Conclusions Lung perfusion MRI has high sensitivity equivalent to perfusion scintigraphy in diagnosing CTEPH but does not require ionising radiation, making it an attractive initial imaging modality to assess patients with suspected CTEPH.

Chronic thromboembolic pulmonary hypertension (CTEPH) occurs in up to 3.8% of patients following pulmonary thromboembolism (PTE) and is a major cause of severe pulmonary hypertension (PH). Patients usually present with breathlessness following PTE or with unexplained PH. The diagnosis may be missed on CT pulmonary angiography (CTPA) by radiologists not experienced in pulmonary vascular disease, so perfusion scintigraphy has been recommended as a screening test, given its high sensitivity. In recent years, MRI has evolved into a holistic imaging modality in PH, allowing morphological assessment of the pulmonary vasculature and structural and functional assessment of the heart. In addition, 3D contrast-enhanced lung perfusion MRI provides insight into regional pulmonary perfusion by tracking the dynamic passage of a contrast bolus. Previous studies have demonstrated the feasibility of this technique in acute PTE. However, there is limited data on the performance of 3D contrast-enhanced lung perfusion MRI in diagnosing CTEPH in a high-risk population.

From a large PH referral centre, consecutive patients with suspected CTEPH or unexplained PH who had lung perfusion MRI, perfusion scintigraphy, CTPA and right heart catheterisation within 14 days were identified from the ASPIRE registry to compare the diagnostic utility of these imaging modalities. 3D MR lung perfusion images were acquired using a time resolved 3D spoiled gradient echo sequence with view sharing and were analysed by two radiologists blinded to the other imaging studies and clinical information. The final diagnosis of CTEPH was based on clinical assessment, right heart catheter and imaging, including MR angiographic studies (but not the MR perfusion images analysed in this study), CTPA and perfusion scintigraphy. The diagnosis of surgically accessible disease was confirmed at the national pulmonary endarterectomy centre at Papworth.

Of the 132 patients fulfilling the inclusion criteria, 78 had CTEPH. Forty-eight patients showed no evidence of CTEPH and included patients with a diagnosis of no PH (n=5), and all major forms of PH, including pulmonary arterial hypertension (n=18), PH associated with left heart disease (n=12), PH associated with lung disease (n=8) and PH multi-factorial (n=5). Six of the MR perfusion image data sets were of non-diagnostic quality. The MR perfusion images correctly identified

Table 1 Summary of diagnostic performance of perfusion scintigraphy, MR perfusion and CTPA in the diagnosis of CTEPH

<table>
<thead>
<tr>
<th></th>
<th>Q scan</th>
<th>MR perfusion*</th>
<th>CTPA</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sensitivity (%)</td>
<td>96 (95% CI 0.89% to 0.99%)</td>
<td>97 (95% CI 0.91% to 0.99%)</td>
<td>94 (95% CI 0.85% to 0.98%)</td>
</tr>
<tr>
<td>Specificity (%)</td>
<td>90 (95% CI 0.77% to 0.97%)</td>
<td>92 (95% CI 0.80% to 0.97%)</td>
<td>98 (95% CI 0.88% to 0.99%)</td>
</tr>
<tr>
<td>Positive predictive value (%)</td>
<td>94 (95% CI 0.86% to 0.98%)</td>
<td>95 (95% CI 0.88% to 0.99%)</td>
<td>99 (95% CI 0.92% to 0.99%)</td>
</tr>
<tr>
<td>Negative predictive value (%)</td>
<td>93 (95% CI 0.82% to 0.99%)</td>
<td>96 (95% CI 0.85% to 0.99%)</td>
<td>90 (95% CI 0.78% to 0.96%)</td>
</tr>
<tr>
<td>Accuracy (%)</td>
<td>94</td>
<td>95</td>
<td>95</td>
</tr>
</tbody>
</table>

*Inter-observer agreement, \( \kappa \) of 0.83.

CTEPH, chronic thromboembolic pulmonary hypertension; CTPA, CT pulmonary angiography.

Figure 1 (A) Selected coronal images of MR lung perfusion (top) and anterior and posterior view of perfusion scintigraphy images (bottom) in a patient with normal lung perfusion. (B) An example of a patient with chronic thromboembolic pulmonary hypertension with bilateral segmental perfusion defects seen on MR perfusion images (top) and perfusion scintigraphy (bottom).
76 patients to have CTEPH giving it a sensitivity of 97% (95% CI 0.91% to 0.99%) and specificity of 92% (95% CI 0.80% to 0.97%) (table 1). The results also showed CTPA, interpreted by the expert pulmonary vascular radiologists, and perfusion scintigraphy to be effective in the diagnosis of CTEPH with a sensitivity of 94% (95% CI 0.85% to 0.98%) and 96% (95% CI 0.89% to 0.99%), respectively (figure 1). None of the imaging modalities missed any of the patients with surgically accessible disease.

Accurate diagnosis of CTEPH is essential to identify a potentially treatable cause of PH. Here it is demonstrated that 3D lung perfusion MRI, as part of a pulmonary–vascular MRI protocol, has very high sensitivity for the diagnosis of CTEPH similar to that of perfusion scintigraphy and CTPA. The 3D data set allows image reconstruction in any plane, enabling better assessment of regional perfusion abnormalities and has superior temporal and spatial resolution to scintigraphy.

In conclusion, 3D lung perfusion MRI has high sensitivity for CTEPH and complements MR angiography and functional cardiac MRI in a single comprehensive radiation-free imaging modality in the evaluation of patients with suspected CTEPH.

Smitha Rajaram, Andrew J Swift, Adam Telfer, Judith Hurdman, Helen Marshall, Eleanor Lorenz, David Capener, Christine Davies, Catherine Hill, Charlie Elliot, Robin Condliffe, Jim M Wild, David G Kiely

1 Academic Unit of Radiology, University of Sheffield, Sheffield, UK
2 Sheffield Pulmonary Vascular Disease Unit, Royal Hallamshire Hospital, Sheffield Teaching Hospitals NHS Foundation Trust, Sheffield, UK
3 Department of Nuclear Medicine, Sheffield Teaching Hospitals NHS Foundation Trust, Sheffield, UK
4 Department of Radiology, Sheffield Teaching Hospitals NHS Foundation Trust, Sheffield, UK

Correspondence to Dr David G Kiely, Sheffield Pulmonary Vascular Disease Unit, Royal Hallamshire Hospital, Sheffield Teaching Hospitals NHS Foundation Trust, Glossop Road, Sheffield S10 2JF, UK; david.kiely@sth.nhs.uk

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