

Pneumocystis pneumonia (PCP, PJP)

Summary

Pneumocystis pneumonia (PCP, also called PJP) is a type of pneumonia that can be life threatening in people with weakened immune systems. People living with HIV whose CD4 counts are below 200 are at risk of developing PCP. The symptoms can include fever, shortness of breath, tightness or pain in the chest, fatigue, night sweats and a dry cough. Fortunately, there are medications that can effectively prevent and treat the illness.

Today, PCP is relatively rare; however, it remains common among people who do not know they have HIV, people who are not receiving ongoing HIV care, and people with severely weakened immune systems.

What is PCP?

PCP is a type of pneumonia caused by a fungus called *Pneumocystis jiroveci*. This fungus is widely distributed in the environment. It usually infects people during childhood but does not make people with healthy immune systems sick. However, in people with a weakened immune system, the fungus can most commonly affect the lungs, causing pneumonia. The original name for pneumonia caused by the fungus was PCP. However, scientists renamed the fungus that caused the pneumonia (*Pneumocystis jiroveci*), and so microbiologists and doctors now also call it PJP. Both PCP and PJP refer to the same form of pneumonia.

PCP is one of a number of infections that can develop in people who are living with HIV, called *opportunistic infections*. These occur only if your immune system is quite weakened and your body becomes vulnerable to infections that would not otherwise affect you. PCP is the most common opportunistic infection among people living with HIV.

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Pneumonia is an infection and inflammation of the lungs. As the infection progresses, the air spaces in the lungs fill with fluid, making it more difficult to breathe.

Who is at risk for PCP?

People who have a weakened immune system, due to HIV, cancer, long-term use of corticosteroid drugs or an organ or bone marrow transplant, are at risk of developing PCP.

People with HIV most at risk of developing PCP are those who:

- have a CD4 count below 200
- have a CD4 cell percentage below 14%
- have had a previous episode of PCP
- have a CD4 count below 300 with signs of a weakened immune system (such as recurrent oral thrush or bacterial pneumonia)

Symptoms

The symptoms of PCP can include:

- shortness of breath
- fever
- tightness or pains in the chest
- dry cough
- fatigue
- feeling of weakness or malaise
- chills/sweats
- diarrhea
- weight loss

In people living with HIV, these symptoms may develop very gradually. At first, the symptoms may be so mild that they go unnoticed for several weeks.

Because PCP can be so dangerous if not treated early, *if you experience symptoms of PCP, such as a persistent dry cough or shortness of breath, it is important that you report them to a doctor as soon as possible.*

Diagnosis

The symptoms of PCP are common to many infections, including the flu and common cold, so lab tests must be done to confirm the diagnosis. Your doctor may order one or more of the following tests:

- Induced sputum test—Breathing in a mist of saltwater causes phlegm (or sputum) from the lungs to be coughed up. The sputum sample is then tested in the lab to look for the fungus that may be causing an infection.
- Bronchoscopy with lavage—The bronchoscope is a very thin, flexible tube that is inserted through the nose, down the windpipe and into the lungs. It allows the doctor to look at the inside of the lungs. A salt-water solution is flushed down the tube (a process called *lavage*). This allows the doctor to collect a sample of cells and fluid from the lungs. When the solution is sucked back up the tube, the samples are sent to the lab for testing. A local anesthetic and muscle relaxing drugs may be given before a bronchoscopy is performed.

If PCP is diagnosed, two other tests may be performed to find out how serious the pneumonia is:

- Pulmonary function tests—These tests give a doctor an idea of how well the lungs are working. They measure the capacity of the lungs to expand and hold air, the rate at which air flows in and out of the lungs, and the amount of oxygen that can pass from the lungs into the blood.
- Blood test (blood gases)—To find out how much oxygen is getting from the lungs into the blood, and how much carbon dioxide is getting from the blood to the lungs, a sample of blood may be taken from an artery so that arterial blood gases can be measured.

The results of these tests can help your doctor classify the PCP as mild, moderate or severe; choose the most suitable treatment; and determine whether medication can be taken at home or in hospital.

Treatment

If you are diagnosed with an acute case of PCP and are not already taking HIV treatment (antiretroviral drugs; ART), guidelines recommend that you start within two weeks of PCP diagnosis. This should strengthen your immune system and help you fight the infection.

The most effective medication for treating PCP is called co-trimoxazole (commonly sold under the brand names Septra or Bactrim and in generic formulations). It is a combination of two antibiotics: trimethoprim (TMP) and sulfamethoxazole (SMX).

Moderate to severe PCP

For moderate to severe PCP, treatment guidelines from the U.S. Department of Health recommend:

- co-trimoxazole (TMP 15-20 mg/kg/day and SMX 75-100 mg/kg/day) intravenously given in divided doses every 6 or 8 hours.

