ANCA-ASSOCIATED VASCULITIS: MICROSCOPIC POLYANGIITIS

What is microscopic polyangiitis (MPA)?

Microscopic polyangiitis (MPA) is a form of vasculitis—a family of rare disorders characterized by inflammation of the blood vessels, which can restrict blood flow and damage 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 an ANCA-associated vasculitis, referring to a blood protein (anti-neutrophil cytoplasmic antibody) that attacks the body's own cells. Other forms of ANCA-associated vasculitis include eosinophilic granulomatosis with polyangiitis (EGPA) and granulomatosis with polyangiitis (GPA). People with these types of vasculitis often test positive for ANCA, although the test is not conclusive.

MPA is a serious but treatable disease. The traditional course of treatment includes corticosteroids such as prednisolone used in combination with other medications that suppress the immune system and reduce inflammation. Even with treatment, MPA is a chronic condition with periods of relapse and remission, so ongoing medical care and monitoring are necessary.

Causes

The cause of MPA is not yet fully understood by researchers. Vasculitis is classified as an autoimmune disorder, a disease which occurs when the body's natural defense system mistakenly attacks healthy tissues. Researchers believe an infection may set the inflammatory process in motion in MPA. Environmental and some genetic factors may also play a role in vasculitis.

Who gets MPA?

MPA can affect people of all ages, but the average age of onset is approximately 55. It affects both men and women, but men may get MPA more often. The disease can affect people of any race or ethnic background.

MPA is a rare disease. It affects approximately 5-15 new people per year in Australia and New Zealand

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 often comes on over a period of months, but may 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 organ systems affected.

- Kidney inflammation, which may be associated with bloody or dark urine. (Note: A patient can have kidney disease without having symptoms; therefore, patients with vasculitis should have regular urine tests.)
- Skin rashes/lesions, especially on the legs
- Cough (coughing up blood, shortness of breath)

- Nerve problems (tingling, numbness, pain, weakness, "foot drop" or "wrist drop"—inability to lift your foot or wrist)
- Joint and muscle pain
- Abdominal pain with eating
- Eye irritation

Complications

Serious, 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 a number of factors, including a thorough medical history, physical exam findings, and results of laboratory tests and imaging studies. A biopsy of the affected tissue is usually obtained to confirm the diagnosis.

- **Urinalysis:** The presence of red blood cells may indicate kidney inflammation. Your doctor may use this test to help diagnosis MPA, and to monitor the kidneys during and after treatment.
- **Blood tests**: The ANCA test can be helpful when positive. Blood tests that can detect inflammation include the erythrocyte sedimentation rate (ESR) test, commonly called the "sed rate," and the C-reactive protein (CRP) test. All these tests may support a diagnosis of MPA, but are not conclusive on their own. A tissue biopsy is typically needed.
- **Tissue biopsy:** This surgical procedure removes a small tissue sample from an affected organ, which is 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 (CT) and magnetic resonance imaging (MRI) scans provide more detailed images of your internal organs and can show abnormalities.

Treatment of MPA

Treatment is based on a number of factors, including disease severity and organ involvement. The cornerstone of treatment for MPA is corticosteroids such as prednisone used in combination with other medications that suppress the immune system and reduce inflammation.

For severe disease, the biologic drug rituximab may be used in combination with prednisone. Rituximab has been approved in Australia and in New Zealand for MPA and GPA in certain situations. Biologic medications are complex proteins derived originally from living organisms. They target certain parts of the immune system to control inflammation.

Another option for severe disease is cyclophosphamide, a chemotherapy-type drug that blocks abnormal growth of certain cells in the body, in combination with prednisone. Prednisolone is typically started at a high dose and then tapered off slowly. Cyclophosphamide can lower the body's ability to fight infection, so it is usually limited to a three- to six-month period and replaced with less toxic

medications such as azathioprine, methotrexate or mycophenolate mofetil. Milder forms of MPA are typically treated with a combination of prednisone and methotrexate.

Some individuals may experience kidney failure, a serious complication that requires dialysis and/or a kidney transplant. Another option for those with very serious MPA affecting the kidneys or lungs may be "plasmapheresis." Plasmapheresis is a dialysis-like procedure that clears proteins from the plasma of the blood and replaces it with plasma from a donor, or with a plasma substitute.

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

Side effects

The medications used to treat MPA have potentially serious side effects, such as lowering your body's ability to fight infection, and potential bone loss (osteoporosis), among others. Therefore, it's 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 vaccinated against influenza, pneumococcal (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 GPA may require the coordinated efforts and ongoing care of a team of medical providers and specialists. In addition to your general practitioner, patients may need to see: a nephrologist (kidneys); rheumatologist (joints, muscles); clinical immunologist (immune system); dermatologist (skin); respiratory physician (lungs); gastroenterologist (digestive system); ENT (ear, nose and throat) surgeon; cardiologist (heart); neurologist (brain/nervous system); or others as needed.

