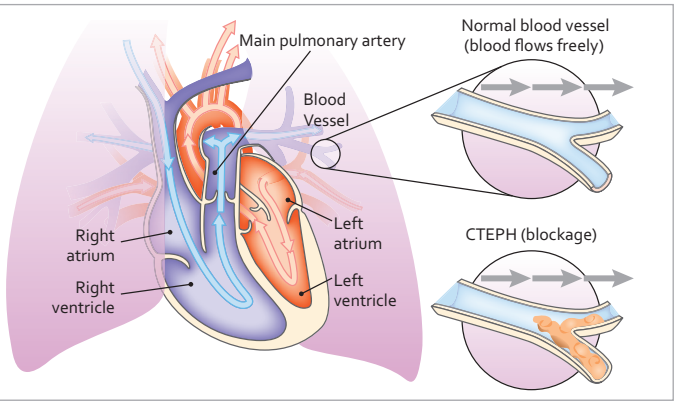
Chronic Thromboembolic Pulmonary Hypertension

Part 2

Chronic thromboembolic pulmonary hypertension (CTEPH) is a condition where there is elevated blood pressure in the pulmonary arteries caused by chronic blood clots (thromboembolic), which obstruct the free flow of blood through the lungs.



This is a special form of pulmonary hypertension that, unlike all the other forms, can potentially be cured with a surgical procedure. This fact sheet will review treatment of CTEPH. For more information about the diagnosis of CTEPH, see ATS Patient Information Series piece *Chronic Thromboembolic Pulmonary Hypertension (CTEPH)* at www.thoracic.org/patients.

What are the treatment options for CTEPH?

Left untreated, CTEPH is a deadly condition. Fortunately, it is perhaps the most treatable form of pulmonary hypertension, and for some people even cure is possible. The treatment of choice for CTEPH is a surgical procedure called pulmonary thromboendarterectomy [thrombo-endar-ter--ek-tomy] (or PTE for short, sometimes referred to as pulmonary endarterectomy, or PEA). This procedure has a proven track record of positive results over several decades, and it offers a definitive cure for many people with CTEPH. This is why, as soon as the diagnosis of CTEPH is made, you should be evaluated at a medical center with experience and expertise in performing PTE surgery.

After this evaluation, some people are deemed to not be good candidates for surgery. For people in whom surgery is not possible or indicated, there are two other treatment options: medications to dilate small pulmonary arteries, and/or a procedure called balloon pulmonary angioplasty (BPA).

How do doctors determine who is a good candidate for PTE surgery?

Doctors look at three main factors to decide who is a good candidate for this procedure. The first one is whether or not the chronic blood clots are located in big enough arteries, so that they can be reached by the surgical instruments. The second question is whether or not the high pulmonary blood pressure is likely accounted for by the amount of pulmonary clots present.

And finally, a person has to be otherwise strong enough to be able to get through and survive a big operation. If the answer to these three questions is yes, then PTE surgery is a good idea. The main challenge however, is that these decisions are complex, largely based on expert opinion, and shaped by the team's experience and expertise. This is why this determination needs to be made at an expert center. Importantly, older age and obesity do not routinely represent absolute barriers for this procedure.

How is PTE surgery done?

This operation is done through a chest wall incision; this is the same type of incision used for heart bypass surgery. Then, doctors stop the heart, drain the blood from the heart and lung arteries, and cool the person's body to about 60 degrees, which prevents brain damage. The surgeon then peels off the scar clot tissue lining and plugging the pulmonary (lung) arteries. Performing a PTE requires a dedicated team and a highly skilled surgeon.

What is the recovery like after PTE surgery?

The usual hospital stay is 10-14 days. Some people notice immediate and dramatic improvements in their breathing function and comfort, while for others it may be a more gradual process. You will be able to gradually resume your usual activities over the following 4-6 weeks. Pulmonary rehabilitation with supervised exercise is often helpful. You may need extra oxygen for some time after surgery. You will be given blood thinner medication, which you will need to keep taking for the rest of your life. By 3-6 months after successful PTE surgery, most people are able to resume normal or near normal levels of activity and lead normal productive lives. Your healthcare providers will want to see you frequently to check for improvements in exercise capacity and pulmonary pressures, as well as for potential complications. You will have follow-up tests

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Am J Respir Crit Care Med Vol. 197, P5-6, 2018

such as the 6-minute walk test, echocardiography, ventilation–perfusion scan and right heart catheterization during the first 3-6 months after surgery. These tests can be done at the PTE center or with your local pulmonary hypertension doctor. For more information on pulmonary rehabilitation, see ATS Patient Information series piece at www.thoracic.org/patients.

