Great Ormond Street Hospital for Children NHS Trust

Pulmonary Arterial Hypertension

Information for families

This booklet will tell you about Pulmonary Arterial Hypertension (PAH) and what to expect when your child comes in to Great Ormond Street Hospital for treatment.

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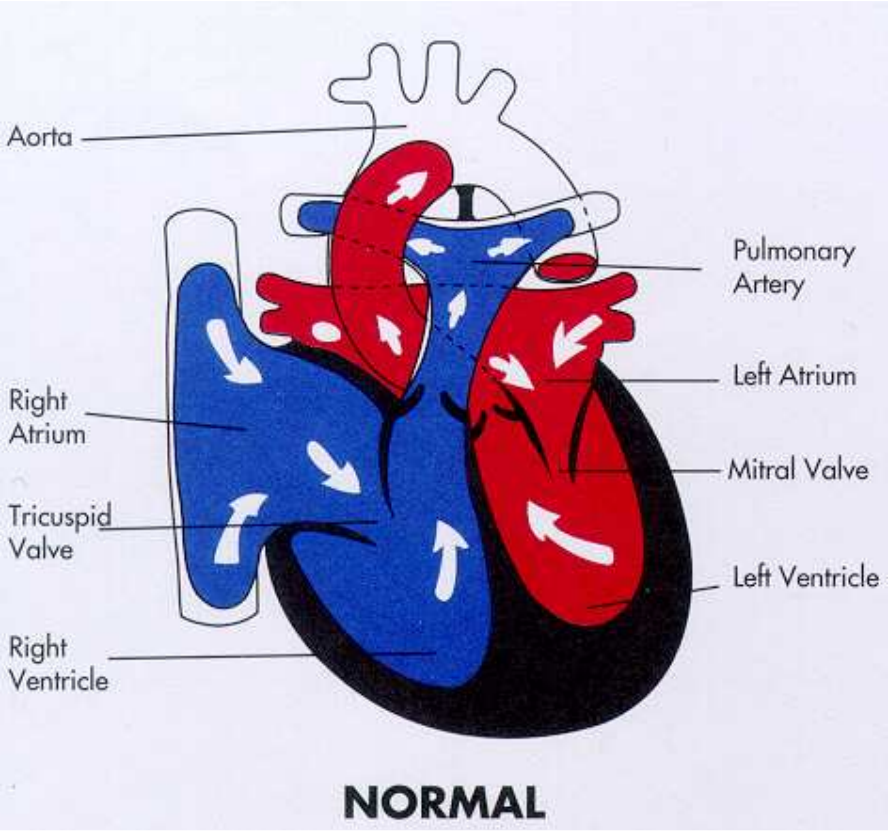
The illness



How do the heart and lungs work normally?

When the heart and lungs are working properly, the blood is pumped at a low pressure by the right side of the heart into the pulmonary artery and then to the lungs where it picks up oxygen. It then travels to the left side of the heart which pumps it around the body at a high pressure, passing the oxygen onto the brain and the muscles so that they all work properly. Once the blood has delivered all the oxygen the body needs, it returns to the right side of the heart and is pumped back to the lungs again.

What is Pulmonary Arterial Hypertension?

Pulmonary Arterial Hypertension (PAH) means that the blood is travelling through the lungs at a higher pressure than normal. The blood vessels that supply the lungs narrow and thicken, so that the heart has to work a lot harder to pump the blood through the vessels. The thick, diseased blood vessels cannot pick up as much oxygen as they should, so the body is not getting as much oxygen as is needed. This makes the person feels dizzy, tired, and short of breath.

There are two types of PAH, depending on whether the PAH is caused by another disease or not. Secondary PAH is caused by another disease, like a heart problem or lung disease. PAH that is not caused by any known disease is called Ideopathic Pulmonary Arterial Hypertension (IPAH). Certain drugs can rarely cause PAH, and this is called IPAH because we do not understand why the person develops PAH.

