

Pulmonary endarterectomy (PEA)
at Papworth Hospital
Patient information booklet





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Introduction

Chronic thromboembolic pulmonary hypertension (CTEPH) is a type of pulmonary hypertension (PH). It can be treated and potentially cured with an operation called pulmonary endarterectomy or PEA. Papworth Hospital is the only hospital in the UK that carries out this specialist surgery.

At Papworth Hospital, we know this can be a distressing time for you and your family but we will do everything possible to help and support you from the moment you are referred to well after your operation. This booklet explains what CTEPH is and what PEA surgery involves. It gives you and your family information about what to expect from your first clinic appointment at Papworth Hospital right through to the surgery and then going home after surgery. There is a lot of useful and important information in this booklet and we don't expect you to read it all in one go but please keep it safe because it will be a useful source of information as you go through your journey. Please remember though that if you have any other questions, then the PEA team at Papworth Hospital will be happy to answer them.

What is the circulatory system?

Learning about the heart, lungs and blood supply to the lungs will hopefully help you understand PH and CTEPH better. The heart pumps blood to your lungs. When blood is in your lungs it is cleaned and given a fresh supply of oxygen. The oxygen-rich blood is then pumped by your heart to the rest of your body. The flow of blood around the lungs is known as the pulmonary circulation. The heart, lungs and pulmonary circulation are all affected in PH.

The heart

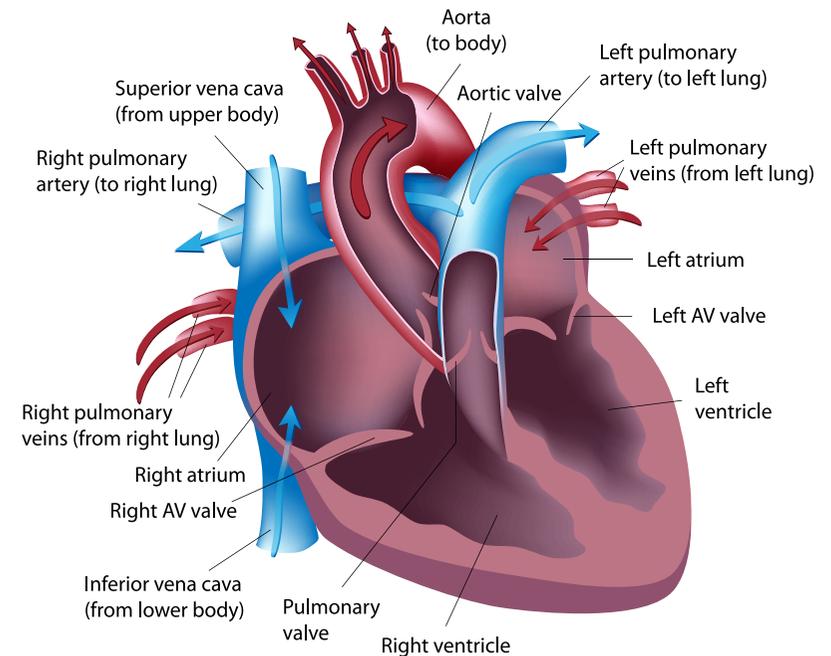


Figure 1. Structure of the heart

The heart is a muscle made up of four parts or chambers. There are two chambers on the right side of the heart and two on the left, working like two separate pumps. The right side of the heart deals with the dirty or impure blood (shown in blue in Figure 1). The upper chamber is called the right atrium and the lower chamber is called the right ventricle. The right ventricle pumps blood to the lungs through a blood vessel called the pulmonary artery. The left side of the heart deals with oxygen-rich blood (shown in red in Figure 1). The upper chamber is called the left atrium and the lower chamber is called the left ventricle. The left ventricle pumps oxygen-rich blood to the rest of the body.

The lungs

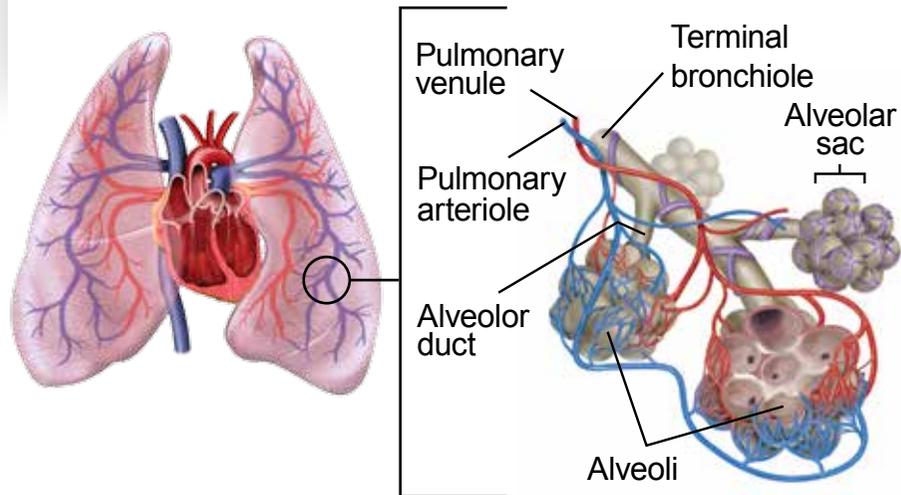


Figure 2. Structure of the lungs

There are two lungs, one on each side of the heart. Each lung is like a sponge made up of sections called lobes. When air is breathed in, it travels down the windpipe or trachea. The trachea splits into a left and right tube leading into each lung. Like the branches of a tree, these tubes keep dividing into smaller and smaller tubes and end in the lungs at tiny air sacs called alveoli, which look like bunches of grapes. There are over 300 million alveoli in healthy lungs, see Figure 2.

The pulmonary circulation

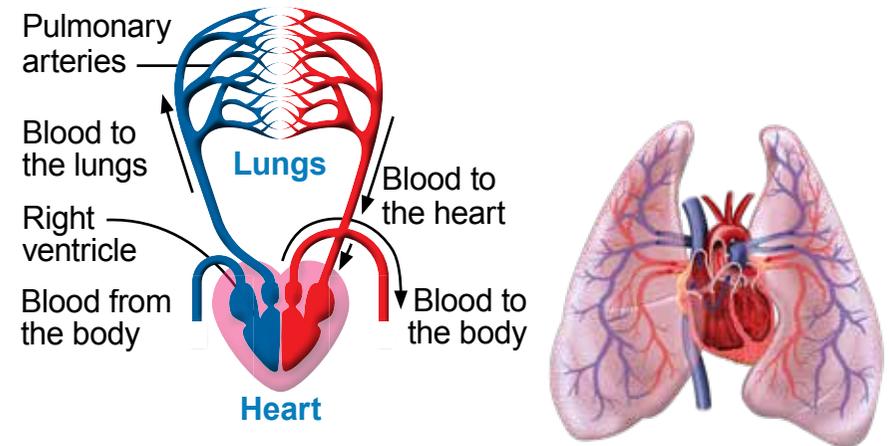


Figure 3. The pulmonary circulation

The pulmonary circulation is made up of blood vessels that carry impure blood to the lungs and oxygen-rich blood from the lungs. Blood that contains waste gas (carbon dioxide) from all over the body is brought to the right side of the heart and is then pumped by the right ventricle through the pulmonary artery. Like the trachea, the pulmonary artery splits into two branches; one going to the right lung and the other to the left lung. These branches continue to divide again and again, becoming very small (see Figure 3). These thin blood vessels surround the alveoli in the lungs and allow the exchange of waste gas (carbon dioxide) and oxygen to take place. The pulmonary veins then take the oxygen-rich blood from the lungs to the left side of the heart and from here it is pumped to the rest of the body.

