



PRIMARY IMMUNODEFICIENCIES

PIDS AND RESPIRATORY DISORDERS



ABBREVIATIONS

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| COPD | Chronic obstructive pulmonary disease |
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| CT | Computed tomography |
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| MRI | Magnetic resonance imaging |
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| IG | Immunoglobulin |
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| PID | Primary immunodeficiency |
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Primary immunodeficiencies and respiratory disorders (1st edition).
December 2014

© International Patient Organisation for Primary Immunodeficiencies (IPOPI), 2014

Published by IPOPI: www.ipopi.org

INTRODUCTION

This booklet explains the respiratory issues that patients with primary immunodeficiencies may encounter and their treatment options.

Primary immunodeficiencies (PIDs) are rare diseases that occur when components of the immune system are either not present or are not functioning normally. Respiratory disorders, which affect the sinuses, throat and lungs, are common in many people with PIDs; recurring ear, nose or throat infections in children or adults are often the first sign that an individual has a PID.

If you have a PID and develop respiratory symptoms, it is important to discuss them with your physician. Early diagnosis will enable appropriate treatments to be prescribed that will improve your health and your respiratory function.

Routine checks of your respiratory system may be beneficial, depending on the type and severity of your PID, and your physician will be able to provide you with information about these tests.



RESPIRATORY DISORDERS

Respiratory disorders in patients with PIDs can be broadly classified into the following three groups. In addition, some patients may experience respiratory symptoms in response to certain treatments.

Respiratory disorders can lead to serious problems in some people with PIDs so it is important that you discuss any new symptoms with your physician as they arise.

1. INFECTIONS

Short-term (acute) and long-term (chronic) infections are usually caused by bacteria, most commonly by these types: *Haemophilus influenza*, *Streptococcus pneumonia* or *Moraxella cararhalis*. Viral infections, usually rhinovirus or enterovirus, can occur at the same time as bacterial infections. In people with more severe PIDs, fungal infections may also occur, e.g. *Pneumocystis jiroveci* or *Aspergillus fumigatus*.

Upper respiratory infections affect the sinuses and throat and typically include:

- Inflammation of the nose (rhinitis)
- Chronic or acute sinusitis
- Common cold
- Croup
- Influenza
- Sore throat (pharyngitis)
- Tonsillitis
- Swollen lymph nodes.

Lower respiratory infections affect the lungs and include:

- Pneumonia
- Chronic or acute bronchitis.

2. NON-INFECTIOUS CONDITIONS

A range of conditions that are not related to specific infections may also develop in patients with PIDs.

- Asthma, allergies and chronic coughs are more prevalent in patients with PIDs.
- Bronchial abnormalities and chronic obstructive pulmonary disease (COPD) occur in some people if infections and inflammation persist and the small airways in the lungs are not working properly.
- Pleurisy is an inflammatory condition that affects the lining surrounding the lungs.

- Chronic lung disease may develop in people who have long-term lung conditions.
- Granulomatosis may occur in a small number of patients as a result of long-term inflammation.
- Pulmonary embolism is a blockage in the pulmonary artery, which carries blood from the heart to the lungs.

3. BENIGN LYMPHOPROLIFERATIVE DISORDERS

These are non-cancerous disorders that occur when the body mistakenly produces too many lymphocytes (a type of white blood cell).

RESPIRATORY SYMPTOMS

The symptoms of many respiratory disorders will be the same in people with and without PIDs. However, the development of the disease is likely to be different in people with PIDs and they often respond differently to conventional therapies. Symptoms vary according to the type of infection, although many are common to a number of respiratory infections.

Your physician will be able to advise you on respiratory disorders that are typically associated with your specific PID and symptoms you may experience.

