About Pulmonary Arterial Hypertension (PAH) and Treatment With OPSUMIT (macitentan)

This booklet is for people who have been diagnosed with PAH and have been prescribed OPSUMIT

This booklet has been written and provided by Actelion Pharmaceuticals Limited

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LIVING BEYOND PAH

First and maybe most important for you to know: You are not alone. Your PAH doctor, nurse, and PAH healthcare team are there to support you.

Learning you have PAH may be hard to accept. It may cause a variety of feelings, from shock and disbelief, to maybe anger, fear, or sadness. However, it may also be a relief to have a name for your condition and know it can be managed with the right treatment. Your focus should be on taking your medicine as directed by your PAH doctor and making sure you have access to the information you need. This will help you manage your feelings, manage your well-being, and move forward with a life beyond PAH.

HAVE CONFIDENCE IN THE FUTURE

Living with and treating PAH is a journey. It will be different for everyone. While PAH is not curable, there are treatments available to help your condition improve and prolong your quality of life.

Think about the things you like to do and the people who are most important to you. Focus on making those activities and relationships a priority. You are still the same person you were before PAH. You still have all the strength, ability, and coping strategies you have been using for years. You can still look forward to planned events, such as family celebrations, social outings with friends and trips abroad. Stay focused on the things that are important to you while taking steps to manage your PAH.

WHAT CAN YOU EXPECT FROM THIS BOOKLET?

Whether you recently learned you have PAH, or have been living with it for some time, information and support is available to help you manage your everyday life. This booklet can help you learn important information about PAH and its treatment. It includes support, guidance, and practical tools to help you. The right information can help you make decisions about how to move forward. It will also help you feel more confident about making choices that fit your lifestyle. This can help you focus on leading an active and fulfilling life.

Your PAH doctor, nurse, and PAH team are the best sources of information for many topics such as medicine, exercise, and family planning. You may also find other useful information from local and national patient associations for example the Pulmonary Hypertension Association UK (PHA-UK) or trusted sources on the Internet.

Feel free to share this booklet with your family, friends, and those who may help care for you.
You learned that you have PAH. PAH is a serious condition that affects the blood vessels that carry blood from the heart to the lungs. To understand more about PAH, first it helps to understand how the heart works.

**THE ROLE OF THE HEART**

The heart pumps blood to the whole body. The heart is a big muscle that is divided into 2 parts—a left and right side. It is responsible for pumping blood to the lungs, back to the heart, and around the body. This system is called blood circulation. PAH causes an increase of blood pressure in the blood circulation system between the right side of the heart and the lungs.

**WHAT HAPPENS TO THE PULMONARY ARTERIES IN PAH?**

As PAH develops, the pulmonary arteries become narrow. The walls of these vessels also become thicker and less flexible. This narrowing of the pulmonary arteries restricts the blood flow to the lungs. This means that the heart finds it harder to pump blood through the arteries, which increases the blood pressure in the pulmonary arteries.

**HOW DOES THIS CAUSE THE SYMPTOMS OF PAH?**

With the increased blood pressure, the right side of the heart has to work harder to pump blood through these vessels and into the lungs. This causes the right side of the heart to become enlarged. In some cases, over a period of time, the heart can become tired and overworked. Less blood will circulate through the lungs, picking up less oxygen. This may make you feel tired and breathless. If PAH is not treated, the heart can wear out from the effort of trying to pump blood around the body.

While PAH is not yet curable, there are reasons to hope. There are treatments for PAH to help you improve your quality of life and live longer.
HOW DO DOCTORS MEASURE THE SEVERITY OF PAH?

You may have heard your PAH doctor or nurse refer to Functional Class. This is simply a way of assessing the effect that PAH is having on your day-to-day living. It provides a useful tool when considering your treatment plan. The Functional Class system has 4 levels to describe how severely PAH affects your life. Your PAH specialist team will use this system to assess how severe your PAH symptoms are. The table below describes the 4 levels.

HOW DOES PAH AFFECT YOU?

You may have first noticed signs when you are physically active, such as when you climb stairs, run, walk uphill, or carry bags. As PAH develops, you may notice signs more often. You may find that your symptoms affect the things you can do. PAH is a disease that can get worse over time. You may have good days and bad days. There are specific treatments that can help you.