What is PH?

Put simply, PH is raised blood pressure in the pulmonary artery. There are different types of PH with different causes, but only patients with CTEPH can be potentially helped with PEA surgery.

CTEPH

What causes CTEPH?

CTEPH is caused by blood clots, known as pulmonary emboli (PE), blocking the pulmonary arteries. In most people who get a PE, the body dissolves the clots within 3 months and then they return to normal. In some people this does not happen properly and the clots turn into scar tissue which can lead to CTEPH developing (see Figure 4). The scar tissue narrows or completely blocks the blood vessels, making it more difficult for blood to flow through them normally. This increases the blood pressure in the pulmonary arteries and in turn causes additional strain on the right side of the heart which causes patients to have symptoms.

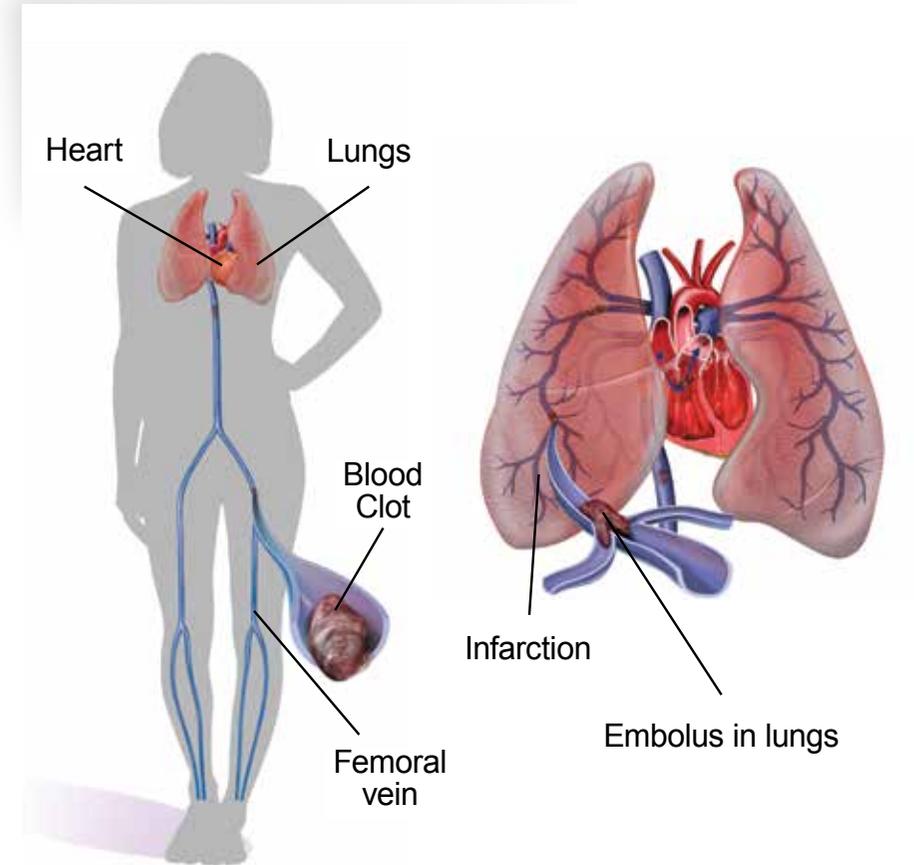


Figure 4. Pulmonary emboli can develop when part of a blood clot from the leg, known as a deep vein thrombosis or DVT, breaks off and travels to the lungs.



Who gets CTEPH?

Some people are more likely to develop CTEPH than others. For example, those:

- Who have had a large blood clot in the lung, also called a PE, or more than one PE
- Who have a blood clot in a deep vein such as the leg, also called deep vein thrombosis (DVT)
- Who have had cancer
- Who have had their spleen removed
- Who have chronic inflammatory diseases, such as inflammatory bowel disease
- With certain blood clotting disorders, such as antiphospholipid syndrome.

What are the symptoms of CTEPH?

If you have CTEPH you will probably feel tired and find it difficult to catch your breath when you work yourself too hard. Over time you may find it difficult to breathe even when you are resting. You may also notice that your legs and tummy are swollen. The early symptoms of CTEPH are the same as those for asthma and heart disease so it can take some time and many tests to get a correct diagnosis.

What happens when CTEPH is not treated?

If CTEPH is not treated, the muscle of the right ventricle in the heart has to work harder and so begins to stretch and get thicker. Over time the heart will become unable to work normally. The high pressure in the lungs can also affect the blood vessels which are not blocked, causing them to get thicker and harder, so that the pressure increases over time.

Pulmonary endarterectomy (PEA)

What is a PEA?

The only cure for CTEPH used to be a heart and lung or lung transplant, but now patients can have PEA surgery to remove the scar tissue which is causing the blockages in the pulmonary arteries.

What are the benefits of PEA?

Most people who have PEA surgery have fewer symptoms, are less breathless, have a better quality of life and live longer after their operation. Some people will have disease in big blood vessels which can be reached by the surgeon and additionally blockages in the smaller blood vessels that cannot be reached by surgery. Therefore, in these cases, the pressure in the pulmonary artery will not completely return to normal, but quality of life should still get better. Unfortunately, a small number of people may not notice any benefit after surgery. If this happens you will be closely monitored by your PH centre.

The operation is discussed in more detail on page 27.

PEA at Papworth Hospital

The first PEA surgery at Papworth Hospital was carried out by Mr John Dunning on 22 January 1997. A small number of people with CTEPH waiting for a heart and lung transplant were thought to be suitable for this new operation and were offered it. These patients were very brave and the current success of the PEA programme owes much to their courage and willingness to undergo the new procedure. PEA is now recognised as the best treatment for many people with CTEPH. It can also be carried out for a few people with a rare tumour in the pulmonary artery.

In 2000 the National Commissioning Group at the Department of Health decided to allow only one hospital in the UK to perform this difficult and high-risk surgery – Papworth Hospital. There are four surgeons doing this operation at Papworth Hospital: Mr David Jenkins (lead surgeon), Mr Steven Tsui, Mr John Dunning and Mr Choo Ng. The number of patients being considered for possible surgery increases every year and at the end of May 2014 1150 patients had undergone PEA surgery at Papworth Hospital.

The following designated PH centres all refer patients to Papworth Hospital for consideration of surgery:

- Golden Jubilee Hospital, Glasgow
- Hammersmith Hospital, London
- Mater Misericordiae Hospital, Dublin
- Papworth Hospital, Cambridge
- Royal Brompton Hospital, London
- Royal Free Hospital, London
- Royal Hallamshire Hospital, Sheffield
- The Freeman Hospital, Newcastle.

What happens when I am referred to Papworth Hospital?

You will already have had a number of tests and scans at your local hospital or PH centre. If these tests show that you may have CTEPH, then the results will be sent to Papworth Hospital along with your medical records. These are reviewed by a multidisciplinary team (MDT) at their weekly meeting. The MDT includes PEA surgeons, PH doctors, radiologists (imaging specialist doctors) and nurses. The surgeon will decide whether PEA surgery may help you.

