The role of MDCT in management of arrhythmias in congenital and acquired pediatric heart diseases

Essay

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Radiodiagnosis

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Introduction

Due to the rapid evolution of the treatment of arrhythmias by catheter ablation which attacks the anatomical foci of arrhythmias by the radio frequency, cardiologist are in need to a road map for the heart structures before intervention which could be afforded by conventional catheterization methods, CT or MRI.

In comparison to the conventional catheterization method MDCT overcomes the limitations such as the overlap of the adjacent cardiovascular structures, difficulties in simultaneously depicting the systemic and the pulmonary vascular systems, the catheter-related complications ***(Haramati et al., 2002),*** especially the prolonged time of exposure in ablation of the arrhythmogenic foci ***(Efstathios et al., 2006),*** also it gives simultaneous evaluation of the cardiovascular structures and the lung parenchyma which is important in patients with congenital heart diseases ***(Leschka et al., 2007),*** obtains functional data about motion of the ventricular wall.

In comparison to MRI, MDCT has the advantages of widespread availability, the shortest acquisition times and the highest spatial resolution, also MDCT has the least cut thickness and less time-consuming, with MDCT there is no requirement of a lengthy period of patient sedation especially in congenital cases, uncooperative or seriously ill patients, MDCT has more accurate evaluation of any associated coronary artery abnormalities ***(Frank et al., 2010), (Manghat et al., 2005),*** MDCT is the appropriate method in patients who are absolutely contraindicated for MRI like patient with pacemaker devices, ICD, old artificial valves and being better in evaluation of patients with metallic stents, coils and clips which can lead to magnetic susceptibility artifacts.

The drawbacks of MDCT include exposure of the patient to ionizing radiation yet its dose could be decreased to submillisivert dose with the dedicated low dose CT protocols, another drawback would include the risks of complications due to exposure to iodinated contrast material.

Arrhythmia now represents one of the most common long term complications in congenital heart diseases patients and adult with congenital heart diseases, posing new challenges in terms of management, rhythm anomalies can be a part of the natural history of the underlying defect itself, but most of them arise from consequences of surgical interventions, from longstanding hemodynamic abnormalities or as an association with the defect.

The most frequent arrhythmia in CHD lesions is Intra-atrial re-entrant tachycardia (IART) which could be associated with atrial septal defect (ASD), tetralogy of Fallot (TOF), Fontan, Mustard and Ebstein anomaly ***(Be´dard et al., 2008).***

Aim of this study is to discuss the role of MDCT in drawing the road map for the catheter ablation for arrhythmogenic foci which come as a complication or association of the congenital heart defects, this will be explained through the discussion, also will be emphasized the ability of MDCT to evaluate and measure the pulmonary veins ostia, mitral isthmus, cavo-tricuspid isthmus (CTI), septal isthmus, thebesian valve and subthebesian pouch, coronary sinus, posterior interventricular vein, left atrium auricle, inferior vena cava (IVC), interatrial septum, phrenic nerves, left atrial size, Koch’s triangle, atrio-ventricular node (AVN) and artery to AVN, sino-atrial node (SAN) and artery to SAN, Crista terminalis.

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9. The relevant radiological anatomy to the electrophysiological studies by the MDCT:

A-supraventricular structures

Pre- and postprocedural examinations with MDCT are frequently performed to detect the anatomy, obtain baseline measurements of the heart to help the cardiologist to select the proper way for ablation, the proper size of catheter, early detection of complications from ablation ***(Ghaye et al., 2003)*** and additional need for certain procedures like left atrial appendage (LAA) exclusion ***(Cabrera et al., 2014).***

1. Pulmonary veins :

Embryology, During the first 4 weeks of fetal development the lungs drain to the systemic veins and the common primitive pulmonary vein comes from the dorsal wall of the atria before separation of them, Then it is shifted to the left to be on the left side of septum primum, When the primitive lungs are fused to the common pulmonary vein, The connections that allow pulmonary venous return to the systemic veins are obliterated ***(Demos et al., 2004), (Dillman et al., 2009).***

Further incorporation of the common PV takes place as the atrial chamber continues to grow, Abnormal resorption of the embryonic structures can lead to (a) Alterations in the diameter or the number of pulmonary veins, (b) Abnormal drainage to the systemic veins or right atrium.

