

## UW HEALTH - ONLINE HEALTH FACT

### Allergic Bronchopulmonary Aspergillosis in Cystic Fibrosis (ABPA)

#### What is ABPA?

*Aspergillus species* is a fungus that often lives in the airways of adolescents and young adults with cystic fibrosis (CF). When people develop an allergic reaction to aspergillus, it is called Allergic BronchoPulmonary Aspergillosis. ABPA affects about 2-11% of the people with CF. It causes inflammation in the lungs and, if untreated, can cause further scarring and fibrosis.

#### What are the symptoms?

**While the symptoms of ABPA are similar to CF symptoms, your health often gets worse without a reason.** This includes:

- Coughing
- Wheezing
- Unable to exercise
- Exercise-induced asthma
- Decline in pulmonary function
- Increased sputum

#### How is it diagnosed?

The diagnosis of ABPA in CF is difficult. It may often take longer because many of the criteria used to diagnose ABPA are the same as the usual symptoms of CF. Below is a list of the criteria.

1. Symptoms that are not related to other problems (see above)
2. Total serum IgE is greater than 500 IU/ml.
3. Immediate positive skin test to *Aspergillus species*
4. Positive laboratory test (Precipitin to *Aspergillus fumigatus* or IgG antibody against *Aspergillus fumigatus*)  
or  
Abnormal chest X-ray or change from previous exam
5. Peripheral blood eosinophilia

#### What factors are linked to with ABPA and CF?

ABPA is more common in males and adolescents. It is also common in people who:

- Have decreased lung function
- Wheeze
- Have allergies
- Have asthma
- Are positive for *Pseudomonas species*

## What is the treatment for ABPA?

Treatment for ABPA involves prevention and treatment of acute (flare up) episodes. There are two parts in the treatment of ABPA. Inflammation in the lung is treated with corticosteroids. Fungal colonization is treated with anti-fungal medicines. Although there is little proof that antifungal agents are useful in CF patients with ABPA flares, reducing the fungal burden in the respiratory tract may reduce long term risk of disease progression. Further research is needed.

### 1. Oral corticosteroid pills or liquid

Names of the medicine

- Prednisolone (Pediapred<sup>®</sup>, Prelone<sup>®</sup>)
- Prednisone tablets
- Methylprednisolone (Medrol<sup>®</sup>) tablets

The anti-inflammatory effects of corticosteroids work well in APBA. The course of medicine is much longer than the short courses used for asthma. The steroids may be used for many weeks and slowly weaned while symptoms and lung function studies are checked. Once all the symptoms have gone away, the steroids are often stopped.

Side effects of taking corticosteroids

- Increased appetite
- Weight gain
- Round moon face
- Red cheeks
- Stomach ache
- Changes in mood

The side effects of steroids are related to the amount and length of time on steroids.

### 2. Antifungal medicines

Itraconazole (Sporanox<sup>®</sup>) reduces the amount of fungus in the lungs. This may lower the chance of the disease getting worse over time.

It is used 1–2 times a day for about 3–6 months or until all symptoms have gone away. It is better absorbed when taken on an empty stomach.

If you are on an acid-reducing medicine, you should take Itraconazole with 8 oz. of a cola drink or orange juice to make sure you are able to absorb all of the medicine.

Side effects

- Stomach upset
- Rash
- Dizziness
- Fever
- Headaches
- Fatigue