Several things may happen after the MDT meeting:

- If the surgeon and the team at Papworth Hospital feel that surgery may help you, we will write a letter to your referring doctor and GP explaining this. You will also receive two letters from us; the first to tell you about the outcome of the MDT meeting and the second with an appointment to come to the PEA clinic to meet with a PEA specialist nurse and surgeon to discuss the potential PEA surgery in detail.
- Sometimes we cannot make a final decision with the information we have and so we write to your referring doctor to arrange further tests.
- If the surgeon does not think that surgery will help you, they will write to your referring doctor and GP to explain why. Your PH team will continue to look after you.

Am I suitable for PEA surgery?

We carefully review the distribution of the scar tissue and clots before deciding whether PEA surgery is an option. The surgeon can only remove disease from the larger pulmonary blood vessels, which is called proximal disease. Unfortunately, some

people cannot be helped by surgery because the blockages are only in the tiny vessels deep within the lungs that are impossible to reach, this is known as 'distal disease'. A proportion of patients have a mixture of proximal and distal blockages making the decision to carry out surgery more difficult. Some of these patients can still be offered surgery if we feel they will benefit.

Clinic at Papworth Hospital

If the surgeon thinks you may be suitable for PEA surgery, you will be invited to Papworth Hospital. This visit is an opportunity for you and your family to find out more about the surgery and the management of your condition to help you make the right decision about whether to have the operation or not. It is important to remember that coming to the clinic does not mean that you will definitely be offered surgery nor does it commit you to having surgery if it is offered.

You will be given a lot of information during your visit so it can be difficult to take it all in. We strongly recommend that you bring a family member or a friend to support you. You may also find it useful to write down any questions you have beforehand and bring these along to your visit. We may arrange for you to have an additional scan but you will be informed about this in advance.

Your visit will be made up of several different sessions.

Group session: the group will be made up of one of the PEA specialist nurses and two or three other potential patients and their family members. The specialist nurse will talk you through your treatment journey from waiting list to a year after surgery.

Individual session: after the group session you will get the opportunity to discuss any personal issues with the specialist nurse on a one-to-one basis.

Clinic with the surgeon: the surgeon will talk through the benefits and risks of surgery specific to you. If the surgeon offers you PEA surgery it is your decision whether to accept or not. He will also give you an information booklet that details the risks and benefits of the operation specific to you, along with a copy of the consent form for the operation. You do not have to give an answer on the day and many people want to discuss it with their family and other medical teams caring for them before making a final decision. We can also put you in touch with other patients who have had the surgery so that you can talk to them about their experience. There is a small chance that you may not be offered surgery after seeing the surgeon. There are many reasons why the surgeon may not think you are suitable for surgery but this will be explained to you at the time. You will still be looked after by your PH centre and you may be offered tablet treatment instead.

What are the risks and benefits of PEA?

A PEA is a big operation and the level of risk versus the benefit will vary from person to person depending on where the blockages are, the pressure in the pulmonary artery, general health, age and other medical conditions. The surgeon will talk through your individual risks and benefits in detail at your clinic visit.

At Papworth Hospital over 97% of patients survive the surgery and are well enough to be discharged home within two weeks.

Additional tests before coming in for surgery

If you accept the offer of surgery after seeing the PEA surgeon your name will be added to the provisional waiting list for the operation. Some people will need a coronary angiogram

(see below) before the operation. When all of the tests have been carried out and reviewed by the Papworth Hospital MDT you will be put on the active list “ready for the PEA surgery”.

Coronary angiogram

All people over 50 (or smokers over 40) and others with risk factors for coronary heart disease may need a coronary angiogram. Younger people with angina (chest pain) or those with a strong family history of heart disease may also need one. During a coronary angiogram a small amount of dye is injected into the arteries of your heart to show any narrowing or blockages in the coronary arteries.

If the coronary angiogram shows you have narrow arteries in your heart (Figure 5), the surgeon may carry out coronary artery bypass grafts (CABG) at the same time as doing the PEA. A CABG is where a piece of vein from the leg or an artery from the inside of the chest wall is used to bypass the blocked part of the artery in your heart, allowing the blood to flow past the blockage to the heart muscle. The decision about the need for the CABG is made at the time of the review of your coronary angiogram at Papworth. A PEA specialist nurse will contact you by telephone to inform you about the decision.

Heart

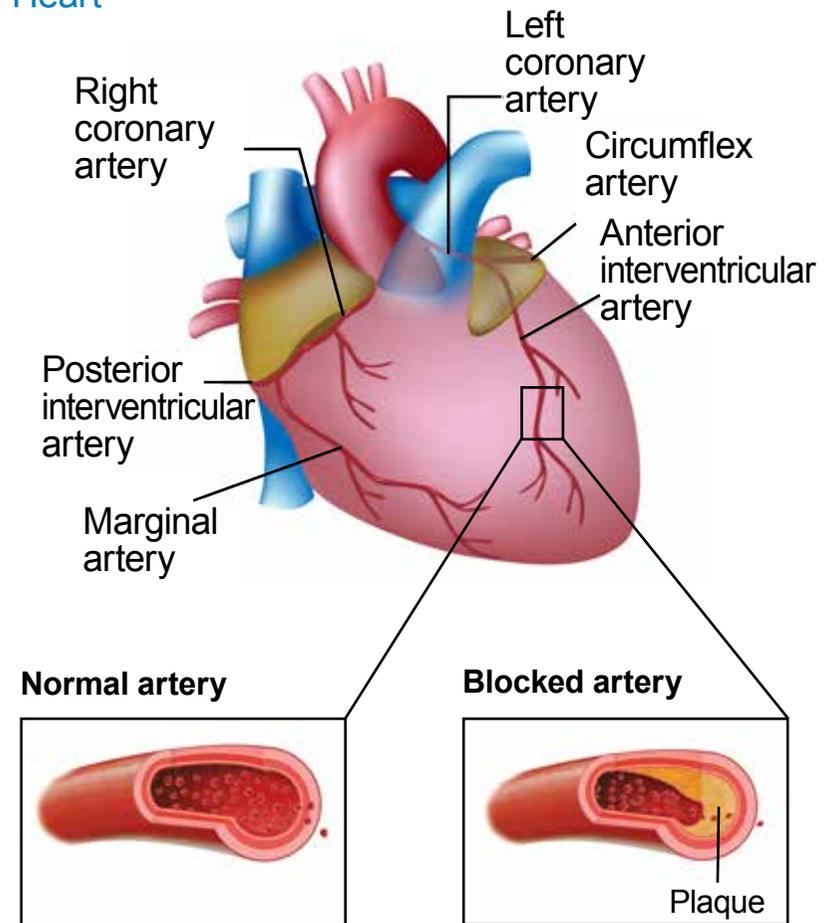


Figure 5: Picture showing narrowing of the coronary (heart) blood vessels that are identified on a coronary angiogram.



Waiting for surgery

How long will I have to wait for surgery?

Once the MDT has reviewed all of your test results, including the coronary angiography, you will be moved to the active waiting list. You will be sent a letter to let you know you've been moved onto this list. The waiting list is managed by the lead surgeon and your waiting time will depend on how many patients are on the list. To make it fair, most patients are operated on in the order that they completed their tests but sometimes people will be moved up the list if they are very sick and need an earlier date for surgery.