You may be experiencing one or more of the following common symptoms of PAH:

- Shortness of breath, especially with physical activity such as walking uphill or climbing stairs
- Feeling tired
- Dizziness
- Chest discomfort or pain
- A fast and/or irregular heart beat (also called palpitations)
- Light-headedness or fainting
- Swelling in your arms, legs, or ankles (also called fluid retention or oedema)

Other symptoms that you may also notice include:

- You have no symptoms with normal daily activities
- You have some symptoms with normal daily activities AND some limits when doing physical activity
- You may feel symptoms when doing less than your normal activity. You may also feel more limits when doing physical activity
- You may feel symptoms with any kind of activity, including while resting

You can learn how to manage your symptoms so you can still do most of the things you want.

Your PAH specialist team will aim to improve your PAH symptoms by getting you to the lowest class possible and then keeping you at that level for as long as possible.
HOW COMMON IS PAH?

- PAH is a rare condition that can affect women, men, and children of any age or ethnic background. This is why many people do not know about PAH.
- Across the globe, only 15 to 50 people in a million are affected by PAH.

DIFFERENT TYPES OF PAH

There are many types of PAH. In some cases, PAH may be associated with other conditions, such as congenital heart disease or connective tissue disease. PAH rarely runs in families. Some of the causes are known and some have yet to be identified. Your PAH specialist team can help you understand what type of PAH you have. Regardless of what type of PAH you have, the treatment options remain the same.

YOU ARE STILL YOU

Regardless of the fact that you have just learned you have PAH, or have been living with PAH for a while, you are still you. Making adjustments to your everyday life, based on how you are feeling and what you want to do, will help you stay in control of your life.
EVERYONE WILL EXPERIENCE THEIR OWN REALITY OF LIVING WITH PAH

Living with and managing PAH symptoms can have an emotional impact. At times, you may feel low, frustrated, anxious, or may be unmotivated. It is important to know: You are not alone. These feelings are very common among people with PAH.

You can develop a support system to help you manage your condition. You can include people close to you. Your PAH specialist team and local and national patient associations, together with the important people in your life, can be a valuable source of support when you need it.

ADVANCES IN TREATMENT

The understanding of PAH and its treatment have improved a great deal over the last 10 years. Having increased research means more effective treatments. There are also improved services and specialised care. This can help people with PAH manage their condition better for longer and to have a more enhanced quality of life. This means that chances for patients to live life on their terms have improved.

SHARE YOUR THOUGHTS AND FEELINGS

People who do not know about PAH may not fully understand how you are feeling. To them, you may look and seem healthy in terms of your appearance. But underneath, you may be experiencing a range of feelings or limitations. You may wish to share some information about how PAH affects you. This can help others understand why you cannot always do what you used to or want to do.

TALKING ABOUT YOUR CONDITION

Although, at first, you may feel embarrassed to talk openly about PAH and its effects on your quality of life, talking to someone may be helpful for you. You may also find comfort in sharing your thoughts, feelings, and experiences with a friend or relative and with your PAH doctor or nurse. A good relationship with others allows you to discuss your concerns more easily. The more your support network knows and understands about you, the more they can help you.

If there is anything you don’t feel comfortable talking about yet, address this openly. You may want to share more at a later point in time.
TALKING TO PEOPLE WHO KNOW PAH
If it is hard for you to talk to your family or friends about everything, ask your PAH doctor or nurse for help in finding someone in your area who you can talk to, like a social worker, counsellor, or psychologist. There are people who deal with PAH every day and can offer help and advice. Patient associations can be a great source of support and comfort in helping you with all aspects of day-to-day living with PAH. Contact details for your local and national PAH patient associations can be found later in this booklet.

HOW YOUR FAMILY AND FRIENDS CAN SUPPORT YOU
If you decide to share information about PAH with your family or friends, this booklet will help them understand your condition. It is as natural for your family and friends to experience a range of feelings as it was when you were first diagnosed. Give them time to ask questions or express their feelings. Like you, the people close to you will want to learn about PAH and understand how it is affecting your life. By explaining the condition you have, the treatment plan you are on, and the changes you will make to your everyday life, you will give others a focus for managing their own feelings and supporting you. For specific advice about explaining PAH to your family, contact your local or national PAH patient association.