In 70% of the general population there are four pulmonary veins (PVs), Two superior and two inferior with four independent ostia, The superior PVs enter the mediastinum downward and anterior to their accompanying pulmonary arteries, The left superior PV being longer than the right, The inferior PVs are oriented upward and located below their bronchi ***(Chung et al., 2002).***

The ostia of the superior PVs are ventral and lateral in relation to the inferior PVs, And the ostia of the left PVs are located higher than those of the right PVs ***(Demos et al., 2004) and (Kato et al., 2003).***

The PVs are not round but oval , The anteroposterior diameter is smaller than the superoinferior diameter, especially in the left veins, The diameter of the PVs generally increases as they approach the left atrium, with the exception of the left inferior pulmonary vein, the diameter of which usually decreases as it enters the atrium, a finding that must be taken into consideration when evaluating possible stenosis at this location ***(Kim et al., 2005).***

The diameters of the superior PVs are larger than the inferior PVs, In addition, variations may occur in the diameter and position of the pulmonary vein ostia according to the phase of the cardiac cycle ***(Lacomis et al., 2007).***

The right superior PV drains the right upper and middle lobes, The left superior PV drains the left upper lobe and lingual and the two inferior PVs drain the lower lobes.

Unlike the intrapulmonary portion of the pulmonary arteries, the pulmonary veins are not located near the bronchi instead, the veins follow the course of the intersegmental septa, So the pulmonary veins can be differentiated from the segmental arteries, which run adjacent to the corresponding bronchus ***(Kirsch et al., 2011).***

The occurrence of more than three PVs on the same side has been described rarely.

Another type of variations involves the return of the middle lobe PV which is drained in right superior PV in 53%–69% of cases, via an independent ostium in 17%–23% of cases or into the right inferior PV in 3%–8% of cases ***(Tsao et al., 2001) and (Yazar et al., 2002).***

Lingual vein in the left side also is seen draining into the left superior PV in most cases while it is also seen draining into the left inferior PV in 2.5% of cases ***(Sugimoto et al., 1998).***

These ectopic variants may be responsible for atrial fibrillation foci that have been successfully ablated ***(Tsao et al., 2001)*** (Figure 2,3).

The left superior PV is the most common focus of atrial fibrillation, and the right inferior pulmonary vein is the least common ***(D'Avila et al., 2003).***

During the embryonic development of PVs, the incorporation of the PV confluence into the dorsal left atrium is followed by the muscularization or atrialization of the venous part of the atrial wall ***(Bliss et al., 1995)***, The resultant myocardium that surrounds atrial-venous junctions extends from the left atrium into the adventitia of the PVs ***(Moubarak et al., 2000)***, These myocardial sleeves are distinct from the smooth muscle in the media of the PVs and are circularly or spirally oriented, forming sphincter-like structures, They are more developed and longer around the superior PVs than around the inferior PVs ***(Nathan et al., 1966),*** The sleeves are thickest at the atrial-venous junctions in the left superior PV, At the veno-atrial junctions, there are abundant autonomic nerve bundles and intrinsic ganglionated nerve plexuses situated in epicardial fat pads ***(Haïssaguerre et al., 1998), (Tan et al., 2006),*** Antigen expression of myocardium similar to that of the cardiac conduction system (the autorhythmic cells) has been shown to occur around the common PV, from which the four PVs originate ***(Blom et al., 1999).***

The point of muscular discontinuity between the PV and atrium muscle is the factor that affect the origin, the risk for arrhythmia and the propagation as the sudden discontinuity leads to an increase in the risk of reentry, While the gradual discontinuation and increased gap of muscular disconnection between the PV and the left atrium decreases the risk for reentry propagation, and this is the idea of PVs ostial ablation and isolation (to increase the gap so prevents the connection between the arrhythmogenic foci in the PVs and the atrium) ***(Tan et al., 2006)*** (figure 1).

