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Case Report

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

Wael El Guindi[1], Michel Dreyfus[2], Gabriel Carles[1], Nidal Alassas[3]

[1]*Department of Obstetrics and Gynecology, Franck Joly Hospital, Saint-Laurent-du-Maroni, France*

[2]*Department of Obstetrics and Gynecology, Caen University Hospital, Caen, France*

[3]*Department of Obstetrics and Gynecology, CHI Redon-Carentoir, France*

***Address for Correspondences:** Wael El Guindi, Department of Obstetrics and Gynecology, Franck Joly Hospital, Saint-Laurent-du-Maroni, France, Paris, Tel. No: 0774253858; E-mail: welguindi@yahoo.com

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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 3 D 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%a (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 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.

DISCUSSION

The introduction of 3- and 4-dimensional (3D / 4D) ultrasound, in particular Spatial And Temporal Image Correlation (STIC) about 15 years ago [4], has added several advantages to fetal echocardiography, in particular the ability to extract a large number of ultrasound plans from 3D / 4D volumes. The integration of 3D / 4D into fetal echocardiography has been used to display images in surface mode with two-dimensional (2D) and color Doppler [5], to extract one or more planes for tomographic view [6], thus allowing the offline examination of the heart 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 fives 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 ventricular septal defect VSD, atrial septal defect ADS, in addition, we have demonstrated the more apical position of the tricuspid valve (figure1 E).

