MICROSCOPIC POLYANGIITIS (MPA)

Vasculitis Foundation Canada
Fondation Vasculites Canada
contact@vasculitis.ca | www.vasculitis.ca
1-877-572-9474

CanVasc
Canadian vasculitis research network
admin@canvasc.ca | www.canvasc.ca

What is MPA?

MPA is an autoimmune disease characterized by inflammation of the small blood vessels in the body. It is also known as an ANCA-associated vasculitis due to the presence of ANCA antibodies in the blood. There are 2 types of ANCAs – p/MPO-ANCA and c/PR3-ANCA. MPA is typically associated with p/MPO-ANCA.

Which organs are affected?

MPA most commonly affects the kidneys, lungs, nervous system, skin and joints. Kidney involvement is present in most patients and is called **glomerulonephritis**. When severe, this can cause kidney failure.

Who is affected? What causes it?

MPA is a rare condition affecting approximately 25-50 per million people. Men are affected slightly more than women. The average age of disease onset is 50 years.

The exact cause of MPA is unknown. As with most autoimmune diseases, there is usually a combination of genetic, environmental and infectious factors.

What are the symptoms?

MPA can cause a variety of symptoms depending on which organs are affected. Possible symptoms include:

- Shortness of breath, cough or bloody sputum
- Sinus and nasal congestion (rarer and less severe than in GPA)
- Skin rash resembling small bleeding spots that appear on the feet and lower legs
- Joint and muscle discomfort
- Burning, numbness or tingling of the extremities
- Weakness or inability to move the wrist or ankle, otherwise known as a “wrist drop” or “foot drop”
- Nausea & abdominal pain
- Unusually bubbly / frothy / dark urine (if the kidneys are affected)
- Fevers, weight loss and unusually severe fatigue

While MPA-related kidney disease and bleeding from the lungs can present quickly and quite severely, other features such as nerve involvement and lung scarring (from interstitial lung disease or fibrosis) can develop over months to years.

How is MPA diagnosed?

Diagnosis is made based on a combination of the following:

1. Compatible symptoms and clinical features
2. Abnormal labs, including:
   a. High inflammatory markers (ESR and/or CRP)
   b. Detection of p/MPO-ANCA in the blood
   c. High creatinine level
   d. High levels of protein and presence of blood in urine
3. Supportive features on biopsy of affected organ(s). The most common biopsy site is the kidney.
4. Imaging of the lungs and other affected sites.
5. Bronchoscopy to assess for inflammation in the airways.
6. Electromyography to assess for nerve involvement.
7. Pulmonary function test.

Remember that any of the listed symptoms by itself can be seen in more common conditions such as infections and cancers. It is therefore important to consider the “whole picture” and rule out other possible conditions when making the final diagnosis.

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How is MPA treated?

There are 2 phases of treatment:

1. **Induction phase** – This phase is aimed at rapidly suppressing the inflammation and "inducing" the disease into a state of dormancy (ie. "remission"). High dose prednisone is started alongside another immune suppressing medication. If your disease is severe, cyclophosphamide or rituximab may be used. The prednisone dose will be lowered over several months' time.
2. **Maintenance phase** – At this point, the disease is inactive and the goal is to maintain remission. Common maintenance medications include rituximab, azathioprine, methotrexate, and mycophenolate mofetil.

Adjunctive therapies include:

- **Pneumocystis pneumonia (PCP) protection** with an antibiotic
- **Plasma exchange,** which can be added for refractory, severe kidney disease or lung bleeding (although no clear benefits were seen in recent studies)
- **Dialysis** if there is severe kidney failure

How is my MPA monitored?

Your rheumatologist often works with other specialists to treat your MPA. Depending on which organs are affected by the disease, you may expect to also see a **respirologist or nephrologist**, among others.

Your family physician also plays an important role in monitoring for high blood pressure & cholesterol, diabetes, and osteoporosis.

Common tests used to monitor the disease include:

- Lab tests for blood counts, inflammatory markers, liver & kidney function, and ANCA levels
- Urine studies for blood & protein
- X-rays, CTs and ultrasounds
- Pulmonary function tests (breathing tests)

Testing is typically more frequent at the beginning or when there is active disease, and less frequent once the disease is stable and in remission.

Biopsies are typically done for diagnostic purposes and do not need to be repeated unless there is a new or worsening feature that needs to be defined further.

Is MPA a fatal disease?

The survival rate for MPA is high when treated – up to 90% at 1 year, and between 65-95% at 5 years. Outcomes depend on a variety of factors, the most important ones being if there is severe kidney disease and any complications (eg. infections) during treatment.

What will happen to me? How long do I take medications for?

Like other autoimmune diseases, MPA is treatable but not yet curable. Symptoms resolve with medications, however may return when they are stopped. When the disease returns ("flares"), it is called a **relapse**.

Currently, we do not know the ideal duration of maintenance treatment for MPA. 1 in 12 patients will relapse within the first 18 months. This increases to 1 in 3 patients after 4-7 years. Because of this, most patients are kept on medications for at least 2-5 years. Patients who have developed organ damage or had multiple relapses already may be kept on longer (or even lifelong) treatment or prevent worsening organ damage.

MPA DISEASE SUMMARY TOOL

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My symptoms included:

- ☐ Painful, red, bloodshot eyes
- ☐ Shortness of breath
- ☐ Cough and/or coughing up blood
- ☐ Skin rash and/or skin ulcers
- ☐ Burning, tingling or numbness of the hands or feet
- ☐ Weakness of the arm or leg
- ☐ Joint pain or joint swelling
- ☐ Abdominal or flank pain
- ☐ Nausea and/or vomiting
- ☐ Unusually bubbly / frothy / dark urine
- ☐ Fevers or night sweats
- ☐ Weight loss
- ☐ Unusually severe fatigue
- ☐ Others: _____

Medications that have been prescribed:

	Date started	Date stopped
IV methylprednisolone _____ mg		
Prednisone: (Starting dose _____ mg)		
Cyclophosphamide _____ mg		
Rituximab _____ mg (INDUCTION)		
Rituximab _____ mg (MAINTENANCE)		
Septra/Bactrim/Sulfatrim		
Methotrexate _____ mg		
Folic acid (only if taking methotrexate)		
Mycophenolate mofetil _____ mg		
Azathioprine ____ mg		
Leflunomide ____ mg		
Plasma exchange (PLEX) (Number of sessions: _____)		
Others:		

Apart from lab tests, I have had the following investigations:

- ☐ Scopes (circle any that apply): Bronchoscopy | Nasal scope
- ☐ Audiology test
- ☐ Pulmonary function test
- ☐ Electromyography (EMG/nerve conduction study)
- ☐ X-rays (circle all that apply): Chest | Abdomen | Joints
- ☐ CT scans (circle all that apply): Chest | Abdomen | Head | Sinuses
- ☐ MRI (circle all that apply): Brain | Spine | Orbits (eyes)
- ☐ Biopsy (circle all that apply): Skin | Nasal cavity | Kidney | Lung
  Other: _____

What do I need to do?

- ✓ Attend **regular** follow-up visits with my rheumatologist and other specialists.
- ✓ **Do regular** blood and urine tests for disease and medication monitoring.
- ✓ Ensure my medical team is monitoring me for diabetes, high blood pressure and high cholesterol.
- ✓ Seek urgent medical attention if you are having daily fevers, coughing up blood or have other concerning symptoms.