Screening for Right-to-Left Shunts With Contrast Transcranial Doppler in Hereditary Hemorrhagic Telangiectasia

Dulka Manawadu, MRCP; Dilini Vethanayagam, FRCP; Maher Saqqur, FRCP; Carol Derksen, RVT; Jonathan Choy, FRCP; Khurshid Khan, FRCP

Background and Purpose—Contrast transthoracic echocardiography (TTCE) is used to screen hereditary hemorrhagic telangiectasia (HHT) patients for right-to-left shunts (RLS) associated with increased stroke risk. We hypothesized that contrast transcranial Doppler (TCDc), shown to be highly sensitive for detecting RLS in patent foramen ovale, will be as comparable to TTCE for screening HHT patients.

Methods—We compared TTCE and TCDc for detecting RLS in 12 patients with HHT who also underwent CT pulmonary studies to determine pulmonary arteriovenous malformation (PAVM) presence. The sensitivity and specificity of TTCE and TCDc in detecting PAVM were determined and the agreement between TTCE and TCDc in detecting RLS was assessed.

Results—Both TTCE and TCDc had 100% sensitivity in detecting underlying PAVM; the specificity was 25% and 38%, respectively. The agreement in detecting RLS between TTCE and TCD was high (κ=0.76). TCD was well-tolerated with no immediate adverse or embolic events over the next 3 months.

Conclusions—TCDc offers a simple office-based alternative to TTCE for screening RLS associated with PAVM in HHT patients. (Stroke. 2011;42:00-00.)

Key Words: arteriovenous malformations ■ hemorrhagic ■ hereditary ■ telangiectasia ■ transcranial Doppler ultrasonography
Table. Disease Characteristics of Study Patients (n=12)

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Number (Percentage)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Epistaxis</td>
<td>12 (100%)</td>
</tr>
<tr>
<td>Gastrointestinal involvement</td>
<td>2 (17%)</td>
</tr>
<tr>
<td>Respiratory symptoms</td>
<td>2 (17%)</td>
</tr>
<tr>
<td>Headache</td>
<td>4 (33%)</td>
</tr>
<tr>
<td>Ischemic stroke or TIA</td>
<td>4 (33%)</td>
</tr>
<tr>
<td>Hemorrhagic stroke</td>
<td>0</td>
</tr>
<tr>
<td>Family history of HHT</td>
<td>11 (92%)</td>
</tr>
<tr>
<td>Telangiectasia</td>
<td>11 (92%)</td>
</tr>
<tr>
<td>Abnormal chest radiograph</td>
<td>2 (17%)</td>
</tr>
<tr>
<td>PAVM on chest CT/CT angiography</td>
<td>4/10 (40%)</td>
</tr>
<tr>
<td>CAVM on brain imaging</td>
<td>0 (1 previous excision)</td>
</tr>
</tbody>
</table>

Two patients did not have chest CT/CT angiogram. CAVM indicates cerebral arterial venous malformations; CT, computed tomography; HHT, hereditary hemorrhagic telangiectasia; PAVM, pulmonary arteriovenous malformations; TIA, transient ischemic attack.

Discussion

These findings suggest TCDc may be comparable with TTCE (existing best practice) in screening for RLS associated with PAVM in HHT patients. In this small series, TCDc had a sensitivity of 100% in detecting RLS and agreed 92% (κ = 0.76) with TTCE. These findings are consistent with a previous report showing that carotid artery ultrasound had 95% sensitivity and 85% specificity of shunt detection in patients with PAVM.6 In addition, TCDc is easy to use, portable, and can be kept within a clinic or office setting to screen for shunts at the time of patient contact. Furthermore.

TCDc, unlike TTCE, does not require extensive technical expertise. TCDc may be particularly helpful in screening during pregnancy because of the higher risk of PAVM enlargement and rupture,7 when TTCE may prove cumbersome because of posture and access.

Our study has limitations, the main one being a small sample size because of the rarity of HHT. The study is open to statistical bias and caution is needed in interpreting findings. The air volume used was 0.5 mL, which was commensurate with local practice but may have compromised sensitivity in shunt detection. This is unlikely because, even with low air volumes, TCDc had 100% sensitivity in detecting RLS. It is possible that, unlike TTCE, TCDc may not be able to distinguish between intracardiac and extracardiac shunts. Our data showed that MES were detected early (within 5 cardiac cycles) in all patients with known patent foramen ovales. This suggests that it may be possible to discriminate between intracardiac and extracardiac RLS with TCDc, but this needs confirmation in future studies. TTCE is the recommended gold standard tool for screening RLS associated with PAVM.8 TCDc offers a simple office-based alternative when TTCE may not be easily available or logistically difficult to perform.

Disclosures

None.

References

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