Diagnostic accuracy of transthoracic contrast echocardiography as a screening method for pulmonary arteriovenous malformations

MWF van Gent¹, MC Post¹, RJ Snijder², JJ Westermann², JJ Mager²
Department of cardiology¹ and pulmonology², St Antonius Hospital, Nieuwegein, The Netherlands

Background

- **Pulmonary arteriovenous malformations (PAVMs)**
  - are abnormal communications between pulmonary arteries and pulmonary veins
  - may cause paradoxical embolism and right-to-left shunting
  - are associated with severe neurological complications (e.g. brain abscess, stroke)
  - efficient endovascular treatment (embolotherapy) is possible and is performed in all PAVMs that are large enough
  - >80% is associated with HHT

- **Hereditary Hemorrhagic Telangiectasia (HHT) (Rendu-Osler-Weber disease)**
  - is an autosomal dominant disorder
  - is characterized by vascular abnormalities varying from small telangiectases to large arteriovenous malformations (predominantly in the lungs, brain and liver)

- **Screening for PAVMs**
  - contrast echocardiography and chest CT
  - disadvantage of chest CT is radiation exposure
  - TTCE recently first choice in guidelines¹

- **Contrast echocardiography (TTCE)**
  - is a simple, minimally invasive technique
  - very sensitive for the detection of pulmonary shunting²
  - grading scale 1-3 depending on magnitude of microbubbles in left ventricle (still-frame)³

Aim of the study

- To determine the diagnostic value of pulmonary shunting on TTCE as compared with PAVMs on chest HRCT, in the largest HHT patient cohort reported so far

Methods

**Study population**
In the period from May 2004 till December 2010, 626 consecutive persons were screened for possible HHT with both TTCE and chest HRCT

**Contrast echocardiography**
10 ml Agitated saline was injected while projecting the AP4CH view without a Valsalva manoeuvre. TTCE was considered positive for a pulmonary RLS if microbubbles appeared in the left atrium after four cardiac cycles. Shunt size was determined (grade 1-3).

**Chest HRCT**
PAVM: a nodular opacity with both an afferent and efferent vessel.

Results

<table>
<thead>
<tr>
<th>Shunt on TTCE −</th>
<th>Shunt on TTCE +</th>
<th>Origin of shunt uncertain</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>PAVM on CT −</td>
<td>357</td>
<td>126</td>
<td>3</td>
</tr>
<tr>
<td>PAVM on CT +</td>
<td>2</td>
<td>118</td>
<td>2</td>
</tr>
<tr>
<td>PAVM on CT uncertain</td>
<td>10</td>
<td>8</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>369</td>
<td>252</td>
<td>5</td>
</tr>
</tbody>
</table>

Table 1. TTCE versus PAVM on chest CT

- The negative predictive value of TTCE for the absence of PAVMs on chest HRCT was 99.4%, and sensitivity was 98.3%.

- In two patients with PAVMs on chest HRCT but no shunt on TTCE, PAVMs were very small and far beyond possibility for treatment.

- The positive predictive value of TTCE for the presence of PAVMs was 48.4%, and specificity 73.9%.

Conclusions

- TTCE has an excellent negative predictive value for PAVMs

- Poor positive predictive value of TTCE relates to detection of small pulmonary shunts (not visible on chest HRCT)

- TTCE did not miss PAVMs amenable for embolisation

- TTCE appears suitable as a first-line screening method for PAVMs

- Important reduction in radiation and costs

- Importance of local experience

Literature cited


Embodisation of PAVM with Amplatzer® vascular plug

Shunt

Large PAVM in an HHT patient

Pulmonary angiography showing two PAVMs