

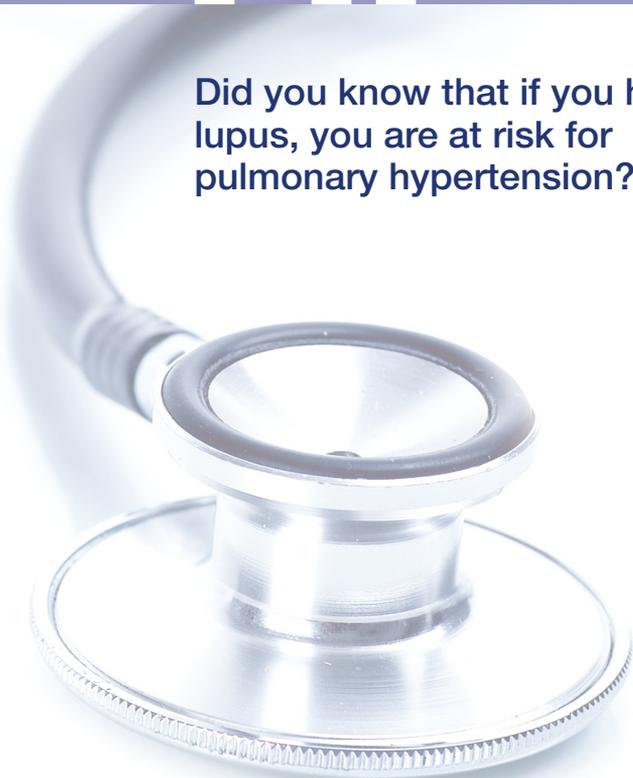
# Lupus

# &

# PH

Pulmonary  
Hypertension

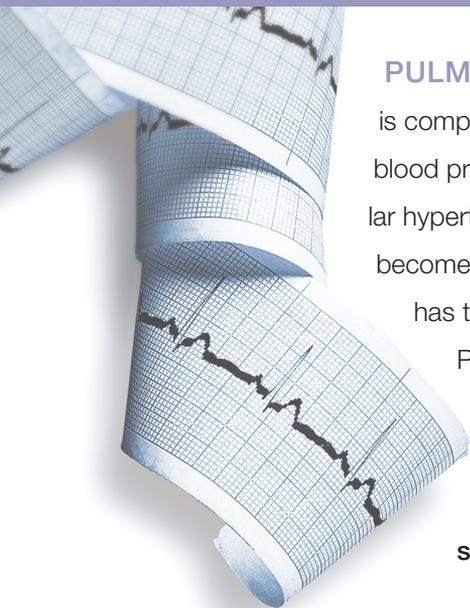
Did you know that if you have  
lupus, you are at risk for  
pulmonary hypertension?



**Pulmonary Hypertension Association**  
Empowered by hope

[www.PHAssociation.org](http://www.PHAssociation.org)

## About Pulmonary Hypertension



### PULMONARY HYPERTENSION, OR PH,

is complex and often misunderstood. PH means high blood pressure in the lungs. PH is different from regular hypertension. In PH, the blood vessels in the lungs become damaged and/or narrowed and the heart has to work harder to pump blood through them.

PH can be caused by many different problems such as heart failure, diseases that damage the lung or multiple hardened blood clots, to name a few. **It is important to understand that not all PH is the same.**

PH affects people of all ages and ethnic backgrounds. The most common symptoms are shortness of breath with physical activity, fatigue, lightheadedness and sometimes fainting. Because these symptoms can be caused by any number of other medical problems, diagnosis is often delayed. Identifying a case of PH can be difficult and may require a specialist. Once the type of PH is diagnosed, however, treatment can begin immediately.

One form of PH is called pulmonary arterial hypertension (PAH). PAH is a complex, progressive type of PH where the high blood pressure in the lungs occurs because tiny blood vessels that

carry blood through the lungs (pulmonary arteries) are narrowed, thickened and stiff. As PAH advances, the heart may lose its ability to pump enough blood through the lungs to meet the needs of the body.

There are several types of PAH. Idiopathic PAH (IPAH) means that the patient develops PAH without any other obvious medical problem leading to high blood pressures in the lung. Heritable PAH (HPAH) comes from abnormal genes that cause PAH. Heritable PAH may be passed on to some members of your family. PAH can be associated with other medical conditions such as connective tissue diseases (scleroderma and

lupus for example), chronic liver disease, congenital heart disease, or HIV infection. Finally PAH can be associated with past or present drug use, such as methamphetamines or certain diet pills. It is not known exactly how these medical problems or drugs cause PAH.

