Living with PAH
At Actelion, we believe that knowledge is power. When you understand your condition, you have the power to ask the right questions, make informed decisions, and get the most from your healthcare team and your therapeutic options. That’s our goal in PAH education and that’s KNOWLEDGE IN ACTION.

Pulmonary arterial hypertension (PAH) is a progressive disease, meaning that, over time, its effects on your blood vessels, heart, and lungs can have an increasing impact on your activities of day-to-day life.¹

Although there is no cure for PAH, there are therapies available that have been specifically developed to help treat the disease. These therapies may help improve some of your symptoms.¹ Working with your doctor to develop a treatment plan is a very important part of managing your PAH.
Chemical imbalances and PAH

There are 3 chemicals that are currently known to be out of balance in patients with PAH: endothelin (en-doe-THEE-linn), nitric oxide, and prostacyclin (pros-tuh-SY-klin). In patients with too much or not enough of these chemicals, the health and function of blood vessels are affected.²

**Endothelin**

En do the le lin

Endothelin causes the blood vessels in the lungs to tighten or narrow. In many patients with PAH, there is too much endothelin in the bloodstream, and the body cannot process it quickly enough. This leaves the pulmonary arteries in a constant narrow state that does not allow for normal blood flow or circulation of oxygen.³

**Nitric Oxide**

Ni tr ic ox ide

Nitric oxide is a gas that is also present in the bloodstream. Nitric oxide promotes the widening of blood vessels, and prevents blood cells from sticking together and forming clots. In many patients with PAH, the body produces less nitric oxide, which causes the blood vessels to tighten. This can lead to stiffening and possible blockage of the arteries.³,⁴

**Prostacyclin**

Pr os ta cy clin

Prostacyclin promotes the widening of blood vessels and increases blood flow by relaxing the muscle cells of the vessels. Some patients with PAH don’t produce enough prostacyclin, which can reduce the ability of their blood vessels to relax and widen.³,⁴
How is PAH classified?

Your PAH status is defined by the symptoms you have and how they impact your ability to engage in daily activities. The symptoms have been organized by the New York Heart Association into 4 levels that define your “Functional Class.”

These Functional Classes are provided to help doctors quickly understand how PAH is affecting your ability to perform daily activities.

<table>
<thead>
<tr>
<th>Functional Class</th>
<th>Description</th>
<th>Activity example</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>No symptoms with ordinary physical activity. Activity example: Walking</td>
<td></td>
</tr>
<tr>
<td>II</td>
<td>Symptoms with ordinary physical activity. Slight limitation of physical activity. Activity example: Walking</td>
<td></td>
</tr>
<tr>
<td>III</td>
<td>Symptoms with less than ordinary physical activity. Significant limitation of physical activity. Activity example: Putting away dishes or folding laundry</td>
<td></td>
</tr>
<tr>
<td>IV</td>
<td>Symptoms with any ordinary physical activity or even at rest. Activity example: Resting or sitting in a chair</td>
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</tbody>
</table>

The lower the Functional Class number, the less serious the symptoms.
Changes in your PAH status

Over the course of time, here’s how your doctor may determine if your PAH is changing or getting worse:

- Change in Functional Class—if your PAH is affecting your daily life more than before, your status may be reclassified to a higher level (for example, from Functional Class II to Functional Class III)
- Hospitalization is required because of your PAH
- Shorter distance is achieved during the 6-minute walk test
- Signs or symptoms of right heart failure
- The need for an “intervention,” such as a surgical or invasive medical procedure
Your healthcare team

Working with your physician on your treatment strategy

Several types of treatments have been developed to help address the imbalances of the naturally occurring chemicals in the body—endothelin, prostacyclin, and nitric oxide—described on page 3.¹

Each patient is different. You and your doctor will work together to plan your therapy and decide what treatment options are best for you, which may include changing or adding to your current treatment. This is not always a sign that your PAH is advancing—it can be part of optimizing your therapy.⁷

If you have other ongoing conditions like depression or diabetes, you may be more likely to be hospitalized.⁸ Remaining healthy enough so you can avoid a trip to the hospital is a specific objective that you should consider discussing with your doctor.

Having an open dialogue about your goals will help your physician plan the treatment strategy that’s best for you.
Help from other experts

You may work with a number of medical experts and supportive care personnel to help manage your PAH. These can include a heart doctor (cardiologist), a lung doctor (pulmonologist), a nutritionist, and possibly a social worker. Each plays a unique role in helping to address your physical, emotional, and practical needs.
Preparing to meet with your doctors

Educate yourself about PAH in advance. One instructive source for patient information is the Pulmonary Hypertension Association website: www.phassociation.org.

Be prepared with health insurance information and records of your medical history.

Write down questions in advance and take notes on your doctors’ answers. Don’t be afraid to ask additional questions about things you don’t understand. Some potential topics:

1. Based on our visit, what is my current Functional Class?
2. What are the recommended therapies for patients like me?
3. Is there any additional information my family or caregiver should know about my illness?
Additional resources

To learn more about PAH, visit the following organizations online.

Pulmonary Hypertension Association
www.phassociation.org
1-301-565-3004

Scleroderma Foundation
www.scleroderma.org
1-800-722-4673
NOTES
Use this space to record notes about PAH, your treatment goals and plan, and any questions you have for your healthcare team.
REFERENCES
