

What Is Hypersensitivity Pneumonitis?

Hypersensitivity pneumonitis (HP) is a lung condition in which your immune system reacts to certain substances in the air you breathe. Repeated inhalation of these substances causes inflammation from the lung's immune reaction, called "pneumonitis." Because this inflammation due to HP can resemble pneumonia from an infection, it may take a long time to arrive at the correct diagnosis. Sometimes this inflammation will resolve, but other times, it can lead to scar formation in the lung and problems with absorbing oxygen into your body. Anyone can be affected, but some people are more at risk than others. This fact sheet addresses diagnosis of HP. For information about treatment, see the ATS Patient Information Series fact sheet "Treatment of Hypersensitivity Pneumonitis."



Image: Freepik.com

While HP is much less common than asthma or pneumonia, the exposures known to cause it occur commonly in the air we breathe. These include high levels of mold spores, dust from pet birds, down feathers, and contaminated water aerosols from indoor hot tubs and humidifiers.

Who is at risk for developing HP?

The following occupations, activities, and substances are associated with HP following prolonged exposure.

Farming and grain processing and use

Inhalation of molds and bacteria commonly found in hay, grains, and animal feeds. The risk seems to be highest when these substances are stored in airtight conditions without first being dried, resulting in bacteria and mold contamination that can be inhaled during baling or unloading.

Indoor air and water contamination

Inhalation of molds and bacteria found in hot, humid conditions, such as hot tubs, whirlpools, humidifiers, dehumidifiers, or areas of water damage.

Birds and dusts from other animals

- Inhalation of airborne dusts generated from birds (mainly their feathers and droppings). This includes pet birds, such as pigeons, parakeets, and canaries, as well as those raised on family or corporate farms (chickens, turkeys). Workers who pluck feathers are also at risk.
- Bedding and other furnishings that contain bird feathers or down products.
- Veterinarians, or others who work with animals, who come into contact with animal droppings, body fluids, or feathers/fur, may also be at risk.

Chemicals, dusts, and other substances used in industrial manufacturing

- Working with certain chemicals has been associated with risk for HP, for example, isocyanates contained in two-part paints and polyurethanes.
- Machine operators who are exposed to machining aerosols or metalworking fluid that contain bacteria may be at risk.
- Lumber workers exposed to moldy wood bark that is being milled may be at risk.

Less common exposures

- Musical instruments (trombones, saxophones) that become contaminated with microorganisms.
- Workers exposed to dusts from oyster shells.

Why do some people get HP, and others do not?

You may develop HP even though your family, friends, or co-workers who share the same environment remain healthy. The reason why some but not other people develop HP is not known, but doctors think that it may have to do with the following:

- Some people inherit genes from their parents that may make them more sensitive to respiratory exposures that can cause HP. This is similar to what happens in the case of allergies.
- The length of time people are exposed and the intensity of exposure to substances that cause HP may be associated with risk for disease.
- Smoking status, viral infections and other unknown factors may play a role in risk for HP, but more research needs to be done to fully understand this.

What are common symptoms of HP?

Symptoms may occur suddenly and be quite noticeable (acute) while others may be more subtle and the onset harder to recognize (chronic). Acute symptoms tend to occur after shorter, higher exposures to substances that cause HP, while chronic symptoms are more often associated with repeated, lower exposures. Symptoms are nonspecific and may include:

- Shortness of breath, especially with activity
 - Frequent dry cough
 - Weight loss
 - Unusual fatigue
 - Recurrent flu-like symptoms of fevers, chills, and sweats
- Sometimes symptoms improve when you remove yourself from the environment in which the exposure is occurring (for example, going on vacation or taking a leave from work). In some people with the chronic form of HP, symptoms do not improve with avoiding a particular environment.

How is HP diagnosed?

Diagnosing HP can be challenging, because it requires that doctors consider the possibility of HP even when symptoms and medical

tests of lung function and chest imaging resemble other conditions, and may include:

Medical History

When your doctor asks about your health history, be sure to talk about:

- Current and former occupations, including use of certain chemicals and exposures to dusts known to cause HP.
- Current and former hobbies and recreational activities (both in and outside your home).
- Use of hot tubs, humidifiers or swamp coolers.
- The age, location, and condition of your home.
- Past episodes of water damage in your home.
- If you are frequently exposed to birds. Most pets do not increase the risk of HP, but birds do.
- Medications you take now or took in the past.
- Family history of HP or other form of pulmonary fibrosis.

Physical Exam

Your doctor will perform a physical exam to evaluate your lungs, neck, heart, and skin to help determine what might be causing the problems in your lungs.

Blood Tests

If the diagnosis of HP is uncertain, blood tests may be helpful. Special antibody blood tests to look for evidence that your immune system has reacted to an HP exposure may be done, but these are often negative in patients with HP.

