reference no: 1620
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Clinical Commissioning Policy:

Balloon pulmonary angioplasty for chronic thromboembolic pulmonary hypertension (all ages) [1620]

Summary

Balloon pulmonary angioplasty is recommended to be available as a routine commissioning treatment option for chronic thromboembolic pulmonary hypertension (all ages) within the criteria set out in this document.

What we have decided

NHS England has carefully reviewed the evidence to treat chronic thromboembolic pulmonary hypertension (all ages) with balloon pulmonary angioplasty. We have concluded that there is enough evidence to make the treatment available at this time.

Links and updates to other policies

This document updates:

- Clinical Commissioning Policy 170031P: Balloon pulmonary angioplasty for chronic thromboembolic pulmonary hypertension (all ages)

Plain language summary

About chronic thromboembolic pulmonary hypertension

Chronic thromboembolic pulmonary hypertension (CTEPH) is a form of lung disease where blood pressure in the lungs is raised due to blood clots in the blood vessels in the lung. This causes right heart strain and may cause premature death. Normally if blood clots travel to the lungs (pulmonary emboli), they are broken down either by the body or by treatments e.g. anti-coagulation therapy. However, in CTEPH the blood clots damage the main arteries (pulmonary arteries) and block the blood flow through the lungs. This causes breathlessness, fatigue, dizziness, chest pain, body swelling, coughing up of blood and an inability to exercise which all impact on patient's poor quality of life. Often patients cannot work and need to take oxygen all the time just to remain comfortable when resting. Usually the disease, if not treated, leads to heart failure and death.

Currently approximately 40% of patients with CTEPH are inoperable and with the limited current alternative treatment options the three year survival rate is only 70% for this group (Delcroix et al 2016a). These patients are currently offered disease modifying therapies

including endothelin receptor antagonists (ERAs) or novel soluble guanylate cyclise stimulators (SGCs) according to an agreed NHS England Commissioning Policy: Targeted Therapies for use in Pulmonary Hypertension in Adults Commissioners PH targeted therapy policy (NHS England A11/P/c), which is continued lifelong. With current medical therapy survival is improved compared with historical controls but is inferior to PEA surgery.

About current treatment

The treatment of choice for CTEPH is surgery to remove the clots; this is called pulmonary endarterectomy (PEA). This is a very complex, high risk operation requiring a lowering of body temperature and stopping blood circulation for a controlled period of time and is delivered at only one specialised centre in the UK. The average operative time is 8 hours. PEA has a mortality of 1.9% (Jenkins et al 2022). The majority of patients improve after surgery and some are cured but 20-40% of patients are not suitable for surgery due to inaccessible disease or prohibitive risk (Mayer et al 2011). These patients are currently treated medically and they have a worse life expectancy and poorer quality of life than those successfully treated by surgery. Patients with inoperable CTEPH have an unmet clinical need. They have a worse life expectancy (70% survival at 3-years) (Delcroix et al 2016a) and poorer quality of life compared to those patients who undergo surgery despite medical treatment with diuretics, digitalis, pulmonary vasodilator therapies and long term oxygen therapy.

Treatment options for patients with inoperable disease are limited:

- Exercise training has been found in small studies to improve exercise capacity and quality of life in patients with inoperable CTEPH who are stable on therapy with pulmonary arterial hypertension (PAH) targeted therapy (Nagel 2012, Fukui 2016). Concern about safety and tolerability has limited its use in routine care for patients with inoperable CTEPH (Fukui 2016).

- Conventional medical treatments to reduce pulmonary vascular resistance (diuretics, digitalis and chronic oxygen therapy) have little effect and do not affect the underlying disease processes in CTEPH (Pepke-Zaba 2016).

- Patients with CTEPH may be treated with medications used against PAH such as prostacyclin analogs (epoprostenol, iloprost), endothelin receptor antagonists (bosentan, ambrisentan, macitentan) and phosphodiesterase-5 inhibitors (sildenafil, tadalafil). Small uncontrolled trials of these medications in CTEPH have had mixed results (Pepke-Zaba 2016). A randomized controlled trial of bosentan in 157 patients with CTEPH (BENEFiT) found improved pulmonary vascular resistance at 16 weeks among patients receiving bosentan compared to patients receiving placebo, but 6 minute walk distance (6MWD) was not significantly different (Mathai 2016).