TYPICAL SYMPTOMS OF RESPIRATORY DISORDERS

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| • Sneezing, nasal discharge, post-nasal drip | • Fever and chills |
| • Difficulty breathing | • Muscle aches |
| • Cough | • Swollen glands |
| • Sinus pain | • Respiratory distress |
| • Headache | • Chest pain |
| • Bad breath | • Weakness and fatigue |
| • Throat irritation | • Generalised aches and discomfort |
| • Decreased appetite | |

RESPIRATORY EVALUATIONS

Depending on your PID and health status, your physician may recommend routine respiratory evaluations, which may include:

- Physical examinations, such as listening to your lungs with a stethoscope.
- Regular lung function tests to check how well your lungs work. They can determine how much air they can hold, how quickly you can move air in and out of your lungs, and how effectively your lungs put oxygen into your blood and remove carbon dioxide from your blood.
- Imaging tests, especially magnetic resonance imaging (MRI) or computed tomography (CT) scans, show detailed images of the inside of your body and play a crucial role in disease detection, diagnosis and treatment monitoring.

It is important that your physician identifies any respiratory problems early so that appropriate therapies can be started to treat your symptoms and reduce the risk of disease progression and further complications.

RESPIRATORY SYMPTOMS AS AN INDICATOR OF PIDS

Respiratory infections, particularly those that recur and do not respond well to antibiotics or are due to unusual or opportunistic (ones that take advantage of a weak immune system) infections, can be an early sign that a patient may have a PID.

It is important that physicians and respiratory specialists look for immunodeficiencies in these patients, especially in children, and particularly if they do not respond to conventional treatments.

If a PID is suspected, your physician/respiratory specialist is likely to initiate specific evaluations, usually blood tests, to look for a diagnosis. They should also refer you to an immunologist.

TREATMENTS

If you receive intravenous or subcutaneous immunoglobulin (IG) replacement therapy, this may afford you some protection against many viruses and bacteria. However, it may not prevent the development of all respiratory conditions and you may require additional support. Antibiotics, for example, may be used to prevent infections developing.

Management of your condition will be aimed at relieving your symptoms and preventing further complications. Your physician may recommend the following treatments:

- Antibiotics to prevent, as well as treat, bacterial infections
- Inhaled steroids, bronchodilators and allergy treatments to reduce symptoms
- Medications to reduce fever and general body aches
- Expectorants to 'water down' mucus secretions so that they are easier to expel
- Decongestants to shrink swollen mucous membranes
- Fluids to maintain adequate hydration; warm drinks may promote nasal drainage and relieve chest tightness
- Small frequent meals of light foods and drinks if the patient has loss of appetite. Once appetite returns, a high-calorie, high-protein diet to replace proteins lost during the acute phase.
- General comfort measures:
 - Mouth rinsing with plain water to relieve dryness and the 'bad taste' that often accompanies illness and mouth breathing
 - Cold-water vaporizer to increase room humidity
 - Lip balm or other coating to protect irritated lips and nose
 - Adequate rest
 - Extra pillows to elevate head and shoulders and provide relief from coughing or nasal dripping
 - Cough suppressant to prevent sleep interruption.

To reduce the risks of passing on infections to other individuals, take the following precautions:

- Cover nose and mouth when coughing or sneezing
- Discard tissues promptly
- Wash hands frequently.

In some cases of bronchitis and pneumonia, coughing and breathing deeply at regular intervals may protect the lungs by removing mucus and foreign particles.

- Deep breathing promotes full expansion of the lungs and reduces the risk of further complications.
- In some cases, other means of clearing mucus may be suggested, e.g. chest or sinus drainage, chest physiotherapy or positive airway pressure.

For people with PIDs who have significant respiratory symptoms, it is essential that your healthcare team includes a respiratory specialist to assist with diagnostic testing and directing treatment.

FURTHER INFORMATION AND SUPPORT

This booklet has been produced by the International Patient Organisation for Primary Immunodeficiencies (IPOPI). Other booklets are available in this series. For further information and details of PID patient organisations in over 52 countries worldwide, please visit www.ipopi.org.

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Supported by an educational grant from Octapharma