What are the potential complications of PTE surgery?

Like with any other major operation, not surviving the procedure is the most serious potential outcome. Currently, in experienced centers, risk of death after PTE surgery is under 5%. Immediately after surgery, fluid build-up in the lungs and bleeding are the most important complications. Pulmonary pressures can remain elevated after surgery. Even if pulmonary pressures return to normal immediately after surgery, they can increase again several months or years later. The CTEPH team will assess how likely these complications are for you, and factor this into the decision to offer surgery.

What is the role of medications in CTEPH?

Everyone who has CTEPH needs to be on a blood thinner for life. This remains true even after successful PTE surgery. Warfarin (brand name -Coumadin) continues to be the preferred blood thinner, as health care providers have decades of successful experience using this blood thinner in people with CTEPH. Whether or not the newer blood thinners available are as effective and safe is not clear at the moment. There is no current blood thinner that addresses the scar clot tissue or the pulmonary pressure elevation. Blood thinners only prevent new blood clots from forming.

Treating this type of pulmonary hypertension with medications is only indicated if you have been properly evaluated by an expert CTEPH center and deemed not to be a good candidate for pulmonary thromboendarterectomy. Currently, there is one FDA approved pill for this indication called riociguat. This medication can also be used if you are left with pulmonary hypertension after PTE surgery. Under these circumstances, this medication is known to improve symptoms and relief pressure elevation in the lungs. Your healthcare provider may also choose to use other medications instead of or in addition to those approved for other forms of pulmonary hypertension.

What is balloon pulmonary angioplasty (BPA)?

In general, angioplasty is a catheter-based procedure well established in its use for treating blocked vessels in the heart and brain. Its application in the lungs is called balloon pulmonary angioplasty (BPA). A rubber tube (catheter) is placed through a blood vessel into the pulmonary arteries. Inflatable balloons are then used to open up blocked vessels. Today, several centers in Japan and increasingly in the USA and Europe are using modern equipment and techniques for this procedure with good results. While BPA is far less invasive than thromboendarterectomy and patients are awake during the procedure, the arteries in the lung are fragile and vulnerable to puncturing (perforation). Moreover, to minimize the risks, BPA must be performed in two to five separate sessions—a process some people might find difficult. BPA is currently indicated only for people in whom PTE surgery is not indicated or not possible.

What is the best treatment option for me?

This is a very difficult decision that requires careful consideration. The stakes are really high. If surgery is feasible, this is clearly the treatment of choice, as it offers the highest likelihood of significant short-term and long-term improvements, even cure in many cases. If surgery is not indicated, medical therapy and/or balloon pulmonary angioplasty are now reasonable treatment options for you. The choice needs to be made after evaluation at an expert CTEPH center with the ability to perform all of these three treatment options. You should not be treated with medicines or BPA in lieu of a surgical evaluation.

Authors: Gustavo A. Heresi MD, Nancy Bair CNS-BC, Raed A. Dweik MD
Reviewers: William Auger MD, Marianna Sockrider MD, DrPH

Rx Key Points

- ✔ The best treatment for pulmonary hypertension due to chronic blood clots (CTEPH) is a surgical procedure called pulmonary thromboendarterectomy (PTE, also called pulmonary endarterectomy or PEA).
- ✔ When surgery is not feasible, medications and/or a procedure called balloon pulmonary angioplasty (BPA) are alternative treatment options.
- ✔ If you have been diagnosed with CTEPH, make sure that you are evaluated at a medical center with experience and expertise in CTEPH, and able to offer all three currently available treatment options.

Healthcare Provider's Contact Number:

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Resources:

Pulmonary Hypertension Association
http://www.phassociation.org/CTEPH

PHA Europe (European Pulmonary Hypertension Association)
http://www.phaeurope.org/dissease-information/what-is-cteph/

American Heart Association (AHA)
http://www.heart.org/HEARTORG/

American Lung Association
http://www.lungusa.org

American Thoracic Society (ATS)
http://www.thoracic.org/patients

National Organization of Rare Diseases (NORD)
https://rarediseases.org/organizations/nihoffice-of-rare-disease-research/