

What Is Allergic Bronchopulmonary Aspergillosis (ABPA)?

Allergic bronchopulmonary aspergillosis (called ABPA for short) is a problem in the lungs that is not very common. It is caused by a severe allergic reaction after being exposed to a type of fungus called *Aspergillus*.



What is *Aspergillus*?

Aspergillus is a type of fungus (also referred to as a mold), that is commonly found in the environment. It can be found in the soil, dust, water, and rotting or decaying vegetation (like dead leaves or compost piles), marijuana, and some foods and ground spices. The fungus forms into spores which are very small particles that can float in the air. Most people inhale its spores from the air, but do not have any problems from being exposed. In most cases, the fungus can live in the mucus in the breathing tubes (airways). This is called *colonization*. Having *Aspergillus* in your airways does not mean you have or will have a sudden (acute), and serious infection by this fungus. Our immune system usually helps protect the body from infections like these. In some people who have a weakened immune system, *Aspergillus* can get into the lungs, causing an acute infection. (For more about *Aspergillus* infection, see the ATS Patient Information Series fact sheet: *Aspergillosis* in the Fungal Disease Series). However, in a few cases, this exposure to *Aspergillus* triggers an allergic immune response without invading the tissues that leads to ABPA (allergic bronchopulmonary aspergillosis).

What is ABPA?

Allergic bronchopulmonary aspergillosis is a form of lung disease that occurs in some people who are allergic to *Aspergillus*. With ABPA, this allergic reaction causes the immune system to overreact to *Aspergillus* leading to lung inflammation. ABPA causes bronchospasm (tightening of airway muscles) and mucus buildup resulting in coughing, breathing difficulty and airway obstruction.

Some people with ABPA will develop bronchiectasis, a form of airway damage that can result in worse lung function and increased risk of infection. Bronchiectasis can also occur in people who have lung diseases that cause chronic inflammation (swelling) in the lungs, such as chronic infections, asthma, or cystic fibrosis. (For more information on bronchiectasis, go to <http://www.nhlbi.nih.gov/health/health-topics/topics/brn/>)

Common signs and symptoms of ABPA

A person with ABPA will have some or all of the following symptoms:

- Coughing frequently
- Coughing up mucus plugs that may be brown in color. You may also cough up blood (called hemoptysis).
- Difficulty exercising
- Wheezing
- Shortness of breath or feeling like it is difficult to get air into or out of your lungs.
- Chest pain or tightness
- Fever that goes away then comes back
- Fatigue

ABPA with asthma

ABPA can be a rare cause of poorly controlled asthma, occurring in less than 1% of asthma patients. A person with asthma who develops ABPA has difficulty controlling their asthma despite using many medications. Because ABPA can be treated, it is sometimes considered in cases of poor asthma control.

ABPA with cystic fibrosis (CF)

Cystic fibrosis is a genetic disease that can affect the lungs. ABPA can be seen in 1-15% of people with CF. ABPA is considered when a person with CF has worsening symptoms and lung function not responding to other CF therapy.

How do I know if I have ABPA?

Your health care provider can find out if you have ABPA. There is no individual test to diagnose ABPA. The diagnosis is made based on your symptoms and results of a set of tests including lung function and allergy tests. As described below, diagnosing ABPA can be difficult.

Blood tests are used to look for signs of an allergic reaction. This includes evaluating your immunoglobulin E (IgE) level. This level is increased with any type of allergy. Many people with asthma have higher than normal IgE levels. In ABPA however, the IgE level is extremely high (more than 1000 ng/ml or 417 IU/ml). IgE levels can also be used to help monitor control of your ABPA. In addition to total

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IgE, all patients with ABPA have high levels of IgE that is specific to *Aspergillus*. A blood test can be done to measure specific IgE to *Aspergillus*. There are some other blood tests that also can help with the diagnosis. Your health care provider can tell you what other tests you may need.

A blood or skin test for IgE antibodies to *Aspergillus* can be done to see if a person is sensitized (allergic) to this fungus. If these skin tests are negative (i.e. does not show a skin reaction) to *Aspergillus fumigatus*, the person *usually* does not have ABPA.

A sputum culture (mucus coughed up from the airway) can be done to see if *Aspergillus* is growing in the airway but is not always reliable. Many people will have *Aspergillus* in their airway secretions (sputum) but not have ABPA. On the other hand, even if the culture is negative, a person can still have ABPA.

Chest X-rays are often not helpful in diagnosing ABPA. A CT scan (computed tomography) however, is a more detailed view of your lungs and therefore can be very helpful. The bronchiectasis is kind of a curve to the reader.

Lung function testing (spirometry) is used to see how severe your lung problem is and to find out how well you are responding to the treatment. (To learn more about spirometry, see the ATS Patient Information Series fact sheet: "*Pulmonary Function Tests*").

How is ABPA treated?

Treatment of ABPA aims to control inflammation and prevent further injury to your lungs.

ABPA is usually treated with a combination of oral corticosteroids and anti-fungal medications.

The corticosteroid (steroid medicine) is used to treat inflammation and blocks the allergic reaction. Examples of corticosteroids include: prednisone, prednisolone or methylprednisolone. Inhaled corticosteroids alone—such as used for asthma treatment—are not effective in treating ABPA. Usually treatment with an oral corticosteroid is needed for months. You should discuss with your health care provider about what possible side effects may occur with oral corticosteroids and how they can be managed.

The second type of therapy used is an anti-fungal medication, like itraconazole (brand name Sporanox®) and voriconazole (brand name VFEND®). These medicines help kill *Aspergillus* so that it no longer colonizes the airway. Usually one of these drugs is given for at least 3 to 6 months. However, even this treatment is not curative (permanently cure you of ABPA) and can have side effects. You will hence be monitored for signs of side effects such as liver damage. Be certain to keep appointments for your blood work to

make sure this monitoring can take place.

Often a person improves with therapy and the illness will go into what is called *remission*. During remission, you will have few if any signs of ABPA. However, repeated flare ups of ABPA can occur that require more treatment. In addition to monitoring you for improvement in symptoms, a combination of X-ray or CT findings, lung function and blood tests are used to help judge if the treatment is working. The IgE level usually gets lower as the allergic reaction is controlled. Your health care provider can help you decide how long treatment is needed, tell you about symptoms of side effects from medicines and a plan for follow-up visits.

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Rx Action Plan

Discuss with your health care provider if the following situations arise.

- ✓ If you cough frequently and bring up "plugs" of brown mucus or blood, see your health care provider.
- ✓ If you or your health care provider think that you may have ABPA, ask to see a specialist who manages ABPA.
- ✓ Take all doses of medicine prescribed to treat ABPA.
- ✓ Continue treatment to control any other lung problem you have (such as asthma or CF).
- ✓ Talk with your health care provider if you are having increased symptoms or any concern about side-effects from the medicines.

Provider's Office Telephone:

For additional information:

The Fungal Research Trust

<http://www.aspergillus.org.uk/abpaframeset.html>

The Merck Manual Home Health Handbook for Patients and Caregivers

http://www.merckmanuals.com/home/lung_and_airway_disorders/asthma/allergic_bronchopulmonary_aspergilliosis.html?qt=&sc=&alt=

Cystic Fibrosis Foundation—Allergic Bronchopulmonary Aspergilliosis

<http://www.cff.org/livingwithcf/stayinghealthy/germs/abpa/>

Centers for Disease Control and Prevention—Aspergilliosis (Aspergillus)

<http://www.cdc.gov/fungal/aspergilliosis/risk-prevention.html>