Patient Resource

Anti-Neutrophilic Cytoplasmic Antibodies (ANCA)-Associated Vasculitis (AAV) with Kidney Involvement

What is it?

Anti-Neutrophilic Cytoplasmic Antibodies (ANCA) associated vasculitis (AAV) is an autoimmune disease that involves the small blood vessels of the body, including those in the glomeruli – or the filters – of the kidney. ANCA-associated vasculitis is one of a larger group of kidney diseases known as glomerulonephritis.

ANCA-associated vasculitis can occur in other organs of the body. This handout will **only** describe treatment for the kidneys.

Here's what happens in ANCA-associated vasculitis:

- ANCA are antibodies produced by the immune system in error (immune dysregulation) that mistakenly target neutrophils. We don't know why this occurs.
- Neutrophils are cells in the immune system that help find and attack harmful things in the body, like bacteria.
- ANCA cause neutrophils to start an inflammatory process that damages the blood vessel wall.
- Damage to blood vessels in the glomeruli causes blood and protein to leak into the urine.
- This leads to an inflammatory response, which destroys the glomeruli and replaces them with scar tissue.
- There are two main types of ANCA: one is called Proteinase 3 (PR3), and the other is called Myeloperoxidase (MPO). About 1 in 10 patients will have the symptoms and features of ANCA-associated vasculitis without an antibody being measured or detected in the blood.
- The specific type of ANCA (PR3 or MPO) may influence how a patient's disease develops or progresses.

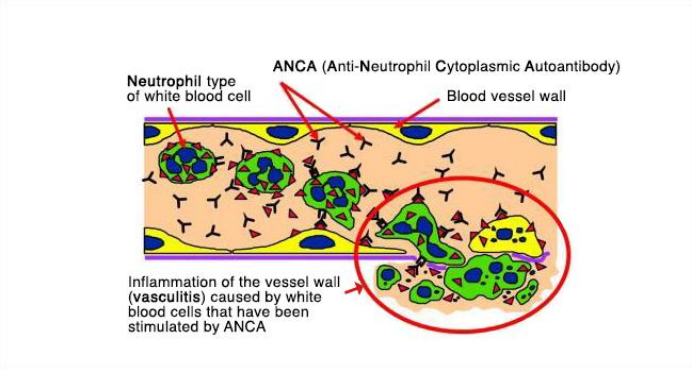

Figure source: https:// unckidneycenter.org/kidneyhealthlibrary/glomerular-disease/anca-vaculitis/

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Changes observed in the kidneys with ANCA-associated vasculitis

The diagram below provides a graphic representation of glomerular inflammation caused by ANCA-associated vasculitis, compared to normal glomerulus.

Normal glomerulus

Glomerulus affected by ANCA-associated vasculitis

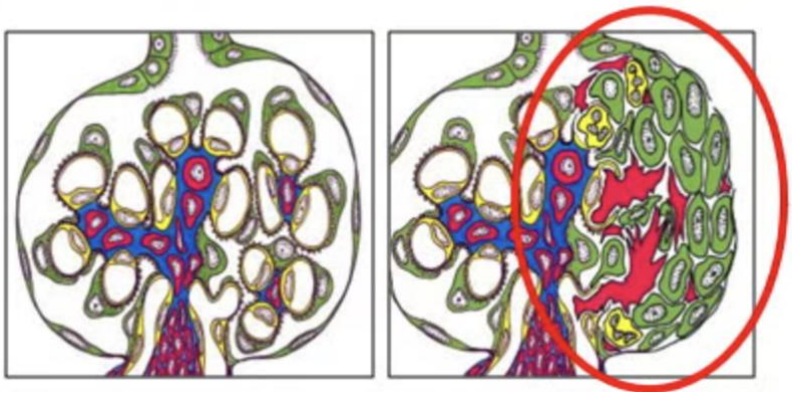

Figure source: https:// unckidneycenter.org/kidneyhealthlibrary/glomerular-disease/anca-vaculitis/

What are the symptoms of ANCA-associated vasculitis?