While you are waiting for surgery, you will need regular follow-up with your PH centre to monitor your breathlessness and they will contact your surgeon if your PH gets worse. It is important that you contact your PH centre if you feel your breathlessness is getting worse between your regular visits.

Medications

Some patients require medicine before surgery to manage the symptoms of PH. Your PH centre will usually decide if you need

it. It is important that you keep taking your medicine and let the PEA nurse at Papworth Hospital know if you start taking any new medicine after your clinic visit at Papworth Hospital. It is important to avoid getting pregnant, as it can be life-threatening for both you and your baby.

Tips for keeping well and exercising

While you are waiting for surgery, it is important that you keep as fit and active as possible to help you to recover after the surgery. Following these tips will help.

- Remember to take your medication as prescribed, including oxygen.
- Stay as active as possible but make sure that you also rest.
- Exercise can include lots of different things, like walking and housework.
- Always talk to your medical team before starting any exercise programme.
- Listen to your body and never push yourself too far. You should have enough breath to talk in complete sentences while you are exercising and not feel dizzy or light headed.
- Don't exercise straight after a meal.
- Don't hold your breath during exercise.
- Avoid any heavy lifting and bending over.
- Follow a healthy diet and avoid weight gain.
- Don't smoke. If you smoke, the most important thing you can do for your heart and lungs is to stop.
- Stop or reduce the amount of alcohol you drink. Remember that alcohol affects the way warfarin works.

- Try to find ways to reduce stress.
- Discuss travelling by plane with your PH team. Being in places higher than 8,000 feet (2,438 metres) can worsen the symptoms of PH.
- Join a support network to get help and support from people going through the same thing as you.

Am I doing too much?

It is important that you don't work yourself too hard. If you feel anything from the list below you need to stop the activity and talk to your medical team:

- Worsening breathlessness
- Light-headedness or dizziness
- Heart flutters (palpitations) and feeling faint
- Extreme weakness or inability to carry out normal activity
- Weight gain (2 pounds in one day or 5 pounds in one week)
- Swelling in your ankles, legs or stomach that has become worse.

If you are struggling with daily activities or you have any social concerns, ask your GP surgery or PH centre for an additional review of your needs. He or she can give you ideas to help make your daily living easier.

Getting a date for surgery

One of the specialist nurses from Papworth Hospital will contact you with a date for surgery. This is usually 2 to 3 weeks before we plan to operate.

Preparing for surgery

Blood thinning (anti-coagulation) medications

It is important that any blood thinning (anti-coagulation) medicines you are taking are stopped before your surgery. It is also important that you follow the instructions below when stopping.

Warfarin

- Take your last dose of warfarin 5 days before the date of your surgery.
- Replace warfarin with subcutaneous (sc) heparin (enoxaparin, tinzaparin, deltaparin or fondaparinux) for the next 3 days. Your GP will receive a letter asking to prescribe the injections for you.
- Do not take any blood thinning medicine on the day you go into hospital.
- If your blood thinning medication is sc heparin, continue with the injections you are on until the day before admission.

If you are on the newer alternative anti-coagulation tablets, please let us know so that we can advise appropriately.

Aspirin and clopidrogel

- Stop one week before the day of surgery.

Other medication

- Metformin: Stop 2 days before the day of surgery.
- Do not stop taking any other medications unless you are specifically told to do so.
- The PEA specialist nurse who calls you to offer a date for surgery will go through all your medication with you again.

Other preparations

Your GP will be asked to test you for a hospital bug called MRSA.

Information on when to arrive at the hospital and who to contact to find out about local accommodation for anyone coming with you, will be sent to you by Papworth Hospital booking office.

Please let the PEA nurse at Papworth Hospital know if your home situation has changed, especially if you live alone, have no-one to be with you when you leave hospital or your home will not be fit for you to be return to.

If you cannot arrange your own transport to Papworth Hospital, please contact your GP. If they cannot help, please call our co-ordinator on 01480 364866 or 364952 and leave a message for someone to call you back. Please note that not all patients can get free hospital transport.

Packing your bag

When you come into hospital for your operation please bring:

- Your PEA patient guide
- Pyjamas or nightdresses (2 or 3)
- Dressing gown
- Wash-bag with toothbrush, toothpaste, soap, shampoo etc. You do not need to bring towels or flannels as these will be provided
- Handkerchiefs or tissues
- Two or three sets of comfortable day clothes. Don't bring anything too tight. A track suit is ideal, if you have one. Remember to take into consideration the weather!

- Underwear (ladies, please bring two or three old bras)
- Flat, outdoor, comfortable shoes, such as trainers
- Socks
- Slippers
- Books or magazines
- Mobile phone and charger if you have one
- Money for the shop
- Anything to help you pass the time. This could include electronic devices, books, notepaper, writing paper and envelopes
- Squash, sweets, fruit or other treats
- Any medicines you are taking. If you are taking Ambrisentan, Bosentan, Sildenafil, Tadalafil, Epoprotenol, Treprostinil or Iloprost, please bring enough for your whole stay. Don't forget to bring accessories if you use a nebuliser or pump.

If someone is coming with you and planning to stay locally, they will also need a supply of suitable clothing and any medicines they are taking. They should also bring the telephone number of their own GP in case they need to be contacted.

Admission for surgery

Most people are admitted to hospital the day before surgery to one of three surgical wards: Mallard Ward, Higginson Ward or Varrier Jones Ward. The surgical wards are at the back of the hospital site, near theatre and Critical Care. Sometimes patients are admitted to the Chest Medical Unit: Baron Ward or Duchess Ward if there are no surgical beds available (see map on page 49).

You will usually be asked to arrive between 10.30 am and 11.00 am to give us time to carry out some final tests. When you arrive at the hospital, please go to the Main Reception (see map on page 49) where you will be checked in, given your medical notes and told which ward you will be admitted to. During the day you may have a 6-minute walk test, chest x-ray, an electrocardiogram (ECG – to check your heart) and blood tests. Some patients may be approached to voluntarily to give a blood sample for research purposes.

When you arrive on the ward, you will be shown around by the nurses. They will tell you about the ward routine and introduce you to some of the other patients. The nurses will also fill in your personal admission forms. This will include asking for information that will help when you are ready to go home (discharge planning).

Who will I see?

On the day you come to hospital you will be seen by a number of people.

- Junior doctor or advanced nurse practitioner: he or she will examine you and ask you about your health and medicines you are taking and ask you to sign the consent form allowing the surgery.
- Consultant surgeon: your surgeon will talk to you again about the risks and benefits of the operation and answer any questions you may have. Your consultant surgeon may not be the same surgeon you met during your clinic visit.
- Anaesthetist: your anaesthetist will also ask you about your health and explain the medicines and equipment which will be used while you are in the operating theatre and Critical Care.

- Ward nurses: will complete your admission paperwork and show you around the ward. They will also discuss the discharge arrangements at this time. If you have any concerns about going home after surgery such as social issues, struggling to manage your daily activities, having carer(s) in place, living alone or requiring any additional equipment, please discuss this with the nurse who can plan ahead of your requirements for discharge.
- PEA nurse: you may not see the PEA nurse on the day of your admission but you can ask the ward nurse to contact them if you or your family have any questions.

Eating and drinking

You will be able to eat and drink normally on the day you are admitted to hospital, but you will be asked to stop eating and drinking (nil by mouth or NBM) from midnight until your operation the next day. You can take your normal medicines unless you are told otherwise. It is important not to eat or drink after this time as it may interfere with the anaesthetic.

