% in adulthood. Progressive prolongation of the PR interval is seen until complete heart block develops [5, 6].

Second, there is gradual dysfunction of the systemic right ventricle after the second decade of life because, histologically and morphologically, the right ventricle cannot sustain the demands of a systemic ventricle [6]. This is explained by different myocytic arrangements in the right and left ventricles as well as ischaemia of the myocardium from abnormals myocardial perfusion [7]. Finally, the left tricuspid valve will progressively become incompetent after the second decade of life [6].

Prenatal diagnosis of this entity is particularly difficult when isolated. It is vital to stress the importance of the four-chamber view and the adequate analysis of the ventricular morphology in order diagnose the anomaly if suspected [8].

**Differential Diagnosis List:** Congenitally corrected transposition of the great arteries or double discordance., Transposition of the great arteries., Double discordance as part of a spectrum of congenital cardiac anomalies.

**Final Diagnosis:** Congenitally corrected transposition of the great arteries or double discordance.

**References:**


Description: Schematic representation of the congenitally corrected transposition of the great arteries or "double discordance": Atrioventricular and ventriculoarterial discordance, restoring normal physiology.

Origin: Helou N, Department of Radiology, Abou Jaoude Hospital, Jal El Dib, Lebanon
Description: The moderator band (asterisk) and the heavily trabeculated wall, hallmarks of the right ventricle (RV) were seen farther away from the chest wall (red line). Note the offset of the valves (dotted lines). Origin: Helou N, Department of Radiology, Abou Jaoude Hospital, Jal El Dib, Lebanon
**Description:** Normal disposition of the ventricles and valves. The right ventricle (RV) with its moderator band (asterisk) is seen closer to the chest wall (red line). **Origin:** Helou N, Department of Radiology, Abou Jaoude Hospital, Jal El Dib, Lebanon
Description: The left atrium (LA), which receives the pulmonary veins, two of which are seen in this image (arrows) is seen connecting through the tricuspid valve (dotted line) with the right ventricle.

Origin: Helou N, Department of Radiology, Abou Jaoude Hospital, Jal El Dib, Lebanon
Description: Normal disposition of the cavities with a left atrium (LA) seen connecting through the mitral valve (solid line) with the left ventricle. Origin: Helou N, Department of Radiology, Abou Jaoude Hospital, Jal El Dib, Lebanon