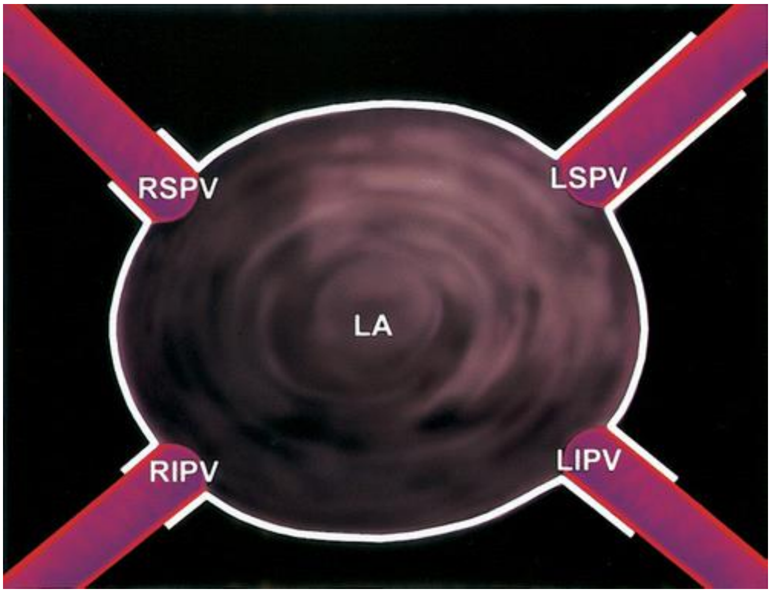

Figure 1: Left atrium and myocardial sleeves of the distal pulmonary veins. Schematic illustrates how the atrial myocardium (white outline) of the left atrium (LA) extends into the distal pulmonary veins. Note that the sleeves of the left-sided veins are longer than those of the right-sided veins and that the left superior pulmonary vein (LSPV) has the longest myocardial sleeve. LIPV = left inferior pulmonary vein, RIPV = right inferior pulmonary vein, RSPV = right superior pulmonary vein. ***(Lacomis et al., 2003).***

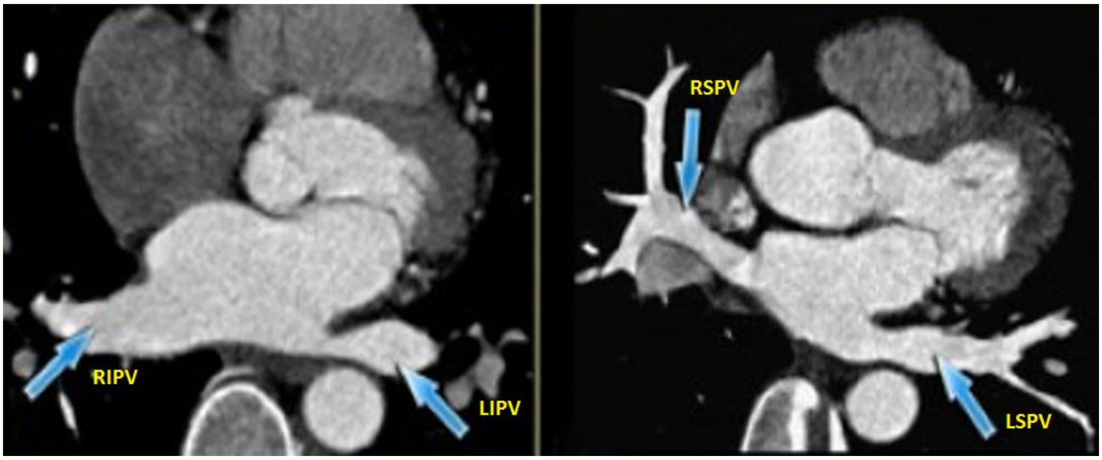

Figure 2: Axial reconstructions showing the pulmonary veins (blue arrows) as they enter the left atrium (Quoted from: ***radiology assistant***).

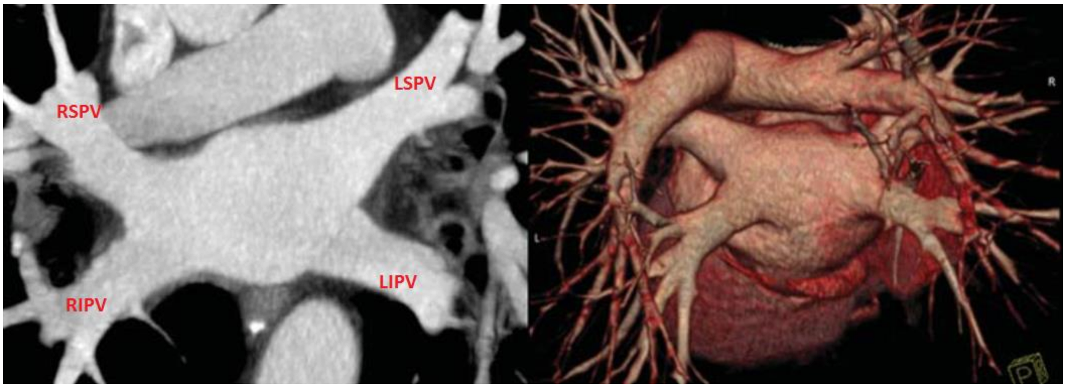

Figure 3: Coronal cut and 3D volume rendering show the 4 pulmonary veins and left atrium ***(Abecasis et al., 2009).***

