



Living with chronic thromboembolic pulmonary hypertension (CTEPH): Supporting you to manage and achieve your goals

early diagnosis • best treatment • better quality of life • finding a cure

PHA EUROPE *for the patients*
European pulmonary hypertension association



About this brochure

- This brochure contains useful information about chronic thromboembolic pulmonary hypertension (CTEPH), a form of pulmonary hypertension (PH), and includes treatment options and practical advice on living with CTEPH
- If you have any questions about CTEPH or your general health, make sure you ask your healthcare team
- This brochure will also be useful to your family, caregivers or friends, so they can support you better
- If you wish to learn from other patients' experiences, or share your own personal process, you can contact your national PH patients' organization



Contents

- 4 What is PH?
- 5 What is CTEPH, and how does this differ from other types of PH?



- 6 What does CTEPH feel like?



- 7 Keeping a note of how you feel



- 9 What treatment options are available?



- 10 Personalizing your treatment plan to achieve your goals



- 11 What is it like to live with CTEPH?



- 12 Practical advice for living with CTEPH



- 14 As a caregiver, how can I support someone living with CTEPH?

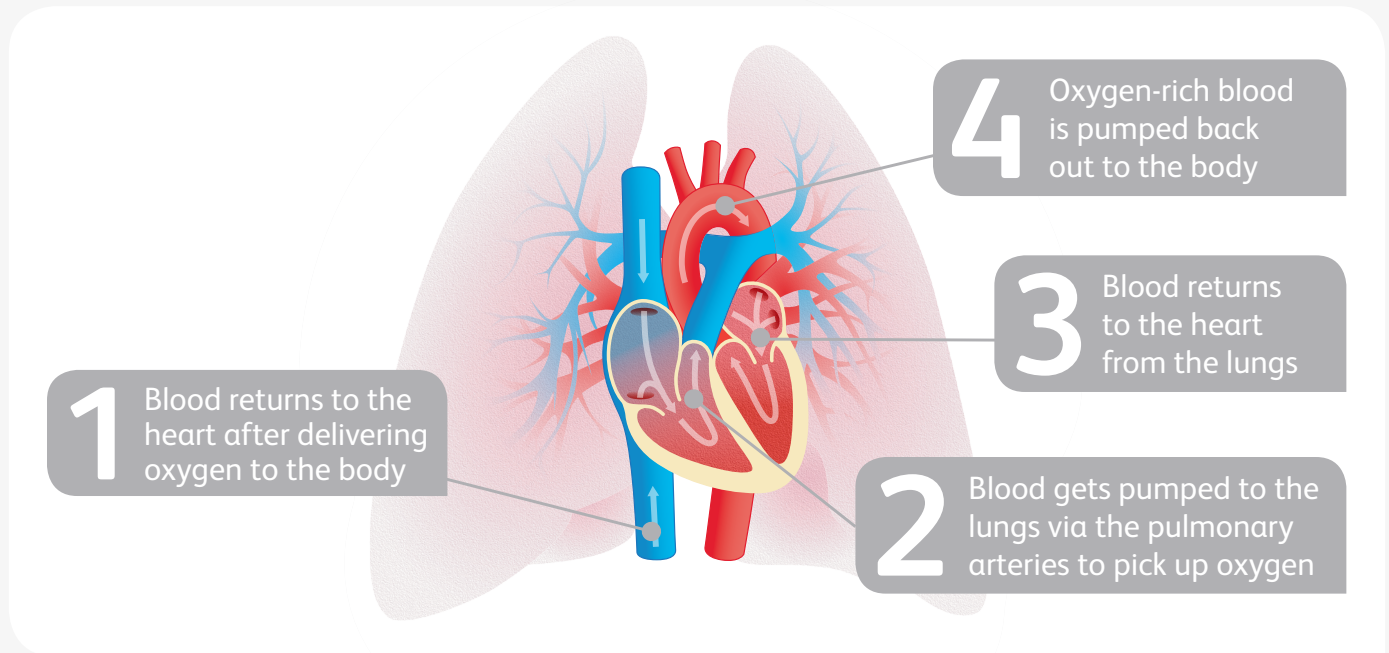


- 16 Notes
- 18 Further information
- 19 References



What is PH?

