Can we Build a House From one Brick?:
Diagnosis of TGV Diagnosis from a Single Stored Heart Volume -

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ABSTRACT

We present a case in which D-TGV was diagnosed by off-line analysis from only one stored 3D volume. To our knowledge, this is the first study to reconstruct the cardiovascular anatomy and establish the diagnosis of complex cardiac malformation by offline analysis of only one stored 3D volume.

INTRODUCTION

The dextro-transposition of the great vessels is a structural heart defect with an atrio-ventricular concordance and ventriculo-arterial discordance, it is the most common type of transposition of, the aorta arises from the right ventricle, and the pulmonary artery arises from the left ventricle. It the second most common neonatal cyanotic congenital heart disease representing 5–7% of all CHDs [1]. D-TGV is a critical heart defect lesion that should be diagnosed prenatally, as allows for scheduling of delivery in a specialist center with the possibility to offer urgent Balloon Atrial Septostomy (BAS) when necessary. The sequelae of TVG could be severe if undiagnosed prenatally with mortality approaching 85% to 90% (85-90%) if not treated [2].

Despite the progress in prenatal diagnosis, antenatal detection rate of TGA/IVS has improved but still remains less than 50% of patients [3].

CASE

A pregnant patient at 22 weeks gestation who complained of uterine contractions or abdominal pain arrived at the hospital emergency department. During the initial ultrasound examination performed by an experienced sonographer who was in charge, interventricular communication was observed, volume data is stored, and the sonographer had to discontinue the examination due to an emergency cesarean section. The patient did not show up for her appointment at the prenatal diagnostic unit and could not be reached after several attempts. We have tried to gather as much information as possible from the single stored volume of the 4-chamber view. Offline analysis of the only volume database that we had showed situs solitus, levocardia with a normal cardiac axis. In retrospective view, the four-chamber view revealed atrioventricular concordance, normal atrioventricular valves, and well-sized ventricles. The Pulmonary Artery (PA) originating from the left ventricle, the aorta of the right, continues upwards forming an arch that gives off branches with loss of the normal crossover, vessel 1 originating from the inferior vena cava through the supra diaphragmatic part of IVC connecting the RA by DV (figure 1 A,B) then we were able to reconstruct the 3-dimensional anatomy allowing remote diagnosis and facilitating scientific cooperation between high and low-income countries [7]. The medical and surgical management of TGA is well-established world is associated with an early survival of 85% [8]. On the other hand, lack of detection may increase morbidity and mortality rate [9]. The diagnosis of fetal TGA is closely related to operator skill and expertise and necessitates thorough knowledge of cardiac anatomy to allow accurate diagnosis and optimal management [10].

The cornerstone of diagnosis of TGV is based on the assessment of ventricular outflow tract and great vessels [11]. By definition TGV means discordant connections between the ventricles and the great arteries, consequently, to establish a diagnosis it is necessary to highlight the anatomic features of both ventricles and great vessels. From the only volume stored we followed the segmental approach described in the literature [10]. We have reconstructed four-chamber views in such a way that the tip of the heart is oriented either to the left, levocardia to the right dextrocardia, i.e. indeterminate situs, (figure 1 A,B) then we were able to reconstruct the 3-dimensional anatomy in a sagittal section clearly showing a situs solitus with the point of the heart facing the left side, gall bladder on the right, stomach on the left and the liver is right-sided. (figure 1 C,D). From the stored volume we were able to demonstrate right ventricular moderator band, fibromuscular structures that traverse the cavity of the right ventricles (figure 1 C,E, figure 2B), left atrial appendage that has a broad-based triangular appearance (figure 1 C,E); in addition the supra diaphragmatic part of IVC connecting the RA by DV (figure 2D), in addition to Eustachian Valve (EV) is an embryonic structure redirecting the blood flow from the inferior vena cava through the foramen oval. (figure 2E).

We identified the left atrial appendage which is characteristically a slender finger-like with the presence of foramen oval flap (red arrow), and pulmonary veins draining into LA (figure 2 C,E).

