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Case Report

Computed Tomography Cardiac Veins Evaluation in Congenital Transposition of Great Vessels Prior to Cardiac Resynchronization Therapy (Crt)

Groudeva $V^{1\ast},$ Chilingirova $N^2,$ Nedevska M^1 and Iovev S^2

¹Department of diagnostic imaging, University Hospital " Prof. Dr. Alexander Chirkov", Medical University Sofia, Bulgaria ²Department of cardiology, University Hospital " Prof. Dr. Alexander Chirkov", Medical University Sofia, Bulgaria

*Corresponding author:

Violeta Groudeva,

Department of diagnostic imaging, University Hospital "Prof. Dr. Alexander Chirkov", Medical University Sofia, Bulgaria,

E-mail: violetagroudeva@gmail.com

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1. Abstract

Adult patients with transposition of great vessels are often candidates for cardiac resynchronization therapy (CRT). Cardiac vein anatomy is of crucial importance in planning optimal CRT therapy. Cardiac veins are very variable. In congenitally corrected transposition of great vessels coronary arteries have an unusual course but coronary veins although much less studied have much more varieties. In such cases cardiac computed tomography (CT) might offer important information prior to electrophysiological procedures. The objective of this study is to present a series of patients with transposition of the great arteries (TGA) both congenitally corrected TGA and D TGA in which imaging was used to help planning placement of leads for CRT. CT findings are thoroughly described as well as the consequent interventional procedure.

2. Introduction

Patients with congenital transposition of great arteries often develop heart failure and need biventricular pacing. CRT is feasible in cases provided favourable venous natomy. The complex anatomy in congenital transposition of the great arteries both L TGA and D TGA is a challenge for treatment. Computed tomography can reveal detailed venous anatomy that can assist decision making in planning CRT therapy. We present 4 cases with TGA in whom cardiac CT was performed prior to the procedure and CRT was successfully implanted.

3. Cases Presentation

We observed 4 patients with TGA for a period between January 2021 and June 2022. Three of them were with congenitally corrected TGA and one with D TGA and single ventricle (large VSD and small rudimentary right ventricle). Three of these patients were

indicated and underwent successful CRT procedure. In all patients cardiac CT with 320 row detector machine Acquillione ONE, Canon was performed. In two of the patient's standard institutional protocol for cardiac CT was performed and in the other two patients additional late venous acquisition phase was performed. Image analyses for coronary arteries and veins was performed on a dedicated workstation with Vitrea software using systolic and diastolic dataset.

3.1. First Patient

35-year-old female with congenitally corrected TGA. Patient complained of palpitations, chest discomfort and fatigue. She reported presyncopal symptoms but no real syncope was realized. Holter ECG revealed cardiac pauses up to 3 secs during the night and single ventricular extrasystoles. ECG at admission showed sinus rhythm, total AV block and ventricular extrasystoles. Echocardiography revealed situs solutes, double discordance corresponding to congenitally corrected TGA.

Cardiac CT findings: Functionally left ventricle had the typical morphology of right ventricle. The ventricle was dilated, trabeculated, with moderator band identifiable. The ventricle was connected with morphologically left atrium. Low position of atrioventricular valve was noted. The aorta was connected with the morphologic right ventricle. Thus atrioventricular and ventriculoarterial disconcordance were present. The aortic valve was located ventrally and to the left of pulmonary valve. Coronary anomaly was detected. A single coronary artery arose from the right located coronary sinus. Right coronary artery (RCA) derived and run with preaortic course towards the left atrioventricular groove and continuesd as posterior descending artery. Left descending coronary Volume 6 | Issue 9

artery (LAD) run anteriorly to join the anterior atrioventricular groove. Left circumflex artery (LCX) was a small vessel located in the right atrioventricular groove. CT cardiac venous anatomy revealed small sized coronary sinus with typical location and connected with the right atrium. Left circumflex vein with small caliber run in the right atrioventricular groove. A large sized posterior venous branch overlying the morphological right ventricle was identified as suitable for lead placement. The anterior interventricular vein drained directly in the right atrial appendage. Small cardiac veins were identified draining directly to the right atrium (Figure 1). PM mode CRT - D was implanted. The lead was located in posterior vein with stimulation rate 1.0V. Right ventricular defibrillating lead was placed in typical location in the apex of the functionally right ventricle (Figure 2).