PAH is a serious condition, and without treatment, symptoms can only become worse, leading to heart failure and even death. Proper diagnosis and therapy from a doctor who understands PAH is essential. Every patient is different. The choice of treatment is based on how sick a patient is and the risks and benefits of any particular therapy. Regardless of risk, it is important that patients and their healthcare providers engage in frequent follow-ups with ongoing discussions about the management of their condition. Current guidelines suggest that changes in therapy should be considered

for patients not reaching their treatment goals.

While no cure has yet been found for PAH, increased research has resulted in treatments that allow patients to live longer, fuller lives with far less interference from the disease. Even more promising research is being conducted every day that is not only advancing our understanding of the PAH disease but also potentially identifying new treatment options for patients in the future.



## Lupus-Associated PH

### What do I need to know about lupus-associated PH?

First of all, it is essential to know that PH and lupus are two separate conditions and that PH does not cause lupus. However, some lupus patients develop PH. As was discussed in the introduction, there are a number of reasons why blood pressure in the lung may become elevated — almost all of which can occur in lupus. For this reason, it is very important that patients with lupus and suspected PH be looked at

very carefully to understand exactly what might be contributing to PH.

Lupus-associated PH may be due to more than just one problem. Other causes of PH include left-heart dysfunction, vasculitis (an inflammation or irritation of small blood vessels in the lung), pulmonary embolism (blood clots in the lungs) or pulmonary parenchymal disease (scarring or irritation of the area surrounding the air sacs and blood vessels in

## Lupus-Associated PH (cont.)

the lungs). Each of these situations would be treated differently. Since this is much more complex than PAH alone, receiving care from a physician who is experienced in the management of lupus and PAH is important.

One of the most serious kinds of PH is PAH. This remains a relatively uncommon complication of lupus, but because PAH can be very problematic, it is important to find and treat this as soon as possible. Current estimates suggest that somewhere between 0.5 and 9 percent of patients with lupus may have PAH.

Lupus remains a very complex process that can affect one or many organs. In particular, lupus can cause kidney, heart or lung dysfunction in some people, which can lead to shortness of breath, fatigue or fluid retention — all of which can also occur in PAH. These additional factors particular to lupus can complicate management of PAH and may require additional testing to ensure the best treatment of lupus-associated PAH.

### **Why do PH and lupus seem to occasionally occur together?**

This is another question that medical science is searching to answer. Medical researchers have not yet been able to say for sure how lupus leads to the development of PAH in some patients. It is not yet known, for example, whether lupus itself can directly cause PH, or whether lupus is simply a trigger for the develop-

ment of PAH in susceptible individuals. Lupus remains a complicated disease that can cause many problems that weaken blood vessels, and these, too, might play into the development of PAH.

### **Who is most susceptible to a lupus-associated PAH?**

Lupus-associated PAH occurs more frequently in females and in a slightly younger age group than idiopathic PAH (IPAH) or scleroderma-associated PAH. There also appear to be racial differences in lupus-associated PAH with increased frequency of non-Caucasian patients compared to IPAH or scleroderma-associated PAH. There are no blood tests able to accurately predict if a lupus patient has PAH or might develop it.

### **Should a lupus patient be tested for PH?**

Lupus patients know that medical supervision is critical to their best possible long-term health and that any change in their condition should be brought to the attention of their doctors. Changes that may have to do with PH might include difficulty when attempting physical activity; swelling of the feet, ankles, legs or abdomen; increased shortness of breath; worsening fatigue; chest discomfort or pain; light-headedness or fainting. These changes should be communicated to your doctor promptly.

### **How is testing for PH carried out?**

A visit to your doctor and a thorough

exam is a good first step for evaluation of potential PH. An echocardiogram is a very useful test that looks at the size and functions of the heart among other things and can suggest whether PH may exist. Unfortunately, it alone is not accurate enough to make the diagnosis of lupus-associated PAH. A test known as right-heart catheterization remains the most accurate way of diagnosing lupus-associated PAH. Additional testing is also typically done to see how much exercise you can do and to check lung function or x-rays to be sure that additional problems in the lungs are not present.

### **Can PH be treated the same way in lupus patients as in those without lupus?**

Through the years, a variety of medications have been shown to slow the damage and relieve the symptoms caused by PAH. These medications are very complex, and choosing the correct initial treatment requires the knowledge and expertise of a physician who specializes in PAH. Professionals at PH centers can help identify PH and choose the correct type of treatment.

