Diagnostic Imaging Pathways - Chronic Thromboembolic Pulmonary Hypertension (Suspected)

Population Covered By The Guidance

This pathway provides guidance on the imaging of adult patients with suspected chronic thromboembolic pulmonary hypertension.

Date reviewed: November 2018

Date of next review: November 2021

Published: March 2019

Quick User Guide

Move the mouse cursor over the PINK text boxes inside the flow chart to bring up a pop up box with salient points. Clicking on the PINK text box will bring up the full text. The relative radiation level (RRL) of each imaging investigation is displayed in the pop up box.

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<th>SYMBOL</th>
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Pathway Diagram
Teaching Points

- A diagnosis of CTEPH was previously associated with a poor prognosis, however it is now potentially curable with surgery or balloon pulmonary angioplasty. Improvements in medical and endovascular treatments have led to some improved survival in inoperable or refractory CTEPH.
- Lung scintigraphy (V/Q or ventilation/perfusion scan) is the first-imaging modality to investigate suspected CTEPH. A normal V/Q scan rules out a diagnosis of CTEPH with high certainty. CTEPH may still be present with an intermediate scan.
- CT pulmonary angiography (CTPA) alone frequently misses the diagnosis of CTEPH so it is not recommended to rule it out.
- Possible cases of CTEPH or pulmonary hypertension due to other causes should be referred to a specialist centre for further evaluation.
- Transthoracic echocardiogram (TTE) may suggest pulmonary hypertension, but invasive measurement of right heart pressures and pulmonary artery wedge pressures with right heart catheterisation is required to confirm the diagnosis.
- The role of CTPA or magnetic resonance pulmonary angiography (MRPA) and cardiac MRI is to assess disease severity, distribution and location to allow planning for surgery or radiological interventions and to assess right ventricular volume and function as a marker for treatment response. The chance of success with surgery is greatest when vascular obstruction is present in the main pulmonary arteries or lobar arteries. Balloon pulmonary angioplasty is successful at treating segmental and subsegmental arteries.
- Catheter pulmonary angiography remains the gold standard for diagnosis and treatment work up and all patients require this to plan intervention.

Chronic Thromboembolic Pulmonary Hypertension (CTEPH)

- Chronic thromboembolic pulmonary hypertension (CTEPH) is classified as group 4 pulmonary hypertension.
- CTEPH is a serious but rare adverse sequela of acute pulmonary embolism (PE) that occurs in about 3% of PE survivors, although up to 38% of patients with CTEPH have no documented history of venous thromboembolism.
- A diagnosis of CTEPH was previously associated with a poor prognosis, however it is now potentially curable with surgery. Improvements in medical and endovascular treatments have led to some improved survival in inoperable or refractory CTEPH.
- Diagnosing CTEPH following PE is challenging because symptoms are generally non-specific; diagnosis is delayed on average for by over a year.
  - Routine screening for CTEPH after PE is not currently recommended.
  - Unfortunately guidelines are lacking for how long patients should be followed up after PE and when diagnostic tests for CTEPH should be performed.
  - Patients with a higher burden of thrombus at time of diagnosis are at increased risk.
  - CTEPH can be suspected in patients who remain symptomatic after 3 months of effective anticoagulation. Risk factors for developing CTEPH include recurrent PE and unprovoked PE.
CTEPH can also develop in the absence of previous diagnosed acute PE. Patients with known clotting disorders and underlying connective tissue diseases such as scleroderma are at increased risk.

- CTEPH is defined as:
  - Mean pulmonary artery pressure ≥ 25 mmHg
  - Pulmonary capillary wedge pressure

**Transthoracic Echocardiogram**

- In patients presenting with dyspnoea following treated PE, echocardiography is recommended as a first-line on-invasive diagnostic investigation for the assessment of pulmonary artery pressures.
  - TTE also gives information on the presence of structural cardiac abnormalities, valvular dysfunction and systolic or diastolic cardiac function that may contribute to symptoms.
  - Cardiac MRI is the gold standard for assessment of the right ventricle, however it is not widely available.

- If TTE findings are compatible with a high or intermediate probability of pulmonary hypertension, a V/Q scan is recommended to exclude CTEPH.
  - However, a positive TTE is not required for diagnosis.