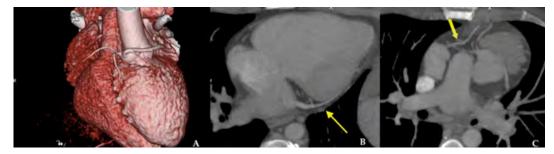


Figure 1: A. Cardiac CT. Volume rendered reconstruction demonstrating all coronary arteries derive from the the right situated coronary sinus. RCA runs with a preaortic course towards to the left AV sulcus to continue as posterior descending artery in the posterior interventricular sulcus. LAD runs in the anterior interventricular sulcus. LCX is a small vessel running in the right AV sulcus.

B. Delayed phase. Large sized posterior venous branch overlying the morphological right ventricle / arrow/

C. Delayed phase. Anterior interventricular vein draining directly in the right atrial appendage / arrow /

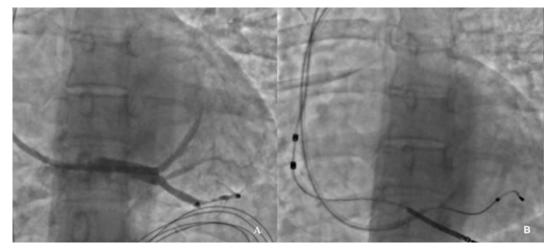


Figure 2: A. Retrograde angiography in LAO - 35 - posterior branch of CS.

B. Bipolar lead placed in the posterior branch of the CS, with good stimulation parameters of the functionally left ventricle (with the morphology of a right ventricle). Defibrillating lead Dx placed in typical position - apex of the functionally right ventricle (with the morphology of a right ventricle).

3.2. Second Patient

38-year-old male with congenitally corrected TGA, large VSD. Past surgery for bending of pulmonary artery at the age of 1 years old. Reoperated with plastic of ventricular septal defect and debanding of the pulmonary artery performed at the age of 12. Covid 19 infection was documented a year prior to study. Patient complained of fatigue and dyspnea during regular physical activities and exertion angina. Limited functional capacity and postprandial malaise were also present. There was evidence of chronic atrial fibrillation Heart failure NYHA III. Echocardiography showed systemic ventricle ejection fraction of 38%. CT findings: The heart was enlarged, with both atria dilated. The atrioventricular valves were situated in the same plane. The systemic ventricle was dilated with morphology of right ventricle - trabeculated with moderator band. Thickness of myocardium was 7,5 mm. Coronary arteries had typical appearance for ccTGA with right coronary artery (RCA) on the left. Right coronary dominance.

Coronary veins - the coronary sinus drained into the right atrium. A venous vessel run in the right atrioventricular groove - left circumflex vein. A large lateral venous vessel overriding the functionally right ventricle was present (Figure 3). A large sized middle cardiac vein running along the base of the heart reaching the apicolateral part of the systemic ventricle. There was no venous vessel along the left ratio ventricular groove. CRT -P was successfully implanted. Quadripolar LV lead is places in the middle cardiac vein (Figure 4).



