Extracardiac pulmonary-systemic connection via persistent

levoatriocardinal vein in adults

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Abstract:

The levoatriocardinal vein is a very rare but clinically important intrathoracic venous anomaly that connects the systemic (cardinal) and pulmonary venous channels. We report four adults with pulmonary-systemic venous communications that can explain the morphology of the extracardiac interatrial shunting through the persistent levoatriocardinal vein. We discuss the imaging features of the two types of such communications: direct connection of the levoatriocardinal vein a) with the left atrium and b) with the left superior pulmonary vein in the absence of obstructive left heart disease. Accurate characterization of these diagnostically challenging cases is important since in addition to hemodynamic imbalance they are at risk of paradoxical embolism. CT and MRI are noninvasive imaging techniques that should play increasingly important roles in the evaluation of these anomalies.

Introduction:

The levoatriocardinal vein is a rare intrathoracic venous anomaly that connects the systemic (cardinal) and pulmonary venous channels. Since early descriptions approximately 55 cases have been reported in the literature mainly in children with left cardiac obstructive congenital heart disease (1-6). Adult cases with no cardiac symptoms are even rarer and only a few cases have been reported (7, 8).

We have encountered four adults with pulmonary-systemic venous communications that can explain the morphology of the extracardiac interatrial shunting through the persistent levoatriocardinal vein. In this report, we discuss the imaging features of the two types of such

communications: direct connection of the levoatriocardinal vein a) with the left atrium and b) with the left superior pulmonary vein in the absence of obstructive left heart disease. Diagnoses of these cases are important since in addition to hemodynamic imbalance they are at risk of paradoxical embolism.

Cases:

Case 1 (Figure 1). A 62-year-old female was admitted for evaluation of end stage renal disease and advanced cryptogenic cirrhosis for possible liver and renal transplantations. Abdominal CT showed cirrhosis, portal hypertension, and moderate ascites and evidence of prior surgeries including hysterectomy and cholecystectomy. Nuclear cardiac stress test was unremarkable. Transthoracic echocardiography demonstrated mild bilateral atrial enlargement and mild right ventricular enlargement. CT angiography of chest showed trace bilateral pleural effusion, mildly enlarged right atrium and right ventricle, normal sized left heart chambers, and moderately enlarged pulmonary arteries. CT images also revealed a left vertical vein bridging between the left brachiocephalic and left superior pulmonary vein consistent with a levoatriocardinal venous anomaly. No intracardiac shunting was seen. The right heart catheterization for evaluation of pulmonary hypertension revealed a pulmonary mean pressure of 29 mmHg (systolic=34 and diastolic=20 mmHg).

Case 2 (Figure 2). 41 year old male admitted for surgical fixation of multiple pelvic and lower extremity fractures after motor vehicle accident. CT angiography of the chest, abdomen and pelvis did not show major vascular injury. A partial anomalous venous return of the left superior pulmonary vein into a left vertical vein with connection of their common trunk

into the left atrium was shown described as the levoatriocardinal vein (8). Cardiac chambers and the coronary sinus were normal and no intracardiac shunt or any other cardiovascular anomaly was by CT or echocardiography.

Case 3 (Figure 3). 36-year-old female with a history of unrepaired congenitally corrected transposition of the great arteries (CC-TGA) first diagnosed in her early twenties. The patient had a reported history of bilateral superior vena cava (SVC) without bridging between the two SVCs as well as partial anomalous pulmonary venous drainage of the right upper pulmonary vein to the right SVC and absence of the coronary sinus.

Both MRI and transthoracic echocardiogram showed CC-TGA, no intracardiac shunt, absent coronary sinus, and normal systolic function of the systemic and pulmonic ventricles with a systemic right ventricle ejection fraction calculated at 55%-60%. All valves were structurally and functionally normal. The inferior vena cava demonstrated normal size and normal respiratory variation suggestive of normal right atrial pressures. MRI also showed bilateral SVCs with the left-sided SVC draining directly into the left atrium and communication between the two SVCs existed through anterior mediastinal collateral veins as well as by the azygos system causing a bidirectional shunt consistent with diagnosis of the levoatriocardinal vein (Figure 3). No bridging brachiocephalic vein was present. The right SVC also received anomalous pulmonary venous drainage from the right upper lobe.