In addition to medications approved for the treatment of PAH, patients may benefit from simple basic measures including attention to fluid balance and therapies that improve the efficiency of the heart. These may include supplemental oxygen or the medication digoxin, as well as anticoagulants (blood thinners).

Once a doctor and patient have been able to establish an effective medical approach, many patients show significant improvement, often in conjunction with doctor-prescribed cardiopulmonary rehabilitation training (CPRT).

A few patients with lupus-associated PH may also benefit from a group of drugs called immunosuppressive medications. These medications treat irritation or inflammation in the body, which can occur in lupus and occasionally can cause additional damage in the blood vessels of the lungs. It is difficult to know in all cases which patient may benefit from these kinds of medications.

### **Does having lupus make PH worse?**

The answer is yes and no. In general, patients who have lupus-associated PAH tend to respond to therapy similarly to IPAH patients who do not have lupus. As was noted above, however, patients with lupus may have other problems such as left-heart dysfunction, blood clots in the lungs, inflammation of the lungs or its blood vessels. All of these could make PH worse for that person.

## PHA Resources



The Pulmonary Hypertension Association (PHA) was founded by and for PH patients. The organization has led the way in bringing pulmonary hypertension into the national and international consciousness. PHA is constantly increasing its services to the medical community through educational programming, membership sections for medical professionals, and much more:

### Website:

PHA's website is a comprehensive source of information for patients, caregivers and medical professionals. Please visit us at [www.PHAssociation.org](http://www.PHAssociation.org).

### Find a Doctor:

The "Find a Doctor" section of PHA's website allows patients and referring physicians to search for PH-treating physicians by state at: [www.PHAssociation.org/Patients/FindADoctor](http://www.PHAssociation.org/Patients/FindADoctor). While PHA does not endorse any of these physicians, PHA strongly recommends that all PH patients see a PH specialist who will be able to provide them with the best PH care.

### *Pulmonary Hypertension: A Patient's Survival Guide:*

This extraordinary 300+ page book was written by a patient and is medically reviewed and updated annually. It presents the illness in a very human and readable way, covering a wealth of topics like the mechanics of PH, the latest treatments, patient care and lifestyle issues. PHA members receive a discount on this resource. The *Survival Guide* is available for purchase as a paperback and an e-book at: [www.PHAssociation.org/SurvivalGuide](http://www.PHAssociation.org/SurvivalGuide).

### Online information about PH:

For information on PH diagnosis, symptoms, treatments and more, visit [www.PHAssociation.org/Patients/AboutPH](http://www.PHAssociation.org/Patients/AboutPH).

### Support Groups:

From the first support group started in 1990 around a kitchen table in Florida, PHA grew to 45 groups in 2001 and to more than 245 in 2013. In many places, patients have the opportunity to meet, learn from and find common understanding with others in similar circumstances. Find a support group in your area at [www.PHAssociation.org/LocalSupportGroups](http://www.PHAssociation.org/LocalSupportGroups).

## About the Pulmonary Hypertension Association



The mission of the Pulmonary Hypertension Association is to find ways to prevent and cure pulmonary hypertension, and to provide hope for the pulmonary hypertension community through support, education, research, advocacy and awareness. PHA's members form a community that is fighting back against this terrible illness.

### PHA fulfills its mission through:

- Funding for research
- Quarterly medical journal *Advances in Pulmonary Hypertension*
- *PHA Online University* offering free CME credits and the latest information on pulmonary hypertension ([www.PHAOnlineUniv.org](http://www.PHAOnlineUniv.org))
- Professional membership sections:
  - ▶ PH Clinicians and Researchers (PHCR)—for physicians and doctorate-level researchers
  - ▶ PH Professional Network—for nurses and allied health professionals
- Educational conferences and materials for medical professionals and patients
- A wealth of information in the *Survival Guide*
- PH patient support groups
- Quarterly newsletter *Pathlight*
- Advocacy and awareness campaigns
- Toll-free Patient-to-Patient Support Line (1-800-748-7274)
- PHA website with PH discussion boards, email groups and online support chats ([www.PHAssociation.org/ConnectOnline](http://www.PHAssociation.org/ConnectOnline))

### More Information on Lupus

Lupus Foundation of America:  
[www.lupus.org](http://www.lupus.org)

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