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Patient Information

Pulmonary Embolism and Chronic Thromboembolic Pulmonary Hypertension

Pulmonary Embolism – introduction

Pulmonary embolism (PE) mostly commonly occurs after a clot in the deep veins (deep vein thrombosis – DVT) in the lower limb or pelvis breaks off, passes through the circulation and lodges in the smaller vessels in the lungs. PE and DVT are different manifestations of the same disease – venous thromboembolism (VTE).
It is an unusual diagnosis in the young, especially if there are no risk factors, but it becomes increasingly common as we age and as the conditions VTE is associated with are more likely.

The major risk factors are:

- Surgery*
  - Major abdominal/pelvic surgery
  - Hip/knee replacement
  - Postoperative intensive care
- Obstetrics
  - Late pregnancy
  - Caesarean section
  - Puerperium
- Miscellaneous
  - Previous proven VTE
- Malignancy
  - Abdominal/pelvic
  - Advanced/metastatic
- Lower Limb problems
  - Fracture
  - Varicose veins
- Reduced mobility
  - Hospitalisation
  - Institutional care

*Where appropriate prophylaxis is used, relative risk is much lower.

PE can present with sudden circulatory collapse and death, sudden chest pain and breathlessness and in some patients be responsible for insidious breathlessness, it can also mimic many other conditions. A missed PE might be a warning for a larger PE and missed PE might also increase the risk of heart failure, and these worries drive a lot of the anxiety to diagnose PE in doctors. However not every PE is clinically relevant, small PE may be unnoticed by the body and circulation, furthermore the treatment for PE is thinning the blood with anticoagulants, and these can increase the risk of bleeding, which can, at worst be fatal itself.

A further complicating fact is that the investigations for PE involve radiation, with long-term health effects that are difficult to quantify. The investigations are not completely accurate, they are very technically demanding to do and to interpret – some small clots are missed and small abnormalities in the circulation and lungs that we all have can be misinterpreted as clots.

It is essential if you consult a doctor who is worried about the possibility of PE that they weigh up all this information quickly and accurately. That they will equally not miss the

possibility of PE if it is likely or reasonably should be considered, but also that they do not put a patient at low risk of PE through unnecessary investigations, where there is a possibility of causing more harm than good. It is a very difficult balance. The ideal doctor will be able to discuss these risks with you as part of the process of consent at each stage of your clinical assessment, diagnostic tests and therapeutic recommendations.

Decisions around anticoagulation cannot receive guidance here, merely to say the patient must make the decisions having been fully informed of the situation by their doctor – this is the process of *informed consent* and it is the bedrock of our clinical practice.

Temporary risk factors causing a single episode of VTE will normally receive a time limited course of anticoagulation. However if the risk factor is permanent, and/or if the VTE is recurrent a risk benefit decision with the patient will often be undertaken about remaining on anticoagulation longer term, sometimes permanently (though an individual risk assessment, weighing up the pros and cons of anticoagulation needs to be made at least yearly).

Chronic thromboembolic pulmonary hypertension

Most patients with PE are told of the long-term risk of recurrence of VTE and the risks associated with anticoagulation, but too many are not warned about the possibility of developing long-term difficulty with blood flow through their lungs causing chronic thromboembolic pulmonary hypertension (CTEPH). CTEPH remains a poorly understood disease with very wide estimates as to how commonly it occurs after PE from 1:1000 up to 1:11 patients! It is reasonable to think CTEPH develops in <1:25 patients. Around a quarter of cases of CTEPH develop in the absence of a history of acute PE.

Clinical symptoms and signs are non-specific or absent in early CTEPH. Only in advanced disease might the patient get signs of right heart failure. The average time from symptom onset to diagnosis is 14 months, in expert centres. The symptoms of CTEPH can mimic acute PE or other causes of pulmonary hypertension. The symptoms can be episodic, the acute PE is followed by a period of relative normality – a *honeymoon* period.

Patients may complain of intermittent or progressive breathlessness and haemoptysis can occur. They should ideally now consult a doctor with sufficient experience of the long-term consequences of PE to appropriately consider the diagnosis.

If the diagnosis is considered an echocardiogram will be the next investigation, there are necessary criteria for right ventricular dysfunction, which if present, should lead to a V/Q scan. Only if V/Q is normal can the possibility of CTEPH be (virtually) ruled out.

If CTEPH is still possible the patient needs very specialist consideration of the pros and cons of right heart catheterisation and pulmonary angiography with or without CT/ MRI, now under a very specialist team to make the diagnosis; lifelong anticoagulation is recommended in CTEPH. The patient should then be referred to a centre with sufficient experience in managing CTEPH – a multidisciplinary CTEPH team – and together, the patient and team can consider whether the condition is technically operable, then, together, what the risk: benefit ratio is. If the risk is unacceptable a second opinion might be necessary.

The surgical treatment of CTEPH is pulmonary endarterectomy (PEA) by a surgeon and team specifically trained in the procedure. It is potentially curative, restoring normal blood flow and improving symptoms in the majority, but the risk of death around the procedure must be considered; the risk is improving as experience in managing the condition gathers in specialist centres performing PEA regularly.

Should my doctor be looking for CTEPH?

I do not recommend routine investigations after a PE for the possibility of CTEPH. With that reasonable estimate that it might occur in < 1:25 patients after PE, in the 18-24 months after the diagnosis it is not desirable or feasible to look for it in asymptomatic patients. However if progressive breathlessness on exercise, or at rest, occurs, with or without unexplained haemoptysis I consider it.

It is important that the patient consults a doctor experienced in considering CTEPH, it is a different diagnostic challenge to the initial suspicion of PE and even the initial diagnosis of PE. The right doctor will have excellent links with echocardiography, nuclear medicine and diagnostic imaging, local haematology and anticoagulation services, cardiovascular medicine, and that team, with the patient, will be able to consider referral of appropriate patients to a CTEPH MDT should that be necessary.