Abnormalities of the thoracic veins

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Learning objectives

To present our experience with CT, MRI and DSA in recognising abnormalities of thoracic veins during 12-year period.

To raise awareness of radiologists regarding numerous findings on this mainly asymptomatic but for surgeons, interventional radiologists and anesthesiologists, sometimes unpleasant occurrence.
**Background**

Congenital abnormalities of the thoracic veins are not common, but they represent significant developmental anomalies. Venous abnormalities in the thorax can be classified into systemic and pulmonary. Systemic venous abnormalities are frequent incidental findings, while the pulmonary venous abnormalities are usually manifested with cyanosis due to frequent obstruction of pulmonary venous flow and abnormal flow of blood. They are also associated with congenital cardiac anomalies, particularly with atrial septal defect (ASD).

Patients with congenital thoracic venous abnormalities typically undergo diagnostic algorithm that begins with contrast enhanced CT, echocardiography, sometimes angiography and finally with the MRI. CT with excellent spatial resolution and anatomic delineation has a disadvantage in use of ionizing radiation and the use of contrast agents. Echocardiography as noninvasive method allows detection of venous abnormalities in the thorax, but the inability of ultrasound waves to penetrate the bone and air greatly reduces the size of the acoustic window and thus reduces the diagnostic potential of the modality. Angiography as very invasive method has all the disadvantages as CT. MRI as a noninvasive method with multiplanar capability and use of gradient-echo sequences, especially the phase contrast variant, is able to display the volume and flow of the direction of blood flow. (1)

Usually venous abnormalities in the thorax represent incidental finding during the evaluation of lungs or cardiac abnormalities.

**Abnormalities of systemic thoracal veins**

*Abnormalities of superior vena cava (superior vena cava - SVC)*

Left superior vena cava has prevalence of 0.3% in healthy individuals, and 4.4% in patients with congenital heart disease. The anomaly represents persistent embryonic left anterior cardinal vein. In most patients left SVC courses lateral to the aortic arch, anterior to the left pulmonary hilum and drains into the coronary sinus. Usually in these patients coronary sinus is enlarged. (2, 3) In a small number of patients the anomalous vein instead in coronary sinus drains into the left atrium, creating a right-to-left shunt.

Left SVC is associated with an absence of the left innominate vein in 65% of patients, and in 10% -18% with the absence of right SVC. In patients with double SVC, the diameter of the right SVC is reduced. (4) Left SVC often occurs in association with atrial septal defect and azygos continuation of inferior vena cava (IVC). (5) Isolated congenital abnormalities involving the SVC are rare and include SVC draining of right SVC in the left atrium, and existence of a congenital aneurysm of right SVC. (6)
**Double SVC**

Occurrence of double SVC in the general population is 0.3%, and reaches up to over 11% of the population with congenital heart diseases. (7) Embryological substrate resides on the contemporary development of normal right anterior cardinal vein and the left anterior cardinal vein. Double SVC has clinical implications only in the case when the branch ends in left atrium. (8)

**Azygos continuation of IVC**

Azygos continuation of IVC is sporadic congenital anomaly characterized by agenesis of the hepatic tract of inferior vena cava and posterior redirection of the blood through enlarged azygos vein that courses the right edge of spine and drains into SVC.(9) Hemiazygos continuation represents even rarer anomaly. In this variant the large venous blood vessel courses the left side of the spine. In some cases enlarged hemiazygos vein drains into SVC. Although asymptomatic this anomaly can be the part of situs viscerum ambiguus condition with left isomerism where liver and stomach are located in the midline, multiple spleens can be found along the great curvature of the stomach ("polysplenia syndrome"), and the minor fissure of the right lung is absent. (10, 11)

Some of the rarest venous abnormalities include the absence of left innominate vein with the flow of venous blood into azygos vein or hemiazygos vein and with anomalous venous connection to the right or left atrium.

**Absence of azygos vein**

Total absence of azygos vein occurs rare when the right cranial aspect of supracardinal vein fails to develop and represents asymptomatic state. Drainage of the right and left intercostal vein in this case happens through hemiazygos and hemiazygos accessory vein where accessory hemiazygos vein converges with the left brachiocephalic vein by using reflently the left supreme intercostal vein

**Double IVC and SVC**

Double IVC and SVC has so far been described in 13 documented cases. In this case the left anterior cardinal vein in the thorax as well as left supra- and subcardinal veins in the abdomen fails to regress. This leads to persistence of symmetrical and bilateral venous system in which the left inferior and superior vena cava drain into the left atrium. This situation results in right-to- left shunt that causes cyanosis. (12)
Pulmonary venous abnormalities

Total APVC

This anomaly account for 2% of congenital heart diseases and is usually manifested during pregnancy. (13) Total APVC results from persistent connection of the embryonic pulmonary venous plexus to the cardinal, cardiac or portal venous systems and the failure of the common pulmonary vein to join primitive venous sinus. (14) Intraatrial shunt (atrial septal defect, or patent foramen ovale) is present. Although the syndrome is usually isolated, an additional finding may be seen such as asplenia syndrome. Most patients present with cyanosis in the first year of life.

