Noninvasive imaging diagnosis of partial anomalous pulmonary venous return

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Learning objectives

1. To describe and classify the types of partial anomalous pulmonary venous return (PAPVR).

2. To provide an overview of the different CT and MRI findings in partial anomalous pulmonary venous return (PAPVR).

Background

PAPVRs encompasses a group of congenital cardiovascular anomalies in which a portion or the entirely pulmonary venous drainage drains into the right atrium or one of its tributaries, resulting in a left-to-right shunt (1, 2).

PAPVRs represent 70% of all APVRs and 0.5% of the congenital cardiac defects. PAPVR is found in 0.4-0.7% of routine necropsy.

In PAPVR the shunt can vary from less than 10% of the global pulmonary flow when only a segmental pulmonary vein (PV) is concerned to more than 75% when all but one lobes are involved. PAPVRs occur on the right side twice as often as on the left.

Although PAPVC can present as an isolated structural abnormality, it commonly occurs in association with other cardiac abnormalities. PAPVR is found in 10-15% of patients with atrial septal defect (ASD) and almost all patients with sinus venosus type of ostium secundum defect.

Multiple case series report PAPVC as an incidental finding and in most of these instances is asymptomatic (1, 3). In symptomatic patients, the clinical findings of PAPVR's are similar to those of atrial septal defect. The extent of symptoms and physiologic disturbances depends primarily on the degree of shunting, the number of anomalous veins, associated valvular abnormalities, and the presence of concomitant cardiac or pulmonary disease (1, 4).

Classification of partial anomalous pulmonary venous return (5):
PAPVR can be classified in four groups based on their drainage:
- **supracardiac type** in which the PV's can be connected to superior vena cava, brachiocefalic venous trunk, azygos vein, persistent left superior vena cava, subclavian vein, hemiazygos vein

- **cardiac type** in which the PV's can be connected to right atrium or the coronary sinus

- **infracardiac type** in which the PV's can be connected to the inferior vena cava (the thoracic segment or abdominal, suprahepatic segment), the hepatic veins, the portal vein, azygos vein, gastric vein, pericardiophrenic vein

- **mixed type** in which any of the types of the above can be mixed together.

**Imaging findings OR Procedure details**

**Material and Methods:**

In this exhibit we will present an overview of the PAPVR's appearances based on a retrospective study of 9815 patients explored by sectional imaging methods at the level of the thorax (computer tomography and/or magnetic resonance) between January 2000 - July 2011 in the Radiology and Medical Imaging Department of Fundeni Clinical Institute.

**CT protocol:**

- unenhanced and contrast enhanced CT (CECT) of the thorax (collimation: 5; pitch: 1-1,5)

- contrast media injection of nonionic iodinated contrast material 1-2 ml/kg at a flow rate of 3-4 ml/sec

- postcontrast CT acquisition:
  - arterial pulmonary phase at 10-12 sec after the start of the injection
  - venous phase at 25-30 sec after the contrast material administration

**MRI protocol:**

- MR imaging was performed at 1.5 T using a Torsopa coil;
- respiratory gating SE sequences with slice thickness 7 mm and spacing 2 mm
- 3D FSPGR with gadolinium injection (0.2 ml/kg, flow 2 ml/s) with bolus detection and multiplanar reconstructions

**Results:**

We detected a number of 13 patients with PAPVR, the majority of the patients explored belonging to the pediatric population aged between 3-18 years (7 cases) with a male to female ratio of 7:6.

Nine of the patients were explored by CT, 3 patients underwent a MRI examination and only one patient was explored by CT and MRI (Table 1 on page 7) and they were distributed according to the classification (Table 2 on page 8).

**Supracardiac type of drainage:**

We detected 8 cases with drainage to the superior vena cava or brachiocephalic venous trunk, from which 6 cases on the left side (into the brachiocephalic venous trunk) and 2 cases on the right side (into the superior vena cava).

- **PAPVR connected to the superior vena cava (SVC)**

PAPVR in the SVC (Fig. 1 on page 9) concern more frequently the right upper pulmonary veins than the lower right pulmonary veins.

The most frequent is the connection of the right upper pulmonary vein at the junction between the superior vena cava and the right atrium.

Its most frequent association is with a sinus venosus type of atrial septal defect, less frequent a ostium secundum type af atrial septal defect.

The surgical indication is formal when there is a sinus venosus type of atrial septal defect; the repair consists in closing the atrial communication and divert the pulmonary vein to the left atrium (1, 5, 6).

- **PAPVR connected to the brachiocephalic venous trunk (BCVT)**

PAPVR can be connected to the BCVT directly or via a vertical vein (Fig. 2 on page 10); it's one of the most commonly recognized PAPVR's on CT.
The anomalous PV usually passes anteriorly to the aortic arch to terminate at the proximal part of the BCVT; in some cases the anomalous PV can pass posteriorly to the aortic arch having a risk of compression between the left main pulmonary artery and the left main bronchus.

If only the left upper lobe is involved, the patient is usually asymptomatic with no signs of right ventricular diastolic overload (6, 7).

**Cardiac type of drainage**

We detected 2 cases with a cardiac type of drainage from which 1 case with drainage to the right atrium and 1 case with drainage in the coronary sinus.

- **PAPVR connected to the right atrium**

PAPVR draining into the right atrium (Fig. 3 on page 11) is usually a consequence of abnormal development of the septum secundum, which is shifted to the left.

PAPVR in right atrium is frequently associated with ASD (a sinus venosus type or a ostium secundum type).

The clinical symptoms are those of a large atrial septal defect, whether there is or not a septal defect associated.

- **PAPVR connected to the coronary sinus**

PAPVR are connected to the coronary sinus (Fig. 4 on page 12) more frequently on the left side than on the right side. It concerns almost always the left lower lobe.