- TTE alone is not sufficient to support a treatment decision or to rule out CTEPH in patients with risk factors.
  - If suspected, a V/Q scan should be performed irrespective of TTE.
    - TTE relies on tricuspid regurgitation of pulmonary arterial hypertension (PAH) and can miss diagnoses. For example, if right heart failure is present, TTE may not demonstrate tricuspid regurgitation despite elevated pulmonary arterial pressures, resulting in false negatives.
    - Alternatively, pulmonary artery pressures should still be confirmed with right heart catheter because TTE may over-diagnose pulmonary hypertension by up to three times.
    - In asymptomatic patients with a high risk of CTEPH, V/Q scan may be the diagnostic test of choice to rule out CTEPH.

- Some patients have chronic thromboembolic pulmonary disease without PAH. These patients may warrant monitoring as they may develop PAH, although there have been no studies to demonstrate this.

**Chest Computed Tomography (CT)**

- Non-contrast CT chest can be useful in the work up of pulmonary hypertension to assess the lung parenchyma, which may reveal a cause for PAH or separate cause for symptoms.
  - Can also rule out extrinsic pulmonary vascular compression from other causes such as neoplasm or fibrosis.
- Multislice CT (or multidetector CT, MDCT) can be reconstructed into thin slices to assess parenchymal and bronchial detail, and continuous thick slices to assess for small nodules. Multislice CT has a fast acquisition time and high resolution when reconstructed in different planes.
Right Heart Catheterisation

- Invasive assessment of pulmonary haemodynamics is still required to confirm the diagnosis and to evaluate operability, as TTE may over- or underestimate pulmonary artery pressures 5
- There is a low associated risk of adverse events, with 1.1% associated morbidity and 0.05% mortality 19
- Pulmonary hypertension is defined as mean pulmonary artery pressure ≤25mmHg and pulmonary capillary wedge pressure

CT Pulmonary Angiography (CTPA)

- May be useful to demonstrate evidence of acute or chronic thromboembolic disease in patients without known history of PE and an indeterminate V/Q scan
- Role of CTPA is well established for the diagnosis of acute PE
- However, CTPA is not recommended to rule out CTEPH due to the high rate of false negatives. CTPA has a significantly lower sensitivity than V/Q scan (51% compared to 96-97.4%) 9
- Signs of CTEPH on CTPA include direct vascular signs of chronic PE as well as secondary signs including:
  - Eccentric, reorganised thrombus
  - Webs in pulmonary arteries
  - Pulmonary artery stenosis, which can be loss of normal tapering or focal overt stenosis. This is difficult to perceive via CT but is one of the commonest findings
  - Pulmonary artery occlusions
  - Paucity of distal vessels despite normal parenchyma. This is the cause for mosaic attenuation
  - Dilated systemic collaterals
  - Evidence of pulmonary hypertension, i.e. central pulmonary artery dilation, right heart dilation and right ventricular hypertrophy with flattening/bowing of the septum to the left, reflux of contrast into hepatic veins

Magnetic Resonance Imaging: Pulmonary Angiography (MRPA or MRA), Pulmonary Perfusion and Cardiac MRI

- MRA is an excellent non-invasive test to whether CTEPH may be amenable to surgical or endovascular treatment
- MRA can demonstrate flow artefact caused by webs and stenoses which are not detectable on CT
- MR is also able to demonstrate perfusion deficits which correspond to abnormal vessels and defects seen on V/Q scan
- Cardiac MRI is the gold standard for the assessment of right ventricular function 24 and can also assess the myocardium and valve disease 18
- Gadolinium contrast-enhanced MRA has shown promise for the diagnosis of CTEPH with one study reporting sensitivity and specificity of 98 and 94%
MRI is especially useful as a baseline for follow-up and monitoring response to treatment. Unlike CTPA and V/Q scan, there is no exposure to ionising radiation.

Disadvantages:

- Limited availability. MR can be misleading or non-diagnostic if not performed in a high-volume, experienced cardiothoracic imaging centre.
- Cost.
- Long acquisition time – dyspnoeic patients may not be able to achieve breath hold.
- Claustrophobic.
- Contraindicated with some ferromagnetic prostheses.

References

References are graded from Level I to V according to the Oxford Centre for Evidence-Based Medicine, Levels of Evidence. Download the document

7. Lang IM, Simonneau G, Pepke-Zaba JW, Mayer E, Ambroz D, Blanco I, et