Pulmonary Function Tests (PFTs)

- HP can cause your lungs to become inflamed and scarred. PFTs are breathing tests your doctor may use to evaluate your lungs for inflammation and scarring by measuring how fast air moves in and out of your lungs, how deep a breath you can take, and how much oxygen in your lungs is absorbed in your bloodstream. Also, your doctor will probably ask you to repeat PFTs at regular intervals to see if your lung disease is changing over time, and to monitor your response to treatment.
- Tests to check your oxygen levels, especially with walking or any kind of exertion, can be helpful to monitor lung inflammation and scarring from HP and to determine whether you need to use supplemental oxygen.

High-resolution Computed Tomography (HRCT)

HRCT, or thin slice chest CT scans, are a special type of lung imaging where your doctors can get detailed pictures of your lungs, heart, blood vessels, and lymph nodes. This is done to look for signs of infection, inflammation, and scarring. Chest HRCT scans are also useful to check your response to treatment and to monitor the status of your HP lung disease.

Bronchoscopy and Lung Biopsy

Bronchoscopy is a test where your doctor passes a tiny camera at the end of a long, flexible tube through your mouth or nose and into your lungs. This is done to check the fluid in your lungs for infection and inflammation, and may also be used to perform a lung biopsy. A lung biopsy is when doctors retrieve a small piece of lung tissue for evaluation under a microscope, which can be an important step in evaluating your lungs for HP. In some cases, a chest surgery may be required to biopsy the lung, but this decision will be made with you and your doctor, and will only be done if necessary to make a diagnosis.

How will HP affect my health?

Most cases of acute HP will get better if recognized early. Some people will have permanent changes, and others may develop a chronic form of lung disease with progressive scarring. Some

forms of HP require treatment in addition to getting away from the triggering agent. For more on treatment, see ATS Patient Information Series fact sheet "Treatment of Hypersensitivity Pneumonitis."

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Rx Action Steps

- ✓ Consult with your doctor if you are concerned you may have HP and to see if you need the help of an environmental/occupational specialist to remove harmful substances from your home or workplace.
- ✓ If you do have HP, take steps to avoid all possible exposure to what you are sensitized to.
- ✓ If you are diagnosed with bird-related HP:
 - You have to avoid exposure to birds.
 - You need to eliminate any remnant bird dust from your home.
 - You cannot keep pet birds in the house (moving a bird to a separate room will not eliminate your exposure).
- ✓ If diagnosed with HP from a workplace exposure, talk with your healthcare provider or HP specialist about:
 - How to avoid further exposure to causative/suspected substances at work.
 - Whether you need to file for worker's compensation benefits.
- ✓ If you have HP from any cause, the following are also essential in addition to avoiding ongoing exposure to the substance that caused your HP:
 - Having regular medical follow up and monitoring of lung function and need for medication.
 - Getting recommended immunizations such as yearly flu vaccine.
 - Stopping smoking and vaping and avoiding exposure.
 - Using oxygen if prescribed.

Healthcare Provider's Contact Number:

For More Information

American Thoracic Society

- <https://www.thoracic.org/patients>

American Lung Association

- <https://www.lung.org/lung-health-and-diseases/lung-disease-lookup/hypersensitivity-pneumonitis/>

CHEST Foundation

- <https://foundation.chestnet.org/lung-health-a-z/hypersensitivity-pneumonitis/>

HPLung

- <https://www.hplung.com>

National Institutes of Health

- <https://www.nhlbi.nih.gov/health-topics/hypersensitivity-pneumonitis>

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Treatment of Hypersensitivity Pneumonitis (HP)



Hypersensitivity pneumonitis (HP) is a lung condition in which your immune system reacts to certain substances in the air you breathe. This fact sheet addresses treatment of HP. For more information about HP, see the ATS Patient Information Series fact sheet “What is Hypersensitivity Pneumonitis” at www.thoracic.org/patients.

First-line treatments

There are both non-medication and medication-based treatments for HP. Overall, the most important treatment is avoiding further inhalation of the particles causing your lung disease.

Please refer to Part 1 of this series for more information on specific causes and diagnosis of HP at www.thoracic.org/patients.

Antigen avoidance

Hypersensitivity pneumonitis (HP) occurs when you breathe in proteins from molds, bird feathers, and other things that cause allergic reactions in your lungs. Breathing in these particles over time can lead to inflammation and permanent scarring in your lungs. The first and most important step in treating HP is to work with your healthcare team to identify what you are inhaling that is causing the problem. This can be challenging and about half the time, it is not possible to identify the problem. If you do find out what is causing it, getting your environment cleaned and staying away from the particles injuring your lungs is the most important thing you can do. Sometimes this may require you to hire a professional cleaner to clean/fix in your home or workplace.

More information can be found on mold-related problems in the ATS Patient Information Series on Mold-Specific Concerns Associated with Water Damage for Those with Allergies and Other Lung Diseases.

Medications

Sometimes the lung injury caused by HP will continue to worsen even after cleaning and removing the inhaled dusts from your environment. When this is the case, your healthcare provider may give you specific medications to help reduce the amount of inflammation in your lungs. The most common type of medications lung specialists use to reduce the inflammation caused by HP are called steroids, such as prednisone.