PH causes changes in the heart and lungs. The layer of cells which line the blood vessels, called endothelial cells, develop changes and become irritated. Doctors

do not know why this happens. When the cells become irritated, they defend themselves by making extra tissue. Over time, this can cause the vessels to become narrowed and scarred. The scar tissue makes the blood vessels less flexible which means that they cannot stretch to allow blood through. To add to the problem, the muscle cells in the wall of the vessels contract more than usual, causing the vessel to narrow even more. Unfortunately, this leads to problems with blood circulation and clots may form which injure even more cells. All these changes constitute ‘pulmonary vascular disease’.

For pictures of the changes that occur in pulmonary vascular disease, please see over the page.

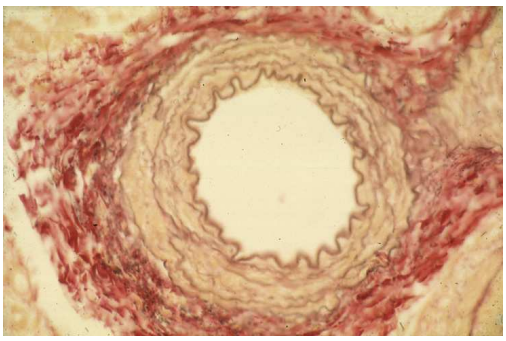


Figure 1:A very thick walled pulmonary artery

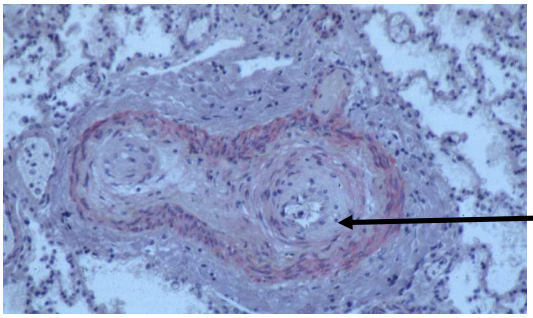


Figure 2: Two small pulmonary arteries
The small one on the left is completely blocked and
the one on the right has only a small lumen

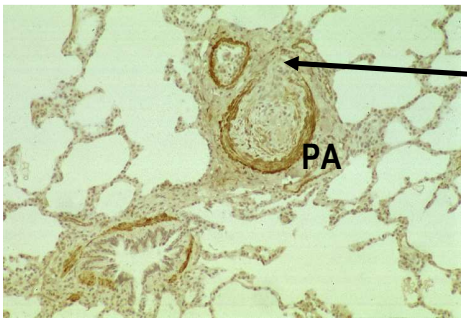


Figure 3: Small, blocked pulmonary artery (PA) with tiny small branch

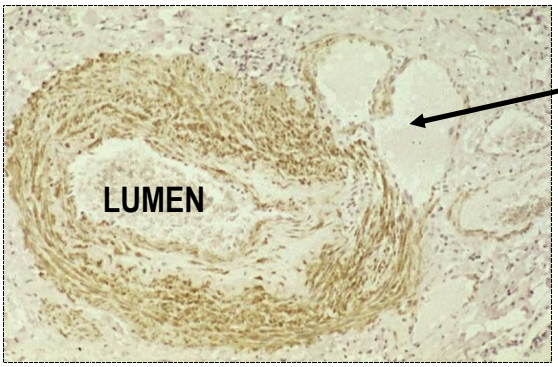


Figure 4: Pulmonary artery getting blocked and
and acquiring abnormal dilation lesions

Progression of disease

The heart develops changes as a result of these problems in the lungs. The heart has to work a great deal harder in a person with PAH to pump the blood through the narrowed vessels, so the right ventricle becomes larger and the walls grow thicker. When treatment lowers the pressure, the heart becomes smaller. However, the right ventricle can become permanently damaged if the blood pressure in the lungs is too high for some time. As the right ventricle becomes less efficient, the pressure builds up behind it and this may lead to fluid retention, especially in the legs, liver and in the space around the lungs. This is called 'congestive right heart failure'.

What are the symptoms of PAH?

The symptoms of Idiopathic and Secondary PAH are much the same, and how bad a person feels usually reflects how badly the disease has affected their body. The disease affects everyone differently, and therefore people with PAH will not necessarily suffer the same symptoms.