- Symptoms may include:
  - Rash or small bruises, usually on the arms and legs
  - Coughing up blood, shortness of breath
  - Nasal congestion
  - Ear pain/drainage
  - Hearing loss
  - Painful, red eyes
  - Swollen or painful joints
  - High blood pressure
  - Numbness, tingling in the hands or feet
  - Muscle weakness
  - Headaches, fever, fatigue

What are the complications of ANCA-associated vasculitis?

- Complications may include:
  - Risk of declining kidney function, which may lead to kidney failure
  - Scarring of the organ involved, including the kidney, which can be permanent causing chronic disease
  - Rapidly progressive disease requiring hospitalization
  - Damage to nerves causing pain, numbness or weakness
  - Increased risk of infection
  - Damage to lungs and bleeding into lungs
  - Higher risk of strokes and heart attacks

Effects of ANCA-associated vasculitis on various areas of the body

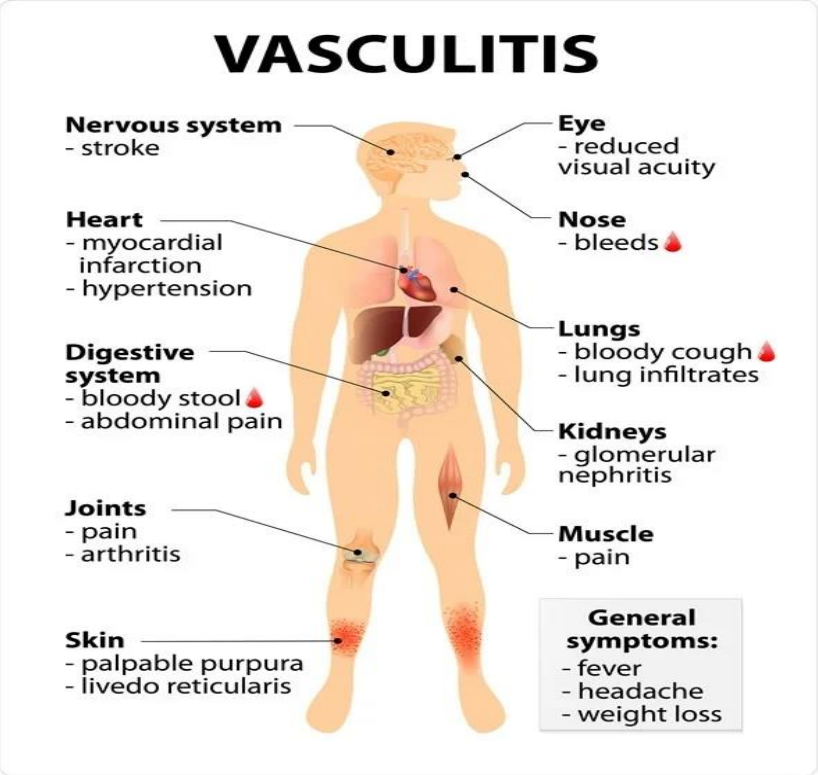

Figure source: https://www.news-medical.net/health/Vasculitis-Affecting-Small-Blood-Vessels-ANCA-Associated-and-Non-ANCA.aspx

What happens after you have been diagnosed with kidney disease due to ANCA-associated vasculitis?

- After your kidney biopsy is reported and a final diagnosis is made, your kidney doctor (nephrologist) and/or kidney care team may order further tests or scans and will discuss appropriate treatment with you.
- You may be referred to other specialists for assessment of blood vessel damage in the lungs, nerves, sinuses or eyes. For example, you may be referred to a rheumatologist or respirologist.

How is ANCA-associated vasculitis treated?

