

What is Microscopic Polyangiitis (MPA)?

Microscopic polyangiitis (MPA) is a form of vasculitis. Vasculitis is a group of rare disorders characterized by inflammation of the blood vessels. Inflammation of the blood vessels may result in narrowing of the vessel, which can block or slow down blood flow to vital organs and tissues.

MPA most commonly affects the small- to medium-sized blood vessels, particularly involving the kidneys, lungs, nerves, skin, and joints. MPA can worsen rapidly, so early diagnosis and treatment are essential to prevent kidney or respiratory damage, or organ failure.

MPA is known as an anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitis, which refers to a blood protein that attacks the body's own cells and tissues. It is a serious but treatable disease.

Causes

The cause of MPA is not yet fully understood. It is classified as an autoimmune disorder—a disease which occurs when the body's natural defense system mistakenly attacks healthy tissues.

It is believed that an infection may trigger the autoimmune response. Environmental and some genetic factors may also play a role in causing it.

Who Gets MPA?

MPA can affect people of all ages, but the average age of onset is approximately 50. It affects both men and women, but men may get MPA more often. The disease is more frequent among Caucasians but can affect people of any race or ethnic background.

Symptoms

The symptoms of MPA and their severity can vary greatly from person to person, depending on which blood vessels and organs are affected. For some the disease is mild, while for others it may be severe or even potentially life-threatening if untreated.

MPA symptoms may come on slowly over a period of months or develop rapidly in a matter of days. People with MPA often feel generally ill, with flu-like symptoms of fatigue, fever, loss of appetite, and weight loss. Other symptoms may be related to the organ systems affected:

- Kidney inflammation, which may be associated with bloody or dark urine
- Rashes or sores, especially on the legs
- Cough (coughing up blood, shortness of breath)
- Nerve problems
 - Tingling, numbness, pain, weakness
 - Inability to lift your foot or wrist (also called “foot drop” or “wrist drop”)
- Joint and muscle pain
- Abdominal pain after eating
- Eye irritation

Complications

Serious and even life-threatening complications can occur with MPA, especially with the kidneys and lungs. If you have symptoms that don't go away, have bloody or dark urine, or are coughing up blood, contact your doctor right away.

Diagnosis

Because there is no single test for diagnosing MPA, your doctor will consider several factors, including your symptoms, a medical history, and physical exam findings, as well as:

- **Urinalysis:** The presence of red blood cells may indicate kidney inflammation. (Note: kidney disease can happen without having symptoms; therefore, patients with vasculitis should have regular urine tests.)
- **Blood tests:**
 - ANCA test can be helpful when positive.
 - Blood tests that can detect inflammation include:
 - “Sed rate,” which is the erythrocyte sedimentation rate
 - C-reactive protein test
 - Complete blood cell count (can show anemia and/or high platelets)
- **Tissue biopsy:** A biopsy is often needed to diagnosis MPA. It is a surgical procedure that removes a small tissue sample from an affected organ and is then examined under a microscope for signs of inflammation or tissue damage. Tissues that might be biopsied for MPA include kidney, lung, skin, nerve, and muscle.
- **Imaging studies:**
 - Chest x-rays may reveal changes in your lungs that are characteristic of MPA.
 - Computed tomography and magnetic resonance imaging scans provide more detailed images of your internal organs and can show other abnormalities.

Treatment of MPA

Treatment is based on disease severity and organ involvement. The traditional course of treatment includes these medications:

- Corticosteroids such as prednisone may be used in combination medications that suppress the immune system and reduce inflammation.
- Milder forms of MPA are typically treated with a combination of prednisone and methotrexate.

For severe disease:

- **Prednisone** is usually started at a high dose and then tapered off slowly.
- **Biologic drugs** such as rituximab are commonly used in combination with prednisone. Rituximab is usually the first option therapy in combination with prednisone. Biologic medications are complex proteins derived from living organisms and they target certain parts of the immune system to control inflammation.
- A **chemotherapy-type drug**, cyclophosphamide, blocks abnormal growth of certain cells in the body, and it is used in combination with prednisone. Cyclophosphamide can lower the body's ability to fight infection and increase the risk of cancer, so it is usually limited to a three- to six-month period and replaced with less toxic medications such as mycophenolate mofetil and azathioprine, or methotrexate, a drug commonly used to treat rheumatoid arthritis.
- **Dialysis and/or a kidney transplant** may be needed if kidney failure occurs.

Once in remission, most patients will likely need to continue taking maintenance medications such as azathioprine, methotrexate, or rituximab, to keep the disease under control. The dose of steroids is usually tapered during remission.

Even with treatment, MPA is a chronic condition with periods of relapse and remission, so ongoing medical care and monitoring are necessary.

Side Effects of Medications

The medications used to treat MPA have potentially serious side effects, such as:

- Lowering your body's ability to fight infection
- Potential bone loss (osteoporosis), among others

Therefore, it is important to see your doctor for regular checkups. Medications may be prescribed to offset side effects.

Infection prevention is also very important. Talk to your doctor about getting a flu shot, pneumonia vaccination, and/or shingles vaccination, which can reduce your risk of infection.

Relapse

Even with effective treatment, MPA is a chronic disease, and relapses may occur. If your initial symptoms return or you develop new ones, report them to your doctor as soon as possible. Regular doctor visits and ongoing monitoring of lab and imaging tests are important in detecting relapses early.

Your Medical Team

Effective treatment of MPA may require the coordinated efforts and ongoing care of a team of medical providers and specialists. In addition to a primary care provider, you may need to see the following specialists:

- Rheumatologist (joints, muscles, immune system)
- Pulmonologist (lungs)
- Nephrologist (kidneys)
- Dermatologist (skin)
- Neurologist (brain/nervous system) or others as needed

The best way to manage your disease is to actively partner with your health care providers and get to know the members of your health care team.

Use a health care journal to track medications, symptoms, test results and notes from doctor appointments.

Make a list of questions before your doctor's visit. Bring along a supportive friend or family member to provide a second set of ears and take notes.

Remember, it is up to you to be your own advocate. If you have concerns with your treatment plan, speak up. Your doctor may be able to adjust your dosage or offer different treatment options.

It is always your right to seek a second opinion.

Living with MPA

Living with a chronic disease such as MPA can be challenging at times. Fatigue, pain, emotional stress, and medication side effects can take a toll on your sense of well-being. This can affect relationships, work, and other aspects of your daily life.

Sharing your experience with family and friends, connecting with others through a support group, or talking with a mental health professional can help.

Outlook

Currently, there is no cure for MPA, but with early diagnosis and proper treatment, many patients can lead full, productive lives. Because relapses can occur with MPA, follow-up medical care is essential.

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