Figure 3: A. Cardiac CT. Delayed phase. A large lateral venous vessel overriding the functionally right ventricle and therefore not suitable for lead placement for stimulation of the systemic ventricle. The vessel drained into left circumflex vein situated in the right AV sulcus

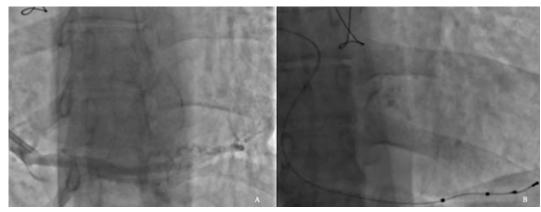


Figure 4: A. Retrograde angiography in LAO - 35 - middle cardiac vein

B. Quadripolar lead placed in the middle cardiac vein with good stimulation parameters of systemic right ventricle

3.3. Third Patient

55 years old female with complex cyanotic congenital heart malformation - D -TGA and double inlet left ventricle. High grade subpulmonary valvular stenosis. Small PDA. Prior haemodilution. Heart failure NYHA III. Patient presented with chronic atrial fibrillation, monotropic ventricular extra systole, secondary repolarizaiton changes, single ventricular extrasystoles. Atrial septal aneurism with small perforations was also identified on echocardiography.

Cardiac CT clearly revealed the anatomy. The atrioventricular

valves were situated in the same plane. Atrioventricular concordance was present with the atria connecting to the corresponding chamber. Large size subvalvular ventricular septal defect was observed making the impression of a single ventricle with partial septum towards the apex. Common ventricle was formed from dilated morphologically left ventricle situated on the left and rudimentary, small RV situated to the right. Common ventricle was with reduced EF - 39%. The aorta was connected with the rudimentary right ventricle. Pulmonary artery was connected with the left ventricle with subpulmonary stenosis present (Figure 5).

Coronary artery anatomy showed non-coronary sinus situated anteriorly and coronary sinuses posteriorly. There was right coronary dominance. Coronary veins - coronary sinus was normally positioned on the base of the heart draining into the right atrium on its posteromedial site. Two posterior venous branches draining together into the coronary sinus. An anterolateral branch was also visible. CRT -P was successfully performed with a LV lead placed in an anterolateral venous branch (Figure 6).

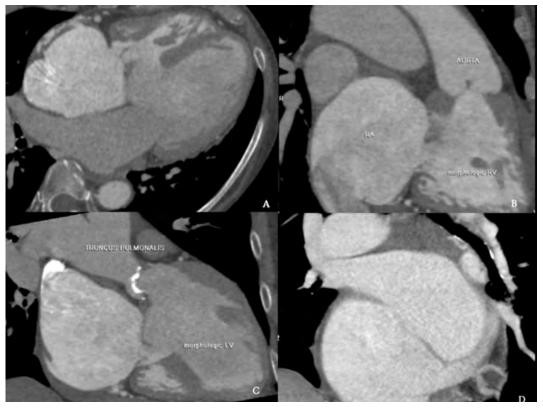


Figure 5: A. Cardiac CT. Four chamber view. Enlarged left situated morphologically left ventricle and a small rudimentary right ventricle. Large inerventricular septal defect.

- B. Morphologically RV is connected to the aorta
- C. Morphologically LV is connected to the pulmonary artery
- D. Coronary sinus draining into the right atrium

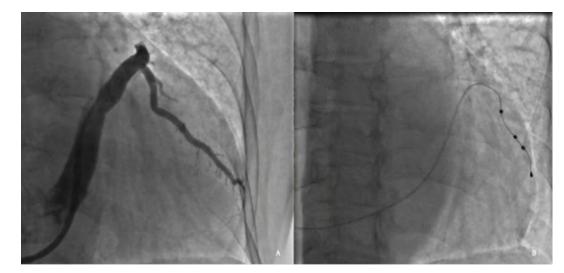


Figure 6: A. Retrograde angiography in LAO - 35 - antero-lateral branch B. Quadripolar lead for placed in the antero-lateral branch with pacing the common chamber with LV function

3.4. Fourth Patient

Female patient with ccTGA and prior surgery - left atrioventricular valve repair. Cardiac CT showed dilated heart with enlarged atria. Atrioventricular discordance. The left ventricle was also dilated. The aortic valve was tricuspid with noncompete coaptation of the leaflets. Ejection fraction was calculated as 51%. Coronary arteries had typical appearance for ccTGA. Coronary veins - the coronary sinus had normal position on the base of the heart and draining into the right atrium. A large posterolateral venous brach overlying the right systemic ventricle was identified. The patient did not meet the criteria for CRT. Conservative medical treatment and strict cardiological follow up were recommended.