ACCEPTING HELP FROM OTHERS
As with any long-term condition, family and friends may want to offer support. Don’t be afraid to ask for help when you need it or accept help when it is offered. Accepting help gives those who care about you a sense of making a contribution at a difficult time. It can also allow you to get on with the things that you can and want to do. While this may not be what you are used to doing, you may find that by accepting help you have more of yourself to give back to others. It may also help to have a partner or family member with you at your PAH doctor or PAH Centre visits. This way they can get a better understanding of PAH and how you and your PAH doctor is managing it. Consider these ways in which others may help you:

SHARE YOUR PLANS
You do not need to give up on your plans. You may want to tell your family and friends what you are looking forward to doing. Only by sharing these things can your family and friends understand how they can support you in achieving your goals.
SETTING GOALS AND MAKING ACTIVE CHOICES
There are many choices you can make to help maintain your health whilst living with the symptoms of PAH. Making these choices will help you to achieve the things you want. The following pages provide some suggestions for how you can help manage your PAH by understanding practical ways to adapt your everyday living.

Eating a healthy diet, being active, learning how to cope with tiredness, and sharing your feelings are just some of the things that can make a big difference to how you feel and how you adjust to living with PAH. As you read through, write down things that come to mind. Also, note any questions you may want to ask your PAH doctor or nurse. This will enable you to focus on managing how you feel, with an aim to make every day a good day.

EATING A HEALTHY DIET
Because everyone is different, there is no set diet to help to reduce PAH symptoms. As with everyone, it is important to eat normally and healthily to keep your energy levels up. You should aim to achieve and stay at a healthy weight. If you notice any significant weight loss or gain, tell a member of your PAH specialist team.

Your PAH specialist team may suggest the following tips to help you maintain a healthy diet:

- Change to a salt-free diet (salt intake causes you to retain water, which may worsen any swelling or oedema you have)
- Avoid packaged and prepared food as these are often high in salt, fats, and sugar
- Eat smaller, more frequent meals
- Eat fresh fruit, vegetables, and whole grains
- Eat fish and lean meat, such as chicken

Drinking alcohol may be acceptable in moderation. Some foods and drinks can affect medicines, so you need to check with your PAH specialist team if there is anything you should avoid.

For further information, contact your local or national patient association. Contact details can be found at the end of this booklet.
WHAT IS THE RIGHT LEVEL OF ACTIVITY FOR YOU?

It may be hard to know how much or what type of exercise is possible with PAH. With help from your PAH specialist team, you can work out the level of activity that is right for you. Choose activities that you enjoy and that make you feel good.

Here are some helpful tips:

- Start out slowly and listen to your body
- When exercising, ensure that you are not too breathless to have a conversation
- Adjust your activity before you become tired
- If you experience any symptoms that concern you, for example dizziness or chest pain, seek medical advice

If you have any concerns or simply want to have reassurance that you are doing the right amount of activity, speak to your healthcare support team. They have experience and knowledge about PAH symptoms. Gym instructors, personal trainers, or people with similar jobs may not understand how PAH can affect an individual, so your best source of advice will be your PAH specialist team.

MANAGE TIREDNESS

A common symptom of PAH can be extreme tiredness. This is caused by a decreased supply of oxygen to the body. It may feel different than feeling run down or tired from lack of sleep. Getting to know your body and understanding the signs it gives are important. This will enable you to rest when your body needs it.

Keeping notes or a diary of when you have felt particularly tired will help you to notice, and then avoid, specific triggers. A diary can help you know the times you are at your best and which activities use the most energy. This can help you adapt to situations and plan for both your everyday activities and the bigger events in your life.

HOW TO LIVE WITH PAH

ABOUT LIVING WITH PAH

EXERCISE AND PAH

Only engage in physical activity and exercise in line with your PAH specialist team’s recommendations.
Everyone with PAH experiences symptoms in different ways. Your PAH specialist team can provide advice based on your individual needs.

ABOUT LIVING WITH PAH

ADDITIONAL AREAS TO THINK ABOUT

STARTING A FAMILY
Pregnancy is associated with an increased chance of severe complications for people with PAH. You must speak to your PAH specialist team and seek medical advice if you plan to start a family or become pregnant whilst on treatment for PAH. Please see page 30 for more information on OPSUMIT and pregnancy.