The best way to manage your disease is to actively partner with your health care providers. Get to know the members of your health care team. It may be helpful to keep a health care journal to track medications, symptoms, test results and notes from doctor appointments in one place. To get the most out of your doctor visits, make a list of questions beforehand and bring along a supportive friend or family member to provide a second set of ears and take notes.

Remember, it's 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, affecting 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

There is no cure for MPA at this time, 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.

Clinical studies are ongoing at multicenter research centers, including the Vasculitis Clinical Research Consortium (VCRC) in the USA, to better understand the risk factors and causes of vasculitis, investigate more effective and safer treatments, and work toward a cure. The Vasculitis Foundation encourages patients to consider participating in clinical research studies to help further understanding of vasculitis. Patients are also encouraged to join the Vasculitis Patient Powered Research Network (VPPRN), where they can provide valuable disease insight and information.

ANZVASC, the Australian and New Zealand Vasculitis Society, was formed in late 2018 to promote collaboration and excellence in vasculitis care and research in Australia and New Zealand. Patients and consumers are welcome to participate and can join as associate members.

For more information on vasculitis and vasculitis research, visit:
www.anzvasculitis.org
www.vasculitisfoundation.org

ANZVASC thanks the Vasculitis Foundation for permission to use this information, with minor changes for use in Australia and New Zealand

About Vasculitis

Vasculitis is a family of nearly 20 rare diseases characterized by inflammation of the blood vessels, which can restrict blood flow and damage vital organs and tissues. Most forms of vasculitis are autoimmune diseases, that occur when the body's natural defense system mistakenly attacks healthy tissues. Triggers may include infection, medication, genetic or environmental factors, allergic reactions, or another disease. However, the exact cause is often unknown.

A Family of Diseases

- Anti-GBM (Goodpasture's) disease
- Aortitis
- Behcet's syndrome
- Central nervous system vasculitis
- Cogan's syndrome
- Cryoglobulinemia
- Cutaneous small-vessel vasculitis (formerly hypersensitivity/leukocytoclastic)
- Eosinophilic granulomatosis with polyangiitis (EGPA, formerly Churg-Strauss syndrome)
- Giant cell arteritis
- Granulomatosis with polyangiitis (GPA, formerly Wegener's)
- IgA vasculitis (Henoch-Schönlein Purpura)
- Kawasaki disease
- Microscopic polyangiitis
- Polyarteritis nodosa
- Polymyalgia rheumatica
- Rheumatoid vasculitis
- Takayasu's arteritis
- Urticarial vasculitis

About the Vasculitis Foundation

The Vasculitis Foundation (VF), based in the USA, is the leading organization in the world dedicated to diagnosing, treating, and curing all forms of vasculitis. The VF provides a wide range of education, awareness and research programs and services. To learn more, and get the most updated disease and treatment information, visit www.vasculitisfoundation.org

The VF on Social Media includes:

- Instagram: vasculitisfoundation
- Twitter: @VasculitisFound
- VF Facebook Discussion Group: www.facebook.com/groups/vasculitisfoundation

Ways to Get Involved

- Participate in research
- Join social media
- Host or participate in an event
- Attend a regional conference and/or symposium

- Give a donation toward long-term solutions
- Become an advocate for yourself, or for others
- Share your journey

Vasculitis Foundation Mission

Building upon the collective strength of the vasculitis community, the Foundation supports, inspires and empowers individuals with vasculitis, and their families, through a wide range of education, research, clinical, and awareness initiatives.

The VPPRN and VCRC

The Vasculitis Patient-Powered Research Network (VPPRN) seeks to improve the care and health of patients with vasculitis by exploring research questions that matter most to patients, and by advancing medical knowledge about vasculitis. For more information, visit: www.VPPRN.org. The Vasculitis Clinical Research Consortium (VCRC) is an integrated group of academic medical centers, patient support organizations, and clinical research resources dedicated to conducting clinical research in different forms of vasculitis. For more information, visit: www.rarediseasesnetwork.org/cms/vcrc

Vasculitis Foundation

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ANZVASC

The Australian and New Zealand Vasculitis Society was formed in late 2018 to promote collaboration and excellence in vasculitis care and research in Australia and New Zealand.
Patients and consumers are critical in the work of ANZVASC and are most welcome to participate and join as associate members.
ANZVASC is a registered health promotion charity with the Australian Charities and not for Profit Commission as has Deductible Gift Recipient Status.

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The Vasculitis Foundation (VF) is a 501(c)(3) nonprofit organization governed by a Board of Directors and advised on medical issues by a Medical and Scientific Advisory Board. VF's educational materials are not intended to replace the counsel of a physician. VF does not endorse any medications, products or treatments for vasculitis, and advises you to consult a physician before initiating any treatment.

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