- Riociguat, a soluble guanylate cyclase stimulator, is the first drug to be licensed for use in inoperable or persistent recurrent CTEPH. The main evidence of efficacy of riociguat against CTEPH is derived from one randomized placebo-controlled trial of 261 patients, CHEST-1, which was followed by a 2-year extension, CHEST-2 (Ghofrani 2013, Simonneau 2016). The detailed results of these trials are described below.

About balloon pulmonary angioplasty (BPA)

Balloon pulmonary angioplasty (BPA) aims to treat CTEPH with the patient awake with a “keyhole” surgical technique. BPA treats the narrowing in the scarred lung arteries with a balloon that is inflated to stretch open the lumen improving lung blood flow. To reduce the risk of complications, the procedure is performed in stages and several sessions (2-6 per patient) are usually required to achieve therapeutic benefit. These are performed at 2-8 week intervals.

Epidemiology and needs assessment

In the general population, the incidence of CTEPH is estimated to be approximately 5 cases per million people per year (Delcroix et al 2016b). Most patients have a known history of massive or recurrent acute pulmonary embolism. It is estimated that between 0.6 to 4.8 % of people who have an acute pulmonary embolism develop CTEPH within the next 2 years (Leopold 2016). This suggests around 272 new cases of CTEPH per annum in England, of whom approximately 60% will be treated with PEA; the remaining 40% will be very heterogeneous group of patients:

1. who are technically operable but not suitable for surgery due to severe co-morbidities (n~18 per annum) or physician/patient choice due to mild symptoms (n~18 per annum)
2. with distal distribution of disease not accessible by PEA and managed medically with PAH targeted therapies at present. There are two potential patient subgroups in this inoperable group:
    a. Patients with lesions suitable for the BPA (main target are considered to be webs and slits lesions. Lesions with severe narrowing or complete obstruction by webs could also be treatable as long as distal run-off is confirmed on imaging), possibly enabling PAH medical therapy to be weaned (n~27 pa).
    b. Patients with lesions inaccessible for BPA who will continue with medical management with targeted PAH therapy or lung transplantation (n~45pa).

These data have been confirmed by National Audit (Health and Social Care Information Centre 2014) provided by Pulmonary Hypertension Centres in the UK. Detailed triaging of inoperable CTEPH patients has determined that 20-30 patients per annum would be clinically eligible for BPA. There is expected to be a small amount of growth in the CTEPH population.

Implementation

Inclusion criteria

BPA will be commissioned for patients with symptomatic CTEPH who have disease not suitable for PEA and angiographic lesions amenable to BPA:

- Mean pulmonary arterial pressure greater than 20 mmHg at right heart catheterisation
- Patient is symptomatic in WHO functional class III or IV
- The pulmonary vascular obstructions (i.e. the disease distribution) is distal, not suitable for pulmonary endarterectomy
- The presence of narrowed or blocked vessels (occluded sub-segmental pulmonary arteries) identified using conventional pulmonary angiography
- There is a Balloon Pulmonary Angioplasty multidisciplinary team (MDT)[1] meeting decision that there are sufficient, accessible, vascular lesion for BPA treatment to improve patient symptoms and haemodynamics.
- The patient has already been discussed by the PEA MDT meeting and assessed as not suitable for PEA.

The documentation will be captured in the data base and patient case notes.

Exclusion criteria

Patients are excluded if they meet **ANY** of the following exclusion criteria:

- Comorbidities that may be contraindications to the intervention (for example, those that increase the risk of the procedure)[2]
- Comorbidities that reduce the likely ability of the patient to benefit, including where life expectancy is likely to be reduced[3]

Starting criteria

Operability is decided by the National Pulmonary Endarterectomy Multi-Disciplinary Team meeting.

Patient pathway

[1] The MDT meeting will be attended by:
- Consultant Interventional cardiologist or consultant interventional radiologist performing BPA,
- Consultant Radiologist with interest into pulmonary vascular diseases,
- Consultant Respiratory Physician,
- BPA coordinator nurse and minute taker

[2] Comorbidities that may be contraindications to the intervention in specific patients include left heart disease: systolic left heart failure, disease of the heart valves, significant parenchymal lung disease (such as interstitial lung fibrosing and severe emphysema).

[3] Comorbidities that reduce the likely ability of the patient to benefit include active malignancy, and extreme fragility of the patient.