WORKING
Your ability to keep working is based on how you feel, the choices you make, and the type of job you have. Your employer may offer you some flexibility as well. Your PAH doctor or your nearest PAH Centre will be able to share information with you on laws relating to your rights in the workplace or what to do if you are self-employed. Your PAH doctor can write a letter to provide proof of your condition for employers or for benefits claims.

SOCIAL SERVICES
Ask at your nearest PAH Centre to learn more about support services that are available to you.

TRAVEL
If you plan to take a flight or to visit a country with a different altitude, ask your PAH specialist team for specific advice and recommendations.
TREATMENT CAN HELP KEEP YOUR PAH FROM GETTING WORSE

It is important to start treating PAH as early as possible. Treating your PAH as directed by your PAH specialist team can help keep your PAH from getting worse. While your medicine is working to help you, you can focus on the things you want to do, not your PAH. Set goals for yourself to remain active and enjoy important events with your family and friends. Looking forward to these things will help you stay positive. While there is no cure for PAH yet, there are treatments that can help you manage your symptoms today and improve your long-term quality of life.

BEING PRESCRIBED MORE THAN ONE MEDICINE

To help you get the best quality of life possible, your PAH specialist team may give you one, or more than one, medicine. Your PAH doctor will choose the medicine (or medicines) that will best help you do things today and help you continue to reach future goals. From time to time, your treatment may be adjusted. This is not necessarily because anything is wrong; it may simply be a way of achieving the best possible results. It is important to take care of yourself and take the medicine or medicines prescribed by your PAH specialist team.

TYPES OF TREATMENT

There are many kinds of treatments and medicines for PAH. Each one may work differently in your body. It can be a lot to learn. But your PAH specialist team are here to help you decide what’s right for you.

Different approaches

A number of treatment approaches may be used to treat your PAH, including:

- Oral therapy—pills that are swallowed
- Inhaled therapy—medicines that are inhaled (breathed in) through the mouth
- Medicines that are delivered continuously to the bloodstream through a vein or a needle placed under the skin (intravenous or subcutaneous therapy)
- Surgery—including lung or heart-lung transplantation (may be considered for people who are unable to perform any physical activity and for whom medication is not successful)

Medicines that treat PAH specifically

There are several medicines that treat PAH specifically. These medicines relax and widen the blood vessels in your lungs. They also reduce and prevent the overgrowth of cells in the walls of the vessels, which makes it easier for your heart to pump blood through your lungs. This should lead to an improvement in the level of your physical activity and well-being.

- Endothelin receptor antagonists (ERAs)
- Phosphodiesterase (PDE5) inhibitors
- Prostacyclin analogues

Other PAH treatments

There are several treatments that can help relieve symptoms of PAH, including:

- Blood thinners to prevent clots
- Diuretics (also called water tablets) to reduce swelling
- Digoxin to increase the strength of your heart and slow your heart rate
- Supplementary oxygen to ensure your body has enough oxygen

WHAT TREATMENTS ARE AVAILABLE?
What is Opsumit and how does it work?

Opsumit is a tablet you take once a day. It was specifically developed for the treatment of PAH. Opsumit is an endothelin receptor antagonist (ERA), which means it blocks the undesirable effects of endothelin.

How does PAH affect your body?

Endothelin is a protein made naturally in the body. When endothelin binds to special molecules (receptors) in the vessels and lung tissue, blood vessels start to narrow. In people with PAH, the amount of endothelin in the body is increased. Over time, this will damage the blood vessels and raise blood pressure. This means you may feel symptoms such as shortness of breath, swollen ankles and legs, dizziness, and possibly chest pain.

How does opsumit work?

Opsumit moves throughout your body and reaches the lungs and vessels. There it binds to and blocks the endothelin receptors. Its chemical properties allow Opsumit to bind to the receptors for a relatively long time. This helps protect your blood vessels from the damage caused by endothelin.

How quickly will your symptoms improve on opsumit?

Each person responds differently to treatment. It may take time for Opsumit to make a difference to how you feel. You may not feel the effect of Opsumit immediately but you may start to notice an improvement over time as it begins to take effect. Your PAH specialist team can explain what you can expect when you start taking Opsumit.