The main symptom is feeling out of breath (dyspnoea), tiredness and feeling dizzy when standing up or climbing stairs. However, these symptoms can be a part of other diseases too. Other symptoms include fainting when exercising (syncope), which is more common in children. It is quite common for people with PAH to suffer from swollen legs and ankles (oedema). Chest pain (angina pectoris) is also quite common, especially during exercise. Some people have heart flutters or palpitations. It is possible for a person with PAH to collect fluid in their neck and abdomen (ascites). As the disease progresses, people may have difficulty breathing when they are lying down (orthopnoea), due to fluid in and around the lungs or they may develop a bluish tinge to their lips and fingernails (cyanosis).

The disease is classified according to how badly the symptoms affect a person:

- Class 1 - PAH patients have no symptoms and their heart functions normally but they have higher blood pressure than normal in their pulmonary artery.
- Class 2 - PAH patients have symptoms when exercising and have a more marked increase in the pressure within their lungs.
- Class 3 - PAH patients have no symptoms when they are resting, but show symptoms when carrying out normal daily activities.
- Class 4 - PAH patients have symptoms even when they are resting, and the heart is showing signs of failure.

How is PAH diagnosed?

As the symptoms of PAH can look like those of many other diseases, your child will need various tests and procedures to rule out those diseases and come to a firm diagnosis of PAH. All the following tests combine to make a complete picture of your child's health. The tests which are always carried out include:

- *Physical examination* - this is a general health check which includes questions about your child's health and also that of the rest of the family. In particular, the doctor will listen to your child's heart, look for swollen veins in the neck (which

can be a sign that the heart is not working properly) and swollen ankles and legs.

- *Electrocardiogram (ECG)* - this assesses how well your child's heart is working. This may be a short test carried out in hospital, or your child may need to wear a portable ECG unit for a few days to get a continuous reading, to show how the heart behaves during normal daily life.
- *Chest x-ray* - This will give the doctor a picture of your child's heart and lungs. He or she use this to tell if the heart is enlarged.
- *Echocardiogram* - This uses sound waves to build up a picture of how your child's heart is working. It is very similar to the ultrasound scans you have when you are pregnant. It can show if the heart is formed properly and if there is any thickening of the heart muscles. It also measures the amount of blood flowing through the valves in the heart.
- *Blood tests* - These tests can tell the doctor how much oxygen is in your child's blood and also how his or her liver and kidneys are working. The amount of oxygen in the blood can also be measured using a pulse oximeter which is like a clothes peg on the end of a finger.
- *Cardiac catheterisation* - This is the most useful test in diagnosing PH and will complete the picture the doctor has built up using the previous tests. This test measures the blood pressure in the heart and lungs, and also measures how your child reacts to various drugs that are designed to lower the pressure, some of which are used to treat PAH. For more information about cardiac catheterisation, please ask for a copy of our leaflet: 'Angiography and angioplasty: information for families'.
- *Lung function tests* - These tests show how well your child's lungs are working by measuring how much air the lungs can hold. For more information about these tests, please ask for a copy of our leaflet: 'Breathing tests for young children'.
- *Exercise tests* - These tests show how well your child can cope with exercise and will involve cycling on an exercise bicycle, walking on a treadmill or simply walking for 6 minutes along a corridor.

Other tests which it may be necessary to carry out include:

- *Perfusion lung scan* - This is a scan which uses a special dye (contrast medium) which shows up on x-rays. The dye is injected into your child and is carried through the lungs by the bloodstream. It will show if there are any areas where the blood is not flowing properly or any blood clots. For more information on these special scans, please ask for our leaflet 'Isotope scans: information for families'.
- *CT scan* - This is a computerised x-ray which can show 'slices' through the body. The doctor will use the results of this scan to get a better picture of your child's heart and lungs.
- *MRI scan* - This uses a strong magnetic field instead of x-rays to give a very detailed picture of the heart and lungs.

You may get the results of some of these tests at the end of the appointment, but you may have to wait for a short while for the others. The doctors will explain the results to you as they get them.

What causes PAH?

Doctors working in this area of medicine are still not sure what causes PAH. In Secondary PAH, where the pulmonary hypertension is caused by another disease, they may understand the disease itself but not how this triggers severe disease of the blood vessels in the lung. However, there are many research programmes being carried out around the world, which will help us to understand the causes of all types of pulmonary hypertension better over the next few years. In the UK this work is funded by the British Heart Foundation.