Figure 4: Segments of the distal pulmonary veins. Slab reformatted image shows the V1 through V3 segments ***(Lacomis et al., 2003).***

2-Left atrium:

Like the right atrium, the left atrium consists of a **venous** component, a **vestibule,** and an **appendage** ***(Morady., 2004), (Quintana and Ho, 2003).*** The venous component, with the pulmonary vein orifices at each corner, is located posteriorly, The vestibular component surrounds the mitral orifice (figure 5).

The left atrial appendage (LAA) (figure 6,12) is derived from the primitive atrium and has a rough, trabeculated surface, The tip of the appendage points anterior and cephalad in most cases and overlaps the base of the pulmonary trunk, the left coronary artery, or its anterior descending branch and the great cardiac vein, Also it is closely related to the left phrenic nerve and its accompanying pericardiophrenic vessels ***(Su et al., 2008) and (Quintana et al., 2009).***

The majority of the left atrium, including the venous component, the vestibule, and the septal component, is smooth walled.

The anterior wall of the left atrium just behind the aorta is usually thin and vulnerable to be perforated. The superior wall, or dome, is the thickest. The posterior wall of the left atrium tends to become thinner toward the orifices of the left and right pulmonary veins ***(Ho et al., 1999)***, Where as the wall is thinner in the middle and between the inferior venous orifices in those with atrial fibrillation ***(Platonov et al., 2008).***

Left atrial isthmus (figure 11) is the area between the inferior left pulmonary vein and the mitral orifice it is a focus of AF and MDCT can accurately demonstrates the boundaries of this area, including the exact location of the mitral valve ring, coronary sinus, and great cardiac vein as well as their anatomic variants ***(Chiang et al., 2006).***

The left lateral ridge (figure 6,7) between the orifices of the left PVs and the mouth of the left atrial appendage is the most relevant structural prominence on the LA endocardium. This structure is actually an infolding of the lateral atrial wall protruding into the endocardial LA surface as a prominent crest or ridge. The ridge extends along the lateral wall of the LA from the anterosuperior to the postero-inferior region and frequently diagnosed as intra atrial mass by the echocardiography. Within this fold, multiple structures run including the remnant of the vein of Marshall, together with abundant autonomic nerve bundles and a small atrial artery (which in some cases is the sinus nodal artery) ***(Cabrera et al., 2008).***

The esophagus is in contact with the posterior wall of the left atrium, Its course may be more toward the right or the left pulmonary venous orifices or more centrally. The esophagus curves to lie beneath the postero-inferior wall as it descends toward the diaphragm and may be only a couple of millimeters away from the pulmonary venous orifices and practically in contact with the coronary sinus ***(Tsao et al., 2006).***

The Coronary sinus is surrounded by a cuff of myocardial extension, Myocardial connections varying in number and morphology leave this coronary muscle cuff and connect to the LA. The ostium of the CS pushes the inferior paraseptal mitral annulus in the pyramidal space.

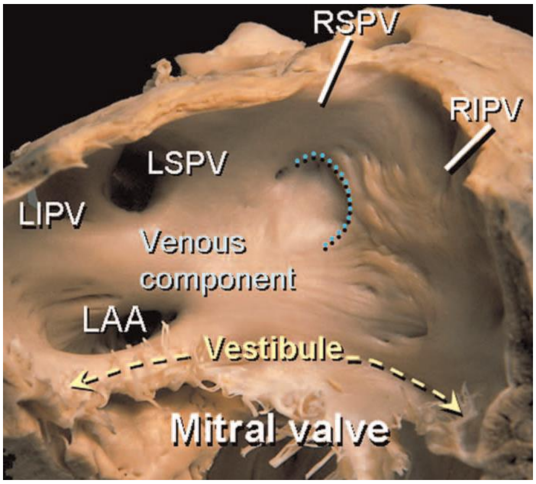

Figure 5: Dissection of the posterior wall of the left atrium (LA) close to the posterior interatrial groove. The smooth walled venous component of the LA is the most extensive. The septal aspect of the LA shows the crescentic line of the free edge of the flap valve (green dotted line) against the rim of the oval fossa (OF). The orifices of the right superior and inferior pulmonary veins (RSPV and RIPV) are adjacent to the plane of the septal aspect of the LA ***(Cabrera et al., 2013).***