1. Opsumit (macitentan) Summary of Product Characteristics, December 2013
PROVEN TO HELP PATIENTS WITH PAH

Your PAH doctor has prescribed OPSUMIT to help manage your PAH over the long-term. OPSUMIT was studied around the world in a large number of patients with PAH for up to 4 years. The study showed that OPSUMIT:

- Improved patients’ quality of life
- Improved patients’ symptoms
- Helped reduce the likelihood of hospitalisation due to PAH

Overall, the results of the study prove OPSUMIT provides long-term benefits for people who have not previously received treatment for their PAH and that it also works well for people who are already taking other PAH-specific treatment. It can be used alone or in combination with other common PAH medicines.

HOW AND WHEN TO TAKE OPSUMIT

GETTING STARTED

It is important that you read the Patient Information Leaflet (PIL) that comes inside your box of OPSUMIT. This patient leaflet explains possible side effects and risks to be avoided when taking OPSUMIT. If you have further questions on the safety profile of OPSUMIT please ask a member of your PAH specialist team. You can also refer to pages 30-32 of this booklet.

ABOUT OPSUMIT

OPSUMIT is a tablet. You take it by mouth once a day. One tablet contains 10 milligrams (mg) of medicine. For best results, it is important that you take your medicine as directed by your PAH doctor.

TAKING OPSUMIT IS EASY

- Swallow 1 whole OPSUMIT tablet with a glass of water each day
- Do not chew or break the tablet
- It is best to take the tablet at the same time each day
- Choose a time that will help you remember, such as first thing when you wake up or when you brush your teeth
- Take OPSUMIT with or without food

ABOUT YOUR TREATMENT HOW OPSUMIT CAN HELP YOU

OPSUMIT can help improve your PAH symptoms AND it can help improve how well your PAH is managed over time.1

1. Opsumit (macitentan) Summary of Product Characteristics, December 2013
**ABOUT YOUR TREATMENT**

**HOW AND WHEN TO TAKE OPSUMIT**

**IT IS IMPORTANT TO TAKE YOUR MEDICINE EVERY DAY**

If you forget to take OPSUMIT, take a tablet as soon as you remember.
- Then continue to take your next tablet at your usual time
- Do NOT take a double dose (2 tablets at once) to make up for the forgotten tablet

OPSUMIT should NOT be stopped (unless a member of your PAH specialist team tells you to do so).
- If you stop taking your medicine, this may cause your symptoms to get worse
- Do NOT stop taking OPSUMIT unless you have agreed to this with your PAH doctor

Don’t let your medication run out.
- If you do run out, contact your PAH specialist team or nearest PAH Centre as soon as possible
- They will help you set up a system to make sure you do not run out again

> Always take this medicine exactly as instructed by your PAH specialist team. Check with your PAH specialist team if you are not sure.

**WHO CAN TAKE OPSUMIT?**

Most people with PAH can take OPSUMIT when prescribed by a doctor. Certain people should NOT take OPSUMIT:
- Women who are pregnant or plan to become pregnant during treatment should not take OPSUMIT
- Women who are breastfeeding should not take OPSUMIT
- People who are allergic to OPSUMIT or any other ingredients in OPSUMIT should not take OPSUMIT
- People with liver disease or who have very high levels of liver enzymes in their blood should not take OPSUMIT

If you belong to one of the above listed groups, please tell your PAH specialist team.
# About Your Treatment

## Who Can Take Opsumit?

### Opsumit and Pregnancy

If you are pregnant or breastfeeding, think you may become pregnant, or are planning to have a baby, ask your PAH doctor for advice before taking this medication.

Opsumit may harm unborn babies conceived before, during, or soon after treatment.

- If it is possible you could become pregnant, use a reliable form of birth control (contraception) while you are taking Opsumit.
- Do not take Opsumit if you are pregnant or planning to become pregnant.
- If you do become pregnant or think that you may be pregnant while you are taking Opsumit, see your PAH doctor immediately.

If you are a woman who could become pregnant, your PAH specialist team will ask you to take a pregnancy test before you start taking Opsumit and regularly (once a month) while you are taking Opsumit.