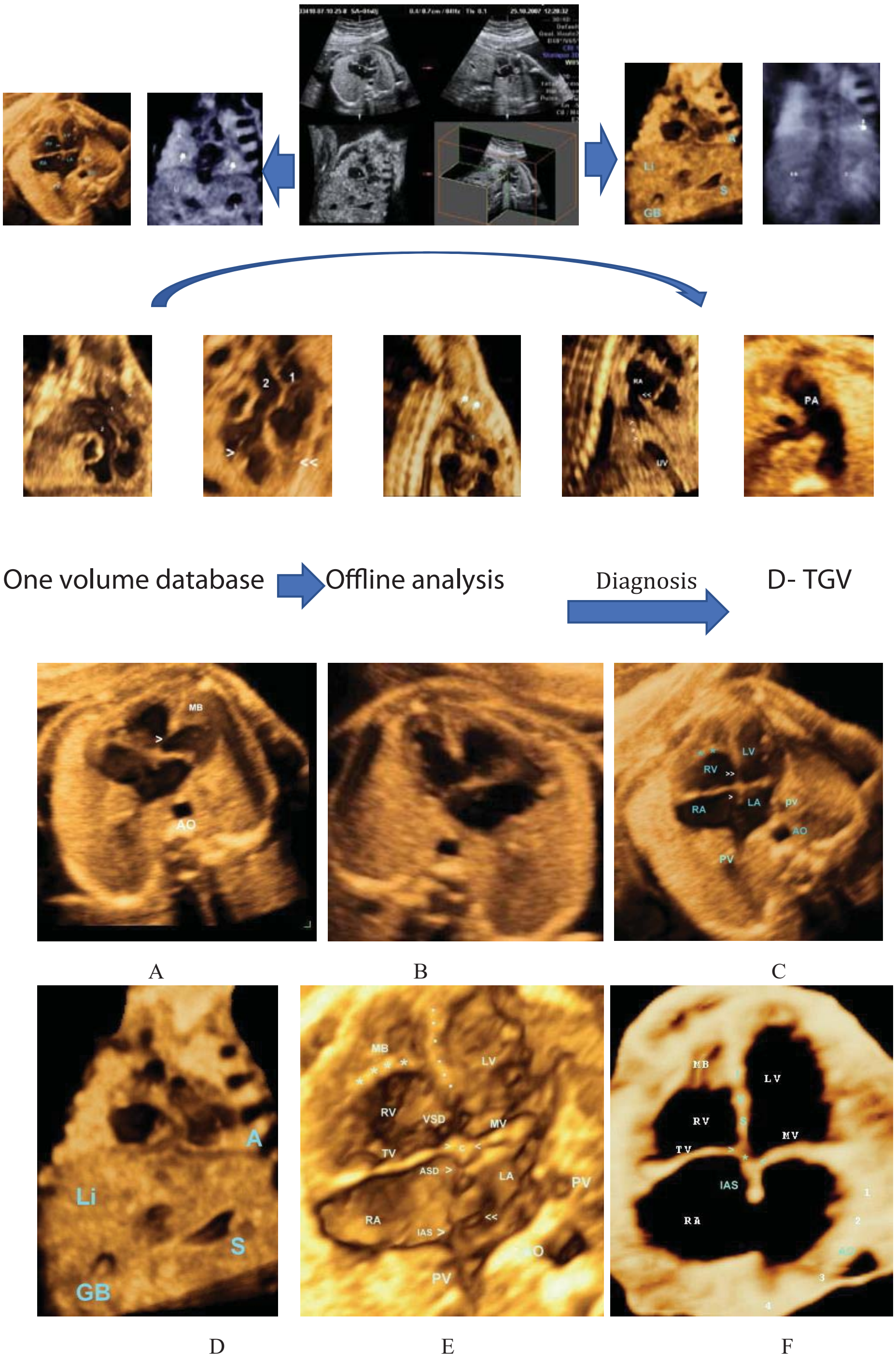

Figure 1: A, B, C, D, E Rendered volume from the same volume dataset. (One volume).
A, B- Situs Cardiac position could levocardia or dextrocardia, i.e. indeterminate situs. See D.
MB** indicates morphologic Right Ventricle. VSD >>, ASD >.
C-Note that the RA appendage is triangular in shape and is connected 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.

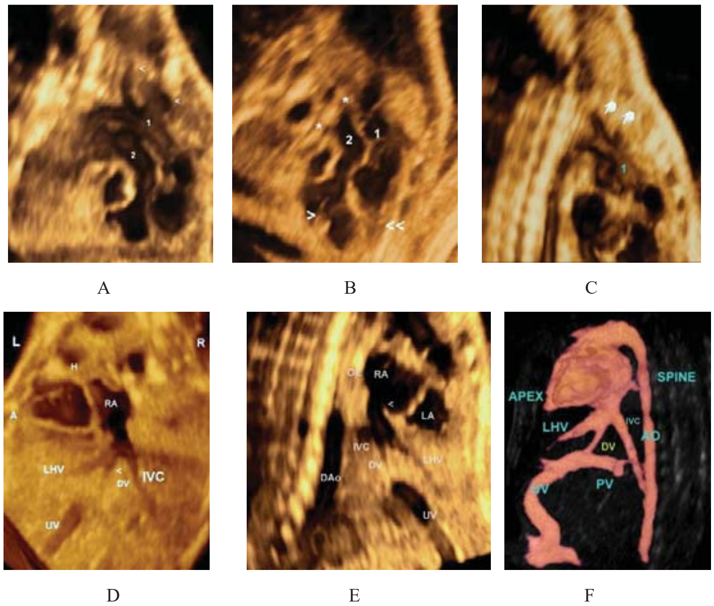

Figure 2: A, B, C, D, E Rendered volume from the same volume dataset
A- Outflow tract views show the parallelism of the great arteries in transposition of the great arteries (aorta [AO], anterior [1] and Pulmonary Artery [PA], posterior [2]), rather than crossing, great arteries arising from the ventricles. 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;

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 antennal 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 4 B, 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 high lightened 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 3 D 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 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).