Case 4 (Figure 4). A 53 year old male, status post repair of tetralogy of Fallot with a prosthetic pulmonary valve, was admitted for further assessment of thromboembolic events. Brain MR and CT images showed chronic cortical brain infarctions related to prior thromboembolic events. MR and CT angiographies as well as echocardiography with agitated bubble showed bilateral SVCs

1 and direct communication of a persistent left SVC with the left atrium with a bidirectional shunt
2 consistent with diagnosis of the levoatriocardinal vein. Although the left brachiocephalic vein
3 was absent, connection between the two SVCs was facilitated by the left azygos system (Figure
4 4). No intracardiac shunt or a patent foramen ovalis was found and the cardiac function was
5 normal. CT angiography of the heart showed an atretic coronary sinus (absent coronary sinus had
6 been documented in the cardiac surgery reports). A right to left shunting through the left
7 levoatriocardinal was diagnosed as the predisposing channel for repeated thromboembolic events
8 and successfully embolized with coils at its entrance into the left atrium.

9 **Discussion:**

10 In embryonic development, while the primitive atrium undergoes septation, the sinus venosus
11 together with its right and left horns comprising of the common cardinal veins from each side
12 become incorporated into the right side of the atrial chamber. The left sinus horn becomes
13 incorporated into the developing left atrioventricular groove as the systemic venous tributaries
14 rotate around the fulcrum provided by the dorsal mesocardial connection (9). Following division
15 of the primitive atrium by the septum primum and vestibular spine, the left sinus horn (with left
16 common cardinal vein) becomes the coronary sinus that opens into the right atrium. The right
17 anterior cardinal venous system eventually develops into the right SVC and the left anterior
18 cardinal venous system remains as the oblique vein of the left atrium draining into the coronary
19 sinus. Within the mediastinal tissues, a mid-pharyngeal strand canalizes to form the primitive
20 pulmonary vein that connects the intraparenchymal pulmonary venous network to the left atrium
21 (10). Four pulmonary venous tributaries of the embryonic pulmonary vein are subsequently
22 assimilated into the atrium. Various abnormal systemic or pulmonary venous connections
23 develop if any of these processes fails to occur properly. The simplest and more common types

are persistent left SVC draining into the coronary sinus (Figure 5) and partial anomalous pulmonary venous return (Figure 3B, right superior pulmonary vein).

Whereas direct connection of the persistent left SVC to the left-sided atrium is not uncommon in hearts with isomeric arrangement of the atrial appendages, it is found only occasionally in hearts that have usual atrial arrangement (situs solitus) and normal pulmonary venous connections. In these, there is usually an associated interatrial communication at the anticipated site of the coronary sinus orifice (coronary sinus atrial septal defect). Rarely, there is direct connection of the left SVC to the left atrium with absence of the coronary sinus and without an atrial septal defect (Cases 3 and 4). This type of connection is classified under "levoatriocardinal vein" (6). The exact mechanism is not clear. Although debatable, Miraldi and colleagues (11) proposed isomerism of the cardinal vein to explain this entity.

The "levoatriocardinal" vein originally is described as a compensatory route for decompression of the pulmonary venous return through the cardinal venous system in patients with obstructive left heart outflow and an intact atrial septum, and that the pulmonary veins are connected to the left atrium but pulmonary venous drainage is abnormal (2, 4-6). Later reports, especially in adults, showed similar anomalous venous connections in patients without intracardiac obstructive lesion (7, 8). Most early case series were in children with congenital heart disease. In the largest series with literature review reported by Bernstein et al. in 1995, mainly in patients less than 2 years old, the most common underlying cardiac anomaly was variations of the hypoplastic left heart syndrome (76%) and cor triatriatum (16%) (6). The atrial septum was functionally intact in most patients. In 68% of their patients the levoatriocardinal vein connected directly to the left atrium whereas the connection was to one of the pulmonary veins in the remaining patients. Drainage of the levoatriocardinal vein was to the left brachiocephalic vein

(48%), right SVC (36%), and jugular vein (4%). Therefore, the levoatriocardinal vein is not necessarily "levo" or "atrial". Earlier case reports (3-5) including the above two articles (2,6) became the basis for the criteria used to define the levoatriocardinal vein as a persistent communication between the cardinal venous system and the left atrium or pulmonary veins in the absence of an atrial septal defect or anomalous pulmonary venous drainage. As shown in Figure 5, a case with a simple persistent left SVC connection to the coronary sinus, the left SVC is closely sandwiched between the left superior pulmonary vein and the left atrial wall. Could defects in the walls or unroofing between the left SVC and the left atrium or left superior pulmonary veins in this region simply open the left SVC into the left atrium or pulmonary vein? The fate of the coronary sinus is not clear and in some cases may not develop normally. In two of our cases (cases #3 and 4), the coronary sinus was not visible and the left SVC directly connected to the roof of the left atrium at the expected site of the left atrial appendage (Figures 3,4). Also shown in cases #3 and 4, was absence of the left brachiocephalic vein to connect the two SVCs, however, SVC connections were shown through alternative routes including the azygos system and mediastinal collateral veins.