### What If You Are Lactose Intolerant?

Opsumit tablets contain small amounts of a sugar called lactose. If you have intolerance to lactose or any other sugars, you should speak with your PAH specialist team before taking Opsumit. Opsumit tablets contain lecithin from soya. If you are allergic to soya, do not use this medicine.

## Taking Opsumit Safely

### Pregnancy Tests

You will be asked to take a pregnancy test before you start taking Opsumit and regularly (once a month) while you are taking Opsumit. This is because Opsumit may harm unborn babies conceived before, during or soon after treatment.

### Blood Tests

Your PAH doctor or nurse will take blood tests before starting Opsumit and regularly during treatment. The required tests will show:

- Whether you have anaemia (a reduced number of red blood cells). This test may be repeated as clinically indicated.
- Whether your liver is working properly. Your doctor or nurse may refer to this as a liver function test (LFT). This test will be repeated regularly during treatment.

### PAH Treatments Like Opsumit May Affect Your Liver

Look out for signs that your liver may not be working properly. If you notice any of these signs, tell a member of your PAH specialist team immediately.

- Feeling sick (nausea)
- Vomiting
- Fever
- Pain in your stomach (abdomen)
- Jaundice (yellowing of your skin or the whites of your eyes)
- Dark-coloured urine
- Itching of your skin
- Lethargy or fatigue (unusual tiredness or exhaustion)
- Flu-like syndrome (joint and muscle pain with fever)

Please read the Patient Information Leaflet that comes with your medicine. It will provide you with important information.

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Like all medicines, OPSUMIT can cause side effects. However, not everyone will get them.

**VERY COMMON SIDE EFFECTS** (may affect more than 1 in 10 people)
- Anaemia (low number of red blood cells) or haemoglobin decreases
- Headache
- Bronchitis (inflammation of the airways)
- Nasopharyngitis (inflammation of the throat and nasal passages)
- Oedema/fluid retention (swelling)

**COMMON SIDE EFFECTS** (may affect up to 1 in 10 people)
- Pharyngitis (inflammation of the throat)
- Influenza
- Urinary tract infection (bladder infection)
- Hypotension (low blood pressure)

**POSSIBLE SIDE EFFECTS**

If you get any side effects, talk to your doctor, pharmacist or nurse. This includes any possible side effects not listed in the package leaflet. You can also report side effects directly via the Yellow Card Scheme at www.mhra.gov.uk/yellowcard. For people living in Ireland, side effects are monitored by the Irish Medicines Board (IMB).

This medicine is subject to additional monitoring. This will allow quick identification of new safety information. You can help by reporting any side effects you may get. See www.mhra.gov.uk/yellowcard for how to report side effects in the UK or for Ireland www.medicines.ie/yellowcardreporting.

**YOUR PAH SPECIALIST TEAM IS HERE TO HELP YOU**

Contact a member of your PAH specialist team or nearest PAH Centre immediately if you:
- Have changes in your breathing during the first few weeks of treatment
- Have signs of an allergic reaction (swelling of the face or tongue, rash, itchy skin)
- Have a side effect that seems to be getting worse
- Notice a side effect not listed here
- Are concerned about how your medicine is affecting you
GETTING TO YOUR APPOINTMENT

It can be difficult to remember all the information that the PAH specialist team tells you. So, think about taking a family member or friend with you to your appointment. If you are not familiar with where your PAH doctor is located, spend some time trying to find out. Then you can plan your route. If you are travelling by public transport, make sure that you are familiar with the times of departure and expected arrival at your destination. Bear in mind the time of day you will be travelling and the weather on the day. Allow enough time for your journey. Aim to arrive 15 minutes early. This way you can be calm and relaxed.

PREPARING FOR YOUR APPOINTMENT

Your PAH specialist team will want to talk to you about your care, provide you with choices, and answer your questions. If there are things about your PAH symptoms or about your treatment that you do not understand, ask your PAH doctor or nurse to explain them to you. Together you can discuss your treatment and future goals. This will help you achieve what you wish in the months ahead.

Your PAH specialist team is there to help and support you. It doesn’t matter how many questions you ask, or what you ask about. Your PAH specialist team will understand and be glad that you are raising your concerns. Don’t be afraid to ask your doctor or nurse to be honest with you or to explain things in simple terms if there is something you don’t understand.