Shaving

The areas where you may have a cut (incision) or drip, such as the chest, forearms and groin, may be shaved before surgery to help reduce the risk of wound infection.

Washing

To reduce the risk of wound infection on the night before and the morning of your surgery you will be asked to wash your whole body, including your hair, with a special antibacterial liquid which the ward staff will give you.

Cancellation of surgery

Very rarely we may need to cancel your surgery because another patient needs an emergency operation or because there are no Critical Care beds available to look after you following your operation. Unfortunately, you will usually be sent home that day and we will have to give you the next available operating slot, usually within a month.

The day of surgery

Preparing for surgery

The theatre team will usually collect you from the ward around 8.00 am. Your family are welcome to stay with you the whole morning until you are taken to the doors of the theatre complex.

In the theatre reception you will be moved onto a trolley and taken into the anaesthetic room, where sticky pads will be put on your chest to connect you to a heart monitor. An oxygen monitor (like a clothes peg) will also be put on your finger. A small drip will be put in each wrist. Medicine will be given through these and you will quickly fall asleep.

During surgery

When you are asleep the anaesthetist will use many different pieces of equipment to carefully monitor you.

- Endotracheal or ET tube: a tube will be put in your mouth and attached to a machine called a ventilator which will breathe for you while you are asleep.
- Central venous pressure (CVP) line or neckline: a drip will be put into a vein in the side of your neck to give you fluid and medicines. This is usually taken out 3 to 5 days after surgery.
- Pulmonary artery or Swan-Ganz catheter: a small tube measures the pressure in your heart and pulmonary arteries. It is put into a vein in the side of your neck and is usually removed the day after surgery.
- Arterial line: a small tube will be put into an artery in your wrist and also in your groin to monitor your blood pressure and oxygen levels. These are removed before you leave Critical Care.

- Urinary catheter: a small tube will be put into your bladder to drain your urine. This usually stays in for around 4 days.
- Oxygen monitor: a small pad will be placed on your forehead to monitor the oxygen levels in your brain. This is removed when you leave the operating theatre.

The operation



Figure 6.
Sternotomy

Before the surgery starts, the skin of your chest and groins will be cleaned with a special pink antiseptic liquid to stop infection.

The surgeon will carefully cut down the length of your breastbone (sternum) to open the chest. When the chest is open the surgeon will attach you to a heart-lung (bypass) machine that will take over the work of your heart and lungs during the operation. Your body temperature

will slowly be brought down to around 20°C. This is a little like putting your body into hibernation and reduces your body's need for oxygen.

The surgeon will then open your pulmonary arteries, one lung at a time, and the inner lining with the attached scar tissue will be carefully peeled away. During this time, the blood flow around your body is reduced or stopped completely for periods of time to give the surgeon a better view. When the pulmonary arteries have been cleared of the scar tissue and closed, your temperature will slowly be brought back up to normal body temperature, around 37°C, your heart will start beating again and you will be taken off the heart-lung machine.

The surgeons will put in two or three tubes at the top of your stomach just below your ribs. These tubes (chest drains) drain away any fluid and usually stay in for 2 to 3 days. They will also put in thin metal wires next to the chest drains (see Figure 6). These are used to control your heart beat after surgery. They are usually removed around 5 days after surgery.

Finally, the surgeons will close your chest using wire to hold your breastbone firmly together. These wires will stay in for the rest of your life. The skin will be closed with a stitch that dissolves as your wound heals. You will then be moved back onto a bed and moved to Critical Care.

Most people are in theatre for 8 to 10 hours but it can take longer.

Critical Care

Everyone having a PEA will be kept asleep for the first six hours and maybe for a few days after the operation. The ventilator will carry on breathing for you and the Critical Care staff will carefully watch you with the help of bedside monitors and other equipment (Figure 7).



Figure 7
Critical Care Unit

A Critical Care nurse will speak to your family, usually within an hour of you arriving in Critical Care. If the family member is not in the hospital, they will give them a call. Your family can visit you as soon as they want and they will be given the number of the telephone closest to your bed so that they can call at any time. We ask that only

one family member or friend calls so that the nurses can spend more time caring for you. Up to two people can visit you at any time and for the first few days we ask that these are members of your family or very close friends.

If you are on treatment for your PH this may be stopped soon after surgery if the surgeon is happy with the amount of material that was cleared from the pulmonary artery and if the pressure in the pulmonary arteries has returned to normal or near normal. Some people may be asked to carry on taking these drugs until they have had their follow-up investigations.

The days after surgery

Everyone having this operation recovers at a different rate and it is impossible to say how quickly you will recover from the surgery. On the morning after your surgery, a team (including doctors, nurses and physiotherapists) will check how your heart, lungs and kidneys are coping and how much fluid is draining from the chest drains.

There are three main paths to recovery. Paths A and B are the most common but patients usually switch between paths during their recovery. Whichever combination of paths is taken, the benefits are equally good in the long term.

Recovery path A

About 7 out of 10 patients follow pathway A, which involves 1 to 3 days in Critical Care or 4 to 5 days if you need continuous positive airway pressure (CPAP) help with your breathing.

If you are warm and stable and need only a small amount of oxygen, the team may decide to stop the drugs keeping you asleep and allow you to wake up. You may wake within minutes



Figure 8. Continuous positive airway pressure (CPAP) mask

but many people take hours to wake up enough to come off the ventilator and breathe by themselves. When you are breathing comfortably the tube in your mouth is taken out and an oxygen mask will be put on. While the tube is in you will not be able to talk, drink or eat. Most people do not remember much about this time.

You are likely to be very tired when you wake up and you will need careful monitoring. You may be helped to sit in a chair by your bed for short periods the day after you wake up and will be able to eat and drink. Your family can visit you at any time.

After the scar tissue has been removed from the pulmonary arteries, fluid in the blood can leak through the blood vessel wall into the air sacs (alveoli) in the lungs; this is known as re-perfusion injury. This makes breathing difficult and you may need more oxygen which is given through a CPAP mask (see Figure 8). The mask is held in place by a special cap or straps and fits snugly over the mouth and the nose. You may need CPAP occasionally for several days.

Sometimes people are woken the day after surgery but are agitated or too sleepy to breathe without the ventilator. If this happens you may be put back to sleep for a few days and become a pathway B patient.

Recovery path B

About 2 in 10 people will spend up to a week in Critical Care.

It is common for some people having a PEA to be kept asleep for several days, while the heart and lungs recover from the surgery. The team will regularly assess you during this time to check your progress. When you are ready you will be woken as in path A.

If you are asleep for more than 24 hours, you will be fed a special high calorie liquid food through a small (nasogastric or NG) tube which goes up your nose and down into your stomach. When you are awake and breathing on your own you will be able to eat and drink normally and the tube will be taken out. Within a day or so of waking, you will be helped to sit out of bed. Your family can visit you at any time.

Recovery path C

About 1 in 10 people will be in Critical Care for longer than a week, especially if they are frail, suffer from other illnesses or have very severe PH.

