Case Report

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

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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% 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 ventricle and the aorta was to the left of the PA confirming the presence of TGA. Ultrasound images were then sent via the Internet to a reference center for prenatal diagnosis (CHU de Caen, France), and the initial diagnosis was confirmed by a multidisciplinary team comprising a pediatric cardiologist, and a pediatric cardiac surgeon. The patient could have finally been reached and she was followed in our antenatal unit and subsequent ultrasound examinations confirmed the diagnosis. The family was informed in detail of the postnatal corrective surgical treatment available. Amniocentesis was performed and revealed a normal male karyotype. The diagnosis of TGV was confirmed after birth. The patient was sent to deliver in a university teaching hospital in Paris. She gave birth to a full-term male baby weighing 3.1 kg. At birth, he had an atrial septostomy. Prostaglandin E1 was administered initially but was discontinued after the septostomy. The child was operated (Arterial Switch (ASO)). The operative consequences were simple. He was followed until the age of 7, he was in good health.
Figure 1: A, B, C, D, E  Rendered volume from the same volume dataset. (One volume).

A, B- Situs Cardiac position could levocardi 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- Four-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).

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

### Figure 2: A, B, C, D, E  
Rendered volume from the same volume dataset
- **A**: Outflow tract views show the parallelism of the great arteries in transposition of the great arteries (aorta [AO], anterior [1] and Pulmonary Artery [PA], posterior [2]), rather than crossing. The great arteries arising from the ventricles. The first vessel (1 AO) gives off a cross with 2 branches seen.
- **B**: Vessel 1 (AO) originates from the morphologically right ventricle recognized by moderator band >>. Vessel 2 (PA) which presents as a bifurcation (star) arises from a morphologically left ventricle as recognized by anterior papillary muscle >.
- **C**: Vessle 1 giving off a cross with branches and a descending branch confirming this as aorta, this means that the morphologic Right Ventricle (RV) supports the transposed aorta.
- **D**: The ductus venosus links the umbilical vein to the inferior vena cava and drains just distal to its connection to the right atrium, left hepatic veins drain into the inferior vena cava.
- **E**: The same anatomy, note the presence of Eustachian Valve, The Eustachian Valve (EV) is an embryonic structure redirecting the blood flow from the inferior vena cava through the foramen ovale.
- **F**: 3D power Doppler ultrasound of a control case of our patients above mentioned anatomical findings.
- **G**: Abbreviations: IVC, inferior vena cava; LA, left atrium; LHV, left hepatic vein;
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.

Figure 3: ‘baby bird’s beak image’
A,B: The pulmonary artery bifurcation is shown, and the left branch pulmonary artery makes a sharp angle with the main pulmonary artery and ductus arteriosus, reminiscent of a baby bird’s head with an open beak.
D,E: Normal bifurcation of PA. (control case).

Figure 4: Same patient on subsequent examinations except B,C.
A- Glass body-rendering mode reconstruction of the outflow tracts off-line ventricular septal defect. Transposition of great arteries (star denotes ventricular septal defect). Note the abnormal arrangement of 3VV, where aorta is anterior and to the right of pulmonary artery.
B- 3D power doppler reconstruction 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).
C, E – Reconstruction of normal heart anatomy note crossover of outflow tracts, normal arrangement of the 3VV the aorta is to the left of PA.), Note leftward convexity of the pulmonary artery arising from the right ventricle and presence of 4 vessels in this view. The fourth vessel is Left Persistent Superior Vena Cava.
D,F - Rightward convexity of the great artery arising from the RV (aorta) was a reliable clue for diagnosing TGA. rightward convex curvature of the RV outflow “boomerang sign”.
G,I - The “big-eyed frog” sign, the main Pulmonary Artery (PA) is situated side by side with the Aorta (Ao), resembling the Sanrio frog character (“Keroppi”)
REFERENCES