There may be a lot of information you would like to cover in your appointment. Preparing in advance for the points you wish to talk about can help.

There are additional sources of support beyond medical treatment. Look for these in your community:

- Engage with your family and friends. They can be a huge source of support and energy and can help you live life with PAH
- Get in touch with your nearest PAH Centre, PAH doctor, and nurses. They are all there to answer your questions, discuss your concerns, and help direct you to other support services
- Join a PAH support group. In this friendly setting you can share with, and learn from, other people living with PAH

By taking an active role in your care, you will stay in control of your life and remain independent. It will give you the confidence to help manage your condition and make it easier for you to make appropriate decisions about your health.
There are some small things you can do to help manage life with PAH. You may find the following tips useful:

**KEEP A DIARY**

Keeping a diary, perhaps for a month before your next appointment, can be very helpful. You may want to record how you are feeling on a day-to-day basis, including your thoughts and feelings, anything about your treatment, or just about everyday life.

You may find it helpful to write down any questions you have for your PAH specialist team to make sure you don’t forget any of them. Consider writing down the answers and ask for correct spellings of words you don’t know. Remember to include questions you have on working, financial issues, housing, and equipment concerns. You can also discuss any emotional or sexual changes you may be feeling.

You can use the space on pages 44-45 to get started.

Sharing this information with your PAH doctor and nurse allows them to better understand you, your PAH symptoms, and your treatment.

**UPDATE YOUR MEDICAL INFORMATION**

Keeping your medical information well organised and up-to-date is important both for you and family and friends involved in your care. Consider making a list of the names of the members of your PAH specialist team, along with contact details and addresses.

It is also important to keep a list of your current medications, vitamins, supplements, and anything else you are currently taking. Be sure to note the dose and how often you take each item. Consider making this information available to the important people in your life.

**LOOK TO THE FUTURE.** Think about what is important to you and set goals for what you would like to accomplish. Tell your PAH team about your personal goals. Together you can make sure you are always discussing the next step in what is best for you.
Finding out that someone close to you has PAH can lead to a variety of emotions. It is important that you take the time to understand and manage your own feelings. Talking about how you feel and considering the practical changes that will be needed are helpful first steps in your journey together with PAH.

Since PAH is a condition family and friends can’t see, it can sometimes be difficult to understand the challenges that PAH can present.

TAKING AN ACTIVE ROLE IN HELPING SOMEONE WITH PAH

Helping to care for someone close to you is an important role. It can also be a very rewarding one. It will be different for everyone depending on the person for whom you are caring. Some people adopt an organising role and carry out many of the physical tasks that can become more difficult for the person with PAH. The type and amount of help a person with PAH needs may change over time.

There are many different ways in which you can help someone with PAH. Here are some practical examples to think about:

- Spending time together
- Understanding when the person with PAH has ups and downs
- Accompanying the person with PAH to his or her PAH appointments
- Organising medicines
- Going grocery shopping
- Running errands
- Doing laundry
- Helping with transportation needs

Taking an active role in helping someone with PAH can be hard and have an impact on your life and relationship.
YOUR EVERYDAY LIFE MAY CHANGE

Caring for someone with PAH can have both a physical and emotional impact on your everyday life over time. You will spend more time with the person with PAH as you learn to manage this new situation together. This may mean a change in your schedule. It may also mean you have less time for yourself and the things you would normally be doing. Although it may be hard, you need to focus on keeping things as normal as possible in your everyday life.

Down the road, as you have more responsibilities, there will likely be more changes. You may need to make changes to your working conditions or living arrangements. In some cases, this can have an impact on household finances. Whatever challenges and changes you face, there is support available for you.

REMEMBER YOUR NEEDS

When you start helping to care for someone with PAH, you can feel overwhelmed and hardly know where to start. That’s okay. You’re not alone. There are resources available to help you.

Getting in touch with a PAH patient association will provide you with a wide variety of support designed just for you. There are support networks that you can contact if you feel you would benefit from speaking to others like you.

Look for ways to continue activities you enjoy together, even if it means doing things for a shorter period of time or in a different way.