The aim of treatment is to lower the production of ANCA and stop the immune system from causing further damage. The treatment for those with kidney involvement is usually conducted in two phases:

- **The induction (or initial) phase** of treatment will start shortly after the diagnosis is made.
- **The medications in this phase may include:**
  - Corticosteroids and/or avacopan
  - In most cases, cyclophosphamide or rituximab may be added to the above medications
- **The maintenance phase** of treatment will start after about 3-6 months of initiating induction treatment. Medications will be chosen to prevent relapses of vasculitis.
- **The medications in this phase may include:**
  - Azathioprine
  - Rituximab

- During this phase of treatment, you are at high risk of bacterial and viral infection. If you develop a fever, please seek medical attention immediately.
    - Mycophenolate mofetil
- Treatment with these medications will continue for at least 2 years and may be extended for a longer period. Length of treatment is individualized based on:
    - Type of ANCA vasculitis
    - Disease severity and frequency of relapses
    - Response to treatment

Other medications may become available for controlling ANCA vasculitis over the coming years as more research is being done in this area.

General therapies:

- You may be started on a combination of trimethoprim and sulfamethoxazole, antibiotics that are used to reduce the risk of very serious infection that occurs in patients on medications that interfere with the immune system.
- Medications to control swelling (diuretics), blood pressure, and blood clotting may be initiated depending on the severity of kidney involvement.
- Plasmapheresis (plasma exchange)[1] may be recommended to remove ANCA antibodies in refractory (resistant) cases.
- Your kidney doctor and/or kidney care team will follow you closely with frequent urine and lab tests to monitor your response to treatment and help you manage any symptoms.
- BC Renal covers the cost of a wide range of medications used for ANCA-associated vasculitis treatment.
- Your kidney doctor and/or kidney care team will support you to learn about the medication options that would be best for you.
- **It is important that patients check with their kidney doctor and/or kidney care team before taking any over-the-counter (OTC) medications and natural health products.**
- It is also important for patients with chronic kidney disease like ANCA-associated vasculitis to keep their vaccinations updated, since some treatments may reduce the effectiveness of vaccination. You are encouraged to discuss with your kidney doctor and/or kidney care team what vaccinations may be appropriate for you.
- Sometimes, people living with ANCA-associated vasculitis may be invited to participate in a clinical trial for new therapies and medications. If you choose to volunteer in a trial, your kidney doctor and/or kidney care team will help you navigate the process.

Living with ANCA-associated vasculitis

[1] Plasmapheresis is a procedure in which a machine is used to separate the plasma (the liquid part of the blood) from the blood cells. After the plasma is separated from the blood cells, the blood cells are mixed with a liquid to replace the plasma and are returned to the body. Plasmapheresis is often done to remove extra antibodies, abnormal proteins, or other harmful substances from the blood. It may be used to treat certain types of blood disorders, autoimmune disorders, nervous system disorders, or other conditions. Also called plasma exchange. Source: https://www.cancer.gov/publications/dictionaries/cancer-terms/def/plasmapheresis.

- BC kidney patients registered with BC Renal have access to a comprehensive kidney care clinic team that includes nurses, dietitians, social workers. In most cases, patients will also have access to a pharmacist.
- It will be important for you to stay active and healthy. The Kidney Foundation's online Kidney Wellness Hub (https://kidneywellnesshub.ca/) has a lot of useful information. It covers staying active, eating well, mental wellbeing, and socially connecting, including peer support groups. It also provides online classes, webinar recordings, and activity suggestions for patients of all ability levels.
- Though most patients treated for ANCA-associated vasculitis with immunosuppression medication improve, the disease course is unpredictable. Relapses are common, even many years after ANCA-associated vasculitis is diagnosed. You will need ongoing follow-up with your kidney doctor and/or kidney care team.
- There is a portion of ANCA-associated vasculitis patients who will end up with kidney failure, despite treatment. If your kidneys fail, your treatment options may include transplant and dialysis. Your kidney doctor and/or kidney care team will be there to educate and support you throughout your journey.

Further information

- There may be a lot of confusing information about ANCA-associated vasculitis and other kidney diseases on the Internet. The following websites are good sources of information for people living with this disease:
    - The Kidney Foundation of Canada - https://kidney.ca/
    - Kidney Wellness Hub - https://kidneywellnesshub.ca/
    - BC Renal GN web page - http://www.bcrenal.ca/health-info/kidney-care/glomerulonephritis
    - Vasculitis Foundation Canada - https://vasculitis.ca/
- If you continue to have questions about your condition or treatment, please keep track of these questions and ask your kidney doctor and/or your kidney care team.