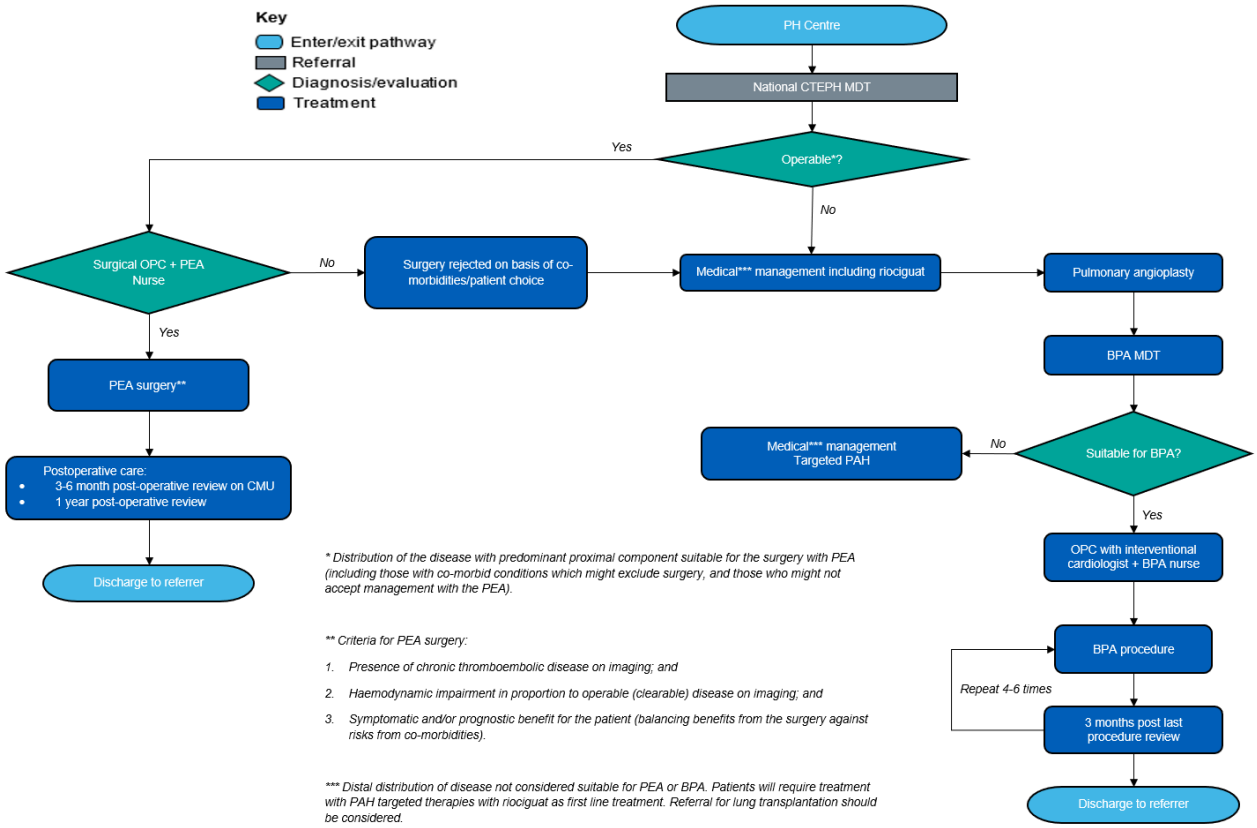
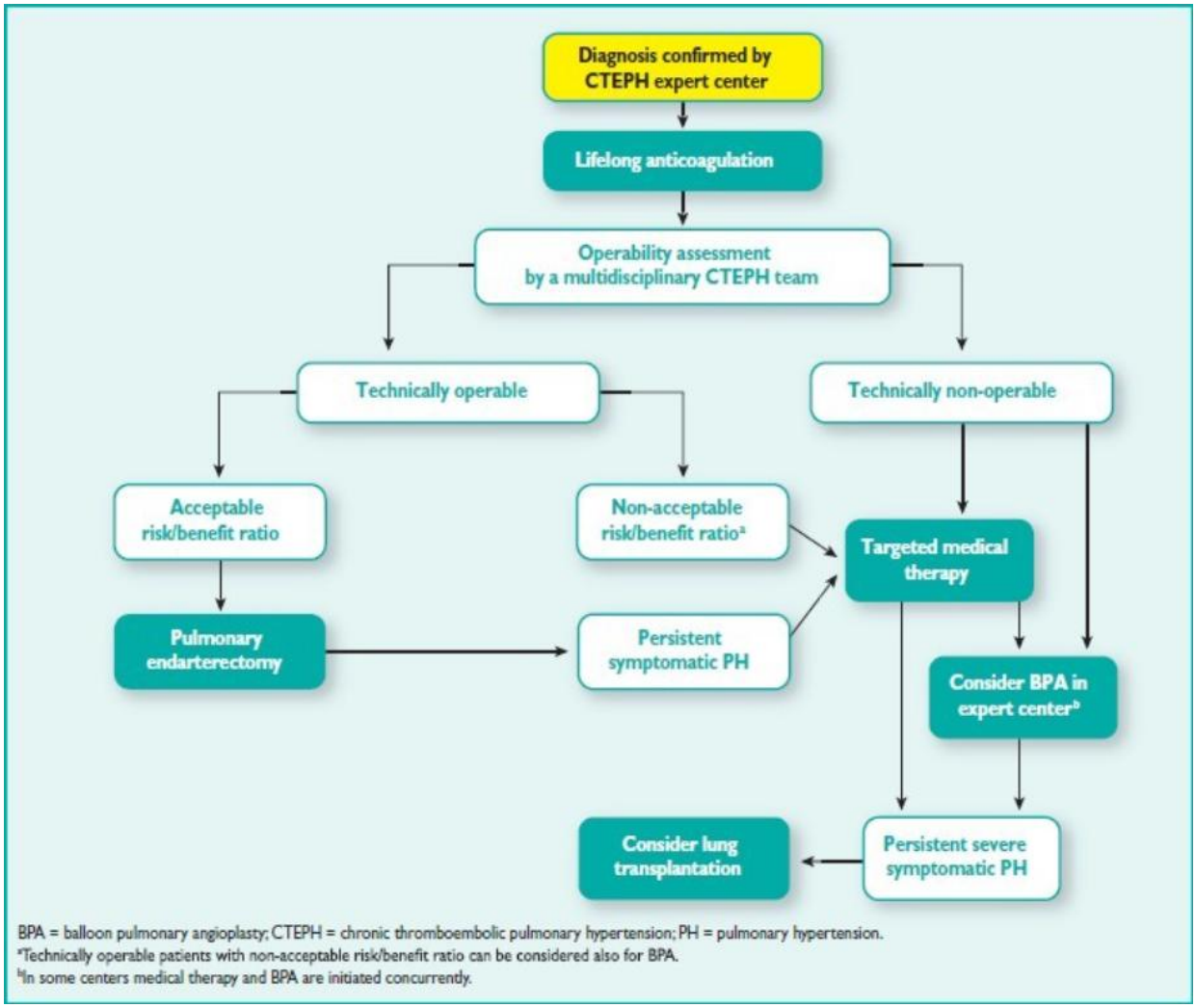