Ventricular Outflow Tract Reconstruction showed parallel great vessels with loss of the normal crossover, vessel 1 originating from the right, continues upwards forming an arch that gives off branches then passes downward (descending aorta) towards the abdominal cavity, along the spine. The other vessel 2 arises from the left ventricle recognized by the anterior right muscle, gives a bifurcation, i.e. the pulmonary artery (figure 2 A,B,C). 4 chamber view have shown a ventriculocavitary septal defect VSD, atrial septal defect ADS, in addition, we have demonstrated the more apical position of the tricuspid valve (figure1 E).
Figure 1: A, B, C, D, E  Rendered volume from the same volume dataset. (One volume).
A, B- Situs Cardiac position could levocardia or dextrocardia, i.e. indeterminate situs. See D.
MB** indicates morphologic Right Ventricle. VSD >, ASD >.
C-Note that the RA appendage is triangular in shape and is connected to anatomically right ventricle recognized by moderator band. whereas the LA appendage is fingerlike, receives four pulmonary veins, PV.
D- Volume-rendered images Situs solitus. The heart and stomach are on the left; the gallbladder is on the right; the liver is right-sided.
E-4 chamber view showing RV recognized by MB connected to RA, LV connected to LA recognized with FO “flap” in the LA <<, and pulmonary veins PV. Note the presence of VSD and ASD. Moreover, the tricuspid atrioventricular valve more apically positioned than the Mitral valve.
F- Four-chamber view showing normal heart anatomy in a normal fetus at 30 weeks of gestation note the intact IVS, IAS, TV more apically positioned than the MV (control case).
Abbreviations: AO, aorta; FO, foramen oval, TV, tricuspid valve, MV, mitral valve, RV, right ventricle, VL, left ventricle, SP, septum premium, SS, septum secundum, SVC, superior vena cava, PA, pulmonary artery, AO, aorta MB, moderator band. PV; Pulmonary vein. LPSVC: Left Persistent Superior Vena Cava.
From a single stored volume we were able to establish the diagnosis of d-TGV, which was confirmed by subsequent ultrasound examinations. In this context we would like to emphasize the importance of properly differentiating between d-TGV which requires immediate postnatal care in a specialized center and corrected L-TGA, in which there is an atrioventricular discordance and does not require immediate intervention as typically unaffected until later in life [12]. From the subsequent ultrasound examinations of this patient we were able to confirm the diagnosis, in addition, displaying cardiovascular anatomy using Power Doppler 3D Modes, moreover comparing with standard ultrasound images of our patients serving as control cases to further clarify the diagnostic usefulness of 3DUS and doppler angiography and help to improve its antenatal diagnosis, which remains modest despite the development in this domain, as half of cases of TVG are still not diagnosed antenatal (figure 1F,2F,3DE,4BD,5B).

From subsequent ultrasound studies of the same patient we demonstrated three signs:

1. "boomerang sign": an abnormal right convexity of the aorta arising from the RV instead of the normal convexity to the left of the pulmonary artery arises from the RV observed in normal hearts rightward convex curvature of the RV outflow "boomerang sign" [13] which is considered a reliable clue for diagnosing TGA (figure 4B, C, D, F).

2. "baby bird’s beak image" described by McGahan JP et al, the pulmonary artery arises from the LV, the left branch makes a sharp angle with the main pulmonary artery and ductus arteriosus with the image of its bifurcation resembling the head of a baby bird with an open beak [14] (figure 3).

3. “big-eyed frog”, the great vessels were disposed side by side, resembling a Japanese fictional character, created by Sanrio, called “Keroppi” [15,16] (figure 4 G, I).

4. At last, we would like to point out that all the images in this study were carried out with a Voluson 730 Pro (General Electric, Milwaukee, WI, USA) with a volumetric abdominal transducer (4–8 MHz), which does not contain the STIC software, i.e., without having access to STIC confirming the inherent capabilities of 3DUS, in addition we highlight the importance of using this means to send a stored volume by internet to experts for remote diagnosis.

CONCLUSION

To our knowledge, this is the first study to reconstruct the cardiovascular anatomy and establish the diagnosis of complex cardiac malformation by offline analysis of only one stored 3D volume. 3D ultrasound offers a high-resolution volume rendering image that provides excellent delineation of cardiac anatomy and add significantly to detection and understanding of the Cardiovascular
anomalies. Offline analysis of cardiovascular anomalies conferred significant diagnostic advantages over standard 2D and represent an invaluable tool for the prenatal diagnosis and optimal management of fetuses with congenital heart diseases. This technology enabling worldwide remote diagnosis especially underserved area not having access to facilities allowing such a diagnosis and enhance scientific cooperation between high and low-income countries.
**REFERENCES**