LEARNING MORE

You may feel like there is a lot you need to learn about PAH and what the future holds. You may have questions and want more information. Support is available from a variety of sources to help you manage what’s ahead. You can seek information for yourself on the Internet. You may even want to share what you find with the person with PAH. However, what you read on the Internet may or may not be accurate. So if you have questions, be sure to ask the healthcare team.

Many friends and relatives of people with PAH join PAH patient associations and become active members. These associations can help you learn more about the emotional and practical aspects of caring for someone with PAH. They can help provide advice on coping with PAH on a day-to-day basis. This includes household finances, changes in sexual relations, and claiming benefits and allowances. You may find it helpful to learn from and share experiences with others like you.

You may also want to receive updates on the person with PAH, such as detailed medical information and what to expect from PAH itself. You may also be interested in talking to PAH doctors and nurses by attending medical appointments.

Details on your local or national PAH patient association can be found on page 47 of this booklet.
HELPFUL DIALOGUE TOOLS
A dialogue tool for family and friends is available from your local or national PAH association. This tool can help prioritise what is important to you. It can help focus discussions with the PAH doctor, family members, patient association, and other relevant parties such as social services and local councils. This booklet will also help you clarify issues and emotions associated with caring for someone with PAH.

HOW OPSUMIT CAN HELP
OPSUMIT is a treatment specifically developed for the treatment of PAH. A long-term study in a large number of patients with PAH showed that it can improve patients’ quality of life, improve symptoms and reduce the likelihood of hospitalisation due to PAH.1 For more information about OPSUMIT, please ask the person with PAH or a member of his or her PAH specialist team.

1. Opsumit (macitentan) Summary of Product Characteristics, December 2013

FOR IMPORTANT PEOPLE IN YOUR LIFE
You may want to share the following pages of the booklet with those people closest to you. These pages have been prepared to help them.

It is good to talk about PAH and to keep a positive outlook.

It is good to talk about PAH and to keep a positive outlook.

PLAN FOR TOMORROW

LIVE FOR TODAY

It is good to talk about PAH and to keep a positive outlook.

It is good to talk about PAH and to keep a positive outlook.

It is good to talk about PAH and to keep a positive outlook.
YOUR QUESTIONS

Please use this section to write down any feelings, questions, or issues that interest or concern you. The more specific you can be, the more your PAH specialist team, friends, and family will be able to support you.

- QUESTIONS ABOUT TREATMENT WITH OPSEMIT
- QUESTIONS ABOUT HOW TO TAKE CARE OF YOURSELF
- QUESTIONS ABOUT EATING, EXERCISING, AND/OR MANAGING FATIGUE
- QUESTIONS ABOUT WORK AND FINANCIAL SUPPORT

YOUR GOALS

Please use this section to write down any goals you have, big or small.

- YOUR GOALS
Congenital heart disease—a general term for a range of birth defects that affect the normal workings of the heart. Congenital means the condition is present from birth.

Connective tissue disease—connective tissue disease refers to a group of disorders involving the tissue that supports organs and other parts of the body.

Endothelin—a protein that tightens the blood vessels and raises blood pressure.

ERA—endothelin receptor antagonist, a medicine that protects against damage to the blood vessels and, in turn, the heart.

Functional Class (FC)—a well-established way of measuring the effect that PAH has on an individual in terms of day-to-day living, developed by the World Health Organization.

Pulmonary arterial hypertension (PAH)—a condition that affects the blood vessels that carry blood from the heart to the lungs.

Pulmonary arteries—blood vessels that carry blood from the right side of the heart to the lungs.

For further information about PAH, managing the condition and the practical aspects of living with PAH, please get in touch with the following organisations:

- Pulmonary Hypertension Association UK: www.phassociation.uk.com
- Pulmonary Hypertension Association Ireland: www.pulmonaryhypertension.ie/pha-ireland/
- The Scleroderma Society: www.sclerodermasociety.co.uk
- Disability Benefits Unit: Warbreck House, Wambreck Hill Road, Blackpool, FY2 0YE
- Carers Allowance (formerly Invalid Care Allowance): www.gov.uk/carers-allowance-unit
  Tel: 0845 608 4321
  Palatine House, Lancaster Road, Preston, PR1 1HB
- Motability: www.motability.co.uk
  Tel: 0300 456 4566
  Warwick House, Roydon Road, Harlow, Essex, CM19 5PX