When the ventilator is breathing for you the muscles in your chest wall are not working and so slowly become weaker. If you take more than 10 to 14 days to become stable enough to wake up, your chest wall muscles may not be strong enough for you to breathe by yourself. The only way to build up these muscles is to wake you up and get you breathing but most people don't like to be awake with a tube in their mouth. If this happens then we will put a small tube into your windpipe, just under your Adam's apple while you are still asleep. We can then connect the ventilator to this tube which is called a tracheostomy or 'trachy', take the tube out of your mouth and wake you up. When you are awake we can change the way the ventilator works so that you gradually do more breathing by yourself and the ventilator does less.

Coming off the ventilator is different for each person and can take anything from a few days to a few weeks. You will be fed through a small tube in your nose (NG tube) until you are able to swallow. When the tracheostomy first goes in, you will not be able to talk as the tube goes through your vocal cords, but you will be able to write messages, use picture message boards and mouth words for others to lip-read. When you are able to cough to keep your airway clear and are off the ventilator, the tracheostomy tube is taken out. The hole is covered with a plaster and it heals itself in 2 to 3 weeks.

During your time in Critical Care your muscles will have become weak and you may need to be lifted in and out of bed. You will need a lot of physiotherapy to help build up your muscles and get you moving again. Pathway C does not mean that surgery has not been successful; it just means that you and your lungs have taken longer to heal. Patients requiring extra-corporeal membrane oxygenation machine (ECMO) support of the heart or lungs fall into this pathway.

Complications

As with all major operations there is a risk of complications. The most common are listed below.

- **Bleeding:** this usually happens in the first 24 hours after surgery and you may need to have blood products such as platelets. Occasionally you may need to return to theatre for the wound to be explored.
- **Cardiac tamponade:** the heart sits in a sac called the pericardium. There is a small amount of fluid in this sac that stops the heart rubbing against it. Sometimes too much fluid collects in the sac and the heart is squeezed. This is called a cardiac tamponade. In many cases the fluid is re-absorbed over time but in some cases people will need a small drain put in or return to theatre to drain the fluid.

- **Renal (kidney) injury:** sometimes after major surgery the kidneys take a little time to recover. If this happens the kidneys will be supported by a type of dialysis called haemofiltration until they recover, usually only a few days.
- **Re-perfusion injury:** Some patients can develop “leaky lungs” or reperfusion injury. Most people can be treated with CPAP but in a few severe cases may need to be supported only by an ECMO.
- **Residual PH:** sometimes the pulmonary artery pressure remains higher than expected after surgery and the heart struggles to cope. ECMO may be used to support the heart giving it time to recover. ECMO is like the bypass system used during your operation. Blood is pumped out of the body, through the ECMO machine picking up oxygen as it goes. The ECMO machine then pumps the oxygen-rich blood around the body. This takes the strain off the heart. ECMO can also be used to rest the lungs when they are filled with fluid following re-perfusion injury and give them time to heal. Patients needing ECMO are usually supported on the machine from around 2 days up to 2 weeks.
- **Mental effects:** when people wake up after surgery they may be confused, irritable, grumpy or aggressive. Other people see or hear things that are not happening (hallucinations), are suspicious and distrustful of their family, friends and staff (paranoia) or have problems with muscle weakness, twitching or co-ordination. There is nothing you can do to stop this from happening so please try not to worry about it. It is due to the operation. You will be back to normal before you go home. Some people remember the times of confusion or hallucinations, others do not. It can also be a very difficult time for friends and family who must understand that the patient is not aware of how their behaviour is affecting others. Please do not take anything they say personally!

Surgical Ward

When you are well enough you will be moved from Critical Care to the surgical ward. You will still have:

- An oxygen mask
- A drip in your neck
- A urinary catheter
- Possibly chest drains.

The nurses will regularly check your blood pressure, heart rate, oxygen levels, fluid intake and weight. They will also help you to wash and dress at first. When all your lines and tubes have been taken out you will be encouraged to wear day clothes and get up and about.

As you recover on the ward you may notice some of the following:

- **Irregular heart beat:** after the operation you may feel your heart beating faster, irregularly (atrial fibrillation) or missing a beat. This is very common and is a reaction to the heart being handled. You may be attached to a heart monitor for a short time and/or started on a medicine to calm the heart. You will need to take the medicine for 6 to 12 weeks.
- **Pins and needles:** some patients get pins and needles in their arms and hands or numb patches on their thighs. This is caused by pressure on your nerves while you are lying on the operating table and should gradually get better.
- **Sickness (nausea) and appetite:** if you feel sick after surgery please let your nurse know. They will give you medicine to control it. Many people have a poor appetite or complain of a funny taste in the mouth after surgery. This will gradually get better but it is important that you eat as you need the energy to heal. Generally, eating little and often is better.

- **Constipation:** it is common to be constipated (not able to go to the toilet) after surgery. Your tummy will gradually return to normal as you eat more and move around. Many people will need laxatives in the first few days after surgery.
- **Pain:** many PEA patients tell us pain is not a big problem. You will be given regular painkillers after surgery whether you ask for them or not. It is really important that you get up and about, cough and breathe deeply as quickly as possible and you will not be able to do this if your pain is not well controlled. Please tell someone if you have pain and we will change or increase your medication.
- **Headache:** can be common after surgery as you are usually a little dehydrated. However, if a headache doesn't go away or is unusually severe you will need a scan of your brain to make sure you have not had a small bleed while your blood has been thinned.
- **Unable to cough and deep breathe:** it is very important that you cough and breathe deeply after surgery to help expand your lungs and cough up any phlegm. If you are not able to do this comfortably because you have pain, please speak to the nurses.
- **Need for oxygen:** most people will still need some oxygen for the first week or two after surgery but you will be weaned off as your oxygen levels improve and when you are walking around. You may be sent home with oxygen and weaned off it more slowly. It can take 2 to 3 working days to arrange oxygen in your home and a family member or friend will need to be at home to let the oxygen company into your house to set up the equipment. You will not be able to go home until the oxygen has been delivered and set up.
- **Tiredness:** it is normal to get tired very easily after surgery

and it may take a few days or even a few weeks to get your energy back.

- **Emotions:** feeling low, emotional, weepy or grumpy is a natural part of recovery and will gradually get better. Mood swings are also common.
- **Wound issues:** your wound will be regularly assessed and re-dressed if necessary. A swab will be taken from your wound if it looks infected and an antibiotic may be started if necessary. When it is clean and dry it will be left without a dressing.
- **Warfarin and internationalised normalised ratio (INR):** enoxaparin (anticoagulation injections) is restarted the day after surgery and your usual pre-surgery anticoagulation, e.g. warfarin, will be restarted when you are transferred to the ward, if there are no bleeding issues. The INR is checked regularly and you will be on both enoxaparin and warfarin until your INR is above 2.

Female patients will need to wear a bra from day 2 of the surgery at least for three months to avoid breasts pulling apart and putting pressure on the scar.

Preventing hospital acquired DVT

People with CTEPH are more likely to develop blood clots and lying or sitting for a long time also increases your risk of blood clots. Warfarin and enoxaparin are given to thin the blood and help stop clots and you may also be given special stockings (TED stockings) to wear. To help stop clots:

- Try to get up and walk about as soon as possible and as much as possible. The physiotherapist and nurses will support you in the early days after surgery. The physiotherapists will also give you leg exercises to do. Try to do them regularly.
- Try not to sit or lie with your legs crossed.

Who will care for me on the ward?

While you are at Papworth Hospital a team of people will look after you. Some of these are listed below.