**Infracardiac type of drainage**

We detected 2 cases with an infracardiac type of drainage both to the inferior vena cava (IVC), the thoracic segment and abdominal, suprahepatic segment.

- **PAPVR connected to the inferior vena cava, the thoracic segment**

PAPVR into the IVC represents 1-5% of all PAPVR's and concerns the whole right lung or a portion of the lung. Drainage is either into the infra-diaphragmatic part of the IVC or at the junction between the IVC and right atrium (Fig. 5 on page 13). Exceptional cases occur on the left side.
The PV that drains into the IVC have been called a "scimitar vein". The anomalous PV can be thin, straight or multiple (5, 8).

- **The scimitar syndrome**

It is also known as the Halasz syndrome or pulmonary venolobar syndrome (Fig. 6 on page 14).

The scimitar syndrome it represents an association of anomalies:

- PV connected to the inferior vena cava, the abdominal segment
- abnormal lobation of the right lung with mediastinal shift to the right, also called hypogenetic right lung syndrome
- "dextrocardia" - dextroposition - dextroversion of the heart
- pulmonary arterial anomalies (small ipsilateral pulmonary artery in half of patients)
- systemic arterial supply originating from the abdominal aorta and less frequently from thoracic descending aorta, which is responsible for further increase of the left-to-right shunt
- other tracheal or bronchial malformations (cysts or diverticula, bronchiectasis)
- accessory hemidiaphragm, diaphragmatic eventration
- horseshoe lung
- other cardiovascular anomalies in 25%, most frequently interatrial communication

Most cases are sporadic, but there have been reported some familial cases.

Surgery is not indicated in well tolerated clinical forms; but if there is an atrial septal defect associated surgery is needed if it cannot be repaired by cardiac catheterism (5, 9).

**Mixed type of drainage**

We detected 1 case with drainage in both the right atrium and the inferior vena cava, supradiaphragmatic (Fig. 7 on page 15).
As a general guideline, surgery is not indicated in an isolated PAPVR, and it is considered if only there is a left-to-right shunt greater than 50% or the patient developed an arterial pulmonary hypertension (10).

**The imaging report** must contain:

- The description of the pulmonary venous drainage (Fig. 8 on page 16):
  - Right upper lobe veins drain into SVC. This type is associated with sinus venosus arterial septal defect.
  - Left upper lobe veins often drain into innominate vein.
  - Right lower lobe veins can drain into IVC (scimitar vein) or to hepatic or portal veins.

  - The evaluation of pulmonary arteries. PAPVR can be associated with hypoplastic pulmonary artery and lung.
  - The quantification of pulmonary flow/ systemic flow ratio (Qp:Qs).
  - The quantification of ventricular volumes and function and assess right atrial dilation.
  - The listing of others possible cardiac or pulmonary abnormalities.

**Images for this section:**
### Table 1: Imaging diagnostic data in the study group.

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<td><strong>CT</strong></td>
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</tr>
<tr>
<td><strong>MRI</strong></td>
<td>3</td>
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<tr>
<td><strong>CT+MRI</strong></td>
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### Table 2: Classification distribution data in our study group.

<table>
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<tr>
<th></th>
<th>SUPRACARDIAC SVC / BCVT</th>
<th>CARDIAC RA / CORONARY SINUS</th>
<th>INFRACARDIAC IVC (SUPRA/INFRA DIAPHRAGMATIC)</th>
<th>MIXED RA and IVC supra diaphragmatic</th>
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<td><strong>RIGHT</strong></td>
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<td>1</td>
<td>2</td>
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</tr>
<tr>
<td><strong>LEFT</strong></td>
<td>6</td>
<td>1</td>
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**Fig. 1:** Supracardiac type of drainage (M, 9 years): two right superior pulmonary veins (blue arrow) drain into the superior vena cava at the junction between the right atrium and the superior vena cava.
Fig. 2: Supracardiac type of drainage (M, 14 years): the left superior pulmonary vein (blue arrow) drains into left brachiocephalic venous trunk.
**Fig. 3:** Cardiac type of drainage (M, 76 years): the right superior pulmonary vein (blue arrow) is connected to the right atrium.
**Fig. 4:** Cardiac type of drainage (F, 3 years): a small left inferior pulmonary vein (white arrow) drains into the coronary sinus.
Fig. 5: Infracardiac type of drainage (F, 30 years): the right inferior pulmonary vein (blue arrow) drains into a dilated inferior vena cava, the thoracic segment. The "scimitar vein" is seen in the coronal reformation.
Fig. 6: Scimitar syndrome (F, 12 years): the right pulmonary vein (blue arrow) is connected to the inferior vena cava (infradiaphragmatic segment). The chest topogram shows the "scimitar sign", dextrocardia, right lung hypoplasia and the diaphragmatic eventration. In lung window visualization of "horse shoe" malformation: the right and left postero-inferior segments are united in the posterior mediastinum.
Fig. 7: Mixed type of drainage (M, 6 years): two right inferior pulmonary veins (blue arrow) drain into the right atrium and one inferior right pulmonary vein (red arrow) is connected to the thoracic segment of the inferior vena cava.
Fig. 8: Schematic drawing of total anomalous pulmonary venous drainage.
Conclusion

1. Cross sectional imaging has a crucial contribution in the diagnosis and characterization of these types of congenital pulmonary vascular malformations and also in detecting any associated abnormalities of tracheobronchial tree, lung parenchyma or systemic vascular malformations.

2. MDCT is now considered the first-line technique in the PAPVR’s work-up.

3. Sectional imaging has an important role in planning the appropriate surgical treatment.

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References


