Navigating the identification, diagnosis and management of pulmonary hypertension using updated ESC / ERS guidelines

Case 2
76 y old male: past history

14 years  deep vein thrombosis (1990)

1 year  gradual onset breathlessness
        ankle swelling
        Rx angiotensin converting enzyme
        inhibitor and betablocker

10 months  CTPA showed chronic thromboembolic
           pulmonary hypertension (CTEPH)
           Rx warfarin since then
           continued to deteriorate
76 y old male: presenting complaint

3 weeks worsening breathlessness
admitted to hospital with gross fluid retention

echocardiogram: peak TR velocity 4.2 m/s
severely dilated right ventricle
moderate pericardial effusion

referred urgently to Hammersmith Hospital
Question: Which one of these statements is correct?

- A significant number of patients with CTEPH do not have any history of pulmonary embolism or deep vein thrombosis
- It is recommended that patients with acute pulmonary embolism should be routinely screened for CTEPH during follow-up.
- CTEPH patients with at least moderate pericardial effusion should have this drained to prevent right heart tamponade
- Patients with CTEPH do not normally require more than 5 years anticoagulation
- All patients with CTEPH should have an inferior vena cava filter inserted
76 y old male: on admission

WHO functional class IV: can transfer bed to chair with difficulty; unable to walk to end of bed

Examination:
Heart rate 85 bpm, blood pressure 111/72, respiratory rate 18 /min, apyrexial, jugular venous pressure to ear lobes, SpO$_2$ 94% on air
Loud pulmonary second sound with right ventricular third sound
Severe fluid retention
Cold peripheries
Resting ECG on admission
Chest radiograph on admission
Echocardiogram tricuspid regurgitation velocity
Echocardiogram parasternal short axis
Echocardiogram apical 4 chamber
Ventilation perfusion scan

ANT/POST Q 22/12/2014
ANT/POST Q 22/12/2014
LPO Q 22/12/2014
RPO Q 22/12/2014

ANT/POST V 22/12/2014
ANT/POST V 22/12/2014
LPO V 22/12/2014
RPO V 22/12/2014

Ant Q 399K Duration: 113sec 256x256 99m Technetium
Post Q 400K Duration: 113sec 256x256 99m Technetium
LPO Q 400K Duration: 128sec 256x256 99m Technetium
RPO Q 398K Duration: 148sec 256x256 99m Technetium

ANT V 381K Duration: 59sec 256x256 81-Krypton
POST V 399K Duration: 59sec 256x256 81-Krypton
LPO V 398K Duration: 87sec 256x256 81-Krypton
RPO V 397K Duration: 75sec 256x256 81-Krypton

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Diagnostic algorithm for CTEPH

1. Symptoms, signs, history suggestive of CTEPH
2. Echocardiographic probability of PH (Table 8)
3. High or intermediate probability of PH
4. V/Q scan
   - Mismatched perfusion defects?
   - Yes: CTEPH possible
     - Refer to PH/CTEPH expert centre
     - CT pulmonary angiography
       - Right heart catheterization
       - Pulmonary angiography
   - No: CTEPH ruled out
     - Work-up for PH/PAH (Figure 1)

CT = computed tomography; CTEPH = chronic thromboembolic pulmonary hypertension; PAH = pulmonary arterial hypertension; PH = pulmonary hypertension; V/Q = ventilation/perfusion.
*CT pulmonary angiography alone may miss diagnosis of chronic thromboembolic pulmonary hypertension.
CT pulmonary angiogram showing enlarged right atrium and right ventricle with septal flattening
CT pulmonary angiogram showing enlarged main pulmonary artery
CT pulmonary angiogram showing chronic eccentric thrombus in right pulmonary artery
Coronal CT and magnetic resonance images of right lower lobe web
Magnetic resonance pulmonary angiogram (maximum intensity projection)
CT mosaic attenuation of lung parenchyma

Hypoperfused lung

Normal lung perfusion
On 2 l/min oxygen
pH 7.47
pCO₂ 4.4 kPa
pO₂ 6.9 kPa

Coronary angiography:
minor coronary artery disease

Cardiac index 1.1 l/min/m²
Pulmonary vascular resistance 21 Wood units
76 y old male

**Diagnosis:** CTEPH deteriorating in WHO functional class IV with severe pulmonary hypertension, severely elevated pulmonary vascular resistance and poor right ventricular function

**Management:**
- Furosemide infusion
- Dopamine infusion
- Continue anticoagulation

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Question: What further treatment would you recommend next?

- Pulmonary endarterectomy
- Balloon pulmonary angioplasty
- Riociguat for long-term treatment
- Riociguat to stabilize him, then pulmonary endarterectomy
- Intravenous epoprostenol infusion
Management algorithm for CTEPH

- Diagnosis confirmed by CTEPH expert center
  - Lifelong anticoagulation
    - Operability assessment by a multidisciplinary CTEPH team
      - Technically operable
        - Acceptable risk/benefit ratio
          - Pulmonary endarterectomy
      - Technically non-operable
        - Non-acceptable risk/benefit ratio
          - Targeted medical therapy
            - Consider BPA in expert center
          - Persistent symptomatic PH
            - Consider lung transplantation
        - Persistent severe symptomatic PH

BPA = balloon pulmonary angioplasty; CTEPH = chronic thromboembolic pulmonary hypertension; PH = pulmonary hypertension.

*Technically operable patients with non-acceptable risk/benefit ratio can be considered also for BPA.

*In some centers medical therapy and BPA are initiated concurrently.
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Pulmonary endarterectomy

Pulmonary artery pressure in the operating theatre:
pre-endarterectomy 121/40 mean 65 mm Hg
post-endarterectomy 51/10 mean 26 mm Hg
76 y old male: post operative outcome

Discharged home post surgery after 10 days

Progressive improvement in breathlessness and exercise capacity

By 3 months post operatively:

- WHO functional class II
- Walking 2 miles per day
- Mean pulmonary artery pressure 25 mm Hg
- Cardiac index 3.1 l/min/m²
- Pulmonary vascular resistance 2.4 Wood units

By 1 year his echocardiogram was within normal limits
Pulmonary endarterectomy is the treatment of choice for CTEPH

- Can be performed in 60% of CTEPH patients
- In hospital mortality 4.7%
- 3 year survival 89% (n=346, age 60 y)
- Bridging therapy with PAH drugs increases risk of death

Delcroix M et al Circulation 2016;133:859-871
A 76 year old male presented with gradual onset of breathlessness and heart failure over 1 year, presented in WHO functional class IV with severe oedema.

Investigations confirmed a diagnosis of operable CTEPH with severe right ventricular impairment.

Three months after pulmonary endarterectomy he had near normal haemodynamics and right ventricular function.
Key messages

- CTEPH is treatable: always consider pulmonary endarterectomy!
- Always consider CTEPH if a patient with a previous pulmonary embolism presents with exercise induced breathlessness
- CT pulmonary angiography cannot exclude CTEPH: perform a nuclear ventilation perfusion scan
- Refer all patients with CTEPH to a multidisciplinary CTEPH team
Thank you