Figure 1 - Patient pathway for CTEPH

Patients with suspected CTEPH will be diagnosed and managed by a specialist PH centre. Patients will be referred from one of these centres (six in England) to the designated provider of PEA, where the patient's suitability for PEA will be assessed. If disease distribution is unsuitable for PEA, the patient will be referred to the BPA provider team. Proposed patient pathway is consistent with current European Society of Cardiology/ European Respiratory Society guidelines (2015 ESC/ERS Guidelines) on the management of CTEPH which is illustrated below.



Source: 2015 ESC/ERS Guidelines

NHS England is only commissioning this service from the current single national PEA provider in the first instance because this is the team that has the expertise to determine whether or not a patient might benefit from PEA or whether BPA is a more appropriate intervention. Once further evidence is gathered and future growth can be determined, NHS England may decide it is appropriate to commission a BPA service from additional providers. If this is the case, it will undertake a market analysis exercise to determine the extent of such expertise.

NHS England has a published policy on the use of Riociguat for the treatment of PAH and CTEPH [Reference: NHS England: 16055/P].

Current ESC/ERS guidelines (2015 ESC/ERS Guidelines) suggest starting Riociguat as the first line therapy and then proceeding to the BPA if indicated.

Rationale to treat patient first with Riociguat and then proceed then to BPA is:

1. There may be a significant delay between presentation to the PH centre and ability to offer BPA.
    1. Adjuvant therapy with PAH targeted therapy may make BPA safer and facilitate BPA treatment (allowing more segments to be treated and therefore fewer procedures).