4. Discussion

Transposition of great vessels (TGA) is rare congenital heart disease. Congenitally corrected TGA is also known as L- transposition and is characterised by both atrioventricular and ventriculoarterial discordance. Thus in ccTGA although the malformation is characterised by normal functioning circulation, the abnormal morphology of the cardiac chambers often leads to dysfunction. Additional malformations in these patients are often found such as ventricular septal defect, pulmonary stenosis or even atresia. Usually patients are diagnosed early in life due to the concomitant anomalies. However, the malformation may remain silent until adulthood especially in cases where there are no additional abnormalities. The main problem in such patients is that because the morphologically right ventricle has to serve as a systemic ventricle over time it is prone to failure due to difficulty to tolerate the increased pressure in the systemic circulation. Apart from increased risk for right heart dysfunction, a big variety of arrhythmias are described [10,13,14]. Typical association in ccTGA is conductive problems such as high degree AV block. Others include AV reentrant tachycardia mediated by left-sided bypass tract, atrial tachvarrhythmias and rarely AV nodal re-entrant tachycardia. Conelly et all in their study showed that 69% of adults with ccTGA have some form of rhythm disturbance [15]. In ccTGA there is a higher incidence of left -sided accessory pathways.

CRT is more and more used to improve quality of life and reduce mortality [6-9]. Consensus statement recommends CRT in patient with EF equal or below 35%, QRS duration equal or above 150ms and ventricular dilatation [1]. However, in cases of congenital heart disease such as TGA guidelines are not conclusive. One of the reason is the scarce amount of reported cases of TGA undergoing CRT therapy. Therefore, European Cardiology guideline on management of grown-up congenital heart disease consider CRT as an 'experimental" therapy [16]. This substantiate the importance of communicating experience with patients with TGA undergoing CRT.

A well-known fact is that optimal CRT requires cardiac venous anatomy assessment. A crucial prerequisite for optimal therapeutical result is lead placement in the region with ventricle dyssynch-

nony. So far the concept was to place the lead along the lateral wall of the systemic ventricle. However, this is possible only in case suitable venous vessel with sufficient calibre is present. Cardiac venous anatomy is very variable. Comparing to the coronary artery morphology, cardiac veins are much less studied. In the last decade the interest in cardiac venous anatomy is increasing with the advance in the interventional techniques using the coronary sinus as an access route to guide interventions. There are different classifications and nomenclature for coronary vein mainly based on the anatomy. The classical anatomic classification divides the cardiac veins into two main groups: tributaries of the greater cardiac venous system (CVS) and tributaries of the lesser CVS, which consists of the Thebesian vessels. The greater CVS is subdivided into two groups: coronary sinus and non- coronary sinus tributaries [2,3]. A supplemental scheme of classification is shown to be even more important in interventions and electrophysiological procedures and can be easily adopted in reporting venous vessels in cardiac CT. In addition to the classical naming of venous vessels on the basis of their origin, they can additionally be described according to the areas of myocardium they overly. In this respect, emphasis is placed on the systemic ventricular myocardium, which is target for ventricular pacing. In such segmental classification the left ventricular myocardium is divided into 9 segments - anterior, lateral, and posterior segments, with additional recognition of basal, mid, and apical portions. The system can then be used to describe the drainage through the primary, secondary, and tertiary branches of the coronary venous system [4]. It is now well recognized that optimal pacing requires appropriate placement of the pacing leads. Clear description of the veins available for leads placement enhances the choice of the electrophysiologist. CT has the potential to reveal detailed morphological information allowing for correlation of the classically described cardiac vein and its unique area of overlying the ventricular myocardium as well as vessels tortuosity and angulation.