- **Nurses and health care assistants (HCA):** the nurses and HCA's will care for you before and after surgery and will help you with washing, going to the toilet and pain control. They work shifts so you usually have two to three nurses per day who are responsible for caring for you and your family.
- **Doctors:** a team of doctors will visit you at least once a day to make sure that you are recovering well from your operation. They will decide what treatment you need to get you better.
- **PH consultants:** the PH consultants review you once a week to monitor your progress and medications. They will also plan your further follow-ups at Papworth Hospital.
- **Physiotherapists:** physiotherapists will help to keep your chest clear and as you get stronger they will help you to get up and about.

- **Housekeeper:** the housekeepers will serve you your food and drink during your stay.
- **Porters:** the porters will move you around the hospital for things like x-rays.
- **Dietitian:** the dietitian will make sure you have enough calories to heal and get better. They will also give you supplements or snacks if you have a poor appetite.
- **Pharmacist:** you will be given different medicines to help your heart and lungs while they recover from surgery. The pharmacist will monitor these. Each person will need to take the medicines for different amounts of time. When you are ready to go home the pharmacist will go through the medicines you are taking with you.
- **Technicians:** the technicians will take your blood samples, do ECGs and carry out walk tests.
- **Occupational therapist:** the occupational therapists can help you with aids and equipment to make things easier for you when you first get home.
- **Social worker or discharge planning team:** if you live alone or have special needs, the social worker or discharge team may help us get you home with a care package or transferred to a local hospital until you are recovered.
- **PEA nurse:** a PEA nurse will visit you most days while you are at Papworth Hospital to monitor your progress after surgery and provide support to you and your family. When you are ready to go home, they will discuss the information required for you and your family on how to manage at home.

Physiotherapy following your PEA

You may be very weak after your PEA and we understand that you may feel nervous about exercise. A physiotherapist will see you the day after your PEA (before you even wake up) to start a gentle exercise programme. All exercises will take into account any other medical conditions and limitations you may have.

- **Gentle arm and leg exercises:** at first you will be encouraged to do some gentle arm and leg exercises to prevent stiffness and help improve circulation.
- **Deep breathing and coughing:** some people become chesty the first few days following surgery and it is important to clear any sputum from your chest before it becomes infected. The physiotherapist will teach you simple deep breathing exercises, coughing and huffing to help clear your lungs.
- **Walking short distances:** over the following days the physiotherapists will help you to walk short distances around the ward and help build up your confidence. It is very common to feel a little unsteady or lightheaded and dizzy when you first start walking around. This will get better.
- **Walking a lap of the ward:** you will be encouraged to gradually increase the distance you walk until you are able to walk a lap of the ward without help.
- **Stair assessment:** when you are more confident and walking independently you will be ready to go up and down stairs with the physiotherapist.
- **Monitoring of oxygen:** the physiotherapists also monitor your oxygen levels at rest and while you are exercising. They will tell the medical team if you need oxygen at home.

Before you leave hospital the physiotherapists will advise you about continuing exercise at home. We do not recommend any exercise regime or rehabilitation programme until you have had your inpatient review 3 to 6 months after surgery.

Ward rounds

- **Surgical:** the surgical doctors will see you each morning to check on your progress, book any tests and plan for your discharge.
- **Consultant surgeon:** you may not see your consultant surgeon every day but their team will update them on your progress. If you or your family want to see your consultant surgeon then the ward staff can arrange an appointment.
- **PH consultant:** your follow-up after the PEA is usually with the PH (medical) team at Papworth Hospital. One of the PH consultants will see you on a weekly ward round while you are recovering. They will monitor your heart function, fluid intake and on-going need for targeted PH therapy.

Going home after surgery

The majority of people go straight home after surgery. You will need someone to stay with you for the first week or two, or you may choose to go and stay with your friend or family member until you are well enough to go home. If you plan to do this, please check that their GP is happy to accept you as a temporary patient.

If you live alone and have no-one to help or your home is not safe for you to be discharged to, we may arrange for you to be transferred to a convalescent home or to your PH centre or local hospital. If this is not possible a care package will be arranged

to get you home. This usually means that a carer will come to your home a number of times every day to help with things like showering and cooking.

The team looking after you will decide when and where you will go having discussed things with you first.

You may be referred to an occupational therapist if you need temporary equipment such as chair raisers, raised toilet seats or shower stools to help you when you first get home. A social worker will be able to help you with other things like benefits.

You will be discharged when:

- Your medical team thinks you are well enough
- Your physiotherapist thinks you are walking safely on your own or with a frame or stick. If you have stairs at home you will also have to pass the stair assessment. The physiotherapist will also talk to you about exercising at home
- The doctors are satisfied your anticoagulation is controlled and you have received the anticoagulation letter for your GP and anticoagulation clinic
- You have someone to be with you and a safe place to go, or you have a care package in place
- When the PEA nurse has discussed the discharge information with you.

Most people will be able to travel home with their family in a car. If no-one can collect you, hospital transport can be arranged. Unfortunately, your family will have to make their own way home as Papworth Hospital cannot arrange transport for them.

If you have been using oxygen before surgery you will still be able to go home with your family, but please bring your own portable oxygen cylinders for the journey. If you have been newly prescribed oxygen you may need hospital transport.

For those on warfarin, the ward staff will arrange an INR test at your GP surgery or INR clinic 2 to 3 days after discharge. It takes time for INR to stabilise after surgery and until then you will need frequent blood tests at your GP or INR clinic. You may also need a blood test for kidney function if you are on water tablets (diuretics) and you are fluid restricted.

If your wound needs redressing the ward staff will arrange for the district nurse or practice nurse to do this when you get home. The pharmacist will discuss the drugs you will be taking at home.

The average hospital stay following a PEA is 10 to 14 days but some people will go home a little earlier than this, while others will be in hospital much longer.

The PEA nurse will call you 7 to 10 days after you go home to check on your progress.

Useful contacts

Papworth Hospital numbers

Hospital switchboard	01480 830541
PVDU co-ordinator for appointments	01480 364708
PEA nurses	01480 364952 (answerphone) 01480 830541, bleep 785 (urgent calls)
Main reception	01480 364625
Duchess Ward	01480 364512
Baron Ward	01480 364227
Mallard Ward	01480 364375 01480 364407
Higginson Ward	01480 364420
VARRIER Jones Ward	01480 364648
Critical Care reception	01480 364400
Bay A	01480 364347
Bay B	01480 364401
Bay C	01480 364404
Bay D	01480 364183
Bay E	01480 366180
Thoracic Day Ward	01480 364194
Thoracic outpatients	01480 364281
Patient advice and liaison service (PALS)	01480 364896
Supportive and palliative care	01480 364154 01480 830541, bleep 680
Chaplain	01480 364121 01480 830541, bleep 722

Social worker	01480 364279
Occupational therapist	01480 834448
Pharmacy help-line	01480 364739
Physiotherapy	01480 834215

Other contacts

The Pulmonary Hypertension Association (PHA) UK is a support group for both patients and their relatives.

Helpline: 01709 761450

Web: www.phassociation.uk.com

AntiCoagulation Europe (ACE)

is a support group for people on warfarin

Telephone number: 02082 896 875

Web: www.anticoagulationeurope.org

Information for visitors

Visiting times

- **Critical Care:** your family members can visit at any time. They need to use the red phone by the door to be let in.
- **Surgical Ward:** visiting time is between 2.15 pm and 8.00 pm.