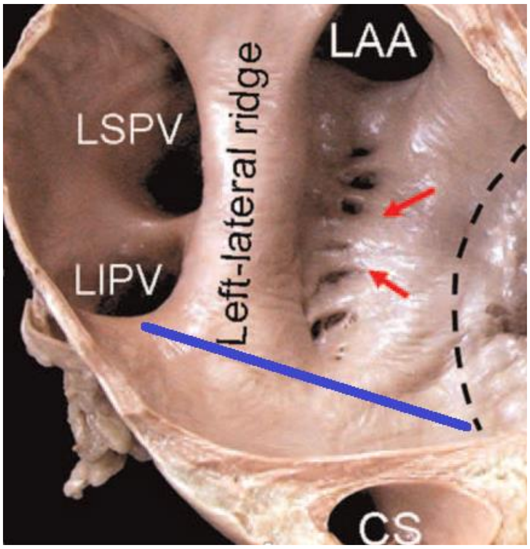

Figure 6: the left atrial appendage (LAA), the mitral isthmus (blue line) with extra appendicular pectinate muscle (red arrows), mitral valve (black interrupted curve), left atrial ridge, coronary sinus CS, left superior pulmonary vein and left inferior pulmonary vein LSPV, LIPV ***(Cabrera et al., 2013)***.

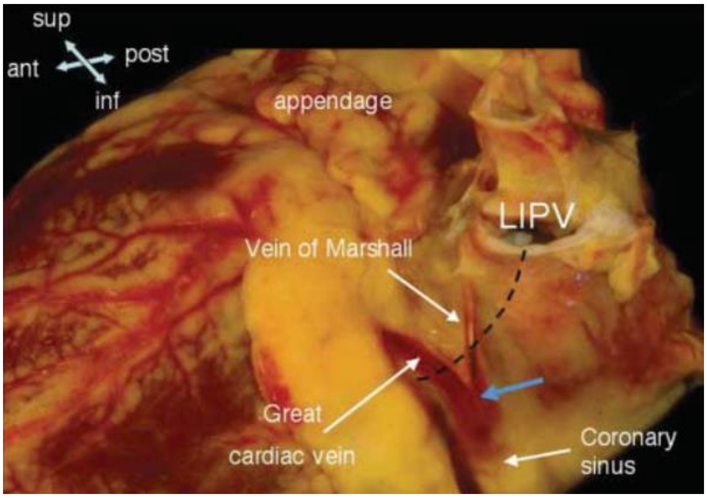

Figure 7: epicardiac view of dissected heart shows the relationship of the mitral isthmus (black dashed line) to the great cardiac vein/coronary sinus transition indicated by the blue arrow (**Wittkampf et al., 2005).**

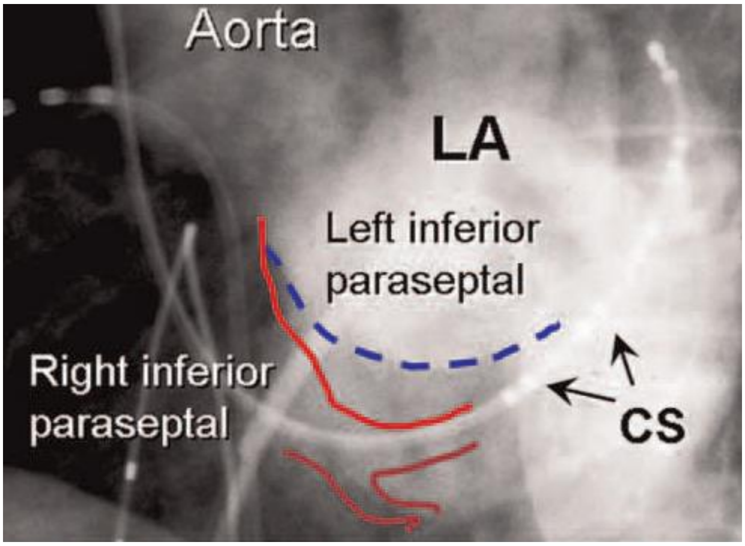

Figure 8: Left atrial angiography throughout a trans-septal puncture in the left anterior oblique projection (LAO). The LAO shows the right and left paraseptal regions, coronary sinus (CS) catheter ***(Cabrera et al., 2013).***