Idiopathic PAH (PAH)- Around 6% of people with Idiopathic PAH have inherited an increased likelihood of developing the disease. A faulty gene was identified in 1999. The disease can skip generations, but it seems to develop earlier in life and to a greater extent if successive generations develop it. The clinical course of inherited PAH is much the same as non-inherited PAH. There are many research programmes being carried out in the UK and USA to find out more about the genetics of PAH.

Secondary PAH - There are many types of congenital heart defect including septal defects (holes in the heart). It is very important to repair these defects surgically before they start to damage the blood vessels in the lungs. However, in some children with congenital heart disease, the PAH appears to be Ideopathic rather than secondary, because the lungs becomes much more damaged, more severely and more rapidly than in most children with the same type of heart defect.

How common is PAH and who develops it?

PAH is a rare disease. The annual incidence of PAH is 2.5 people in every million. It affects males and females. Males tend to develop the condition in later life, whereas females tend to develop it between the ages of 20 and 50. However, PAH can occur in newborn babies and children too. It is not limited to a particular race or ethnic background. It can affect anyone.

How can PAH be treated?

As research into PAH continues, new and better treatments are discovered and developed. There are now many ways of treating PAH, but which method works best depends on the individual child. In Secondary PAH, the main aim is to treat the disease which is causing the PAH. This may involve an operation or drug treatment, depending on the original disease.

The following are some of the options for treating PAH:

- *Oxygen therapy* - Low levels of oxygen in the blood can make a person with PAH feel worse and therefore most people have oxygen therapy at some point in their treatment. This therapy can be continuous, where the person has to breathe in oxygen from a gas cylinder at all times, or can be occasional, where the person only needs to breathe in oxygen when moving around or sleeping. If a person with PAH has sleep apnoea (stopping breathing for short periods while asleep), they can use a machine to give them oxygen in short bursts throughout the night.

- *Medications* - Various medications can help a person with PAH:
    - *Diuretics* (water tablets) - PAH can cause a person to retain fluid which causes swelling. If the fluid retention is not too severe, adjusting the amount of salt eaten in the diet may help. If this does not help, the doctor may prescribe a diuretic. This is a medication which encourages the kidneys to remove more fluid from the body in the form of urine.
    - *Anticoagulants* (for example, warfarin and Asprin) - These are medicines which stop the blood becoming sticky and clotting too much. They thin the blood and encourage it to flow smoothly and easily through the lungs.
    - *Digoxin* - This medication was developed many years ago from the foxglove family of plants. It can help PAH patients as it helps a weak heart to squeeze better.
    - *Calcium-channel blockers* - These medications help the blood vessels in the body stay open as they relax the muscle layer in the walls of the vessels. This means that blood can be pumped through them much more easily. Some types of calcium-channel blockers can also slow down the nerve impulses passing through the heart muscle, which can correct certain types of heart flutter or palpitations.
    - *Vasodilators* - These are medications which help widen blood vessels, by relaxing the muscle in their wall. Epoprostenol (formerly known as prostacyclin) is the vasodilator commonly used. It is useful for people with PAH because it makes it easier for the heart to pump blood through the vessels. This medication is usually given directly into the bloodstream via a vein (intravenously) using a central venous access device called a Hickman catheter. Iloprost, another form of prostacyclin can also be given by inhalation using a special ultrasonic nebulizer device, experience is limited in children at present.

- Transplantation - Lung transplantation can often improve the quality of life for people with PAH. A heart-lung transplant is usually carried out.

The doctors will discuss with you the various options for treatment as and when they are needed. They will give you more information about each option, including any risks or side effects and whether they think it would benefit your child. Further information about the various types of medicines used are available in our series of medicines information sheets which are available on the ward and from the Pharmacy.

What is the outlook for people with PAH?

In most cases, how your child feels is the best indicator of how well his or her body is coping with the disease. In previous decades, the outlook for people with PAH was poor, with most people dying from heart failure within a few years of being diagnosed. However, now the outlook is much better due to new and improved methods of treating PAH.