Guidelines suggest that once patients improve they can be switched to an oral formulation of co-trimoxazole. Treatment is for 21 consecutive days.

In addition to co-trimoxazole, corticosteroids are often prescribed to reduce inflammation in the lungs. Although long-term use of corticosteroids can weaken the immune system, short-term use can help to reduce injury to the lungs. Corticosteroids should be started as soon as possible—within 72 hours of starting co-trimoxazole. People with severe PCP and other conditions that make it difficult to swallow medications may need to take the antibiotics intravenously in the hospital. (Co-trimoxazole also protects against another opportunistic infection called toxoplasmosis.)

In severe cases of PCP, patients may also be given oxygen to inhale as part of their treatment.

Treatment for PCP usually lasts 21 days. The way a patient responds to the treatment depends on the drugs used, whether or not they have had previous episodes of PCP, the severity of the illness, the state of their immune system, and when the treatment was started.

Mild-to-Moderate PCP

Guidelines recommend that co-trimoxazole (TMP 15-20 mg/kg/day and SMX 75-100 mg/kg/day) be given orally in three divided doses. Or two double-strength tablets of co-trimoxazole taken three times daily. Treatment lasts for 21 consecutive days.

Your doctor will monitor your treatment carefully. Common side effects from co-trimoxazole include rash, fever, nausea, vomiting, loss of appetite, low white blood cell count and low platelet count. Your doctor may recommend additional medications to manage these side effects.

Some people with HIV are allergic or hypersensitive to this medication. In these cases, alternative medications can be prescribed. There is evidence to suggest that in some cases where people are hypersensitive to co-trimoxazole, starting with a small amount of co-trimoxazole and increasing the amount until a full dose can be tolerated can help a person overcome adverse reactions, or help “desensitize” a person who is hypersensitive to the medication.

Pregnancy

When co-trimoxazole is taken during the first three months of pregnancy, U.S. authorities have stated that there may be a small but increased risk of birth defects. Overall, U.S. PCP treatment guidelines note that the benefits of treating PCP greatly outweigh the risks, as untreated PCP results in severe illness and, in many cases, death. U.S. guidelines note that supplements of the B-vitamin folic acid (4 mg/day) during the first three months of pregnancy may reduce the risk of birth defects. They add that the dose of folic acid should be reduced to 0.4 mg/day after the first three months of pregnancy.

Because a pregnant person with PCP also faces a higher risk of preterm labour and delivery, pregnant people who have PCP after 20 weeks of pregnancy should be monitored for early contractions.

If, after four to eight days of anti-PCP treatment, the pneumonia has shown no sign of improvement or has worsened, your doctor may recommend another treatment. Other anti-PCP treatments, such as dapsone and trimethoprim, primaquine and

clindamycin, or atovaquone, offer alternatives to people who cannot tolerate co-trimoxazole.

Once the pneumonia has resolved, your doctor may recommend that you take medication to prevent the infection from coming back. This preventive medication should be taken until your CD4+ count stays above 200 cells/mm³ for at least three consecutive months. Talk to your doctor before you start or stop taking any prescribed medications.

Prevention

Once you have recovered from PCP, you will need to take antibiotics for a period of time to prevent a recurrence. This is called secondary prophylaxis (prevention). Guidelines recommend either one double-strength co-trimoxazole tablet taken daily or one single-strength tablet taken daily for secondary prevention of PCP.

Ultimately the best way to stay healthy and prevent future episodes of PCP is to keep your immune system strong and your CD4+ count well above 200 cells/mm³. Taking ART exactly as prescribed can help keep your CD4+ count above 200 cells/mm³ and reduce your risk for future episodes of PCP. U.S. guidelines state: "Secondary prophylaxis should be discontinued in adult and adolescent patients whose CD4+ counts have increased from less than 200 cells/mm³ to 200 more cells/mm³ for at least three consecutive months as a result of ART."

Regular visits to your HIV clinic and lab are important so that your health can be monitored.

If you smoke, another way you can help reduce the risk of getting PCP is to quit smoking. Research has shown that smokers living with HIV are far more likely to develop PCP than non-smokers living with HIV. Talk to your doctor, nurse or pharmacist about ways to help you quit smoking.

Preventive medication should be taken by HIV-positive adults and adolescents, including pregnant people and people on HIV treatment, whose CD4 count is below 200 cells/mm³ or have a history of thrush (oral yeast infections). The medication used to treat PCP can also be taken to prevent it. The most effective preventive medication is co-trimoxazole. However, if you cannot tolerate this,

there are other medications that your doctor can recommend.

References

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