The lungs and heart work together to supply the body with oxygen, as described below:

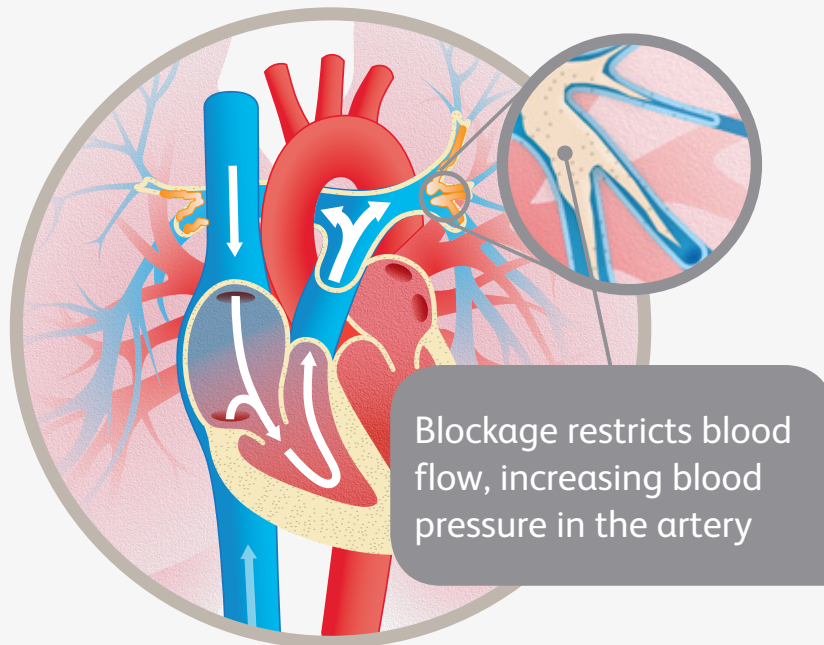


- PH is a disease of the lungs and heart, in which the blood pressure in a person's lungs is higher than the normal range¹
- PH may be caused by narrowing or blockage of the pulmonary arteries.¹ This reduces the space in which blood travelling to the lungs can flow, causing the blood pressure in the pulmonary arteries to increase, just like the rise in water pressure that happens when you squeeze a garden hose
- These changes put a strain on the right side of the heart,¹ which means that it needs to work harder than normal to pump blood to the lungs
 - If your lungs receive less blood, less oxygen will be picked up and circulated around your body. This might leave you feeling breathless



What is CTEPH, and how does this differ from other types of PH?

- There are several forms of PH, which differ in terms of how they develop, and how they are diagnosed and treated^{1,2}
- CTEPH is one form of PH, and is caused by a blood clot sticking to the wall of a blood vessel in the lungs. This restricts blood flow, leading to high blood pressure in the lungs³



What does CTEPH feel like?

As someone living with CTEPH, you may be experiencing symptoms such as shortness of breath during exercise, feeling tired, weak or dizzy, fainting, chest pain, and swelling of ankles, arms or stomach.²



Shortness of
breath with
exercise



Feeling tired,
weak or
dizzy



Swelling in the
ankles, arms or
stomach area



Fainting



Chest pain

- These symptoms may become more noticeable over time, making it more difficult for you to do normal daily activities or chores
- Talk to your doctor if you notice that your symptoms are getting worse



“I’m finding it more difficult to keep up with my dog on our walks together. He often has to wait for me so I can catch my breath. Our walks aren’t as fun as they used to be.”

All the quotes within this brochure are fictitious accounts based on real-life experiences of people living with CTEPH.



Keeping a note of how you feel

- It is important that you see your doctor on a regular basis to review and discuss your health
- Your doctor wants to know how you are feeling now, as well as how you have been feeling since your last visit. So, before you see your doctor, think about how you have been since your last visit
- Your doctor may talk about your 'functional class'. This is one way of describing how severe your symptoms are and whether they affect your daily life (for example, your ability to do exercise or regular chores)
 - Classifying the severity of your CTEPH will help your healthcare team understand better how it is affecting you

Functional class²

I	Symptom-free when physically active or resting	• Make sure you tell your doctor if you think your symptoms have improved
II	No symptoms at rest, but normal activities such as climbing the stairs, grocery shopping or making the bed cause some discomfort and shortness of breath	• If you feel your symptoms are worse, or are not improving, it is important to tell your doctor, so you can work together to find the best way forward
III	Resting may be symptom-free, but normal chores around the house are greatly limited because of shortness of breath or feeling tired	
IV	Symptoms at rest and severe symptoms with an activity	



Keeping a note of how you feel

Here is a list of some of the things you might wish to talk to your doctor about at your next visit. Some people find it easier to write things down, so there is space for you here.

Since your last visit:



How have your general energy levels been?

.....
.....



Have you felt normal daily activities (like washing, dressing, or moving around the house) are noticeably more (or less) challenging?

.....
.....



Have you experienced any new symptoms?

.....
.....



Have you experienced dizziness, weakness or feeling faint?

.....
.....



How many minutes can you walk for before you need to take a break?

.....
.....



How many flights of stairs can you walk up before you feel breathless?

.....
.....