Accommodation

There is only a small number of places to stay in the village closest to the hospital, so we ask that where possible, those with their own transport should stay in one of the surrounding villages just a few minutes away. This allows others who are unable to drive to stay within walking distance of the hospital.

If accommodation is required in the village, please contact the accommodation office through the switchboard **01480 830541**. Unfortunately, there is no accommodation available at Papworth Hospital.

Visitors are asked to make sure that they bring enough supplies of their own medicines with them, especially if they do not live locally. There is no facility at the hospital to dispense any medicines to visitors. It is helpful to bring with you the telephone number and fax number of your own GP.

Papworth Everard village

There is a small general store, Post Office, parish church, library and restaurant in Papworth Everard village. Other services can be found in the nearby towns, which are about 15 minutes' drive away. There is a limited bus service for those who do not drive.

Please also see the separate booklet called 'Visitor handbook - General Information'.

Travelling to Papworth Hospital

By air

Stansted Airport is about 40 minutes' drive from Papworth Hospital and has regular scheduled flights to all parts of the UK, Europe and elsewhere.

By rail

Cambridge railway station is about 20 minutes' drive from Papworth Hospital. Huntingdon and St. Neots stations are both about 15 minutes' drive away. All are main line stations with regular service connections to London.

By bus

For details of bus and coach services please contact Whippet Coaches on 01480 463792 or visit www.go-whippet.co.uk or Traveline on 08712 002233.

By road

Papworth Hospital is in the village of Papworth Everard, which is close to the M11 motorway. If you are using Sat Nav please use the postcode CB23 3RE.

From the M11 northbound:

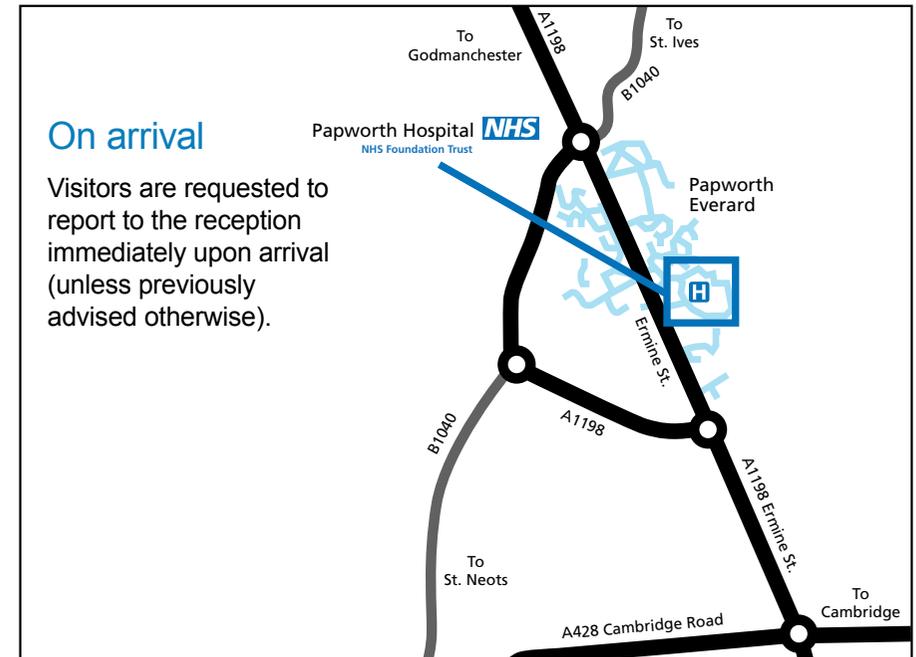
Leave the motorway at junction 13 and join the A1303 westwards, continue along this road and join the A428. Once on the A428 continue to the roundabout at the junction with the A1198 and take the third exit onto the A1198, signposted Papworth Everard. Continue along the A1198 and take the second exit at the next roundabout into the village of Papworth Everard. Shortly after entering the village the entrance to Papworth Hospital is on the right-hand side.

From the A1/A14 southbound:

Leave the A14 at the intersection with the A1198 at Godmanchester. Take the third exit from the roundabout onto the A1198 (signposted Royston). Continue along this road for approximately 4.5 miles into the village of Papworth Everard. Continue through the village for 1 mile. The entrance to Papworth Hospital is on the left-hand side.

From the A14 westwards:

Follow the A14 around Cambridge and follow the signs for Bedford. After passing the slip-road to the M11/A14 (westbound) join the A428 (signposted Bedford). Continue on the A428 for approximately 7.5 miles and at the roundabout at the junction with the A1198 take the third exit onto the A1198 (signposted Papworth Everard). Continue along the A1198 and take the second exit at the next roundabout into the village of Papworth Everard. Shortly after entering the village the entrance to Papworth Hospital is on the right-hand side.

**On arrival**

Visitors are requested to report to the reception immediately upon arrival (unless previously advised otherwise).

Further information

Further information about CTEPH and Papworth Hospital can be found at <http://www.papworthhospital.nhs.uk/docs/leaflets/> including the following:

PEA information (number PI-26)

PEA consent form (number PI-131)

CTEPH frequently asked questions (FAQs) (number PI-87)

Accommodation around Papworth Hospital

Papworth Hospital

NHS Foundation Trust

Bernhard Baron Building 1 2 3

- CF Unit (1)
- Chest Medical Unit (Baron, Princess, Duchess Wards) (3)
- Respiratory Physiology (1)
- Main Reception (2)
- Patient Travel & Car Parking Office (2)
- Thoracic Day Ward (2)
- Thoracic Oncology (1)
- Thoracic Outpatients (1)

Cardiothoracic BiIncubator (CTBI) 9

- Research & Development

Chapel & Multi-Faith Rest Centre 8

Christopher Parish Building 11

- Cardiac Day Ward
- Critical Care
- Echocardiography (ECHO) Department
- Hemingford Ward (HDU)
- Higginson Ward
- Hugh Fleming Ward
- Mallard Ward
- Nuclear Medicine
- X-ray Department

Corporate Affairs & Chairman's Office 20

Diagnostic Centre 18

Ellen Kemp Building 7

- Chief Executive
- Patient Advice & Liaison Service (PALS)
- Restaurant
- Switchboard

Heritage Centre 16

Lakeside Crescent 10

- Cardiac Consultants Offices

Lakeside Lodge 19

- Booking Office

Lecture Theatre 13

Mortuary & Chapel of Rest 15

Papworth Direct 12

Papworth Hospital Charity 21

The Papworth Shop 22

Patient Hostel 17

Pharmacy Department 4

PPCI Entrance 18

Respiratory Support & Sleep Centre (RSSC) 5

Respiratory Support & Sleep Centre (RSSC) 6

- Outpatients

Sims Woodhead Building 14

- Pathology Department (Laboratory)

UKLS

UKLS CT Appointments 18

UKLS Clinic Parking 23

UKLS CT Appointment Parking 24

UKLS Clinic 25

Transplant Outpatients (TCCU) 13

Varrier Jones 12

- Cardiac Outpatients & Pre-Admissions Clinic
- Physiotherapy Gym
- Varrier Jones Reception
- Varrier Jones Ward

P Patient & visitor pay and display parking

Papworth Hospital NHS Foundation Trust
Papworth Everard, Cambridge, CB23 3RE

Tel: 01480 364168

Fax: 01480 364161

www.papworthhospital.nhs.uk

A member of Cambridge University Health Partners



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Tel: 01480 364657

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