Other medications your healthcare team may use

include (but are not limited to) mycophenolate mofetile, azathioprine, or rituximab. These medications are used on a case-by-case basis depending on your body's response to steroids or the development of steroid side effects. There is some evidence to suggest that patients who may benefit the most from these medications are those with signs of significant inflammation on chest imaging.

If your lung scarring continues to worsen despite these medications, there are two medications approved to help slow down decline in lung function. These medications are nintedanib and perflinone, and you should talk to your healthcare team about whether they are right for you.

Supplemental oxygen

The lung inflammation and scarring caused by HP can sometimes make it difficult to get enough oxygen into your body. People with HP should have their oxygen levels routinely evaluated with a device used to detect oxygen levels in your blood, called a “pulse oximeter.” The prescription of extra oxygen (“supplemental oxygen”) is required when oxygen levels fall below a certain level in your blood. Your healthcare team may recommend that you have oxygen that you wear all day or just at night. Over time, the amount of extra oxygen you need may increase or decrease depending on how your body responds to other treatments. There are a number of devices that can help provide oxygen to you, and more specific information can be found in the ATS Patient Information Series fact sheet on Oxygen Therapy.

For more information on pulse oximetry and ways in which you can safely monitor your oxygen levels at home, please refer to the ATS Patient Information Series on Pulse Oximetry.

Pulmonary rehabilitation

If you feel very short of breath and are not able to do all your activities at home, your healthcare team may recommend a pulmonary rehabilitation program. This is typically an outpatient, exercise and education-

based program that can help you to feel less short of breath and improve your ability to do physical activity. It includes physical exercises and related education that are individualized to you. Education on medications, nutrition, smoking cessation, and counseling on oxygen are also important parts of the program. You will work with a pulmonary rehab specialist 1 to 2 times per week for an average of 4 to 12 weeks, followed by at-home exercises you can do on your own.

The American Thoracic Society has partnered with the COPD Foundation and the Gawlicki Foundation to create a public awareness website for pulmonary rehabilitation located at www.livebetter.org and <https://www.copdfoundation.org/Learn-More/Pulmonary-Rehabilitation/What-is-Pulmonary-Rehabilitation.aspx>. Additionally, more information on pulmonary rehabilitation, including a brief ATS Video on Pulmonary Rehabilitation, can be found at www.thoracic.org/patients.

Clinical trials

There are many scientists who are studying new types of treatment for HP and other similar forms of lung disease. You should ask your healthcare team if they are aware of any ongoing research for treatment of HP and whether any clinical trials are right for you.

Please visit www.clinicaltrials.gov for more information related to new treatments and active research in the field of HP.

Lung transplant

Some people with severe, end-stage lung disease from HP may qualify for a lung transplant. Lung transplantation is a procedure where healthy lung(s) are donated to a person with end-stage lung disease. These surgeries are performed by specialized surgeons and managed by transplant pulmonologists, which are specialized lung doctors who evaluate candidates for transplant and who help keep your new lungs healthy after transplant. Not all hospitals perform lung transplantation, and lung transplant programs are typically located at highly specialized centers throughout the United States. If needed, your healthcare team will refer you to a lung transplant program where a group of doctors will perform extensive testing to determine whether you are a good candidate for a lung transplant.

For more in-depth information, the American Thoracic Society has an extensive Patient Education and Information Series on lung transplantation for both adults and children, which can be found at the following web address: www.thoracic.org/patients.

Palliative care

Unfortunately, end-stage lung disease from HP can lead to chronic and severe breathlessness, cough, pain, and other symptoms. Palliative care is a team of healthcare providers who specialize in helping improve patients' quality of life when they experience a significant symptom burden from life-threatening and end-stage conditions. Palliative care

specialists can also help patients with spiritual, emotional, and psychological well-being. Palliative care can begin at any stage of your illness and in virtually any healthcare setting (your home, clinic, rehabilitation facility, or even during hospitalization). It has been shown that early involvement of a palliative care team can actually help you to live longer with a terminal condition.

Hospice care is different than palliative care in that hospice providers typically become involved at the very end of life to when life-prolonging treatment is failing. Hospice specialists are experts in relieving suffering at the very end of life, typically when there is less than 6 months to live.

For more information, please see the ATS Patient Information series on Palliative Care for People with Respiratory Disease or Critical Illness.

Action Steps:

- ✓ Be sure to speak with your healthcare provider about help with antigen avoidance
- ✓ Ask your healthcare team if there are any medications needed for your lung disease or if you would benefit from enrollment in a clinical trial
- ✓ Ask your healthcare provider if pulmonary rehabilitation is right for you
- ✓ Ask if there are lung transplant doctors where you live and if they have any suggestions to help ensure you are a good candidate for transplant
- ✓ Ask your healthcare team if they believe you may benefit from being evaluated by a palliative care team

Healthcare Provider's Contact Number:

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Resources:

American Thoracic Society

- www.thoracic.org/patients

ATS Livebetter Website

- <http://www.livebetter.org>

<http://www.clinicaltrials.gov>

National Institutes of Health

- <https://www.nlm.nih.gov/health-topics/hypersensitivity-pneumonitis>

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