People who have PAH can still go to school, college or work and have a normal family life. In most cases, they do not look ‘sick’ and only feel ill when they overdo it. Like everybody else, people with PAH can help themselves by eating a healthy diet, not smoking and resting well. Be careful about other medications which might be prescribed for your child by other doctors. For example if your child is unlucky enough to have Attention Deficit Hyperactivity Disorder (ADHD), it is better not to take Ritalin. Also herbal medicines are not a good idea as we understand too little about them, and some have been harmful. Do not take Aspirin, this thins the blood (as well as relieving pain) and your child is already taking an anticoagulant which indeed mat be in the form of Asprin already.

Pregnancy is not recommended in women with PAH as it puts an extra strain on the heart. Oral contraceptives (the Pill) are not recommended for women with PAH because they can encourage clot formation in the lungs. If you and your daughter would like to talk to someone about this, the doctor can arrange this for you.

Frequently asked questions

What sort of support can I expect when I get home?

In the first instance your GP should be called, it is important that he or she become familiar with your child's condition, additionally many areas now have good community paediatric nursing support and we will ensure they are briefed about your child's condition and treatment..

If your child needs intravenous Epoprostenol, once your child's treatment has been started, a home care nursing team will be appointed to help you. The team will try to visit you in hospital, before you leave us.

If your child needs nebulized Iloprost, there is a special protocol you will be given, and all equipment and supplies will be provided by Health Care at Home.

Depending on the treatment chosen for your child you may have lots of new skills to learn but we will make sure you are confident before you go home. Although it will be daunting at first, it will become easier with time. Remember that there is always someone on the end of the telephone at Great Ormond Street Hospital and in your local area to advise and reassure you. Before you go home, you should feel confident about all of the things on the following checklist - tick them off as you understand them.

- About PAH ❒
- Drugs needed and how they are given ❒
- Oxygen therapy and how it is given (if applicable) ❒
- Caring for the Hickman catheter (if applicable) ❒
- What to look out for ❒
- Trouble shooting ❒

Can my child have a shower or a bath?

The only restrictions on this are if your child has a Hickman catheter. In this case, you should talk to the doctor before you leave Great Ormond Street Hospital and read the information booklet called "Hickman Lines and Intravenous Epoprostenol"

Will my child be able to go to school?

This depends on how well your child is feeling. In most cases, he or she will be able to go to school, but the school staff may be worried because of your child's illness. The staff at Great Ormond Street Hospital are always willing to talk to them to put their minds at rest.

Your child is likely to need a 'Statement of Special Educational Needs', but the ward teacher will discuss this with you.

What about swimming or PE?

Again, this depends on how well your child is feeling. Gentle exercise is good for most patients, but energetic games like football or rugby may not be advisable. Your doctor will give you an idea of what your child can manage, but he or she will become expert in this very quickly! If your child is receiving Epoprostenol due to the risk of infection swimming is not recommended.

Will we be entitled to any extra benefits?

You should talk to the hospital social worker before you leave Great Ormond Street Hospital. He or she will explain about the benefits system and whether you are entitled to extra benefits.

If your child is having oxygen therapy, this may mean you need to live in a ground or first floor flat, rather than higher up in a block of flats. This is mainly due to the weight of the oxygen cylinders and the difficulties in carrying them up many flights of stairs in a building with an unreliable lift!

Can we go on holiday?

Again, this depends on how well your child is feeling. If you are planning to travel, remember to carry your child's medications in your hand luggage with this booklet in case your suitcases go missing. If your child needs extra equipment like syringes, you will need a letter from your doctor explaining why your child needs them to avoid any problems at security checks and customs. Always remember to carry your child's PAH fact file with all his or her details in it, whenever you travel.

If your child is having oxygen therapy, this may cause problems when flying abroad as oxygen cylinders are require special permission. The amount of oxygen available is less at altitude and your a child with PAH may not tolerate this well, even if not usually on home oxygen. There may be ways of getting around this, for instance, organising oxygen at your holiday destination, and travelling by rail or sea. Do talk to the airline which you would like to use, and your consultant and ask for their help. The Pulmonary Hypertension Association are always happy to help and advise with travel issues.

The staff at Great Ormond Street Hospital will be able to give you advice about holidays also, and a fact sheet of organisations that can help is available from the Health Information Centre.

How are we all going to feel about PAH?