It has been shown that coronary arteries are most often inverted in TGA with the morphologically right coronary artery on the left, arising from the left located coronary sinus and the morphologically left coronary artery respectively on the right. The non-coronary sinus in such cases is located anteriorly as opposed to the normal anatomy in which it is found posteriorly [10, 11, 12]. Also coronary artery anomalies are not unfrequent. In a study Ueura et all found that coronary anomalous course was evident in 24% of the cases. In one of our patients we also observed a coronary anomaly, namely a single coronary artery giving rise to both left and right coronary branches which is reported in the literature as the most common coronary anomaly in this patient population [5].

In TGA - the coronary vein is more anomalous [5]. One of the largest study at the coronary venous circulation in ccTGA is publish by Bottega et al. [10]. They found that in ccTGA, the CS develops with the morphologic atria and therefore lies on the side of the

left atrium, the venous branches, including the great cardiac vein, lateral branches, etc., develop with the morphologic ventricles and overly the corresponding morphological ventricle [10]. The coronary sinus is generally located on the side of the morphologically left atrium and drains in the posteromedial aspect of the right atrium as present in all our patients. It receives the morphologically right ventricular and inferior interventricular veins as opposed to the normal hearts in which the CS receives the left ventricular and interventricular veins. The later in ccTGA often drain directly to the right atrium or some of them may sometimes drain through the coronary sinus. The anterior interventricular vein in normal hearts is part of the great cardiac system and drains through the CS. In TGA the anterior interventricular vein may continue in the right atrioventricular groove, between the morphologically right atrium and morphologically left ventricle, often named as left circumflex vein. In such case from left circumflex vein originate the lateral veins. This anatomy was present in our second patient in whom a large sized lateral vein was present. However, in such cases as the lateral vein originating form left circumflex vein overlies the morphologically left, but nonsystemic ventricle, this vein is of no interest for lead placement. Another possibility is the anterior inteventricular vein to drain directly in the right atrial appendage as in patient number one. Although coronary sinus in TGA is draining into the RA, there are reports in the literature of difficulties in its cannulation mainly due to abrnomal location of the CS orifice relative to the eusthachian valve The morphologiclly right ventricular veins which are the target for lead placement in CRT can be readily identified on cardiac CT as in all of all patients. In the literature the morphologiclly right ventricular veins are reported to be less, or smaller caliber and length [4]. Most often they drain via the coronary sinus. Detailed description of number and position adjacent to the morphologically right ventricle as well as size, tortuosity and vessel angulation is crucial before CRT and further might decrease significantly procedural time. All this makes cardiac CT a strategical tool for preprocedural planning.

Imaging evaluation of cardiac veins are by far overshadowed by the coronary arteries. Cardiac CT angiography has been developed firstly as a noninvasive assess coronary artery stenosis and occlusions in the last decades there is a continuously growing interest for noninvasive imaging modalities prior to procedures for planning interventions. Cardiac CT can assess normal anatomy and variations of the vascular structures, assess normal anatomy and variations of the vascular structures. The utility of CT has been evaluated to optimize planning and lead placement during cardiac resynchronization therapy. Although optimal venous opacification needs adjustment of protocol with a little delay from the standard arterial angiography, even standard cardiac CT is generally able to reveal the morphology of the cardiac venous system. With the increase number of CT performed for different clinical indications it is important for radiologists to be familiar with the different pattern of cardiac venous system as in TGA and to be able to identify the venous vessels on cardiac CT.

5. Conclusion

Cardiac CT is a noninvasive tool able to demonstrate not only the cardiac structural abnormalities, the atrioventricular and venticuloarterial disconcodrance in patients with transposition of great vessels, but also the anatomy of the coronary arterial and venous vessels. The last one is a valuable information prior to planning CRT therapy.

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