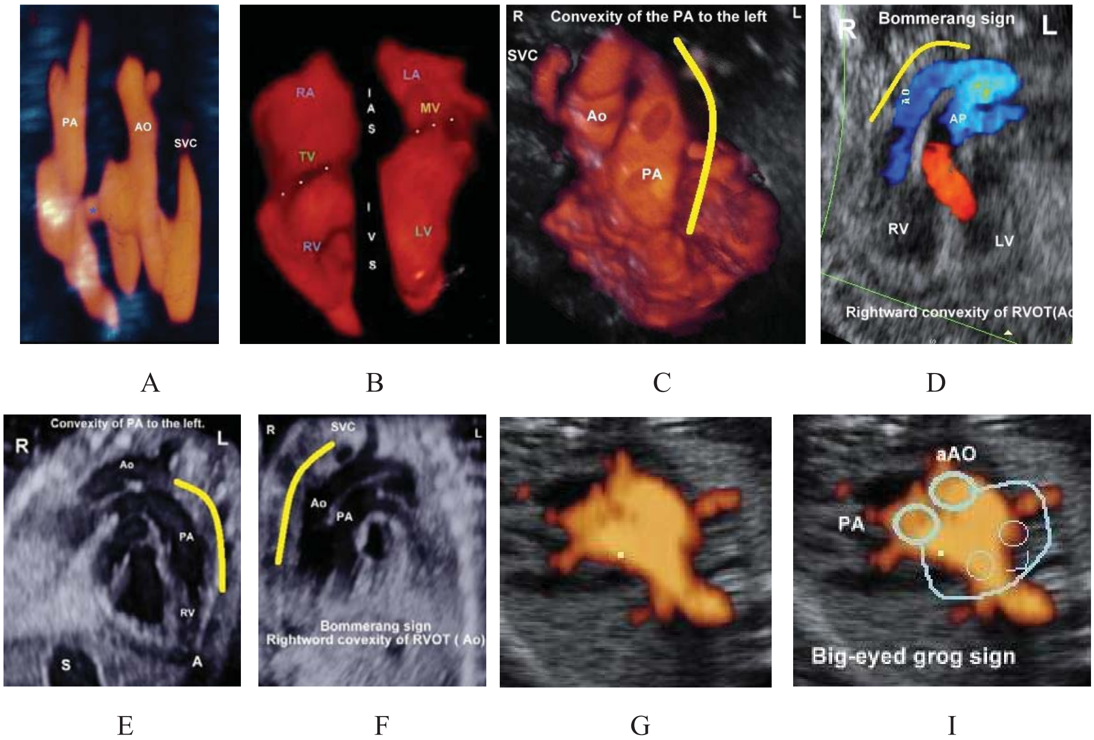

Figure 4: Same patient on subsequent examinations except B,C.
A- Glass body-rendering mode reconstruction of the outflow tracts off-line ventricular septal defect. Transposition of great arteries (star denotes ventricular septal defect). Note the abnormal arrangement of 3VV, where aorta is anterior and 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")

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.

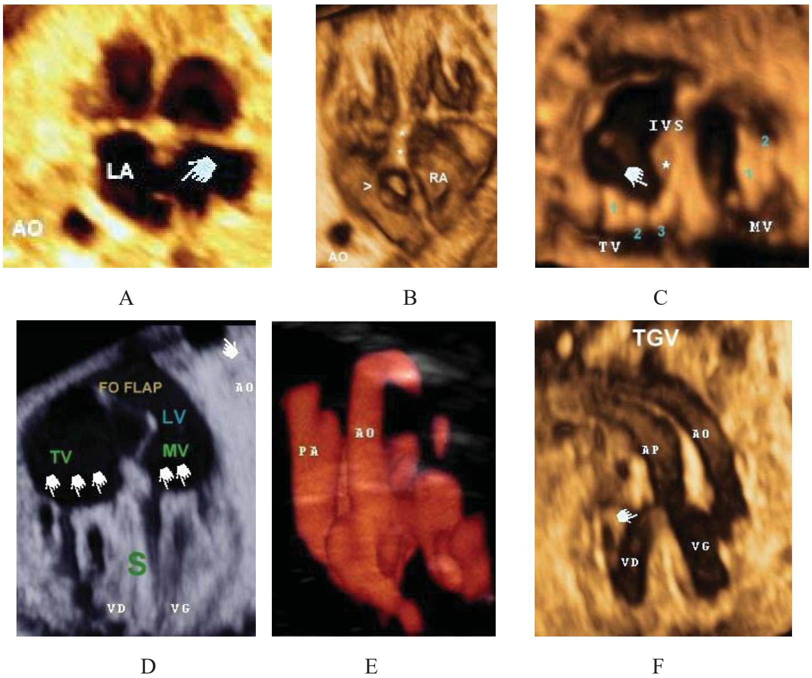

Figure 5: Same patient on subsequent examinations excep B,D (case control).
A-Volume-rendered image from a subsequent US examination showing 4 chamber view with FO flap (hand) in the LA.
B - Four-chamber view showing foramen ovale in a normal fetus at 30 weeks of gestation. (case control).
C,D-Volume-rendered image of the same patient in a subsequent ultrasound examination (C) and a control patient (D) showing the Tricuspid Valve (TV) that consists of 3 leaflets , with the characteriatic septal leaflet (hand-star), the mitral valve has 2 leaflets with no attachments to the septum.
E,F Volume-rendered images from the same volume dataset showing showing transposition of the great vessels (TGV), hand indicates VSD confirmed by color doppller ,F.
LA, left atrium; MV: mitral valve; LV: Left Ventricle; IVS: Interventricular Septum; R, Right Side; RA, Right Atrium; T, Tricuspid Valve , RV: Right Ventricle. FO: Foramen Oval. RA: Right Atrium.

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