Total APVC is classified according to the drainage site. (15) In the supracardiac form which is the most common type, drainage occurs through the vertical vein to the left brachiocephalic vein (80% of cases) or directly to the right SVC or azygos vein (20%). In the cardiac type anomalous blood vessel or several individual vessels drain into the coronary sinus (80% of cases) or right atrium (20% of cases). In subdiaphragmatic type venous drainage usually occurs through the portal vein. (16) There is a mixed form of APVC where the pulmonary veins drain to more than one site.

Partial APVC

Partial APVC can occur either as an isolated case, or in combination with intracardiac defects. (17) In patients with congenital heart disease, some authors report the frequency of up to 0.5%-0.7%. (18, 19).

The most common form of partial APVC is anomalous right upper lobar vein that drains into the SVC or right atrium. In the less frequent form of partial APVC, left pulmonary vein drains into the IVC. Anomalous vessel has a characteristic "scimitar" appearance as it descends towards the base of the right lung. (20) Partial APVC in the left upper lobe of the lung is often confused with persistent left SVC since the appearance of these conditions is similar on cross-sectional images. Vertical vein courses cranial to the innominate vein in partial APVC of left upper lobe (drainage in the vertical vein).

Pulmonary abnormalities related to APVC

Several abnormalities can be associated with pulmonary APVC. These include bronchopulmonary sequestration, pulmonary arteriovenous malformations, cystic adenomatoid malformation. (21)

Pulmonary varix
Pulmonary varix is described as a rare lesion that consists of focal enlargement of segmental pulmonary veins. This rare lesion is usually asymptomatic, clearly delineated parenchymal or mediastinal mass on conventional chest X-ray. Due to similarities with other lesions like malignant mediastinal and paramediastinal masses, accurate diagnostics is very important because there is an absence of cardiac problems that would indicate the existence of abnormality, thus filling of the varices usually occur in venous phase. (22)
Imaging findings OR Procedure details

From 01.01.2000 until 01.01.2012 at our Radiology Clinic, a total of 4669 Thorax CT scans were performed. All patients underwent native CT scanning, and afterwards suspicion on the abnormality of the thoracic veins was either confirmed or rejected with Contrast enhanced CT (37 patients), I.V.DSA (5), or MRI (1).

Venous abnormalities were found in 43 patients out of 4669 examinations (0.92%). There were 27 male (62.79%), and 16 female patients (37.21%). Average age was 47.9 yrs (youngest was 20, and oldest 87 years). The most common abnormality was Double Vena Cava Superior which was found in 13 patients (0.28% of all patients scanned). The second most common abnormality Dilatation of Azygos Vein with Continuation was found in 7 patients (0.15%), while the third was Pulmonary Varix which was found in 4 patients (0.08%), followed by other not so frequent abnormalities.

According to the literature, venous abnormalities in the thorax are quite rare. In our study abnormalities occurred in 0.92% of cases.

The most common abnormality was double superior vena cava with incidence of 0.26% of scanned patients which is slightly less than other authors have been found (0.3% -0.5%). Persistent left superior vena cava with the absence of the right was found in 0.04% of patients, and its frequency in our case is smaller in comparison with other studies.

The next most frequent abnormality in our study was azygos vein dilatation (0.15%) followed by dilated azygos vein continuation (0.06%), and pulmonary varix (0.09%). Dilatation of azygos vein was 4 fold rarer in our study in comparison with the study of Parikh et al. (23)

Two other abnormalities were also detected and they are somewhat rarer in comparison with other authors.

Other abnormalities were in line with the average found by other authors.

It must be noted that very few authors have thoroughly studied the frequency of occurrence of these abnormalities in the population.
Images for this section:

Fig. 1: Left SVC. MIP image.

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Fig. 2: Left SVC. DSA image.

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**Fig. 3:** Double SVC.

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Fig. 4: Double SVC. DSA image.

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Fig. 5: Double SVC - axial CT scan.

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Fig. 6: Azygos continuation of SVC

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Fig. 7: Anomalous flow of right superior pulmonary vein in SVC. MRI scan.

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Fig. 8: Axial CT scan. Pulmonary varix.

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Fig. 9: Dilated anomalous connection between left brachiocephalic vein through enlarged left superior intercostal vein and azygos vein. Connection courses dorsal to aorta. MRI scan.

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Fig. 10: Tibutary vein next to right principal bronchus.

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Fig. 11: Enlargement of left superior intercostal vein.

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### Table 2: Frequency of venous abnormalities.