To what extent do you feel that your CTEPH is affecting your overall quality of life and emotional well-being? For example, have you noticed that your mood, motivation and/or energy levels are lower than usual?

.....
.....



What treatment options are available?

There are several treatment options for CTEPH:



PEA surgery

- CTEPH can potentially be cured, through an operation called a pulmonary endarterectomy (PEA). This surgical procedure removes the blockage in the pulmonary arteries⁴
 - Even after PEA surgery, some people's CTEPH symptoms continue, or they come back at a later date, because of other changes in the lungs that CTEPH can cause⁴
 - If CTEPH still causes problems after the operation, your doctor will discuss alternative treatments with you



Medical treatment

- If surgery is not a suitable option for you, or if your CTEPH remains or returns after the operation, you can receive medication:
 - An oral medication that is used specifically to improve symptoms of CTEPH⁵
 - A blood-thinning medication (anticoagulant) to prevent new blood clots from forming in the lungs. This should be taken by all patients with CTEPH for the rest of their lives²



Balloon pulmonary angioplasty

- Balloon pulmonary angioplasty (BPA) is a procedure that uses an inflated balloon to increase the width of the blocked arteries (as opposed to removing the blockage as done in PEA surgery)⁶
- BPA may not be offered at all hospitals because the procedure is relatively specialized,² and its long-term outcomes are not yet known

Your healthcare team will discuss the available treatment options with you, and your doctor will recommend the most suitable choice for you.



Personalizing your treatment plan to achieve your goals

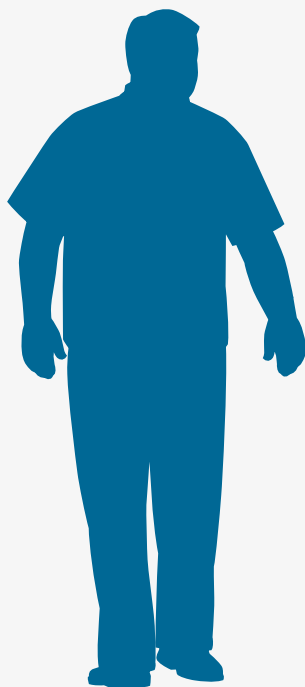
‘Making do’ with how you are feeling is not good enough; you should hope to be symptom-free when resting or while doing physical activities, or you should feel that your symptoms are improving.

- It may be useful to keep a written record of how you feel. If your symptoms have changed, or have not improved as much as you would have liked, please discuss this with your healthcare team



Claire:

“I felt that my symptoms were not getting better after having been on the same treatment for years. After talking to my doctor about my concerns, I was given another drug to take and I have started feeling better.”



Claire’s doctor:

“Regular follow-up with Claire is an important part of her care. As a result of these visits, we found that her symptoms were not improving with the medication she’d been taking for several years, so I suggested starting another treatment.”



What is it like to live with CTEPH?



Chris:

“I had surgery to remove the blockage from the arteries in my lungs. The surgery has changed everything and I have had no symptoms since. Being outside with my dog is one of the things I love, and I am so happy I can now enjoy our walks together again.”



Claire:

“I had heard that there was an operation that could potentially cure me of my CTEPH, but was told that it would be too risky for me. Thankfully, there was a medication for CTEPH that my doctor recommended I try; it has made a huge difference and I am now back to feeling more like my former self – my work schedule is now more manageable again.”



Craig:

“I had an operation that I was told could potentially cure me of my CTEPH. Unfortunately, it didn't work for me, and my symptoms came back. I discussed with my doctor the next best option, and started to take a drug for CTEPH. In the last few weeks, I have been able to ride my bike again without getting quite as breathless.”



Practical advice for living with CTEPH



Emotional well-being

Your emotional, spiritual and psychological well-being is as important as your general health. Your doctor can recommend support groups and other experts who can help with your emotional well-being.



Social support

In addition to support from family and friends, your doctor can provide you with details of local support groups and patient organizations where you can talk to other people with CTEPH. It is important to talk about how you are feeling, to reduce stress and to make sure that you do not feel alone.



Work

You might need to think about changes to your day-to-day activities at work, especially if they are leaving you breathless. Once your symptoms start to improve, you could increase your activity level at work.



Education

If you or someone you are caring for is at school, college or university, you may wish to talk to someone at the institution to let them know how CTEPH might affect studies or extracurricular activities. Changes to the timetable, extensions on assignments or examinations, and fewer or lighter activities outside of study might be helpful.



Reporting side effects

When starting treatment, it is important to speak to your doctor about any side effects that you may experience. If you think you may have experienced a side effect after using a medicine, contact your doctor who will be able to tell you if you need any alternative care. In the European Union you can also report side effects or adverse reactions directly through various methods (e.g. online patient reporting forms) without going through your physician.