Your child (and you and the rest of your family) may feel fed up with PAH from time to time. The disease affects every family in a different way, but the following are problems that commonly crop up. If you would like to talk through any of the following, please contact your child's team at Great Ormond Street Hospital who will be very happy to help.

- Keeping things normal

  Try to treat your child as normally as you can. Children who are over-protected or treated as sick can become demanding. Your child is only 'different' in that he or she has an illness. Your child will be happier if rules stay the same and life carries on much the same as before.

- Behavioural problems

  When you leave hospital, you may find that your child is more demanding than usual. This is a common reaction to being in hospital, so you should expect it. Your child may become more clingy or may revert to earlier behaviour, for instance, bed wetting, until he or she is used to being at home with you again. If you are worried about your child's behaviour, please talk to the nurses or play specialists. They may be able to offer you help and advice about settling back into a normal routine once you return home.

- Body image

  If your child is having oxygen therapy or has a Hickman catheter, he or she may feel self conscious about it. Talking to other people can help – you could try contacting the support organisations listed at the end of this leaflet for support and advice.

- Feeling tied down by the illness

  Your child, you and the rest of your family may feel that everything has to revolve around your child's illness and treatment. Keeping to your normal routine as far as possible may help.

- Sibling rivalry

  If you have other children, they may feel upset at the attention their brother or sister is receiving. Having 'special time' with your other children may help. If relatives and friends seem to focus on your child with a Hickman catheter and pay less attention to your other children, you could ask them to treat all your children equally.

Your guide to trouble shooting

Use this section to deal with any problems that may occur. It will help to familiarise yourself with it beforehand, so you know what to do if a problem crops up.

What if …	***Action***
Your child: • has chest pain • has a rapid or irregular heartbeat • faints or is feeling faint • has bronchitis or chest congestion • is coughing up coloured mucus • is coughing up blood	• Contact your local consultant, GP or Great Ormond Street Hospital.
Your child: • has a high or prolonged temperature of 38℃ or more • has diarrhoea	• These are signs that your child has an infection which can make PH feel worse • Contact your GP contact Great Ormond Street Hospital for advice or failing that take your child to your local hospital.
Your child: • has unusual shortness of breath • has unusual fluid retention	• These could be signs that your child's PH is getting worse • Contact Great Ormond Street Hospital for advice and an appointment to see the doctor
• Your daughter becomes pregnant	• Contact Great Ormond Street Hospital for advice and an appointment to see the doctor immediately.
• If changes are planned in medications (especially if taking over the counter medicines or vitamins)	• Contact Great Ormond Street Hospital • You will need to discuss this with the doctor
• If your child is due to visit the dentist	• Your child may need antibiotic cover to help prevent infection of or around the heart
• If a doctor thinks your child needs an operation	• They must discuss this with your consultant at Great Ormond Street Hospital or one of the nurses in the

	PAH Service.

Side Effects of Medications: Calcium channel blockers (e.g. Nifedipine)	
Ankle swelling Hypotension (low blood pressure) with dizziness, flushing, lethargy Tachycardia, palpitations Headaches (which tend to improve with time) Rash, itching Constipation or Diarrhoea Gum overgrowth Nausea and vomiting	If you are concerned about any of these side effects, please contact your doctor. Do not stop taking the medications suddenly. Do not chew the tablets, as they are designed to dissolve slowly in the stomach, releasing the calcium channel blocker over a period of time. Avoid taking these tablets with grapefruit or grapefruit-based drinks (like Sunny Delight). Grapefruit makes the drugs more powerful which could cause side effects.
Side Effects of Medications: Anti-coagulants (e.g. Warfarin)	
Signs of an overdose or that indicate the dose might need to be reduced include Prolonged bleeding from cuts; Bleeding that does not stop by itself; Nose bleeds; Bleeding gums; Red or dark brown urine; Red or black stools; In girls- increased bleeding during periods. Anticoagulants may also cause unusual bruising. Fevers, diarrhoea, nausea and vomiting Rash Signs of internal bleeding, such	Any of these side effects should be reported to the doctor who is prescribing your anticoagulant therapy

as a swollen, painful abdomen, back pain, dizziness, joint pains with stiffness or prolonged headaches.	