The lack of any major intracardiac obstructive lesion in our series who are all adults, and also in some other reported cases (7,8) indicates that decompression of pulmonary venous return to the heart is not the only mechanism for development of this anomaly. Owing to the variable relationship of the anomalous vein dorsal or ventral to the left pulmonary artery, variable origins and insertions, a more appropriate term for these anomalies would be a 'pulmonary-to-systemic collateral vein' regardless of embryonic origins (5). Nevertheless, levoatriocardinal vein, unroofing of the coronary sinus, persistent left SVC probably are the spectrum of a common

developmental defect and are important to distinguish from a vertical ascending vein that exists in the setting of totally anomalous pulmonary venous connections.

In summary, diagnosis of these rare types of the pulmonary-systemic communication has important clinical implications. The potential bidirectional shunt may have hemodynamic consequences. Additionally these patients are predisposed to paradoxical embolism even if the anomalous connection is small. This complication occurred in case#4 that had been missed for years. CT or MR angiography is very important in diagnosis of these cases since it can easily be missed in routine echocardiography studies.

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Figures:

Figure 2. Case#2. 41 year old male admitted for motor vehicle accident. Volume rendered CT angiography views show the levoatriocardinal vein (LACV) connected the left brachiocephalic vein (BCV) with the left atrium via the left superior pulmonary vein (LSPV). There is normal drainage of the left LIPV into the left atrium (LA). The concept of the bidirectional flow of the LSPV is shown by yellow arrows in B. No intracardiac shunting was seen. LIPV= left inferior pulmonary vein, SVC=superior vena cava, LV=left ventricle, RV=right ventricle

Figure 3. Case#3. Cardiac MRI of a 36-year-old female with a history of unrepaired congenitally corrected transposition of the great arteries (CC-TGA). A-C. Coronal MR angiography (MRA) in early (A) and late (B) phases as well as volume rendered left anterior descending (LAO) view

1 (C) of the great vessels are shown. There is direct communication of the levoatriocardinal vein
2 (LACV) to the left atrium (LA) causing right to left shunt. Connections between the between
3 the right SVC (R-SVC) and the LACV are made through anterior mediastinal collateral (Collat.)
4 veins shown in A. Connection of the LACV to the left azygos (L-Az) system as shown in E.
5 provides additional route between the right and left sides. Right to left direction of flow of the
6 LACV is shown by red arrows in A-E. This direction can be reversed in case of increased left
7 heart pressure. Note, partial anomalous pulmonary venous return (APVR) of the right upper lobe
8 into the R-SVC in panel B. F. Four chamber cine MR image shows posterior location of the
9 systemic right ventricle (RV) and discordant atrioventricular connection consistent with CC-
10 TGA. AA=ascending aorta , PA=pulmonary artery, RA=right atrium, LV=left ventricle.

Figure 4. Case#4. 53 year old male, status post repaired tetralogy of Fallot. A. and B. Coronal MR angiography images showing direct communication (red arrow) of the levoatriocardinal vein (LACV) with the left atrium (LA). The communication was coil embolized as the patient had multiple episodes of embolic brain infarctions due to right to left shunting (proved by contrast echocardiography and MRI). Although there was no communication between the right SVC (R-SVC) and LACV at the level of the brachiocephalic vein, this communication existed through the left azygos (A-Az). This is shown in sagittal CT angiography (C) when contrast injection was made in the left arm. The injected contrast entered into left atrium directly (right to left shunt)

and into the right atrium (not shown) indirectly by the left azygos venous channels. D. Posterior view of a color coded CT angiography shows atretic coronary sinus with the inferior interventricular vein (IIV) draining into the right atrium (RA) and the great cardiac vein (GCV) draining into the LA. Ao=aorta, LV=left ventricle.

Figure 5. Volume rendered and sagittal CT angiography showing a typical case of bilateral superior vena cavae (SVC) with the left SVC (L-SVC) draining into the coronary sinus (CS). Note, close association of the left SVC with the left superior pulmonary vein (LSPV) and the left atrium (LA) as it passes between them. Hypothetically, defects between the walls of the left SVC with either of the two structure walls create a systemic to pulmonary circulation connection with the potential for development of a bidirectional shunting. LV=left ventricle, R-SVC= right superior vena cava , RA=right atrium.