Practical advice for living with CTEPH



Travel

Take care not to over-exert yourself while traveling or on holiday. Find out about any CTEPH clinics where you are going, and take information about your CTEPH and your medication with you in case you need medical support. High altitudes (for example, >1500 meters) should also be avoided, unless you can take supplemental oxygen. Some people might also need supplemental oxygen to maintain their oxygen levels during long flights.²



Diet and exercise

It is important to eat a balanced diet, and to maintain a healthy weight. You can stay active while living with CTEPH, but it is important to seek advice on exercise from your healthcare team. Carefully supervised physical activity can form part of an effective treatment strategy for CTEPH. For some patients with CTEPH, even the mildest forms of activity might be too exhausting. For others, moderate exercise might be beneficial.



Sexual health and family planning

You may experience a decline in sexual intimacy, perhaps because of concerns relating to over-exerting yourself. Talk openly with your partner about your feelings. Pregnancy poses a substantial risk for women with CTEPH, because it will put extra demands on the heart and lungs. Therefore, women with CTEPH are recommended to use reliable contraceptive methods to avoid becoming pregnant.²

For further information and support, please refer to the websites listed on page 18 at the end of this brochure.



As a caregiver, how can I support someone living with CTEPH?

Being emotionally supportive is probably the most important thing you can do as a caregiver. But there are also some practical things that you may be able to do to help the person you care for, to ensure that their emotional, psychological and physical needs are being met by helping to identify where they may need additional support.



Keep an eye on symptoms

Keep note of any changes you have noticed since the last check-up. This includes any changes in emotional well-being and behavior, as well as physical changes. It is very important that the healthcare team knows what the person you care for is going through daily and how they are progressing with their treatment goals.



Help prepare for and attend doctors' appointments

To get the most out of doctors' appointments within the often-limited time available, it is important to prepare beforehand. Have a chat with the person you care for and ask them to:

- **think carefully about what they want to get out of the appointment (see page 8)**
- **write down any concerns or questions they (or you) might have**

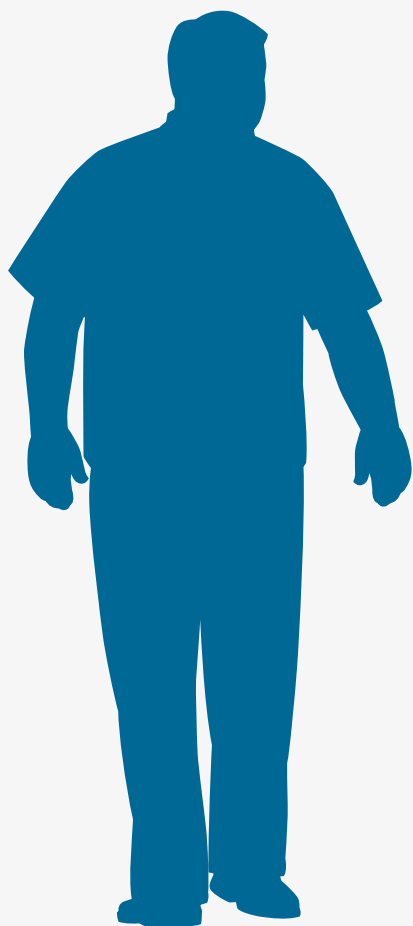


Make sure the person you care for feels valued and included

CTEPH is likely to have an impact on a person's social life. Encouraging other friends and family members to visit can stop people living with CTEPH feeling isolated, as can including them in activities of normal day-to-day life where possible (with consideration of the limitations of their disease).



As a caregiver, how can I support someone living with CTEPH?



Claire's caregiver:

“Claire sometimes isn't aware of how her symptoms are gradually changing. It is sometimes easier for me to spot things, so I note down the changes so we can discuss them with her doctor.”

As a caregiver, it is vital to take steps to preserve your own health and well-being, because when your needs are taken care of, the person you care for will benefit too. For further information on caring for someone with CTEPH, please refer to the caregiver brochure, which is free to download via the PHA Europe website:

<http://www.phaeurope.org>



Further information

The following websites offer additional information and support for people living with CTEPH and their carers:

Pulmonary Hypertension Association Europe

www.phaeurope.org

CTEPH resources for patients and their carers

www.cteph-info.com

Bayer is not responsible for the content of external websites.



References

The following publications support the guidance and advice within this brochure.

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6. Lang I *et al.* Balloon pulmonary angioplasty in chronic thromboembolic pulmonary hypertension. *Eur Respir Rev* 2017;26:160119.





Bayer AG
13353 Berlin, Germany

www.bayer.com

PP-ADE-ALL-0110-1
Date of preparation: November 2018