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<table>
<thead>
<tr>
<th>Entity</th>
<th>M</th>
<th>F</th>
<th>Total</th>
<th>Mean age</th>
<th>%</th>
</tr>
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<tbody>
<tr>
<td>Persistent left SVC</td>
<td>1</td>
<td>1</td>
<td>2</td>
<td>49.2</td>
<td>~ 0.04</td>
</tr>
<tr>
<td>Double SVC</td>
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<td>3</td>
<td>11</td>
<td>51.2</td>
<td>~ 0.26</td>
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<tr>
<td>Aberant SVC</td>
<td>1</td>
<td>0</td>
<td>1</td>
<td>56.0</td>
<td>~ 0.02</td>
</tr>
<tr>
<td>Dilated ayglos vein continuation</td>
<td>5</td>
<td>2</td>
<td>7</td>
<td>46.1</td>
<td>~ 0.15</td>
</tr>
<tr>
<td>Dilated ayglos vein continuation</td>
<td>2</td>
<td>1</td>
<td>3</td>
<td>47.0</td>
<td>~ 0.06</td>
</tr>
<tr>
<td>Pulmonary varix</td>
<td>1</td>
<td>3</td>
<td>4</td>
<td>48.5</td>
<td>~ 0.09</td>
</tr>
<tr>
<td>Anomalous inflow of innominate vein into ayglos vein</td>
<td>2</td>
<td>0</td>
<td>2</td>
<td>37.5</td>
<td>~ 0.04</td>
</tr>
<tr>
<td>Dilated right superior pulmonary vein</td>
<td>0</td>
<td>3</td>
<td>3</td>
<td>51.0</td>
<td>~ 0.06</td>
</tr>
<tr>
<td>Dilated left intercostal vein</td>
<td>3</td>
<td>0</td>
<td>3</td>
<td>50.3</td>
<td>~ 0.06</td>
</tr>
<tr>
<td>Anomalous inflow of right superior pulmonary vein into SVC</td>
<td>0</td>
<td>1</td>
<td>1</td>
<td>49.0</td>
<td>~ 0.02</td>
</tr>
<tr>
<td>Dilatation of SVC</td>
<td>1</td>
<td>0</td>
<td>1</td>
<td>47.0</td>
<td>~ 0.02</td>
</tr>
<tr>
<td>Dilatation of left superior pulmonary vein</td>
<td>0</td>
<td>1</td>
<td>1</td>
<td>37.0</td>
<td>~ 0.02</td>
</tr>
<tr>
<td>Aberant pulmonary vein located dorsal from the right principal bronchus</td>
<td>1</td>
<td>0</td>
<td>1</td>
<td>46.0</td>
<td>~ 0.02</td>
</tr>
<tr>
<td>Anomaly of left internal jugular vein</td>
<td>0</td>
<td>1</td>
<td>1</td>
<td>53.0</td>
<td>~ 0.02</td>
</tr>
<tr>
<td>Aberant tributary vein of SVC</td>
<td>1</td>
<td>0</td>
<td>1</td>
<td>79.0</td>
<td>~ 0.02</td>
</tr>
<tr>
<td>Dilated connection between left innominate vein with ayglos vein</td>
<td>1</td>
<td>0</td>
<td>1</td>
<td>19.0</td>
<td>~ 0.02</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td>27</td>
<td>16</td>
<td>43</td>
<td>47.9</td>
<td>~ 0.92</td>
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</table>

<table>
<thead>
<tr>
<th>Age groups</th>
<th>Male</th>
<th>Female</th>
<th>Total number of diagnosed venous abnormalities</th>
<th>%</th>
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<tbody>
<tr>
<td>0-10</td>
<td>0</td>
<td>0</td>
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<td>0.00</td>
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<td>11-20</td>
<td>1</td>
<td>0</td>
<td>1</td>
<td>2.32</td>
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<td>21-30</td>
<td>6</td>
<td>3</td>
<td>9</td>
<td>20.94</td>
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<tr>
<td>31-40</td>
<td>4</td>
<td>3</td>
<td>7</td>
<td>16.28</td>
</tr>
<tr>
<td>41-50</td>
<td>3</td>
<td>4</td>
<td>7</td>
<td>16.28</td>
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<tr>
<td>61-70</td>
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<td>2</td>
<td>7</td>
<td>16.28</td>
</tr>
<tr>
<td>71-80</td>
<td>2</td>
<td>1</td>
<td>3</td>
<td>6.98</td>
</tr>
<tr>
<td>81-90</td>
<td>1</td>
<td>0</td>
<td>1</td>
<td>2.32</td>
</tr>
</tbody>
</table>
Table 1: Age groups of patients with venous abnormalities in the thorax.

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Conclusion

According to our experience venous abnormalities in the thorax are relatively rare. Our results mostly match results of other authors who have been interested in the same problem.
Most frequent venous abnormalities in the thorax according to our study are double superior vena cava, and azygos vein continuation followed by pulmonary varices.
Venous abnormalities are subtle and sometimes difficult for detection on chest X ray, and can simulate a pathological substrate in the thorax.
Venous abnormalities are pseudo lesions, therefore non invasive CT and sometimes MRI examination is essential for proper diagnosis of these conditions.
It is essential for radiologist to be aware of the existence of these relatively rare abnormalities that should not be overseen during imaging interpretation. In cases there patient has to undergo interventional or surgical treatment radiologist plays critical role in alerting surgeons, interventional radiologists, anesthesiologists on the existence of abnormalities of thoracic veins.
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