2. Successful BPA may allow patients to function without targeted pulmonary hypertension therapies, e.g. Riociguat. (See flowchart below for further detail on the patient pathway with the legend further describing clinical criteria at each point in the pathway).

Governance arrangements

BPA will be commissioned from a single provider with experience of PEA.

Mechanism for funding

The funding and commissioning will be managed through the relevant local NHS England Specialised Commissioning Team from 1st April 2018.

Audit requirements

All patients with CTEPH are already included in the national arrangements for audit of PH. The following information will be collected for all patients treated with BPA as recommended by NICE in its Interventional Procedures Guidance 554, and uploaded to the NHS England QSIS portal:

Summary of data for BPA for chronic thromboembolic PH.

https://www.nice.org.uk/guidance/ipg554/resources

This tool helps clinicians using BPA for CTEPH to review clinical outcomes. Data should be reviewed at appropriate intervals and practice should be changed if the results suggest the need to do so.

The tool contains a data collection sheet containing drop down options and free text boxes. A summary of the data is shown in the tables below:

Consent
A discussion has taken place about the uncertainties surrounding the procedure's safety and efficacy
The patient has received written information explaining that there are uncertainties about the procedure's safety and efficacy
Written consent to treatment has been obtained

Outcome measures of benefit
Improved functional activity (WHO-functional class / NYHA class)
Improved exercise tolerance (six minute walk test)
Improved pulmonary haemodynamics (pulmonary vascular resistance)
Reduction in the use of oral medication
Other outcome measure of benefit

Adverse outcomes
Mortality resulting from procedure
Perforation
Aneurysm
Reperfusion pulmonary oedema
Haemoptysis / haemosputum
Desaturation
Acute kidney injury
Other adverse outcome

To ensure that any valuable insight regarding the consequences of this procedure is shared among clinicians, serious or previously unrecognised patient safety incidents should be documented and information submitted to the National Reporting and Learning System (NRLS), operated by the National Patient Safety Agency.

Policy review date

Add text in 'Paragraph' style

This document will be reviewed when information is received which indicates that the policy requires revision. If a review is needed due to a new evidence base then a new Preliminary Policy Proposal needs to be submitted by contacting england.CET@nhs.net.

Our policies provide access on the basis that the prices of therapies will be at or below the prices and commercial terms submitted for consideration at the time evaluated. NHS England reserves the right to review policies where the supplier of an intervention is no longer willing to supply the treatment to the NHS at or below this price and to review policies where the supplier is unable or unwilling to match price reductions in alternative therapies.

Equality statement

Promoting equality and addressing health inequalities are at the heart of NHS England's values. Throughout the development of the policies and processes cited in this document, we have:

- Given due regard to the need to eliminate discrimination, harassment and victimisation, to advance equality of opportunity, and to foster good relations between people who share a relevant protected characteristic (as cited under the Equality Act 2010) and those who do not share it; and

- Given regard to the need to reduce inequalities between patients in access to, and outcomes from healthcare services and to ensure services are provided in an integrated way where this might reduce health inequalities.

Definitions

Balloon pulmonary angioplasty (BPA)	The use of percutaneous catheter directed balloon dilation treatment in stenotic and sub-totally occluded sub-segmental pulmonary arteries.
Chronic thromboembolic pulmonary hypertension (CTEPH)	A rare form of PH, is categorized by the World Health Organisation as Group IV PH (see below). It is defined as mean pulmonary arterial pressure ≥25 mmHg and pulmonary capillary wedge pressure ≤15mmHg in the presence of multiple chronic/organized occlusive thromboemboli in the elastic pulmonary arteries (main, lobar, segmental, sub- segmental) after at least three months of effective anticoagulation.
Pulmonary endarterectomy (PEA)	The surgical removal of accessible intravascular occlusive material, which is the first choice therapy for CTEPH.
Pulmonary hypertension (PH)	A disorder of the blood vessels in the lung, characterised by raised pressure in the pulmonary artery, which results in a range of symptoms and may be life threatening.
Medical therapy	Pulmonary vasodilators can be used to control symptoms of pulmonary hypertension. These are only prescribed by designated Pulmonary Hypertension Centres.
World Health Organisation (WHO) functional classes	A standard classification of disease severity in PH. Patients in class I have no symptoms; patients in class IV are symptomatic even at rest. Classes II and III are intermediate.

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